A TEXTBOOK

OF

NERVOUS DISEASES

FOR STUDENTS AND PRACTISING PHYSICIANS

IN THIRTY LECTURES

BY

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ONLY AUTHORIZED TRANSLATION

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WITH ONE-HUNDRED AND ELEVEN ILLUSTRATIONS IN THE TEXT

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Preface

When it was suggested by my publishers that I should write a text-book of nervous diseases adapted to the needs of students and practical physicians; in size between the summary compendiums and outlines and the large reference works and handbooks of neurology, I found upon inquiry among my colleagues so general an agreement that such a book was desirable, that I no longer hesitated to undertake its preparation. When I decided to present my material in the form of lectures, it was because of the recollection that in my student days, the reading of text-books in lecture form was to my fellow students and myself like an oasis in the midst of the tiresome work of preparing for examinations. On the other hand, I did not conceal from myself that the systematic arrangement of the material, as compared to its presentation in lectures, had in general the advantage of greater clearness and better adaptation for quick reference. On this account I have endeavored to combine the advantages of both methods of arrangement, by the introduction of titles, summaries in tabular form, and recapitulatory headings. I have also laid great stress upon a complete index.

In the arrangement of the subject matter, I have subordinated the customary topographical and pathologico-anatomical classification almost entirely to the etiological and pathologico-physiological, as a glance at the table of contents will show: I was so enabled, for example, by grouping together the syphilogenic diseases of the whole central nervous system, through the consideration in common of the most varied disturbances of conduction in the territory of the peripheral nerves, by the clinical synthesis of the diverse varieties of spinal "Transverse lesions," by handling in common the "Dyskinesias," the "Dysglandular Symptom-complexes," etc., to avoid much repetition. This was all the more necessary, since in spite of my plan of limiting the size of my text-book, I found it necessary to treat somewhat at length, subjects of special importance to the practicing physician; in fact in some instances to go more into detail than has so far been done in the larger works: so for example, as to the nervous manifestations of arteriosclerosis, the treatment of tabes, of neuritis and neuralgia, the psychology of the neurasthenic, etc.

Further in favor of this plan, was the fact that with regard to localization; by referring to my "Compendium of Topical Diagnosis of Diseases of the Brain and Spinal Cord" from which a few loans have been obtained, I have been enabled to treat the matter in a manner very condensed, but adequate to the scope of this book.

With all justice to my endeavor to present particularly my own experiences and my own views, I believe that I have been careful enough in imparting the

observations and opinions of other authors to hope that my book may present to the non-neurologist a well-rounded view of our specialty, which unfortunately still suffers in some places from insufficient consideration in the clinical courses of instruction.

In this connection I have refrained from forcing to the front the views of any particular school, rather have I endeavored to exercise a certain eclecticism, which is justly considered as the scientific signature of our country, in which the German and the Latin manner of thought and investigation blend, and reinforce one another in harmonious fashion. With regard to the illustrations of my book, it was not my intention to prepare an atlas of rare and atypical cases, but rather to bring before the reader the instructive and characteristic of the more important morphological anomalies. Where my own photographs and sketches have been insufficient, I have been aided in the most friendly manner by my colleagues who have put their material at my disposal. I am indebted in the highest degree to Doctors *Eduard Hagenbach*, *F. de Quervain*, *R. Stähelin*, *E. Wieland*, *E. Villiger*, *B. Bloch*, *P. Knapp*, and *H. Iselin* of Basle, *K. Frey* and *E. Bircher* of Aarau, *E. Ruppaner* of Samaden. All the photomicrographs were kindly prepared by my Assistant *Dr. Leuenberger* from preparations of my collection.

Basle THE AUTHOR.

In translating this thoroughly useful, practical and up-to-date book, it has been endeavored to present the author's views as nearly in his own language as is consistent with clear and readable English. An occasional foot-note of explanation, or of reference to matters which have come up since the German Edition was published, has been added.

TRANSLATOR.

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LECTURE I

GENTLEMEN: In this course of lectures I will endeavor to guide you in a way differing somewhat from the customary one, through the extensive and interesting subject of clinical neurology. I will not treat separately the general symptomatology and diagnosis of nervous diseases, neither will I precede the account of individual affections by a special consideration of their pathology as is customary in most text-books. It seems to me more correct didactically, to proceed directly, without the usual introductory remarks, to the study of the clinical pictures and to describe the methods of examination and the interpretation of the different neurological symptoms and syndromes, as digressions at the points where they fit most naturally and with least compulsion, into the general presentation of the subject. The same considerations apply to the anatomical, physiological and pathological data, which are indispensable for an understanding of the clinical material. However, care will be taken that the treatment of these matters is sufficiently restricted not to interfere with the clinically useful character of the book.

Since practical neurology is to be treated, and above everything the point of view of the non-specialist is to be borne in mind, rare affections are brought forward and described it is true, but those diseases of the nervous system which on account of their frequency are met with in daily practice are more particularly considered and carefully described. We will also lay great stress upon questions of treatment and will endeavor to prevent occupying ourselves so far with the "School Cases" and their great symptom-complexes, that the "minor" neurology of office practice, an important and thankful field for the development and exercise of professional ability and therapeutic tact, is not forced into the background.

Diseases of the Peripheral Nerves

The affections of the peripheral nerves agree so generally in their diagnostic relations, that we can readily consider their symptoms from two clinical points of view. Without doing violence to the facts, we will hence consider in common all disturbances of conduction, whether of traumatic, neuritic or neoplastic origin, and after that will take up peripheral nervous irritative symptoms, which in part have their origin in neuritic processes, in part as neuralgias, can claim a certain clinical autonomy.

Considered on the contrary from the standpoint of pathology, the diseases of the peripheral nerves present a great multiplicity of clinical pictures, as many

in fact as there are peripheral nerves, and their recognition and treatment presupposes intimate familiarity with anatomical and physiological data.

In order therefore to adhere to our plan and to avoid burdening you with tiresome repetition and unnecessary details, I will consider the material under the following divisions:

1. The peripheral disturbances of nerve conduction.
   a. In the spinal nerves.
   b. In the cranial nerves.
2. Special remarks on neuritis and polyneuritis.
3. The neuralgias.
4. The treatment of the diseases of the peripheral nerves.

A. Disturbances of Conduction

Etiology

Traumatic influences of various kinds play a very important rôle as starting points for destructive lesions of the peripheral nerves. In the first place we would mention wounding or severing of the nerve trunks as they occur in the extremities in stab or incised wounds, from penetrating projectiles, sometimes from the splinters of a fractured bone, etc. A single severe compression if it affects nerves exposed in a superficial position may also cause interruption of conduction, so for example in one of my cases a musculo-spiral paralysis was caused by the patient being struck over the region where this nerve passes around the outside of the humerus, by a falling boiler. Most frequently, however, the duration of the compression is longer. So, paralyses of the brachial plexus are produced by the pressure of a crutch, by the nerve plexus being caught under the clavicle in over-elevation of the arm in prolonged narcoses, or as a result of dislocations of the shoulder. Paralyses of the musculo-spiral nerve are sometimes produced in prisoners by too tightly tying their arms.* Peroneal paralysis has been known to arise in agricultural laborers from prolonged squatting; in fact falling asleep with the knees crossed has been sometimes known to seriously injure the compressed peroneus. Other "sleep-paralyses" are seen not infrequently in the musculo-spiral and ulnar nerves, in persons who sleep at night, the head upon the outstretched arm, or in those who enjoy a noonday nap their elbows resting upon the table. On account of constantly supporting the elbows against the hard table edge, "professional paralyses" of the ulnar nerve occur in glass workers, xylographers, telephone operators, etc.

A cervical rib can produce paralysis of the dilator pupillæ and tarsalis superior muscles, by pressure upon the sympathetic nerve in the neck. Too tightly put in plaster, in extension apparatus, compression by growing bone callus or by a tumor, the application of the *Esmarch* bandage in checking hemorrhage, may, on account of their relative frequency, be mentioned from

* This refers to the habit of the Continental Police of pinioning the prisoner's arms behind his back. (Translator.)

an almost endless list of causes of compression paralysis. Finally, strain and overstretching of nerve trunks can cause a break in conduction, which, not to speak of obstetrical paralysis in children forcibly extracted, plays a part in accident practice. For example, I saw in a stableman a serratus paralysis produced by a horse whose hind leg he was holding to be shod, suddenly pulling it loose; in a plasterer, median paralysis due to the sudden jerking out of his hand of a rope by the fall of a heavy box of plaster to which it was attached, etc.

"Neuritic processes" represent the second subvariety of disturbances of peripheral nerve conduction. Indeed, the term neuritis, though sanctioned by clinical use, cannot stand a strictly scientific criticism. Rather do we find in this group of diseases, not only inflammatory, but also primarily degenerative processes, and we can assure ourselves that even in histopathological relations, as we will see directly, the lines between these two categories cannot be too sharply drawn. The list of causes which have been held responsible for neuritis is varied enough. Particularly often are there in the history, statements about exposure to cold. Vague and unsatisfactory as this idea is, we cannot get rid of it. To select one example: in the so-called rheumatic facial paralysis, in which, according to the findings of *Minkowski*, *Dejerine* and *Theohari* and others, there is a neuritic or degenerative process affecting particularly the peripheral portions of the facial nerve, *Remak* found in 45%, I in 58%, and other neurologists in as high as 70% of the cases, a history of exposure to cold, which not rarely appeared to have acted specially upon the affected side (a draft). Neuritis from overuse has undoubtedly been demonstrated. *Frank Smith* first described a "Hammer Palsy" which he saw develop in the right arms of smiths in the Sheffield Iron Works, after excessive exertion; *Coester* called attention to an atrophy of the small muscles of the hand, with pain in the median distribution, which occurs in cigar-makers and a similar involvement has been observed in milkmaids, drummers and locksmiths; musculo-spiral paralysis is found in weavers, ulnar paralysis in oarsmen, etc.*

Now, however, in these neuritides from cold and from overfunction, it is very much of a question if these factors are exclusively responsible for the production of degenerative processes in the affected nerves. Very often further inquiry develops the fact that the real cause of the disease lies deeper, that for example alcoholism, disturbance of nutrition, or infectious diseases, have so modified the nervous system of the patient that these accidental noxious influences can exert a pathogenic action. When on any account, metabolism in nerves is so altered that material for the replacement of the constituents used

* These neuritides from overuse have been grouped, together with the pressure paralyses of certain nerves, in persons of definite callings as "professional pareses." To these must be added certain "atrophies from overuse," which probably do not depend upon neuritic processes, but are of myopathic nature, since the affected muscles do not correspond in their grouping to those supplied by a definite nerve, the electric examination gives no indication of the neurogenic origin of the symptoms, neither are there pain, sensitiveness to pressure in the nerve, nor sensory disturbances. For example, in glass-blowers, an atrophy of the muscles of the cheeks has been observed, and I saw in a somewhat anemic dentist an atrophy of the deltoid muscle, apparently from constantly holding up the electric drill.

up in functionating, is available in less quantity than normal, as *Edinger* has shown, the addition of noxious influences in themselves slight, can so disturb the already unstable trophic equilibrium, that clinical and anatomical manifestations of disease are produced.

On the other hand, in the cases in which, in a previously healthy person, after severe chilling, a neuritis develops, as in other rheumatic troubles, the supposition that the injurious influence of the refrigeration through reduction of the general and local resistance of the organism has opened the door to some sort of infectious process, and has only indirectly acted in producing organic lesion, is altogether plausible.

In toxic and infectious neuritides, as well as in those occurring in the course of general nutritional and metabolic disturbances, we can base our views as to etiology upon a firmer foundation. Of all exogenic poisons alcohol stands at the head, indeed not in the sense that a single alcoholic intoxication, even the most intense, is able to set up neuritic processes, but rather that chronic alcoholism in part causes neuritides by itself alone, and in part produces a very decided predisposition to them, on which account, for example among the victims of "professional pareses," drinkers are present in very high percentage. *Oppenheim* hence speaks with justice of "toxico-professional" paralyses. Chronic lead poisoning also plays an important rôle; type-setters, painters, glaziers, type-founders, and polishers, etc., are, as is well known, particularly exposed, and these callings also furnish their contingent to the toxico-professional cases. Contrary to alcohol and lead, arsenic can set up neuritides after acute poisoning. (I saw a case after an attempt at suicide.) Other poisons which may be mentioned here are, ethereal oils, carbon bisulphide, carbonic oxide, dinitro-benzol, anilin, phosphorus, mercury, copper and silver. In one of my cases, in a worker at a factory for chemicals, combined intoxication, with the vapors of bromide of methyl and methyl alcohol had preceded the outbreak of the polyneuritic symptoms. Among infectious diseases which can set up neuritis are to be mentioned typhoid fever, influenza, pneumonia, erysipelas, diphtheria, gonorrhea, septicemia (puerperal), malaria, relapsing fever, syphilis, amebic dysentery, and tuberculosis. In these, the respective bacteria are not to be found in the affected nerves, so that only toxine action can come into question. Lepra alone produces multiple foci in the peripheral nervous system by penetration and proliferation of bacilli; since, however, the clinical picture of this disease in no way agrees with that of the other neuritides, but presents considerable similarity to that of syringomyelia, we will put off its consideration to the chapter devoted to this disease of the spinal cord (Lecture X).

Cachexia and autointoxication also furnish a frequent basis for the development of neuritic phenomena: carcinomatosis, diabetes, gout, anemia, senile marasmus, pregnancy,* leucemia, chlorosis. By local poisoning, neuritis occurs after ether injections in the neighborhood of a nerve, by local infection (with bacterial invasion which often ascends along the nerves) in connection with

* The very frequent accompaniment of the neuritides of pregnancy by pernicious vomiting (hyperemesis gravidarum), speaks for their autotoxic origin.

septic wounds, phlegmons, etc., by local disturbance of nutrition, in the distribution of vessels with arteritic or arteriosclerosic lesions.

It is unexplained whether the form of polyneuritis known as Beri-beri or Kakke, endemic in southeast Asia, Japan and Brazil, is to be attributed to a poison which is formed outside the body and introduced with the rice eaten, or to a toxine formed in the body itself by microbes which have entered it, that is, whether this affection is to be counted among the toxic or among the infectious neuritides.

Finally, after traumatic and neuritic influences, the third and last category of causes of disturbances of conduction in the peripheral nerves, tumor formation, must be considered. In this, mainly true neoplasms, seldom the infectious granulomata—for example, gummata—are concerned. Carcinomata frequently involve the neighboring nerve trunks, cancer metastases in the axilla for example attacking the brachial plexus. Sarcomata, fibromata, myxomata, lipomata, may arise from the nerve itself. These are designated as "neuromata," though it is not entirely proven that the nerve tissue itself takes part in the tumor formation. However, the nerve connective tissue plays the chief part in the neoplastic proliferation.

Besides solitary "neuromata," * there are plexiform neuromata which develop along the whole of a nerve trunk or branch, and finally a "general neuromatosis" (*Recklinghausen's* disease). While these latter often cause pain, it is remarkable that they only infrequently cause any considerable disturbance of conduction in the affected nerves, though on account of the deformity which they produce and also for the reason that fibromatous growths show considerable tendency to undergo sarcomatous degeneration, they are of surgical importance.

Pathogenesis and Pathological Anatomy

The injury of a nerve at any part of its course, has as a result degenerative changes in its parenchyma: the axis cylinder shows the earliest changes, it becomes tortuous, varicose, and at length breaks up into very fine granules. Soon after, the medullary sheath begins to undergo changes, the myelin of which it is composed breaks up into fatty droplets which are at length absorbed. Certain proliferative changes in the interstitial tissues proceed parallel with the degeneration of the parenchyma; the sheath of *Schwann* shows considerable increase in its nuclei, which arrange themselves in columns.

According to the law of *Waller*, "A nerve fiber can only preserve its anatomical and physiological integrity, while it remains in uninterrupted connection with its living cell of origin"—the degeneration described occurs throughout the peripheral portion of the severed nerve. The same changes are usually found, also, for some distance in the central stump.

Of course, the same lesions are found in man after the severing of a nerve; simple squeezing, or compression which lasts longer, produce in the nerve

* Those which have their seats upon the smallest skin nerves seldom give any other symptoms than more or less sensitiveness to pressure; these are the so-called "tubercula dolorosa" of the surgeons.

changes of the same character, but quantitatively less severe. However, the anatomical picture produced by toxemic injury, refrigeration, etc., is often so similar to the traumatic nerve degeneration, that *Babinski* could remark, that apart from tumor-formation, all diseases of the peripheral nerves are to be attributed to neuritis. In spite of this, I cannot decide, like other neurologists, to denominate the disturbances of conduction after external violence as traumatic neuritis, and on this account, in speaking of etiology, I kept the two categories sharply separated. For the rest there are acute infections and toxic neuritides, in which the inflammatory nature of the process is quite plainly marked by extensive infiltration of the connective tissue endoneurium, epineurium and perineurium, by hyperemia of the nerve, diapedesis of white corpuscles, etc. Of clinical importance is the tendency of the sheath of *Schwann* to undergo proliferatory processes as already mentioned; the nuclei arranging themselves in rows, form in the affected parts columns, along which from the central stump regeneration of the axis cylinder, followed by that of the medullary sheath, can occur. In severed nerves, regeneration is possible even years later, as is shown by the results of reunion of the cut fibers by nerve suture undertaken long after the injury.

The General Symptomatology of Interruption of Conduction

Since the peripheral nerves have to conduct stimuli of varied kinds, centripetally the different forms of sensation, centrifugally motor, trophic and vasomotor impulses, and since disturbance of their capacity to conduct can impair any of these, it is necessary here to present to you a short review of the clinical physiology of each of these functions.

I. SENSIBILITY

This term in no way expresses a simple conception. The clinic, however, does not go so far in its analysis as experimental physiology which differentiates a sense of pressure, sense of position, sense of cold, sense of heat, sense of pain, sense of movement, etc. It limits itself rather in the testing of "general sensibility" (*i.e.*, the afferent impulses which do not come from the eye, from the ear and from the gustatory and olfactory apparatus) to the investigation of four chief qualities.

a. Touch sense. We test this by stroking the part with a wisp of cotton, a brush, the finger, etc. Its diminution is called tactile hypoesthesia, its loss, tactile anesthesia. In pathological increase of sensitiveness to touch, whereby this is found painful, we speak of tactile hyperesthesia.

b. Temperature sense. To study this, we test the ability of the subject to distinguish between cold and warm objects. The condition of impaired or absent temperature sense is called thermohypoesthesia or thermoanesthesia as the case may be. The opposite condition is thermic hyperesthesia.

c. Pain sense. Pinpricks, pinching of a fold of skin, etc., furnish information as to this. Diminution = hypalgesia. Loss = analgesia. Increased sensibility to pain is called hyperalgesia.

d. Deep sensibility (also "Bathyesthesia"). In contradistinction to the previously enumerated "Superficial-sensibilities," whose seat is in the integument, we understand under the above name the sum of the centripetal stimuli, which stream to our central nervous organs from the muscles, tendons, bones, joints, etc. A part of these pass the threshold of consciousness in the brain, and inform us as to the position of our limbs, the angles at which our joints are bent (sense of position), the extent of a movement which is being carried out (sense of movement), etc. Here belongs also vibration-sense (pallesthesia), which is felt when a vibrating tuning-fork is placed upon a superficially seated part of the skeleton. Another part of these influences, however, does not come into consciousness, but regulates, subconsciously, the motor mechanisms, which come into action in all complicated and combined movements, such as walking and standing. It makes possible in this way the maintenance of equilibrium, the harmonious course of locomotion, the synergistic action of related muscle groups, etc.

More or less marked defects in deep sensibility manifest themselves by the symptoms of ataxia (hypotaxia), incoördination, asynergy, etc. Since in recognizing an object by feeling it with closed eyes, the sense of position and sense of movement of our fingers plays the chief part, stereoanesthesia (inability to recognize form by palpation) is also an expression of disturbed deep sensibility. Loss of vibration-sense is called pallanesthesia. As *Head* has shown, the fibers conveying deep sensibility run in the muscle nerves, on which account they are uninfluenced in lesions of the cutaneous nerve branches.

II. MOTILITY

Under motility in the broader sense, we understand, not only voluntary movements, but also the phenomena of the tonus and the reflexes.

a. Voluntary movement. Complete abolition of the ability to perform voluntary movements, we denominate paralysis, partial disability, paresis. We speak of a paralysis of individual muscles when a few muscles, of plegias when whole limbs, or at least portions of limbs, are affected; if a single extremity is involved we call it a monoplegia, if two extremities on the same side, a hemiplegia, if two symmetrical extremities, a diplegia (when it is the two legs the term paraplegia is used); an extension to three or to all four limbs becomes respectively triplegia and tetraplegia.

b. Tonus and Reflexes. A mechanical stimulus (striking or stroking) applied to certain parts of our bodies (certain tendons, bones, skin regions) under normal conditions produces certain definite motor phenomena (a contraction of certain muscles); we speak of them as reflexes. Besides this, there stream (probably from the whole surface of our bodies and from all the skeleton) to our central organs, continued subconscious centripetal stimuli of little intensity, which by reflex action keep up a moderate continuous contraction of all our muscles. This is the tonus. We can define this in the following terms: "The definite degree of tension which gives our muscles the ability to reply promptly with a contraction to voluntary impulses which reach them." That in normal life, the stimuli which produce the tonus are continually at

work is proved by the fact that in healthy persons the tonus is never relaxed, even in sleep. The muscles of the sleeper are never so completely relaxed as in the corpse; and only in an advanced stage of narcosis does the tonus give way. Abolition of the reflexes and of the tonus we call respectively areflexia and atonia; diminution of these, hypo-reflexia and hypotonia; for the exaggeration of these phenomena on the other hand the expressions hyper-reflexia and hyper-tonia are used.

I have prepared a table in which the different reflexes and the methods by which they are elicited are exposed, in which the differentiation between skin, tendon, and bone reflexes is shown, and the most important reflexes are emphasized. One can expect to elicit all the reflexes only in very young persons. After puberty the great majority of them cannot be obtained even in perfectly healthy individuals. As to the condition of tonus, we estimate it partly by determining the tension and resistance of the muscles by palpation, partly by noting the resistance which is opposed to passive movements.

Tendon and Bone Reflexes.	Skin Reflexes.	Method of Eliciting.	Result.
1.	Scapular reflex	Irritating the skin over the scapula	Contraction of the muscles over the scapula
2. Biceps reflex	.....	Striking upon the biceps and tendon	Flexion of the forearm
3. Triceps reflex	.....	Striking upon the triceps tendon	Extension of the forearm
4. Scapulo-humeral reflex	.....	Striking upon the inferior internal angle of the scapula	Adduction of the arm
5. Radius reflex	.....	Striking upon the styloid process of the radius	Supination of the forearm
6.	Palmar reflex	Irritation of the palm	Flexion of the fingers
7.	Epigastric reflex	Stroking from the mamma downward	Retraction of the epigastrium
8.	Upper abdominal reflex	Stroking the skin of the upper abdomen	Retraction of the abdomen
9.) 10.)	Middle abdominal and lower abdominal reflex	Stroking the skin of the abdomen in the middle and lower parts	Retraction of the abdomen
11.	Cremasteric reflex	Stroking the inner side of the thigh.	The testicle is jerked upward
12. Patellar reflex	.....	Striking upon the quadriceps tendon	Extension of the leg
13.	Gluteal reflex	Stroking the nates	Contraction of the gluteal muscles
14. Achilles reflex.	.....	Striking upon the Achilles tendon	Extension of the foot
15.	Plantar reflex	Stroking the sole	Flexion of the toes
16.	Anal reflex	Irritating the perineum	Contraction of the external sphincter ani muscle

III. TROPHIC FUNCTIONS

The Law of *Waller* already mentioned applies also to the muscles, for whose anatomical integrity an uninterrupted connection with the cells of the anterior horns of the spinal cord, or in the case of those innervated by the cranial nerves, with the motor nuclei of the brain axis, through their nerves

of supply, is presupposed. If, in a motor nerve, conduction is broken, the muscles supplied by it undergo a degenerative process which we call neurogenic degenerative atrophy. Histologically this is characterized by the disappearance of the contractile element (by granular, albuminous and fatty, less frequently by hyaline and vacuolar, degeneration), and its replacement by fatty and connective tissue. These degenerative processes, under conditions which admit of a reëstablishment of the connection between the muscles and the trophomotor cells of the central nervous system, can be repaired by regenerative processes; the histological criteria of these last are, among other things, the proliferation of the nuclei of the sarcolemma surrounding the muscle fibers, the appearance of polynuclear giant cells, "myoblasts," from which muscle fibrils are formed anew, the presence of very voluminous muscle fibers, etc. A clinical sign of neurogenic atrophy is the notable electro-physiological alteration, first carefully studied by *Wilhelm Erb* in 1872, and called by him "Reaction of Degeneration" (R. D.). To the symptomatology of this we will return farther on in this lecture. Besides this, the central nervous system through the peripheral nerves exerts a trophic influence upon the skeleton; when this is cut off in growing individuals, there is more or less limitation of further bone growth in the affected territory. Abnormal fragility of bones can also occur under these conditions.

IV. VASOMOTOR FUNCTIONS

Experimental physiology teaches us that two sorts of fibers pass from the sympathetic, through the peripheral nerves to the blood vessels, those which narrow ("vasoconstrictors") and those which increase ("vasodilators") the calibre of the vessels.

Dilator fibers indeed are only recognizable in a few nerves, for example, in the sciatic. Vasoconstrictor fibers on the contrary are quite generally recognized as constituents of peripheral nerves. Nevertheless, in man, only rarely (that is only in the earliest stages) are we able to observe the picture which the physiologist sees in animals after destruction of the vasoconstrictor elements, namely, redness and heat of the skin. Much more common is the apparently opposite picture which can follow such an injury, or even without one being definitely determined. Namely, the integument of the parts cut off from vasomotor innervation becomes cyanotic and cold. The cyanosis is explained by the chronic inhibition of the capillary circulation, on account of the removal of the vascular tonus, the coldness, by the changes in nutrition of the skin resulting from this cyanosis. The mechanism of the sweat secretion is very similar to that of the vasomotor innervation, the secretory fibers being mostly closely mixed in with the vasomotor fibers. Their destruction has as a result, lessening or inhibition of the sweat secretion (hypoidrosis or anidrosis). The opposite anomaly is called hyper-idrosis.

After this general diagnostic excursion, we will proceed to the description of the sensory symptoms which accompany the disturbances of conduction in peripheral nerves.

Apart from traumatic severing of nerves, there is seldom total anesthesia

of the integument, more usually there is a general reduction of superficial sensibility. Very frequently, however, the different qualities of sensation are affected in different degrees, so, for example, touch sense is intact, while pain and temperature senses are diminished. In neuritides, very often the disturbances of sensibility are not equally distributed over the whole area supplied by the affected nerves, but increase in intensity toward the periphery, which is perhaps connected with the increased distance of the parts there situated from the trophic centers. In diseases of a number of nerves, as in neuritis of plexuses and in polyneuritis, bathyanesthesia or bathyhypoesthesia can make itself felt in disturbances of sense of position or of movement, namely as ataxia or incoördination. A disturbance of vibration-sense is recognizable when certain nerves are put out of function, particularly over the corresponding bones. Disturbances of deep sensibility are usually accompanied by reduction or loss of tendon reflexes.

The motor symptoms in destructive lesions of nerves, consist in losses of function, varying from slight paresis to complete paralysis. They are always accompanied by reduction of muscular tonus with regional hyporeflexia or areflexia, with more or less marked neurogenic muscular atrophy and with the reaction of degeneration already mentioned, which will now be discussed.

THE REACTION OF DEGENERATION (*Erb*, 1872)

Let us first get clearly the normal electric reactions of the human muscles as they are brought out in the usual clinical-neurological examination. In this, we use the so-called unipolar method of stimulation, in which a large ("indifferent") electrode is placed upon the chest or back of the person being examined, a smaller one (as a rule the 3 cm square electrode, *Stintzing's* "Normal electrode") upon the muscles or nerves to be tested. Contraction of muscles occurs with the Galvanic current, not when it is slowly raised, but on sudden variations of current strength (that is, on making or breaking the galvanic circuit, naturally also, upon conducting through them the faradic current with its rapid alternate making and breaking). With a weak galvanic current, a contraction is obtained only when the negative pole is used as stimulating electrode, and the current is closed; Kathodal closing contraction (KCC). If, now, the current is made stronger and stronger, we obtain next an Anodal closing contraction and an Anodal opening contraction (AnCC and AnOC), (the sequence of these two reactions is different in different individuals, AnCC usually appears before AnOC), only with much stronger current is a Kathodal opening contraction (KOC) obtained. If, now, with the current strength which has sufficed to produce KOC, the current is closed, the negative pole being on the point to be stimulated, we get, not a short contraction, but one which persists while the electrode is applied, Kathodal closing tetanus (KCTe). A still more powerful current is needed to produce Anodal closing tetanus (AnCTe), while in normal man, Anodal opening tetanus (AnOTe) does not occur.

It is self-evident that KOC can only occur when the muscle was not just before the interruption of the current in a condition of tetany; KCTe can be

avoided even with a very strong current, however, if with careful avoidance of sudden current variations, the number of milleamperes necessary to produce KOC are gradually introduced by use of the rheostat. Finally it must be added that between the results of "indirect" (through the nerve) and "direct" (to the muscle itself), galvanic stimulation, there is this difference, that in the last method, the effect of opening the circuit is generally less.

In "Reaction of degeneration" (the clinical physiological criterion of disturbed or interrupted trophic connection between nervous system and muscle), we find in contradistinction to the normal relations described above, the following phenomena. The galvanic and faradic irritability through the nerve grow less and less, until, in case there is complete interruption of conduction in the nerve supplying the affected muscle, by the end of fourteen days it has completely disappeared. Direct faradic irritability of the muscle acts sim-

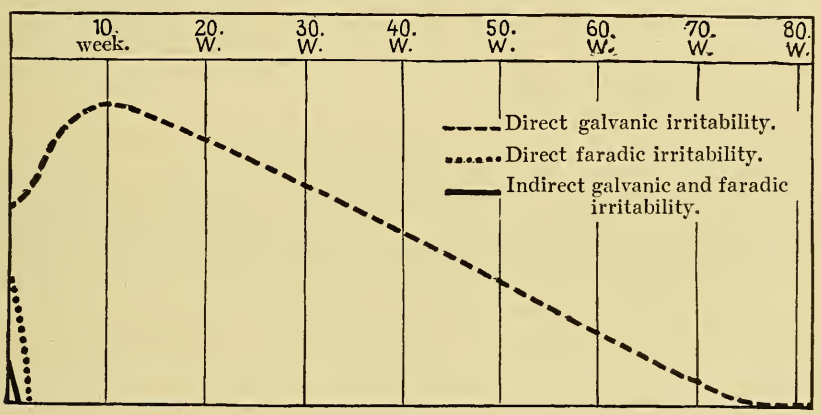


Fig. 1.

Electric Irritability in "Reaction of Degeneration."

ilarly. Direct galvanic irritability, on the contrary, first increases, to fall again at the end of about two months, in case the trophic connection is not in the meantime reëstablished. If conduction is later renewed, galvanic irritability and later faradic irritability of the muscle direct, come back, and at length electric stimulation of the nerve is again followed by contraction. If conduction is not restored, however, after from one to one and a half years, even galvanic irritability of the muscle is permanently lost.

With these quantitative anomalies of irritability, qualitative changes go hand in hand. There is an alteration of the character of the contraction on galvanic stimulation; instead of the normal quick contraction we get a slow "vermicular" response, further a modification of the so-called contraction formula. While normally KCC appears with a weaker current than AnCC in reaction of degeneration this relationship is reversed. We indicate this by the formula AnCC>KCC. Instead of complete reaction of degeneration, we find in slight neurogenic-trophic disturbances of the muscles different varieties of "partial R. D.," to consider all of which would take us too far

afield. *Stintzing* has distinguished no less than thirteen varieties of partial R. D. We must content ourselves with a description of the most frequent of these varieties. In this nerve irritability is preserved, usually faradic irritability of the muscle also, but there is galvanic overirritabilty, reversal of the polar formula and as the most important criterion of the R. D., slow character of the contraction. While in complete R. D. slow return to the normal—in case this occurs—requires from three to seven months, in partial R. D. under proper treatment restitution can begin in six weeks. Very rapid recovery can only be expected in such peripheral paralyses in which R. D. does

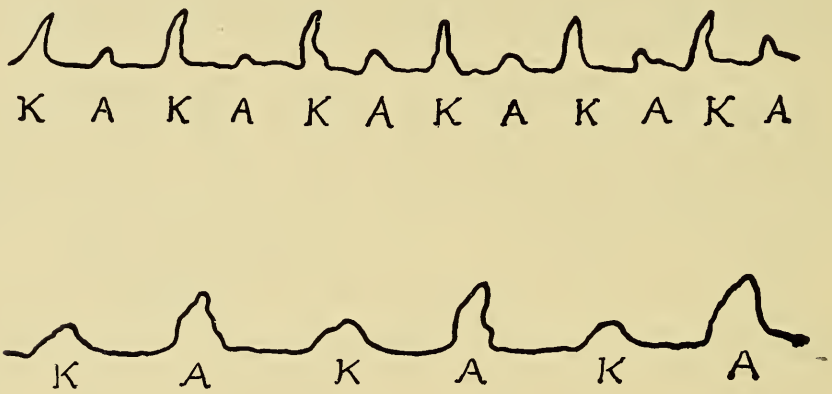


Fig. 2.

Myographic Curves on Direct Stimulation of a Muscle with the Galvanic Current. K = Cathode; A = Anode. Upper curve, normal reaction; lower curve, "Reaction of Degeneration."

not appear, however. The normal irritability of the most important nerves and muscles is shown in the following tables.

According to *Stintzing*, the nerves can be arranged according to their galvanic irritability in milleamperes as follows:

Lower Limit.		Upper Limit.		Average.	
1. N. musculo-cutaneous	0.05	1. N. musculo-cutaneous	0.28	1. N. musculo-cutaneous	0.17
2. N. accessorius	0.10	2. N. accessorius	0.44	2. N. accessorius	0.27
3. N. ulnaris upper	0.2	3. N. ulnaris upper	0.9	3. N. ulnaris upper	0.55
4. N. peroneus	0.2	4. R. mentalis	1.4	4. N. medianus	0.9
5. N. medianus	0.3	5. N. medianus	1.5	5. R. mentalis	0.95
6. N. cruralis	0.4	6. N. cruralis	1.7	6. N. cruralis	1.05
7. N. tibialis	0.4	7. N. peroneus	2.0	7. N. peroneus	1.1
8. R. mentalis	0.5	8. R. zygomaticus	2.0	8. R. zygomaticus	1.4
9. N. ulnaris lower	0.6	9. R. frontalis	2.0	9. R. frontalis	1.45
10. R. zygomaticus	0.8	10. N. tibialis	2.5	10. N. tibialis	1.45
11. R. frontalis	0.9	11. N. facialis	2.5	11. N. ulnaris lower	1.6
12. N. radialis	0.9	12. N. ulnaris lower	2.6	12. N. facialis	1.76
13. N. facialis	1.0	13. N. radialis	2.7	13. N. radialis	1.8

TABLE SHOWING THE IRRITABILITY OF THE MUSCLES ACCORDING TO STINTZING.

Muscle.	Galvanic Irritability in M. A.	Size of the Electrode in cm^2
M. trapezius	1.6	12
M. deltoides	1.2–2.0	12
M. pectoralis major	0.4	6
M. pectoralis minor	0.1–2.5	6
M. serratus magnus	1.0–8.5	12
M. brachialis anticus	1.1–1.7	3
M. extensor communis digitorum	0.6–3.0	3
M. extensor carpi radialis	0.8	3
M. extensor brevis pollicis	1.5–3.5	3
M. pronator radii teres	2.5–2.8	3
M. flexor sublimis digitorum	0.3–1.5	3
M. flexor carpi ulnaris	0.9–2.9	3
M. abductor minimi digiti	2.5	3
M. rectus femoris	1.6–6.0	20
M. vastus internus	0.3–1.3	20
M. tibialis anticus	1.8–5.0	12

Finally as concerns the trophic and vasomotor disturbances, in destructive diseases of the peripheral nervous system, they are less regularly found than might be expected from the fact that through the "gray rami communicantes," sympathetic fibers run into all the peripheral nerves. As already said, redness and local raise of temperature occur only as fleeting symptoms, later in the place of hyperemia, there is cyanosis and abnormal coolness of the integument. Local hyperidrosis is not infrequent. I will mention now the œdema which occurs in infectious polyneuritis. In beri-beri this symptom is so frequent, that we can speak of a hydropic form of this tropical disease, the "moist" form of beri-beri. The skin in all forms of breach of conduction can be shiny and atrophic (the "glossy skin" of the English), and shows then an increased vulnerability, so that the smallest injuries will not heal. Further, vesicular eruptions in the skin areas deprived of innervation, also falling off of nails, collections of air in the nail substance, hypertrichosis or falling out of the hair, and retardation of growth or thinning of bony parts are sometimes seen.

SPECIAL SYMPTOMATOLOGY OF THE DEFECTS OF FUNCTION IN THE DIFFERENT PERIPHERAL NERVES

In order to understand the motor and sensory symptoms corresponding to lesions in the territories of distribution of definite nerves, an accurate acquaintance with the functions of the various peripheral nerves, as well as with those of the muscles supplied by them is, of course, necessary.

A synoptic table which will present this information most expeditiously and in most condensed form is hence introduced at this point.

I. The Spinal Nerves

1. *Motor Functions*

A. Plexus cervicalis

Nervi cervicales	Musculi profundi colli	Flexion, extension, turning the neck
	Mm. scaleni	Raising the ribs on inspiration
N. phrenicus	Diaphragm	Inspiration

B. Plexus brachialis

N. thoracicus ant.	M. pect. maj. and min.	Adduction, and drawing the arm downward and forward
N. thoracic. long.	M. serratus magnus	Fixation of the scapula on raising the arm
N. dorsalis scap.	M. levator scapulæ	Raising the scapula
	Mm. rhomboidei	Drawing the scapula upward and inward
N. suprascap.	M. supraspinatus	Elevation and outward rotation of the arm
	M. infraspinatus	Outward rotation of arm
N. subscapular	M. latissimus dorsi M. teres major	Inward rotation and adduction of the arm toward the back
	M. subscapularis	Inward rotation of arm
N. circumflex (axillaris)	M. deltoid	Raising the arm to a horizontal position
	M. teres minor	Outward rotation of the arm
N. musculo-cutan.	M. biceps bracchi	Flexion and supination of the forearm
	M. coraco-brachialis	Elevation and adduction of the forearm
	M. brachialis anticus	Flexion of the forearm
N. medianus	M. flexor carpi radial	Flexion and drawing to the radial side of the hand
	M. palmaris longus	Flexion of the hand
	M. flexor sublim. digitor	Flexion of the middle phalanges of fingers II-V
N. medianus	M. flexor long. pollicis	Flexion of the end phalanx of the thumb
	M. flexor prof. digit (radial half)	Flexion of the end phalanges of fingers II and III
	M. pronator radii teres	Pronation of hand
	M. abductor brev. pollicis	Abduction of metacarp. I
	M. flex. pollicis brev.	Flexion of proximal phalanx of thumb
	M. opponens pollicis	Opposition of metacarp. I
N. ulnaris	M. flexor carpi ulnaris	Flexion and drawing to the ulnar side of the hand
	M. flexor digit. prof. (ulnar half)	Flexion of end phalanges of fingers IV and V.
	M. adductor pollicis	Adduction of metacarp. I
	Mm. hypothenares	Abduction, opposition, flexion of little finger
N. ulnaris	Mm. lumbricales	Flexion of proximal, extension of other phalanges
	Mm. interossei	Same action as preceding, also spreading apart and bringing together fingers
Nervus radialis (musculo-spiral)	M. triceps bracchi	Extension of forearm
	M. supinator longus*	Flexion of forearm

* The term "supinator longus" is, in fact, incorrect, since, as shown by electric stimulation, the muscle has no effect in causing supination, but produces rather slight pronation. The designation "brachioradialis" would hence be preferable, but though used by anatomists, it has never been adopted by clinicians.

B. Plexus brachialis	M. extensor carpi rad.	Extension and drawing to radial side of hand
	M. extensor digit. comm.	Extension of proximal phalanges of fingers II-V
	M. extensor minim. digit.	Extension of proximal phalanx of little finger
	M. extens. carp. ulnar	Extension and drawing to ulnar side of hand
	M. supinator brevis	Supination of forearm
	M. abductor pollicis long.	Abduction of metacarp. I
	M. extensor pollicis brev.	Extension of proximal phalanx of thumb
	M. extensor pollicis long.	Abduction of metacarp. I and extension of end phalanx of thumb
	M. extensor indicis prop.	Extension of proximal phalanx of index finger
C. Nervi thoracales	Mm. thoracis et abdominis	Raising the ribs, expiration, abdominal pressure, etc.
D. Plexus lumbalis		
N. cruralis	M. ileo-psoas	Flexion of the thigh
	M. sartorius	Inward rotation of the thigh
	M. quadriceps	Extension of leg on thigh
N. obturatorius	M. pectineus M. adductor longus M. adductor brevis M. adductor magnus M. gracilis	Adduction of the thigh
	M. obturator extern.	Adduction and external rotation of the thigh
E. Plexus sacralis		
N. gluteus sup.	M. gluteus med. M. gluteus min.	Abduction and inward rotation of the thigh
	M. tensor fasciæ latæ	Flexion of the thigh
	M. pyriformis	External rotation of the thigh
N. gluteus inf.	M. gluteus max.	Extension of the thigh
N. ischiadicus (sciatic)	M. obturator intern. Mm. gemelli M. quadratus femoris	External rotation of the thigh
	M. biceps femoris M. semitendinosus M. semimembranosus	Flexion of the leg
(a) N. peroneus (*a*) prof.	M. tibialis anticus	Dorsal flexion and supination of the foot
	M. extens. long. digit	Extension of the toes
	M. extens. halluc. long.	Extension of great toe
	M. extens. digit. brev.	Extension of the toes
	M. extens. halluc. brev.	Extension of great toe
(*b*) superf.	Mm. peronei	Dorsal flexion and pronation of foot
(b) N. tibialis (ant.)	M gastrocnemius M. soleus	Plantar flexion of the foot
	M. tibialis posticus	Adduction of the foot
	M. flexor long. digit.	Flexion of the end phalanges II-V
	M. flex. halluc. long.	Flexion of end phalanx I
	M. flex. brev. digit.	Flexion of the middle phalanges II-V
	M. flex. halluc. brev.	Flexion of middle phalanx I
	Mm. plantares pedis	Spreading, bringing together and flexion of the proximal phalanges of the toes
N. pudendus	Mm. perinei et sphincteres	Sphincter muscles of the pelvic organs, co-operation in the sexual act

In the first column of this table will be found the names of the different nerves, in the second, those of the muscles supplied by them, in the third the movements which are produced when these muscles act. The ability or inability to carry out these movements informs us as to the state of the respective muscles, whether they are paralyzed or not. The investigation of course must include a study of the electric contractility as well as that of voluntary movement. For this, an acquaintance with the electric motor points of the muscles is necessary. These are shown—as they usually occur in normal persons—in Figures 3 to 8. In the investigation of pathological

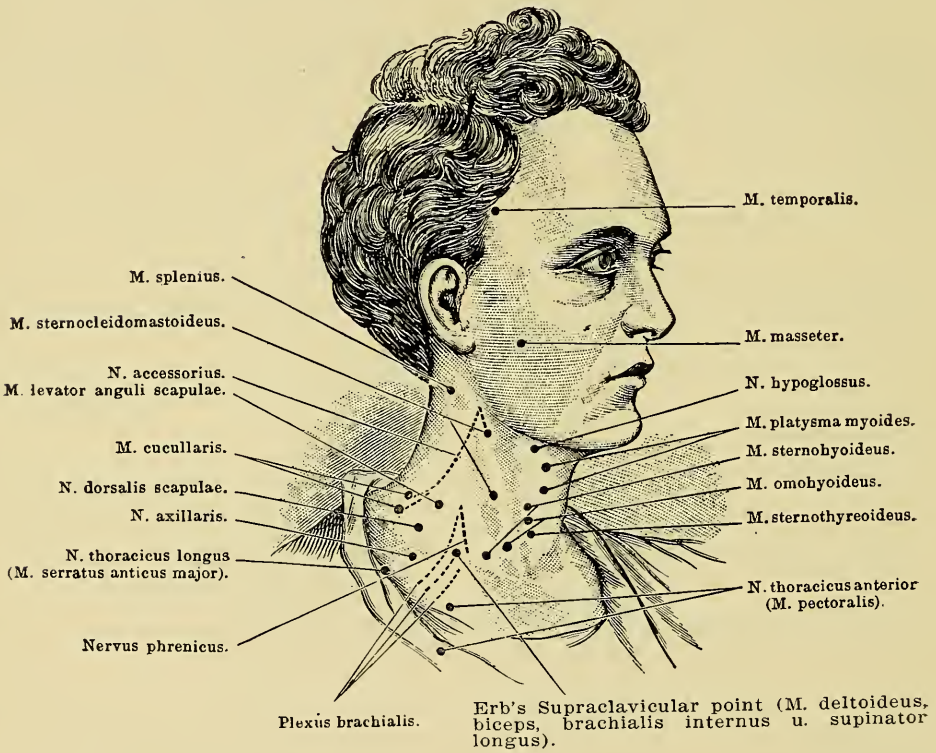


Fig. 3.

Motor Points of the Neck and of the Trigeminus. *Erb's* Supraclavicular Point.

cases the fact, first discovered by *Wertheim-Salomonson*, that in muscles showing RD, there is "transposition of the motor points" should not be forgotten. The affected muscle reacts from points other than those given in the tables, usually more distally located.

2. *The Most Important Types of Paralysis*

We will now pass briefly in review the clinical pictures of the most important paralyses of the spinal nerves.

a. In total paralysis of the brachial plexus, the whole arm hangs flaccid and immovable, the muscles of the shoulder with the exception of the trapezius,

are also incapable of functionating. In the upper arm type (also called the *Duchenne-Erb* type) of paralysis, as a rule only the biceps, deltoid, brachialis anticus and supinator longus are affected: elevation of the arm and flexion at the elbow are hence impossible. Only exceptionally does the paralysis affect one or more of the following muscles: Infraspinatus, latissimus dorsi, teres major, pectoralis major, in which event the arm hangs down in a position of adduction and rotation inwards. The lower arm type of paralysis (*Klumpke's* paralysis) is rare. It is often limited to the muscles of the ball of the thumb and little finger and to the interossei; occasionally, however,

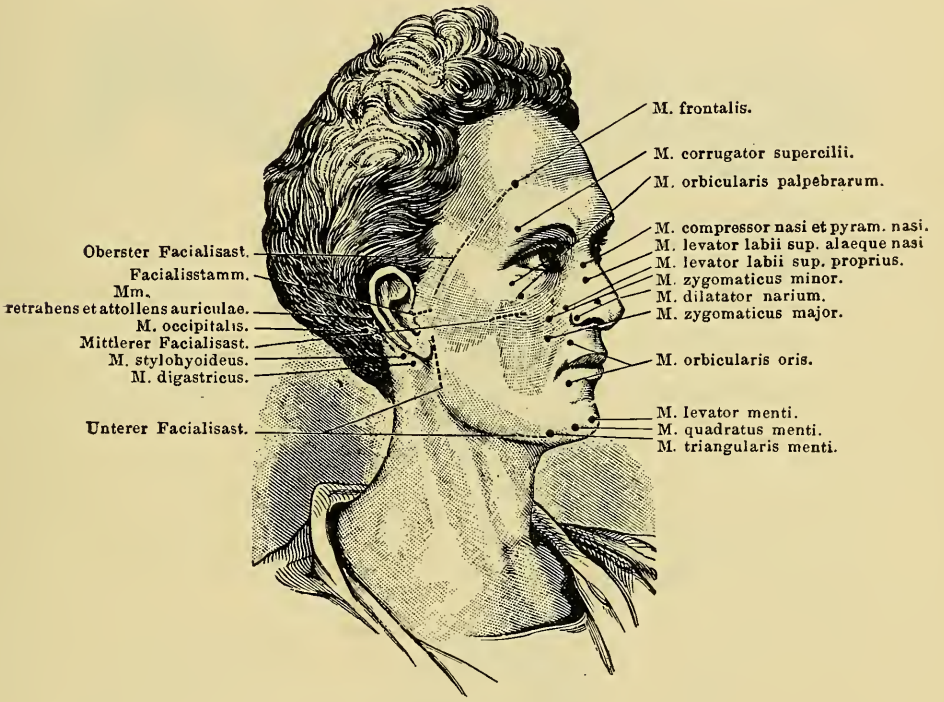


Fig. 4.

Motor Points of the Face.

affects some of the flexor muscles of the forearm and on account of accompanying lesion of the sympathetic fibers (the ramus communicans of the first dorsal nerve) is apt to be accompanied by a narrowing of the pupil on the affected side (paralysis of the dilator pupillæ muscle).

b. In paralysis of the long thoracic nerve, the arm can no longer be raised above the horizontal by the action of the serratus magnus muscle; if the patient extends it forward, a wing-like deviation of the scapula from the thorax is noticed ("scapula alata" or winged scapula, see Fig. 9).

c. Paralysis of the suprascapular nerve, on account of the loss of function of the supraspinatus and infraspinatus, hinders external rotation of the arm, which can be very disturbing, for instance in writing; besides this, the atrophy of the affected muscles is very apparent. (See Fig. 10.)

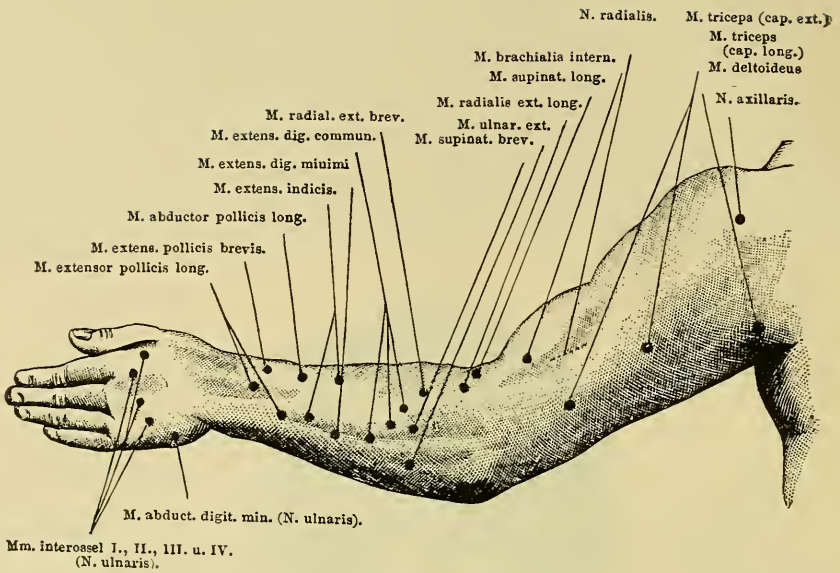


Fig. 5.

Motor Points of the Upper Extremity, Outer Side.

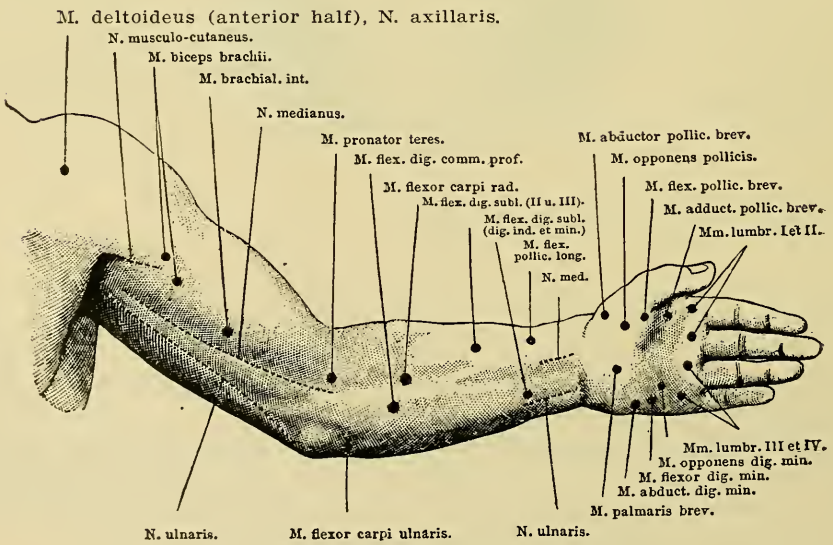


Fig. 6.

Motor Points of the Upper Extremity, Inner Side.

d. Paralysis of the circumflex nerve puts the deltoid out of function; the arm cannot be raised either laterally, anteriorly or posteriorly. The rounding of the shoulder disappears and the contours of the head of the humerus and the acromion process become visible.

e. Paralysis of the musculocutaneous nerve causes great reduction of the ability to flex the forearm at the elbow. Flexion is only possible with the

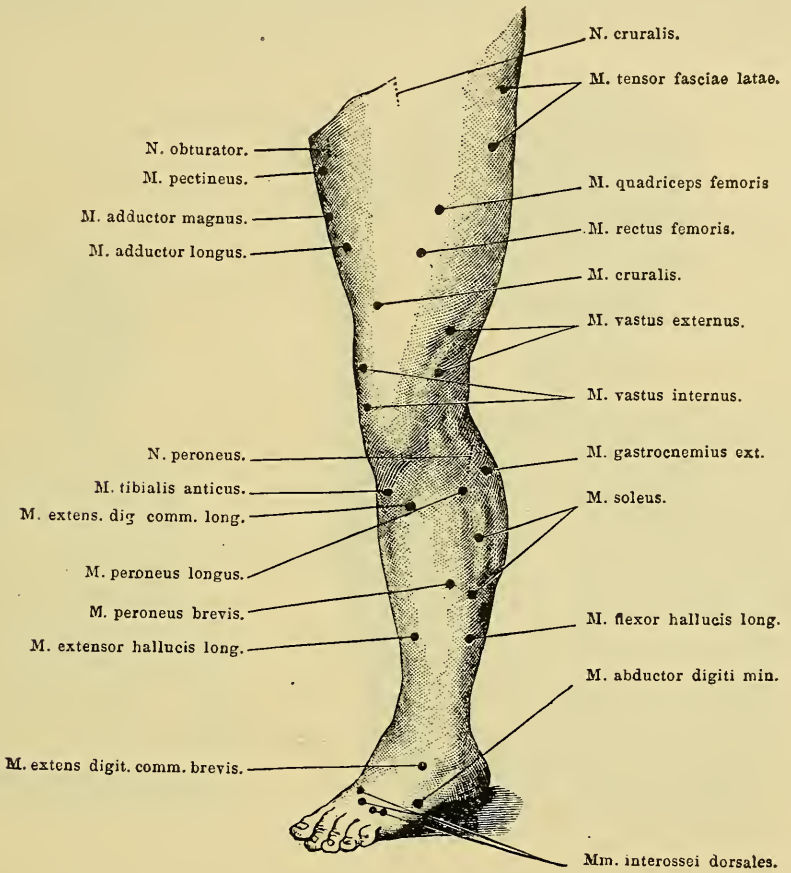


Fig. 7.

Motor Points of the Lower Extremity, Anterior View.

forearm in pronation, in which position, the supinator longus and the flexors of the fingers can to a certain extent take the place of the paralyzed muscles (biceps and brachialis anticus).

f. Paralysis of the median nerve is made evident by the following symptoms: weakness in flexion at the wrist with deflection toward the ulnar side; inability to bend the thumb or to exert its action as opponent of the fingers; substitution of inward rotation of the arm for pronation of the forearm; interference with flexion of the index finger (in that this can only be accomplished in the metacarpo-phalangeal joint by the action of the interosseus);

considerable impairment of the power of grasping and holding on to objects, for which only the three fingers on the ulnar side can be used.

g. Particularly typical, and to be diagnosed at the first glance is ulnar paralysis. (See Figs. 11 to 13.) Flexion, especially of the ulnar side of the hand, is weak, on account of the lack of action of the flexor carpi ulnaris muscle, the movements of the little finger are impossible, in the index, middle and ring fingers, the proximal phalanges cannot be flexed, the distal phalanges cannot be extended, on which account, when the trouble lasts long, the so-called

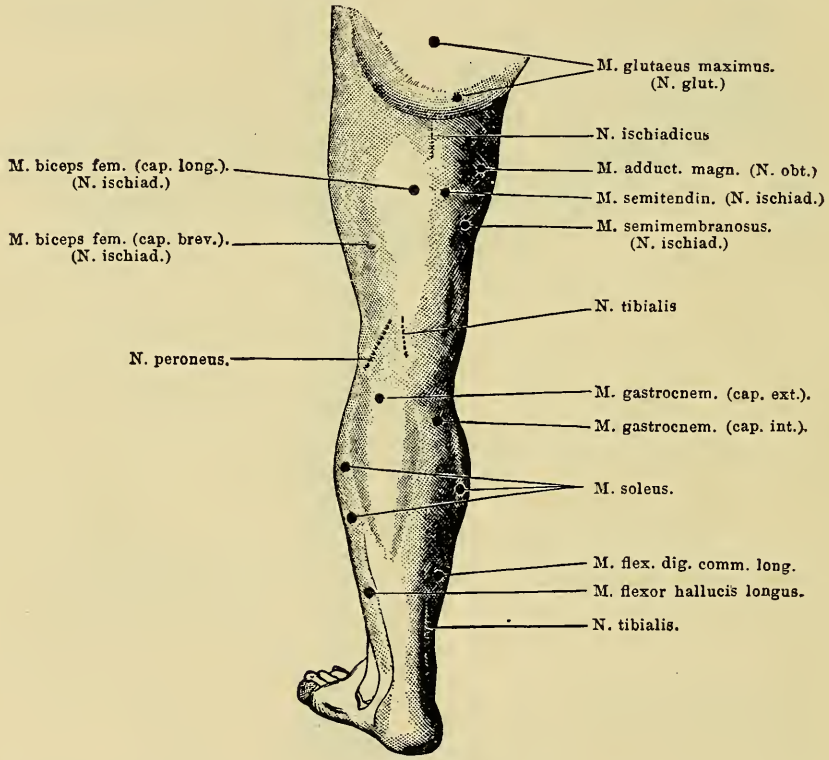


Fig. 8.

Motor Points of the Lower Extremity, Posterior View.

"claw hand" is produced by the action of the antagonists (extension at the metacarpo-phalangeal—flexion at the inter-phalangeal joints). The paralysis of the interossei also causes inability to spread the fingers and to bring them together again. On account of the preponderance of the action of its abductors over that of the paralyzed adductor pollicis, the thumb is continuously separated from the index finger. When atrophy sets in, the sinking in of the interosseous spaces and the thinning of the hypothenar region are particularly noticeable.

h. Paralysis of the musculo-spiral nerve results in inability to extend the hand at the wrist-joint, the hand hence hangs helpless down and cannot be raised ("wrist drop," see Fig. 14). When the seat of the lesion is high up,

extension at the elbow, by the action of the triceps is also impossible; of course in testing this function, the arm must be put in a position eliminating the action of gravity on the elbow-joint. Since normally, complete flexion of the fingers (as in squeezing the hand) is aided by extending the hand at the wrist, in musculo-spiral paralysis the strength of the flexors of the fingers is considerably diminished, unless the wrist is passively extended beforehand. The thumb is held in a position of opposition and falls somewhat forward. The forearm usually takes the position of pronation. The paralysis of the supinator

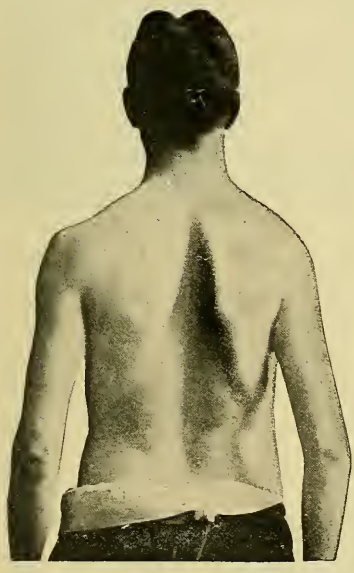


Fig. 9.

Paralysis of the Serratus on the Right Side ("Winged Scapula").

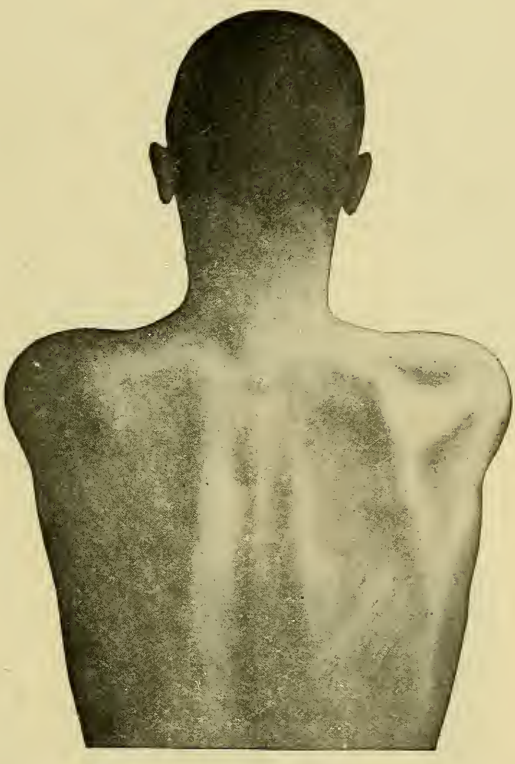


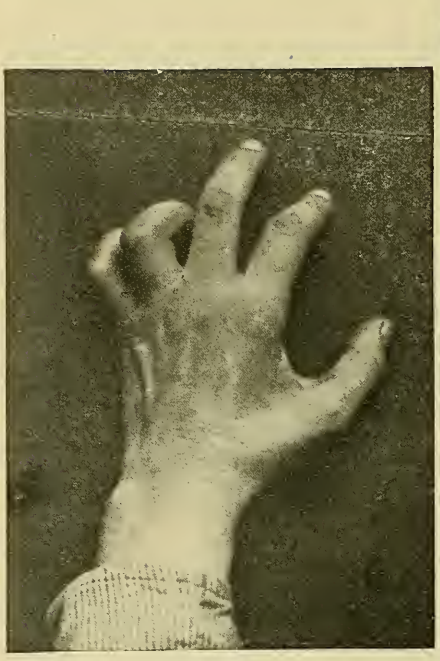
Fig. 10.

Paralysis of the Suprascapular Nerve on the Right Side, Atrophy of the Supra- and Infraspinatus.

longus causes a weakening of flexion at the elbow joint, also when this movement is attempted, the normally prominent outline of the supinator no longer shows up.

i. In paralysis of the anterior crural nerve, the thigh cannot be flexed, the leg cannot be extended; the thigh muscles atrophy. Walking with extension at the knee is indeed possible, in that the patient uses his leg extended by gravity like a stilt, but he cannot climb the stairs with it.

j. Paralysis of the tibialis posticus nerve prevents flexion of the toes, the patient cannot raise himself on his toes and if the paralysis lasts long, talipes calcaneus may develop. The toes can neither be spread apart nor bent.



Figs. 11 and 12.

Ulnar Paralysis (trauma), "Claw-hand."

Fig. 13.

Ulnar Paralysis (neuritis).

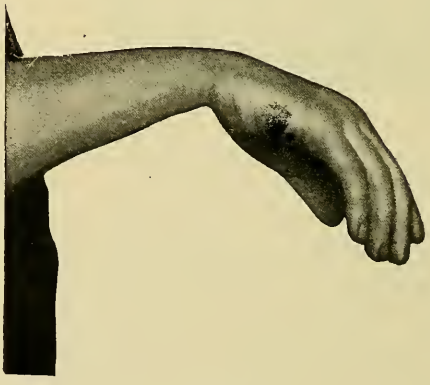


Fig. 14.

"Wrist Drop" in Paralysis of the Radial Nerve; the patient is unable to extend the hand and fingers.

Sometimes there develops the analogue of what is observed in the hand in ulnar paralysis, a "claw foot."

k. In peroneal paralysis the foot hangs in the so-called "drop foot" position and at the same time somewhat supinated (pes equino-varus). The toes stand flexed at the interphalangeal joint. Since the patient must lift his foot abnormally high in order to prevent the toes dragging on the ground, there is a characteristic gait, which *Charcot* has called "steppage" from its resemblance to the action of an English thoroughbred horse.

Fig. 15.

Traumatic Paralysis of the Peroneal Nerve.

The tables given on pages 14 and 15 will furnish sufficient information about the paralyses of such spinal nerves as have not been specially described here, and also with regard to cases in which the territories of the nerves mentioned are only partially affected.

3. *Areas of Distribution of the Spinal Sensory Nerves and Peripheral Anesthesias*

Figs. 16 and 17 show the skin areas of the different sensory nerve trunks and their branches. It must, however, be remarked that the distribution of

single skin nerves is quite variable; the limits can show considerable shifting in favor of the one or of the other nerve. It is to be emphasized also that in the etiologically most different varieties of interference with conduction, even

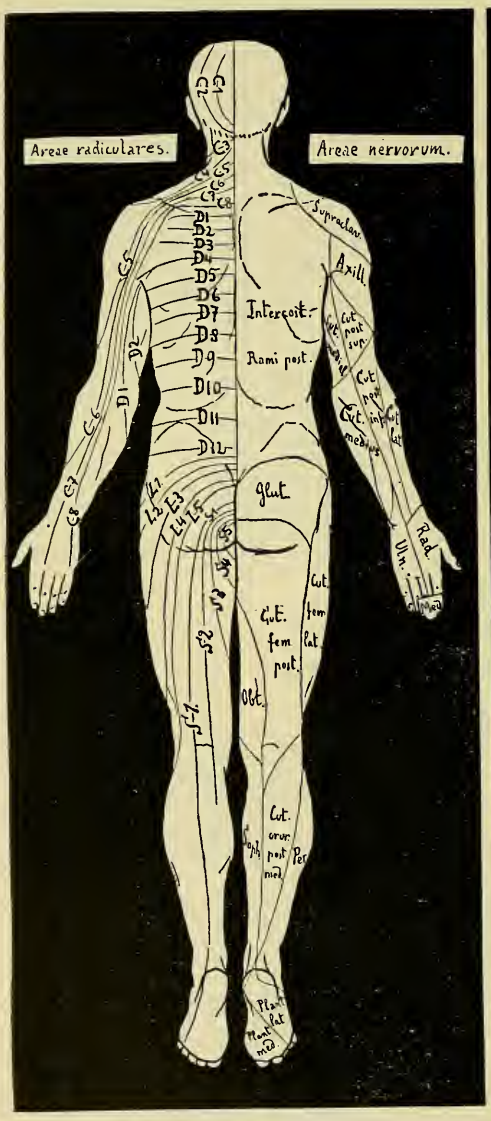


Fig. 16.

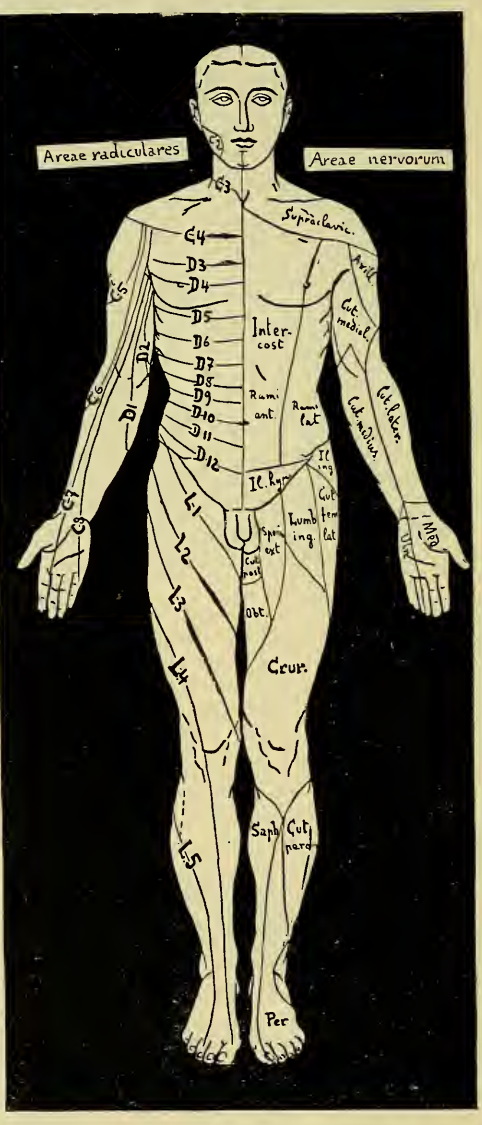


Fig. 17.

when they affect not purely motor, but "mixed" nerves, sensation may remain very little disturbed or even intact. As far as they refer to the types of peripheral paralyses already considered, the sensory disturbances are as follows:

Paralyses	Disturbances of Sensation
Total brachial plexus paralysis	Usually present; the inside of the arm is almost always unaffected, otherwise the distribution varies greatly
Upper plexus paralysis	Less frequent: as rule located on the outside of the arm
Lower plexus paralysis	Are found in the majority of cases, particularly in the ulnar distribution, on the inner side of both arm and forearm
Nervus thoracic. long.	None
Nervus suprascapularis	None
Nervus axillaris (circumflex)	Sometimes on the outside of the arm (see Figs. 16 and 17)
Nervus musculo-cutan.	On the outside of the forearm (nervus cutan. lateral) (see Figs. 16 and 17)
Nervus medianus	In typical cases on the radial parts of the palm (as in Fig. 17); also dorsally on the terminal phalanges of fingers II, III and IV (as in Fig. 16); the dorsum, however, often remains unaffected, and also the palmar area may be only partially involved; occasionally no sensory disturbances at all
Nervus ulnaris	Almost always present; in the ulnar part of palm and dorsum of the hand they occupy the region indicated in Figs. 16 and 17
Nervus radialis (musculo-spiral)	Often absent; most frequent, however, on the radial half of the back of the hand; only exceptionally are the territories of the cutaneous post. infer. and cutaneus post. sup. involved (see Fig. 16)
Nervus cruralis	Sometimes sensory disturbances corresponding to the areas marked "Crur." and "Saph." in Fig. 17
Nervus tibialis	Sometimes sensory disturbances in the areas marked "Cut. crur. post. med.," "Plant. lat." and "Plant. med." in Fig. 16
Nervus peroneus	Chiefly sensory disturbances in the areas marked "Cut. peron." and "Per." in Figs. 16 and 17. Their extent varies greatly, however

LECTURE II

Diseases of the Peripheral Nerves

A. Disturbances of Conduction (*Continued*)

SPECIAL SYMPTOMATOLOGY OF THE DEFECTS OF FUNCTION IN THE DIFFERENT PERIPHERAL NERVES

II. The Cranial Nerves

In the first lecture, after we had considered the etiology, the pathogenesis and the pathological anatomy of disturbances of conduction in the peripheral nerves, we took up the general symptomatology of those pathological conditions and finally considered the defects of function which occur in lesions of the spinal nerves. We must now discuss in their proper sequence the symptoms of defective function in the cranial nerves when diseased.

1. *The Olfactory.*—Loss of sense of smell is called anosmia; its reduction hyposmia. To test the sense of smell only such reagents should be used as do not (for example like acetic acid or ammonia) act as irritants to the branches of the trigeminus which are distributed to the mucous membrane of the nose, *e.g.*, turpentine, oil of rose, tar, benzene, tinct. valerian.

2. *The Optic.*—The symptoms of defect upon the part of the optic nerve are manifold and for their determination and clinical interpretation we are largely dependent upon the assistance of the ophthalmologist. Hence we can give only a summary account of them here. Total destruction of one optic nerve causes complete blindness in the corresponding eye (amaurosis); hence throwing a beam of light into the amaurotic eye can no longer cause reflex contraction of the pupil. This can, however, be provoked by the action of light upon the sound eye. Slight alterations of the optic nerve cause dimness of vision, amblyopia.

Partial, lacunar defects in the visual field are called scotomata, according to their location, central or peripheral scotomata. When the perception of single colors is lost in a limited portion of the field, we speak of color scotoma. If the radii of the field of vision of one eye are comparatively evenly shortened, we have concentric narrowing of the visual field. So far the symptoms of defect which occur in peripheral diseases of the optic nerve; half-sided loss of vision, the so-called hemianopsia is found only in lesions of the visual tract back of the chiasma; hence it will not be discussed until we come to diseases of the brain. Of all the cranial nerves, the optic occupies a unique position, in that it alone is accessible to direct inspection by the aid of the ophthalmoscope. Three sorts

of pictures may be presented to us in disturbance of conduction in the peripheral course of the optic fibers. If the optic nerve is affected by a tumor or an injury, it undergoes "simple atrophy"; the papilla becomes paler, eventually very white and at the same time there is cupping (atrophic excavation) and the gray points of the lamina cribrosa are specially prominent; from the surrounding retina it is abnormally sharply delimited, the finer blood vessels of the nerve head disappear more and more. "Optic neuritis," on the other hand, provided that the papilla is also affected, which is not necessarily always the case,* gives the following ophthalmoscopic changes: The papilla is more or less prominent in the eye ground, it is reddish gray in color, sometimes appears

FIG. 18.

Left-sided Oculomotor Paralysis. Ptosis.

mottled, its boundaries are not clear, its diameter appears increased, its veins are swollen and tortuous, the arteries, on the contrary, narrow.

As a result of intense and advanced optic neuritis, finally, there is a third pathological ophthalmoscopic picture, the so-called "inflammatory atrophy." In this the papilla is at the start gray white and shows indistinct outlines; later it becomes, like in simple atrophy, very white with sharply defined boundaries, but nevertheless appears irregular and shrunken; also the lamina cribrosa does not show up and the neighboring choroid in many cases presents irregular decolorization.

3. *The Oculomotor Nerve.*—The third cranial nerve innervates both the

* When inflammation of the papilla is absent or very slight, we speak of "retrobulbar neuritis."

external and the internal muscles of the eye. Namely, to begin with the first, the levator palpebræ, which raises the upper lid, the rectus internus, which draws the eyeball inward, the rectus superior and rectus inferior, which draw it respectively upward and downward, and the inferior oblique, whose action consists in rolling the ball upon its axis so that the lower portion of its periphery is turned inward toward the nasal border of the orbit. The internal muscles innervated by the oculomotor are, the ciliary muscle, through whose contraction the zonula of the lens relaxes (from which greater convexity of the lens and

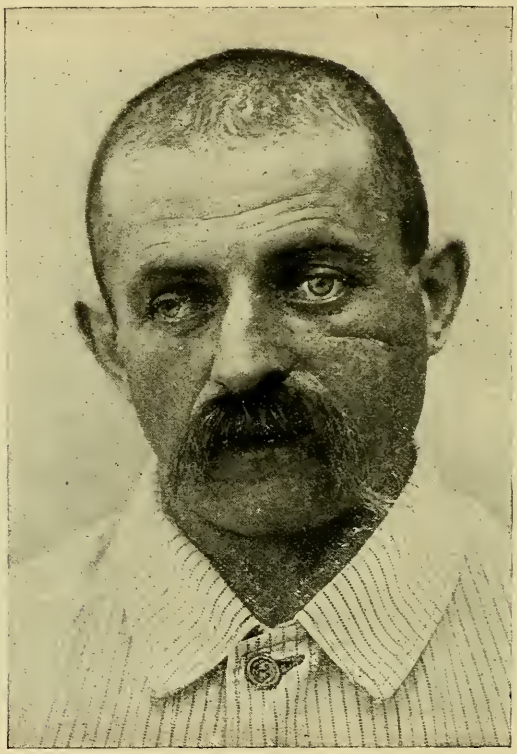


Fig. 19.

Paralysis of the Internal Rectus on the Right Side (Fracture at the Base of the Skull).

accommodation for near objects results) and the ring fibers of the iris, the sphincter pupillæ. In complete paralysis of the oculomotor, the upper lid hangs flaccid (ptosis) (see Fig. 18) and the eye is drawn outward and downward, this through the unopposed action of the rectus externus and superior oblique muscles, which are innervated by the other eye muscle nerves (the abducens and trochlearis). Besides this, the pupil is dilated and the eye remains in a position of accommodation for distance (paralysis of the sphincter pupillæ and ciliary muscles). When the disturbance of conduction in the oculomotor nerve is caused by progressive processes at the base of the brain, for example, tumors, ptosis is usually the first symptom. The distinction between an oculomotor paralysis from lesion of the nerve itself and one due to disease of its

nucleus in the brain axis is not always easy; it is well to remark, however, that in nuclear paralyses the internal muscles of the eye usually escape. If on the contrary these alone are involved, we speak of "ophthalmoplegia interna."

4. *Trochlearis.*—This nerve supplies exclusively the superior oblique muscle, which as antagonist of the inferior oblique, rolls the eyeball inward, its upper periphery toward the nasal border of the orbit.

5. *The Trigeminus.*—The trigeminus possesses a large sensory, a smaller motor portion, of which the first divides into three large divisions, the Ramus ophthalmicus, R. maxillaris and R. mandibularis. The following table presents the sensory distribution of these three branches.

Ramus primus (I), seu ophthalmicus	Ramus secundus (II), seu maxillaris	Ramus tertius (III), seu mandibularis
(a) The skin area marked 1 in Fig. 20	(a) The skin area 2, in Fig. 20	(a) The skin area 3, in Fig. 20
(b) Conjunctiva, cornea and iris	(b) The mucous membrane of the antrum and the lower parts of the nose	(b) The mucous membrane of the cheeks, the lower jaw, the floor of the mouth, the tongue
(c) The mucous membrane of the frontal sinuses and the upper parts of the nose	(c) The mucous membranes of the upper jaw and of the palate to the palatopharyngeal arch	(c) The lower teeth
	(d) The upper teeth	

In total or partial destruction of one of the three branches, there is of course anesthesia or hypoesthesia in the corresponding parts. Further, in lesion of the ophthalmic branch, there is loss of the conjunctival and corneal reflex (closing of the eye on touching the conjunctiva or cornea with a blunt object, for instance, the head of a pin, also of the reflex of sneezing produced by tickling the upper part of the nasal mucous membrane. In lesion of the maxillary branch the sneezing reflex is absent when the lower part of the nasal mucosa is irritated, as is also the palate reflex, that is the movements of swallowing or of retching produced by tickling the soft palate.

It is to be remarked here, however, that all of these three reflexes are inconstant and subject to great individual variations. Hence only the absence of these phenomena is of diagnostic importance. It is well to recall, too, that the sensation of pricking perceived on inhaling ammonia or acetic acid is produced not through the olfactory nerve but by irritation of sensory terminations of the trigeminus.

If the trigeminus is out of function these do not occur, neither do the reflex symptoms produced through it (watering of the eyes, changes in the pulse, arrest of respiration).

Besides conducting common sensation the trigeminus contains also fibers for special sense. The mandibularis, through one of its branches, the lingualis collects the taste fibers from the anterior two-thirds of the tongue; these proceed then through the chorda tympani to the facial trunk, to return again to the trigeminus after they have run some distance in the facial. Since disturb-

ances of taste (ageusia, hypogeusia) occur most commonly in facial paralysis, we will postpone the discussion of the methods of testing taste, until we come to speak of the facial nerve. The motor trigeminus fibers are included in the third branch and are distributed to the muscles of mastication, the anterior belly of the digastric, the mylohyoid, the tensor tympani and the tensor palati muscles. Unilateral paralysis of the muscles of mastication (masseter, temporal, pterygoids) is called monoplegia masticatoria. In this, lateral movement of the lower jaw is only possible toward the paralyzed side, since only on the sound side are the pterygoids still able to contract. By palpation the absence of contraction in the masseter and temporal of the affected side can also be noted. In diplegia masticatoria, the lower jaw drops and all lateral motion is impossible. Further, there is no jaw reflex—the contraction of the masseters on striking upon the finger laid upon the lower row of teeth, which can be obtained in most normal individuals.

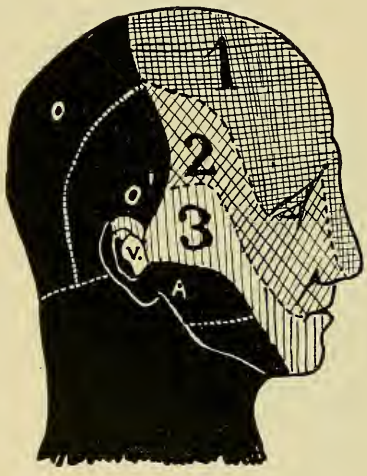


Fig. 20.

The Sensory Nerve Supply of the Head

Hatched: Trigeminus
1. The ophthalmic branch
2. The maxillary branch
3. The mandibular branch

White: Vagus
v. Nervus auricularis vagi

Black: Cervical nerves
O, Great occipital nerve
o, Lesser occipital nerve
A, Great auricular nerve

The paralysis of the anterior belly of the digastric and of the mylohyoid, sometimes produces a sense of relaxation on palpation in the muscles of the floor of the mouth on the paralyzed side; we have little information as to the symptoms of loss of function on the part of the tensor tympani and the tensor palati. It is certain that in most cases they remain latent; here and there, however, there appear to be anomalies in the position of the palatopharyngeal arch and poor hearing for low tones.

Besides the motor and sensory fibers mentioned, immediately after their exit from the skull all of the three divisions of the trigeminus have sympathetic fibers added to them, and these fibers accompany these divisions and their branches in their further course. The meeting points of these sympathetic fibers (they arise altogether from the plexus accompanying the head arteries) with trigeminus neurones, are at certain nodes or ganglia (ganglion ciliaris, sphenopalatinum, oticum, linguale). Hence, under some circumstances, a lesion of the first division can have as a result a narrowing of the palpebral fissure and a myosis of the pupil on account of paralysis of the tarsalis superior and dilator pupillæ muscles innervated by the sympathetic, a lesion of the second division, by paralysis of the orbital muscle at the base of the orbit, an "enophthalmus," that is, a sinking in of the eyeball. All of the three branches carry sympathetic fibers to the blood vessels and sweat glands, on which account in interruption of conduction we almost always find in the anesthetic parts heat

and redness (in fresh cases) or cyanosis and coldness (in older cases), as well as anidrosis. Other neurones originally sympathetic, but running in the trigeminus, which must be mentioned, are fibers regulating the secretion of the nasal mucus, contained in the ophthalmic and maxillary divisions. Upon their failure to functionate depends the abnormal dryness of the nasal mucosa which occurs in paralyses of these nerves and leads secondarily to reduction of the sense of smell. Finally, there are other secretory fibers coming from another cranial nerve, the facial, the already mentioned lingual nerve containing those for the salivary secretion, the lacrymal nerve (a branch of the ophthalmic) those for the tear secretion. Hence these secretions can sometimes be affected, in troubles of the fifth nerve.

6. *The Abducens.*—To the sixth nerve belongs the motor innervation of

Fig. 21.

Bilateral Abducens Paralysis.

the external rectus muscle which turns the eye about a vertical axis outward, abducts it. Isolated abducens paralysis has hence as a result a deviation of the affected eye inward (see Fig. 21). When the abducens is affected, together with the oculomotorius and trochlearis (ophthalmoplegia totalis), the eye remains immovable, directed to the front.

7. *The Facial.*—This nerve supplies all the muscles of the face (including the buccinator, excepting the levator palpebræ superioris, which is innervated by the oculomotor) from the frontal muscle to the platysma myoides, and besides these, the stylohoid, the posterior belly of the digastric and the stapedius. The failure of the motor function of the facial nerve produces the clinical picture of facial paralysis (or prosoplegia) which when unilateral is denominated monoplegia facialis (see Figs. 22 and 23), when bilateral, diplegia facialis. It is also called *Bell's* paralysis. The paralyzed half of the face is devoid of the mimic movements, masklike, immovable, expressionless; the naso-labial fold

is obliterated, the forehead cannot be wrinkled, the eye—on account of the paralysis of the orbicularis palpebrarum—cannot be closed (lagophthalmus), the angle of the mouth hangs down. The falling downward of the back portion of the tongue betrays the paralysis of the stylohoid and the posterior belly of the digastric, an abnormal acuteness of hearing and a sensitiveness to deep tones (oxyakoia, hyperacusis), that of the stapedius whose function it is to close the fenestra ovalis of the drum cavity by drawing on the stapes. From the neighborhood of its origin also, the facial trunk carries along with it other sorts of centrifugal fibers, namely those for the tear secretion and those for the salivary secretion; both of these sets of fibers as we have already heard, enter

Fig. 22.

Left-Sided Peripheral Facial Paralysis.

the trigeminal nerve, while on the other hand the taste fibers from the anterior two-thirds of the tongue leave the trigeminus for some distance and join the facial. Fig. 24 will bring before you the anatomical relations of this somewhat complicated exchange of fibers. They contribute chiefly to the fact that according to where a break in the conduction in the peripheral course of the facial has occurred different clinical symptoms develop. I have presented these in tabular form on page 33.

Testing of sense of taste on the tongue is carried out in the following manner. The subject is instructed to tell the taste of the solutions which are applied to his outstretched tongue without drawing it back into his mouth again. These precautions are to prevent the falsification of the results by their perception by the mucous membrane of other parts of the mouth which are innervated by the glossopharyngeal. A card bearing the words "bitter, sour,

salty, sweet," is placed before the patient and he is asked to indicate upon it what he tastes as his tongue is touched successively by cotton brushes wet with tinct. gentian, vinegar, salt solution, and syrup. After each test the tongue must be washed off with a wisp of wet cotton. The eyes are best covered during these tests.

BREACHES OF CONDUCTION IN THE FACIAL NERVE.

a. Outside of the cranial cavity.

Almost exclusively monoplegia facialis (Exception: facial paralysis from double otitis).

a. Distal to the origin of the chorda tympani. (Area 1, Fig. 24.) Symptoms: paralysis of the facial muscles; if the lesion is very far toward the periphery, that is beyond the pes anserinus, some of the branches may escape.

b. In the Fallopian canal, between the chorda and the origin of the stapedius (Fig. 24, 2). Symptoms: facial paralysis; loss of taste on the anterior two-thirds of the tongue; salivary secretion altered.

c. In the Fallopian canal between the origin of the stapedius and the geniculate ganglion (Fig. 24, 3). Symptoms: facial paralysis, loss of taste on the anterior two-thirds of the tongue, salivary secretion altered, hyperacusis.

d. Between the meatus acusticus internus and the geniculate ganglion (Fig. 24, 4). Symptoms: facial paralysis, no taste disturbance, alteration of the salivary secretion, often nerve deafness (see below) through involvement of the auditory, only when this is not present, hyperacusis, loss of the affective and reflex tear secretion.

b. Within the cranial cavity—"basal" lesions of the facial. Not infrequently diplegia facialis (basal gummatous meningitis). Symptoms: as above under *d* generally involvement of a number of basal nerve roots (abducens, glossopharyngeus, vagus, accessorius, hypoglossus); general brain symptoms (vertigo, vomiting, headache).

8. *The Auditory.*—The eighth cranial nerve is made up of two portions each of different function: the nerve of hearing proper, the cochlear, and the nerve of orientation in space, the vestibular. The first stands in connection with the cerebrum, the latter with the cerebellum, the organ for the preservation of the equilibrium.

Loss of function of the cochlear nerve produces impairment of hearing to complete deafness (hypacusis or anacusis). Since, however, these disturbances may also be produced by affections of the sound-conducting apparatus, of the middle or of the outer ear,* the chief characteristics of "hardness of

* A lesion in the perceptive apparatus in the cochlea of the labyrinth (that is, in the organ of *Corti*) gives the same symptoms as a breach of conduction in the cochlear nerve.

hearing or deafness of nerve origin" must be considered. These are: 1. The reduction or loss of hearing through bone conduction. 2. Partial loss of perception for the notes of the scale.

The first of these phenomena is determined, a, by *Schwabach's* test. In this the time during which a vibrating tuning-fork placed upon the parietal bone, the teeth, or the mastoid process, can be heard, on the one hand, by the person under examination, and on the other by the normally hearing examiner, is compared. In impairment of hearing from nerve lesion, there is shortening of the period during which the sound can be perceived through bone conduction; in

Fig. 23.

Left-Sided Peripheral Facial Paralysis, Upon Attempt to Close Both Eyes.

nerve deafness bone conduction may be lost. In middle ear affections on the contrary the tuning-fork applied to the bone is heard longer than normal. This method is naturally only applicable in bilateral disturbances of hearing.

b. By "*Rinne's test.*"—In this, a vibrating tuning-fork is placed upon the mastoid process; when the patient ceases to hear it there (that is, bone conduction is no longer perceived), the fork is transferred to opposite the ear. The normal person now hears the sound anew and this is denominated a positive result ("*Rinne* positive"). In disease of the sound-conducting apparatus (where the perception through air conduction is diminished, but through bone conduction unaltered) in the second part of the test, sound is no longer perceived, it is negative.

In nerve "hypoacusis" on the contrary, there is, as a rule, "positive *Rinne*"

provided, of course, that there is not a high degree of defect of hearing. In this latter case, as in nerve deafness, hearing through air conduction is considerably reduced or lost.

c. By "*Weber's* test." If in a normal individual a vibrating tuning-fork is placed on the vertex it is heard in both ears; if now one ear is plugged up, the sound is "lateralized" on the side upon which air conduction is in this manner

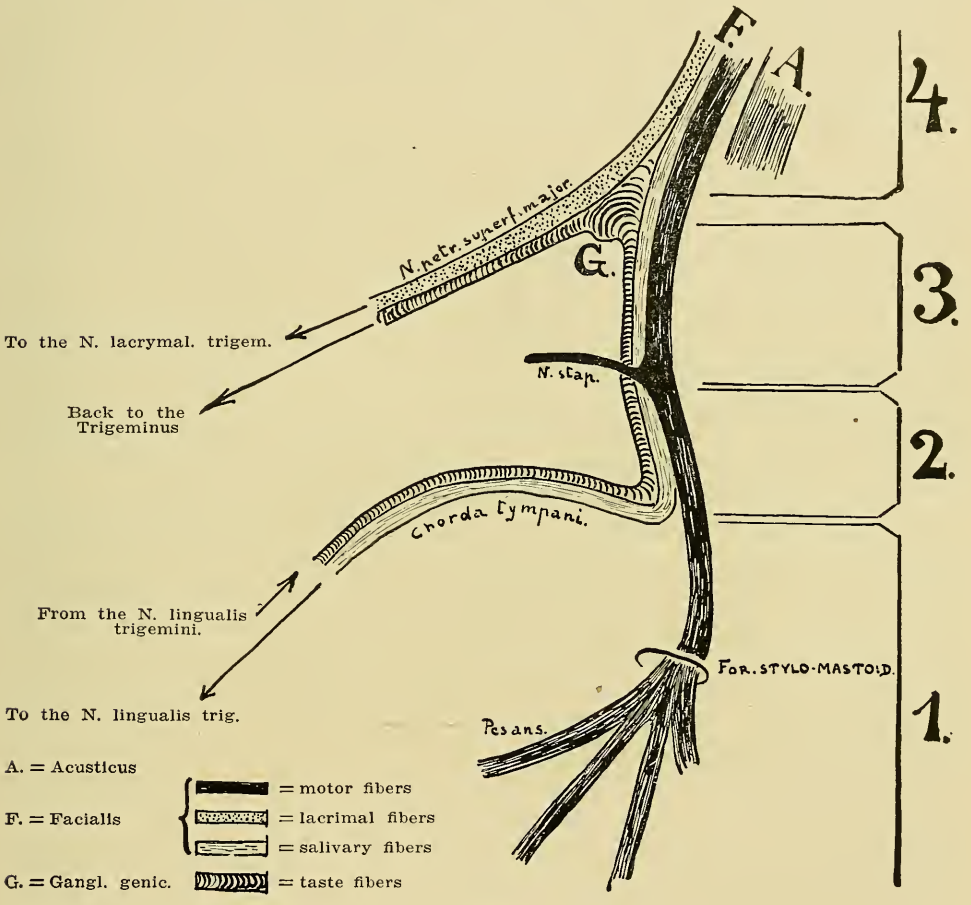


Fig. 24.

Diagram of the Different Categories of Fibers in the Facial Nerve.

interrupted. This "lateralization" of the sound occurs spontaneously on the side of impaired hearing in persons with diseases of the sound conducting apparatus ("positive *Weber*"), while the person with nerve deafness lateralizes to the sound side ("negative *Weber*").

Defects in the perception of the notes of the scale are tested by the so-called *Galton's* whistle. Deafness for the upper notes of the scale are particularly characteristic of affections of auditory nerve tracts as opposed to those of the middle and outer ear. In "nervous hardness of hearing" the perception of words with sharp consonants and clear vowels (as sheriff, swagger, fish, wasp),

suffers especially, while those with dull consonants and vowels (as brooder, hunger, ore, worm*) are much better understood. The opposite is the case in non-nervous hypoacusis.

The diagnosis of nerve hardness of hearing and deafness must be made in general by exclusion, after examination of the drum membrane, when the hearing is not improved after Pollitzerization of the ear, etc. As to the value of that peculiar phenomenon, the so-called paracusis of *Willis* for the diagnosis of nerve deafness, opinions are divided. Many otologists are of the opinion that nerve deafness paradoxically makes itself less felt in a noisy place (in a railway car) than in a quiet one; according to others still, in middle ear disease "Paracusis Willisii" can also occur.

The vestibular nerve transmits to the cerebellum information as to the position in space of the body, especially of the head. The sensory epithelium of the ampullæ, the utricle and the saccule of the labyrinth are stimulated to function through the hydrostatic relations of the fluid in the semicircular canals (which are arranged according to the three planes of space). If now there is a contradiction between the condition of stimulation and the actual position in space of the body, the resulting confusion gives to the patient the illusion of rotary movements, either of his own body or of surrounding objects, which we denominate rotary vertigo. The last can arise from interruption of conduction in the vestibular nerve and then shows itself sometimes by moderate uncertainty of gait and difficulty in turning, as well as by holding the head stiffly.

We denominate "galvanic vertigo," a manifestation which appears, when in the person under investigation, a constant current of gradually increasing intensity is conducted through the head at the level of the ears. When a current strength of about 5 to 6 milleamperes is reached, the normal person has the feeling that he is going to fall toward the side on which the positive pole is, and usually inclines his head to that side. If one vestibular nerve is out of function, however, in many cases the patient in this experiment has each time the feeling that he is sinking toward the side of the injured nerve regardless of the position of the anode or the cathode.

More applicable to the clinical investigation of the vestibular apparatus is *Bárány's* test, that is, the examination for caloric nystagmus. Each vestibular apparatus is in anatomical and physiological connection with the eye muscles through the fasciculus longitudinalis posterior, on account of which, irritation of the ear by syringing with hot or cold water produces rhythmical horizontal movements of the eye. (Probably through the differences of temperature there are produced movements of the endolymph, which irritate the nerve structures of the labyrinth.) In healthy persons, the syringing of the ear with cold water produces nystagmus toward the opposite side; if hot water is used, the nystagmus on the contrary is toward the side of the syringed ear. If the vestibular nerve is paralyzed, no caloric reaction can be produced from the corresponding ear. The healthy side generally reacts normally, though occasionally irritability on this side is also somewhat reduced.

9. *The Glossopharyngeal Nerve.*—This nerve is mainly sensory, but has a

* For the German words given by the author, English words of as similar sound as possible have been substituted.—Translator.

motor division which supplies the stylopharyngeus muscle, an elevator of the pharynx. The glossopharyngeal transmits sensory impressions from the uppermost part of the pharynx and from the middle ear, as well as taste perceptions from the palate and from the posterior third of the tongue. On this account, ageusia in the last-mentioned region is the most important symptom of its loss of function; also an anesthesia of the pharynx can be recognized, as well as loss of the pharyngeal reflex. This last, however (in correspondence with what was mentioned in connection with the palate reflex), can only be considered as a sign of breach of conduction in the glossopharyngeal when there is a difference between the reflexes on the two sides. The stylo-pharyngeus muscle, as elevator of the pharynx—in which function it unites itself with the pharyngopalatine, supplied by the vagus—is of too little importance for motor symptoms to occur from its elimination alone.

10. *The Vagus (or Pneumogastric) Nerve.*—The vagus is a mixed nerve. On the one hand it supplies the muscles of the palate, pharynx, larynx, trachea and bronchi, as well as those of the œsophagus, stomach and small intestine, with motor fibers, and carries inhibitory fibers for the heart and vasomotor fibers for numerous vessels; on the other, it is the sensory nreve for the dura mater, the external auditory canal,* the lower portion of the pharynx, the larynx, the trachea and bronchi, the œsophagus and the stomach. Total double paralysis of the vagus has no symptomatology, since it is incompatible with further existence. On the other hand, a partial double, or a total single vagus paralysis, can occur clinically. In the last case there is a unilateral paralysis of the uvula, pharynx and larynx. One-half of the velum palati hangs flaccid down, on which account the voice becomes nasal. One vocal cord is immovable in the middle or the cadaveric position, since both the closing and the opening muscles of the glottis are put out of action. Still, the voice may remain normal through compensatory overaction of the other vocal cord. Usually, indeed, it is somewhat hoarse, changing to falsetto. On the other hand, disturbances in swallowing are almost always minimal, since hemipharyngoplegia, on account of the mutual interlacing of the muscular fibers of the pharynx, is without great functional importance. An increased frequency of the heart beat—a tachycardia—has very seldom been observed in unilateral loss of function of the vagus; the same thing is true with regard to disturbances of respiration (in the sense of slowing and irregularity of the breathing). In complete paralysis of the vagus, only a few of the above-related symptoms are present, sometimes these, indeed, are only partially developed. So, instead of total paralysis of the vocal cords, there is only paralysis of the posterior cricoarytenoid, the opener of the rima glottidis. (See Fig. 25, C.) From this "posticus paralysis," when it is bilateral, there results an interference with respiration, while phonation is normal.

11. *The Spinal Accessory Nerve.*—This nerve alone supplies the sternocleidomastoid muscle, while in the innervation of the trapezius it is assisted by

* The researches of *J. R. Hunt* seem to show that the facial nerve has also a sensory root which supplies a cone-shaped area including the external auditory canal and a small portion of the outer ear.—*Translator.*

the upper cervical nerves. If the spinal accessory is destroyed, there is a complete paralysis of the sternocleidomastoid. Hence, in one-sided lesion, it is impossible to turn the chin completely toward the opposite side, while in bilateral paralysis there is also a tendency for the head to fall backward. On the other hand, the paralysis of the trapezius is incomplete and makes itself evident only in impaired force and extent of the elevation of the arm.

12. *The Hypoglossal Nerve.*—The hypoglossus can be described briefly as the nerve supplying the tongue muscles. Indeed, its part in the innervation of the lower muscles attached to the hyoid bone (sternohyoid, sternothyroid, omohyoid), which serve in part to fix the larynx, is only apparent. In the so-called "ansa hypoglossi," through which this innervation occurs, there run fibers from the cervical nerves, which only enter the hypoglossus through

FIG. 25.

Image of the Vocal Cords on Inspiration.

A. Normal.
B. Left-sided vagus paralysis (Recurrens paralysis).
C. Bilateral posterior arytenoid paralysis.

anastomoses. Bilateral paralysis of the hypoglossus, of course, produces a total "glossoplegia"; following the influence of gravity the tongue lies immovable upon the floor of the mouth; speech is unintelligible, eating is considerably interfered with. In unilateral paralysis (hemiglossoplegia), on the other hand, the motor disturbance is relatively small; indeed, talking and eating are usually not at all, or scarcely, disturbed. This is on account of the multiple interlacements between the muscle fibers of both sides of the tongue. If, on the contrary, the patient is asked to show his tongue, a deviation of the tip of the tongue toward the side of the paralysis occurs, on account of the preponderance of action of the genioglossus on the sound side.

THE OCCURRENCE OF PERIPHERAL INTERRUPTIONS OF CONDUCTION IN THE CRANIAL NERVES

Although in Lecture I (in speaking of the etiology of disturbances of conduction in peripheral nerves) we had the cranial nerves also in mind, and for example, considered rheumatic facial paralysis, I will, in this connection, bring before you the conditions under which paralyses of a peripheral nature of individual cranial nerves come under observation. According to the localization of interruption of conduction of this nature, in a given cranial nerve, we must take into consideration certain etiological factors.

The olfactory nerve is involved most frequently in injuries to the head with

or without fractures of the skull, but also in tumors, abscess and meningitis in the anterior fossa of the skull.

The optic nerve can be affected in syphilis, acute infectious diseases, poisoning (*e.g.*, with lead, arsenic, nicotin, santonin, iodoform, quinine, alcohol), in severe anemias (*e.g.*, after hemorrhages from the stomach or metrorrhagia), in auto-intoxications (diabetes, nephritis), also by neoplasms, whether these arise from the nerve itself or from neighboring structures (*e.g.*, from the orbit); finally, it is not seldom wounded in attempts at suicide (by a shot in the temple).

Etiological factors for disease in the course of the eye-muscle nerves are: fractures at the base of the skull (the abducens is especially endangered on account of its exposed position at the point of the pyramid of the petrous bone); compression by aneurisms of the internal carotid or by tumors; otitis media (abducens paralysis); acute infectious diseases; diabetes, syphilis, nephritis, gout, alcoholism, chronic lead poisoning.

Peripheral interruptions of conduction in the trigeminus rarely affect single branches only; apart from injuries of the bones and the soft parts of the face, in the main only tumors, tuberculous and syphilitic processes of the bones which contain the foramina of exit of the nerves, and of their periosteum come into consideration as causes. The Gasserian ganglion and the common trunk of the nerve are, however, much more frequently affected and, indeed, almost always unilaterally (aneurisms of the internal carotid, tumors of the internal carotid, transverse basal fractures behind the sella turcica, hypophysis tumors, etc.).

The facial, as said before, is by far most frequently affected by "rheumatic" influences; that is, without any other recognizable etiology than exposure to cold. In "idiopathic facial paralysis" even this etiological factor is sometimes not to be made out. Other causes are: otitis media, meningitis, tumors in the posterior fossa of the skull, diseases of the petrous bone, syphilis, erysipelas, diphtheria, protective inoculation against rabies. Finally, are to be mentioned traumata, which are quite common in the course of the facial nerve (fall against a window-pane, sabre cuts in duels).

The auditory nerve is affected by tumors, diseases of the meninges and caries of bone in the posterior fossa of the skull, as well as by inflammations of the middle ear, corresponding to its close relations with the facial nerve, quite as frequently as this latter. Also fractures of the base of the skull sometimes involve it. Besides different acute and chronic infectious diseases (typhoid fever, diphtheria, influenza, scarlet fever, parotitis, tuberculosis, syphilis), leucemia has been specially mentioned as an etiological factor in auditory neuritis.

Isolated paralysis of the glossopharyngeal nerve scarcely ever occurs. Other nerves are usually involved at the same time, particularly the vagus, especially where the interruption of conduction is of traumatic origin (fractures of the base, tumors, the pressure of aneurisms, or sinus thromboses, etc.). The vagus can be affected in many kinds of poisonings (*e.g.*, carbonic oxide, morphine, atropin, phosphorus, lead, arsenic, alcohol), as well as in connection with different acute infectious diseases. Paralysis of the recurrent laryngeal nerve is, as a rule, caused by local anomalies in the neck or in the mediastinum (struma, tumors, enlarged glands, etc.).

The spinal accessory and hypoglossus are particularly exposed to traumatic injury and upon the basis of the same causal factors as the glossopharyngeal and vagus. Hypoglossal neuritis occurs also in intoxications (alcohol, lead, arsenic, carbonic oxide).

B. Special Remarks on Neuritis and Polyneuritis

I. IRRITATIVE SYMPTOMS

While we might consider disturbances of conduction, whose general and special phenomenology we have already learned, as a general characteristic, as well of traumatic and neoplastic, as of neuritic diseases of the peripheral nerves, we must now enumerate the irritative symptoms, which play a not less important rôle in the clinical picture of the last-mentioned nosological group.

They manifest themselves to by far the greatest extent as disturbances of sensation: that is, as pain along the course of the affected nerves. They may have a shooting, stabbing, or boring character, are sometimes described as a burning on the surface of the skin (the "causalgia" of *Weir-Mitchell*). From these pains proper, they pass by imperceptible degrees to the "paresthesias," the more or less painful abnormal sensations which are often felt in the territory of the affected nerves: formication, prickling, tickling, subjective feeling of cold or heat, etc. Also hyperesthesia to touch, on account of which the pressure of the clothing or of the bedclothes may be unbearable, is not infrequent. Here and there, also, spontaneous pains are projected into skin regions in which, by objective tests, touch and pain sense have been proved to be reduced (hypesthesia dolorosa, hypalgesia dolorosa). Further, it sometimes occurs that pains, at least for a time, are located in the joints, and so arthritic diseases are simulated. The irritation of fibers conducting centripetally can exceptionally be accompanied by exaggeration of reflexes. The conditions under which the influence of the disease in exaggerating the reflexes, overcomes the effect of lesions of the afferent or efferent part of the reflex arc in destroying the reflexes, are not clear to us. Perhaps in such cases the perineuritic processes predominate anatomically over the parenchymatous, since the spontaneous pains may well depend upon perineuritic changes. That in general, the irritative phenomena in the centripetal fibers (pain, hyperreflexia) occur most usually in acute neuritides and polyneuritides, agrees with what was *a priori* to be expected.

Motor irritative phenomena are, on the other hand, very infrequent, and have been described almost exclusively in multiple neuritis, *e.g.*, cramps in the calves in localization of the disease in the legs. *Babinski* asserts that there is often a latent predisposition to cramps in the calves which can be provoked by faradization; I have so far not been able to convince myself of this. On the other hand, I saw a peculiar muscle wave ("myokymia"), which has also been corroborated by *Remak*, once in weak development in the peroneal muscles in a tuberculous polyneuritis, very plainly in the biceps brachii in an alcoholic neuritis.

II. PALPATORY FINDINGS

Abnormal findings by palpation of superficially situated nerves are quite frequent. In the first place, there is more or less marked sensitiveness, so that pain is produced by moderate pressure—upon the brachial plexus above the clavicle or in the axilla, upon the nerve structures in the internal bicipital sulcus, upon the ulnar on the epicondyle, upon the musculo-spiral in the groove on the outside of the humerus, upon the peroneal behind the head of the fibula, upon the sciatic between the flexors of the knee, etc. A certain familiarity with the intensity of the pressure which normally is necessary to produce pain in these different places is needed in testing for this phenomenon; for example, when it is necessary to elicit it by comparison with the healthy corresponding nerve on the opposite side (in polyneuritis). Less frequent than abnormal sensitiveness to pressure, but by no means infrequent, a thickening of the nerve trunks can sometimes be felt upon palpation. This can well be the expression of a perineuritic process; such a process appears to be the cause of the sensitiveness to pressure of nerves afflicted with neuritis, which can occur in purely motor branches and is in this case attributable to the irritation of the "nervi nervorum." In order to determine by palpation that there is a definite swelling of a nerve, industrious practice in palpation of normal peripheral nerve trunks is even more necessary than in the testing of sensitiveness to pressure. It is well, also, not to lose any opportunity which is presented to control one's palpatory findings anatomically. For example, I have found on autopsy, or on operative exposure, nerves which appeared swollen by palpation actually two or three times as thick as the corresponding portions of normal nerves.

III. THE DIFFERENT CLINICAL FORMS

The neuritides may be grouped according to different principles; for example: 1, according to their course into acute, subchronic and chronic; 2, according to the nature of their symptoms into motor, "mixed" and sensory; 3, according to their extent, into mononeuritides, disseminated neuritides, plexus-neuritides and polyneuritides.*

The acute neuritic diseases belong for the greater part to the etiological

* Of late attention has been directed to the "root-neuritides" ("radiculitides"). These conditions differ anatomically from the ordinary neuritides by being located proximally to the plexuses for the extremities—that is, in the region of the spinal nerve roots; clinically, by the fact that the pains arising in consequence of them are located, not in the territories of the peripheral nerves, but in the spinal segmentary areas of innervation. (See, farther on, Lecture XI.) Etiologically, infections and intoxications play the chief rôle, as in ordinary neuritides. Ordinarily the disease manifests itself in two phases: a stage of irritation with attacks of pain, hyperesthesia of the skin, and sometimes exaggeration of reflexes, and a stage of destruction with anesthesia, paralysis and absence of reflexes. The pains have the "lancinating" and specially severe character typical for radicular affections (*e.g.*, for tubes or carcinoma of the vertebræ—see below). They are often set up by movements or by over-exertion—in specially characteristic manner by sneezing. Sensitiveness to pressure, or upon palpation of accessible peripheral nerves, is not present or scarcely perceptible. Painful points over the spinous processes are recognizable, however.

group of infectious and rheumatic neuritides; they also arise, however, upon the basis of intoxications; for example, in lead, arsenic, and carbonic oxide poisoning. In my first lecture I mentioned a case due to the combined action of bromide of methyl and methyl alcohol, which had an acute course. The so-called "acute curable ataxia" of drinkers is a by no means seldom form of alcoholic polyneuritis; an "apoplectiform alcoholic paralysis" was first shown on autopsy, by *Eichhorst*, to be a polyneuritis beginning hyperacutely. Certain polyneuritides which ascend from the lower extremities in "foudroyant" manner and rapidly cause death by paralysis of the heart and respiratory nerves are denominated the "neuritic form of *Landry's* paralysis"; if an infectious basis is here the rule, this unfavorable course can also be a manifestation of alcoholic polyneuritis. Chronicity distinguishes in general the arterio-sclerotic, senile, anemic, cachectic, diabetic, and also the majority of alcoholic and tuberculous neuritides and polyneuritides.

In carrying out the division into motor, "mixed," and sensory neuritides, it is at once apparent how infrequent the last are; apart from the neuritis of

Fig. 26.

Tuberculous Polyneuritis. General Muscular Atrophy.

the external cutaneous nerve of the thigh, the so-called "paresthetic meralgia," we find in the literature very few descriptions of sensory neuritides (*e.g.*, isolated neuritis of the lateral cutaneous peroneal, of the internal cutaneous of the thigh, of the chief sensory branches of the radial), and also those polyneuritides in which every electrodiagnostic and dynamometric sign of an involvement of the motor fibers is absent, form a quite small minority. Pure motor neuritides are, on the other hand, more frequent and most remarkable, even when the affected nerves contain sensory fibers. A certain "electivity" in the action of the disease-producing cause, which we will consider more fully farther on in this lecture, is evident in these cases. Nevertheless, I have found, that by the use of finer methods of testing sensibility (*e.g.*, by the tuning-fork), a large number of mononeuritides and polyneuritides, which at first sight appear to be purely motor, later turn out to be of the "mixed" form.

The above mentioned "meralgia paræsthetica" is a not so rare clinical picture described by *Roth* and *Bernhardt*. The peculiar relations of the external cutaneous nerve in its passage through the fascia of the thigh, appear to predispose this nerve to destructive lesions. As exciting causes, infectious diseases, overexertion, abnormal static relations (flat foot) diabetes, arteriosclerosis

and nicotinism, have been mentioned. The disease is usually unilateral. The patients complain of the most varied paresthesias and spontaneous pains in the region of distribution of the external cutaneous nerve; often certain points are also very sensitive to pressure. The hyperesthesia of the skin can cause the friction of the clothing to be painful. Objectively hypesthesia or anesthesia in a more or less extensive portion of the territory in question is found. Meralgia is in general persistent and shows a tendency to relapses.

We speak of mononeuritis when the territory of only one nerve is affected, of total or partial plexus neuritis when a nerve plexus, for example the brachial plexus, is attacked in its entirety or with the escape of single branches. In a disseminated neuritis single nerves are attacked at the same time without any regularity in the distribution and development of the paralysis. In contradistinction to this, we reserve the term polyneuritis for cases of more or less diffuse disease of the peripheral nervous system, which present a nearly symmetrical distribution and run a regular course. We will now devote some space to these forms.

According to their extent, polyneuritides may be divided into those of paraplegic or tetraplegic type, those which run their course as symmetric arm plexus neuritis without involvement of the legs, and finally those in which the cranial nerves are exclusively attacked or are affected along with the others. It is noteworthy that according to the special etiology, different topographical and physiopathological peculiarities are regularly present. Lead neuritis avoids the sensory element and affects (where by overexertion of special groups of muscles the localization is not influenced in the sense of the "toxico-professional" paralysis of *Oppenheim* (see above), particularly the muscles of the forearm supplied by the radial nerve; very rarely, generalized lead paralyses, in which loss of motor power affects the territory of one nerve after another symmetrically, either in exacerbations, or in rapid succession, sometimes under febrile movement, occur. In these, however, the predisposed muscle groups are more seriously affected than the others. Arsenical polyneuritis, which usually produces both motor and sensory symptoms, as far as its motor components are concerned, presents in so far the contrary of saturnine polyneuritis in that the lower extremities are attacked by preference. Among 72 cases of *Brouardel's* they were affected first in 69 cases, exclusively in 35 cases. However, when the arms are affected, the atrophic paralysis of the ulnar and median nerves by far predominates, while the radial paralysis typical of lead intoxication occupies the background. Characteristic of diphtheritic polyneuritis is, on the one hand its preference for certain centripetal nerve fibers from the lower limbs, which lead to a "post diphtheritic ataxia"; on the other a decided affinity for motor nerves from the proximal parts of the cerebrospinal apparatus. Here the paralysis of the palate and the muscles of accommodation of the eye (the ciliary) by far predominate. Fortunately the pharyngeal, diaphragm, and heart paralyses, so dangerous to life, are rare; the extension of the polyneuritic process to the innervation of the neck and external eye muscles is also rare. I only once observed a symmetrical accessorius paralysis. However, diphtheritic paralyses of the muscles of the extremities, which are quite inconstant in their topographical distribution, also occur. The

polyneuritides from the inhalation of sulphide of carbon affect mainly the flexors of the hand and the extensors of the foot. These few examples may serve as instances of the characteristic choice which certain nerve poisons exercise, with noteworthy regularity, in the production of polyneuritic symptom-complexes.

This "electivity" speaks in the sense of *Ehrlich's* doctrine of "organotropy," for the idea that certain poisons possess certain chemical affinities for certain cell substances. According to the chemotherapeutic investigations of *Ehrlich* the different arsenic preparations, for example, may be arranged in an almost continuous scale as regards their "neurotropy." In spite of this, in the assumption of an elective chemical action of the poisons causing neuritis, we come up against the fact that clinically as well as anatomically, the symptoms manifest themselves preponderantly or exclusively in the periphery of the neurones in question. A second factor comes here into the question: the greater the distance from the trophic centre (that is, from the spinal cord and the spinal ganglia) the less the power of resistance of the neurone to the action of toxic substances. Here the "Ersatz" theory of *Edinger*, already mentioned in Lecture I, can with advantage be taken into consideration.

LECTURE III

Diseases of the Peripheral Nerves

C. The Neuralgias

Pains which occur in attacks and radiate throughout the territory of distribution of peripheral nerves we call neuralgia.* As to the material basis of this characteristic symptom, we are very poorly informed; however, in the vast majority of cases in which the nerves attacked have been examined microscopically, they have been found entirely normal. *Oppenheim* suggests with justice, that fine disturbances of nutrition of the nerve (perhaps also of its nervi nervorum) lie at the base of the neuralgias, disturbances which only exceptionally lead to visible alterations, which then approach the anatomical picture of neuritis. The cause of these disturbances of nutrition is only in a minority of the cases of mechanical nature; for example, a stasis of the veins accompanying the nerve, through which the latter is compressed in a bony canal or in some other channel (*Bardenheuer*), as perineuritic adhesions, osteitis, periostitis, tumors, cicatrices, etc., which exert a traction or compression upon the nerve, or, finally, a trauma; usually, however, general toxemic influences are to be held responsible. Of the infectious diseases, influenza, malaria and syphilis play the greatest rôle in the etiology of the neuralgias; of diseases of metabolism, diabetes and gout, of other auto-intoxications, chronic constipation, of exogenic poisonings, alcoholism. Undoubtedly the "nervous" (viz., neurasthenics and hysterics), are just as much predisposed to neuralgias as anemic and chlorotic individuals. Further, the influence of refrigeration, especially of circumscribed nature, as, for example, a draught on one side in trigeminus neuralgia, sitting on cold stones in sciatica, is not to be denied. I have already indicated what can be said about the rheumatic origin of diseases of the peripheral nerves.

Finally, the so-called "reflex" neuralgias are to be considered; problematic is this method of origin for such cases in which facial neuralgias have been brought into causal connection with a retroflexion of the uterus. Better founded, on the contrary, are the very frequent connection of trigeminal neuralgia with uncorrected anomalies of refraction of the eye, caries of the teeth, suppuration of the middle ear, catarrhs of the frontal sinus and of the antrum of *Highmore*, that of sciatica with flat foot, etc. Puberty and the period of retrogres-

* There are, indeed (as complement to the root neuritides described in the footnote on page 41), radicular neuralgias (root neuralgias, radiculalgias) in which the pain radiates, not in the anatomical distribution of the peripheral nerves, but presents the character of the lancinating pains of tabes dorsalis, to be described in Lecture XI.

sive change, seem the times of special predisposition for neuralgias. Men are affected much more frequently than women; nevertheless, during pregnancy and in the puerperium, the latter are decidedly more susceptible.

THE GENERAL SYMPTOMATOLOGY OF NEURALGIA

The attack of neuralgic pain either appears with lightning-like suddenness, or unpleasant sensations (a feeling of heat, of tension, of formication, etc.), in the distribution of the nerve affected, precede it by some time. Between the single attacks there is either entire freedom from pain, a dull pain of little intensity, or a feeling of irritation and soreness of the neuralgic nerve is complained of. Sometimes definite exciting causes can be held responsible for the outbreak of the attack of pain; for example, sudden movements, a cold draught, meteorological influences, mental excitement; often, however, we seek in vain for such exciting factors.

The pain has usually a tearing, dragging character, sometimes it is described as burning, cutting or boring; it is usually not continuous, but consists of paroxysms rapidly following one another; frequently these are so intense that the patients grow pale, break out in a sweat, groan, or even scream. The duration of the attack is varied, sometimes very short (a quarter or half minute), mostly a few minutes; sometimes, however, lasting for hours. Characteristic of neuralgia is the fact that the pain is always quite sharply localized by the patient so that he sometimes can demonstrate to us the whole course of the painful nerve with anatomical correctness; in severe cases, indeed, the irradiation of the pain into a neighboring nerve territory is a frequent occurrence at the height of the attack. Objective accompanying symptoms are only to be made out in part of the cases; most frequent appears to me to be the reddening of the integument in the region of the affected nerve; this can, indeed, outlast the attack and can be observed between the attacks. The same remarks apply to the much more infrequent finding of a slight œdema of the skin. I would mention, further, the motor symptoms accompanying severe attacks of neuralgia: a tension of the muscles of the region during the attack, sometimes, also, clonic contractions. In neuralgias of the extremities of long duration, in consequence of the position which the patient causes the affected limb to assume in his effort to avoid the pain, there is often a decided degree of emaciation.

A hyperesthesia of the skin in the affected territory can quite frequently be made out and, on account of this, pressure and the friction of the clothing often become unbearable. Less frequently there is slight hypoesthesia. More important for diagnosis is the sensitiveness to pressure of the affected nerve, which is mainly limited to definite points in its course (where it leaves a bony canal, perforates a fascia lies upon a firm tissue); exceptionally, however, affects the whole of its course accessible to palpation. These so-called "*Valleix* points" can, as a rule, also be demonstrated independent of the attacks of pain; pressure exerted upon them can, indeed, provoke the attack, while, on the other hand, in many cases firm pressure upon these points during the attack is perceived as ameliorative. In describing the special forms of neuralgia we

will further consider the *Valleix* points, which, however, are no necessary accompaniment of neuralgias.*

THE MOST IMPORTANT FORMS OF NEURALGIA

a. Trigeminus Neuralgia

Among the general causative factors of this very frequent form of neuralgia, for which, also, the names of prosopalgia and "*Fothergill's* face pain" are occasionally used, diseases of metabolism and infectious diseases play a great rôle. Among these there is in so far a certain electivity in that, for example, malarial and influenzal neuralgias affect the first branch, diabetes and syphilis the third branch, by preference. This last in contradistinction to the first branch which is almost without exception affected on one side alone, can be bilaterally involved.

(Specially characteristic of syphilis is the bilateral *Seeligmueller's* neuralgia of the auriculotemporal nerve, in which the pain "like a child's comb" runs over the vertex from one ear to the other.) Among local causes, caries of the teeth, pyorrhœa alveolaris, osteitis alveolaris in people without teeth ("névralgie des édentés"), empyema of the antrum of *Highmore*, or of the frontal sinus, diseases of the nose, ears and eyes, must particularly be considered and in obscure cases there should be no hesitation in enlisting the services of the dentist, the oculist or the oto-rhinologist.

Not infrequently the attack of pain comes on at some definite time of day, for instance, on awakening in the morning, or at night (nocturnal neuralgia of *Oppenheim*). Chewing, yawning, sneezing, pressing the teeth together, blowing the nose, speaking, often set up the attack, so that many of these patients live in continual anxiety with regard to these "provocative agents," withdraw from all society, nourish themselves only insufficiently with liquid food, etc. Sometimes they discover for themselves certain procedures which can cut short the attack; for example, in neuralgia of the mandibular branch, the production of negative pressure in the mouth by movements of sucking, the mouth being closed, in that of the ophthalmic branch by energetic pressure upon a *Valleix's* point. The whole trigeminus is seldom affected, by far most frequently the ophthalmic branch, particularly its division supplying the forehead ("supra orbital neuralgia"); there is also a "ciliary neuralgia" which has its location behind the eyeball. Neuralgia of the maxillary branch affects by preference the infra-orbital nerve; it is somewhat more frequent than that of the mandibular.

On page 48 the *Valleix* pressure points in trigeminus neuralgia are exposed in tabular form.

* It is somewhat impressive that pressure upon *Valleix's* points causes local pain instead of pain projected into the terminal branches of the nerve. We know, for example, that irritation set up in an amputation stump is often perceived as pain in fingers or toes. Probably the sensitiveness of the pressure points depends upon an irritation of the nervi nervorum.

I Branch	II Branch	III Branch
1. "Supraorbital point," over the supraorbital foramen	1. "Infraorbital point," at the exit of the infraorbital nerve in the canine fossa	1. "Mental point," over the mental foramen
2. "Nasal point," somewhat inside of the inner canthus	2. "Malar point," at the exit of the malar branch on the zygoma (For. zygomatico-faciale)	2. "Auriculotemporal point," in front of the ear on the zygomatic process
3. "Palpebral point," laterally from the upper eyelid (exit of the lacrymal nerve)	3. "Dental points" on the upper gum	3. "Temporal point"
		4. "Parietal point," both in the course of the auriculotemporal nerve, between the auriculotemporal point and the vertex
		5. "Dental points," on the lower gum

Of the occasional, but in no way regular, accompanying symptoms of trigeminal neuralgia, the sensible and sensory disturbances must next be mentioned; most frequently a hyperesthesia in the skin territory of the diseased nerve is to be found. This is usually particularly marked, on touching the part with a cold object (a piece of metal); many patients evince great pain from this test. In inveterate neuralgias we find, however, here and there a slight reduction of cutaneous sensibility, while cases with actual anesthesia, are hardly any longer to be classed as trigeminal neuralgia. The attacks are often accompanied by photophobia, less frequently by narrowing of the visual field, difficulty in hearing or disturbances of taste. Vasomotor, secretory and trophic disturbances have been observed. I will content myself with mentioning the most important and interesting: heat and redness of the skin of the face, of the conjunctiva and of the mucous membrane of the mouth (upon which, indeed, small extravasations of blood may occur), increased salivation, lacrymation, rhinorrhœa, œdema, chemosis of the conjunctiva, the eruption of herpes vesicles; in inveterate cases, circumscribed falling out of the hair, decolorization of the hair, and atrophy of the integument. Finally, accompanying motor symptoms, are the chronic contractions of the muscles of the face, less frequently of the masticatory muscles occurring with severe attacks, which justify the denomination "tic douloureux" or "spasmodic epileptiform neuralgia" (*Trousseau*).

The prognosis of trigeminal neuralgia cannot be made on general principles, but only through intimate acquaintance with the clinical peculiarities of the individual case. There are forms which are cured completely and finally in a few weeks, while, on the other hand (particularly in old age), there are those which are refractory to every kind of therapy and which drive the patients to despair, so that suicides have been frequently observed. Even in mild cases the tendency to recur is very common, so that the history of the disease lasting years and decades is nothing unusual.

The clinical peculiarities by which the prognostically more favorable cases are distinguished from the more severe ones are the following:

"Neuralgia major" has the most marked paroxysmal character in that

the pain is not present between the single attacks; these, however, begin with lightning-like suddenness and usually with great severity. In the most severe cases, however, they follow so close upon one another that they make the impression of a continued pain. Typical is the provocation of these attacks by movements, by speaking, by chewing, by swallowing. The pressure points are very often not at the points of exit of the nerves, but upon the tooth-alveoli. In general, only one branch is affected, usually the maxillary; later, however, the ophthalmic and mandibular may be involved. Sometimes in the course of the disease there occur spasms which are now of involuntary character ("tic douloureux"), and again are voluntary (chewing movements, grimacing); secretory and vasomotor disturbances occur.

In "neuralgia minor," on the other hand, even between the attacks there is a certain amount of pain, from which the attacks distinguish themselves as exacerbations. *Valleix's* points are to be found particularly at the points of exits of the nerves, even between the paroxysms. The localizations of choice are the first and third branches, the second is less frequently affected, only very exceptionally the trouble involves the whole of the trigeminus. Motor, secretory and vasomotor symptoms are absent. Peripheral etiological factors (affections of the nose, the eye, the teeth) can often be made out.

b. Occipital Neuralgia

This is less frequent than trigeminal neuralgia and attacks the nervus occipitalis major, oftener than the nervus occipitalis minor. *Valleix's* points are, for the first, midway between the mastoid process and the upper cervical vertebræ on the linea nuchæ superior laterally from the insertion of the ligamentum nuchæ; for the latter nerve, the region between the insertions of the sternocleidomastoid and the trapezius, as well as over the mastoid process itself. The pains radiate over the occipital region to the vertex and are often bilateral. Etiologically, besides traumatic influences affecting the region of the neck, gout, malaria, influenza and typhoid fever come into consideration.

c. Neuralgia of the Phrenic Nerve

This rare neuralgia depends particularly upon local causes, pleurisy, pericarditis, aortic lesions, fractures of the clavicle. The attack of pain is often initiated by coughing, swallowing or drawing a deep breath; the pain radiates from the base of the thorax toward the neck, sometimes also toward the shoulder, the mastoid process, or the hand (which is explained by the anastomoses of the phrenic with the other branches of the brachial plexus). The following pressure points have been described: 1. At the insertion of the diaphragm into the tenth rib, somewhat lateral from the linea alba ("bouton diaphragmatique" of the French). 2. In the neck, in front of the scalenus anterior. 3. At the border between the bone and cartilage of the fifth rib.

d. Brachial Neuralgias

Apart from ulnar neuralgia in which gout is always to be considered in the etiology, true brachial neuralgias or "brachialgias" are rare. (On this account be cautious in making this diagnosis.) Neuralgiform pains in the arms should always arouse a suspicion of disease of the vertebræ or the meninges and tumors in the region of the plexus. Pressure points are, for the musculospiral the place where it runs around the humerus in the sulcus spiralis; for the ulnar, back of the internal condyle; for the median, the bend of the elbow and the palmar surface over the radio carpal joint.

e. Intercostal Neuralgia

This localization of neuralgia is again a much more frequent one. Together with the causative factors for neuralgias in general, a number of pathological conditions of the thorax and its contents must be specially mentioned; such are scoliosis, kyphosis, fractures of the ribs, pleurisies, aortic aneurisms. The great majority of cases, however, can be referred to general deleterious influences (infectious diseases, anemia, cachexia, etc.). It must be especially pointed out that tumors of the spinal cord for a long time may have their true character concealed under the harmless mask of an intercostal neuralgia. As concerns the prognosis, many cases of intercostal neuralgia are characterized by extreme persistence, almost all, by a tendency to relapse. The pressure points are quite characteristic: the "vertebral point" lies just along the spinal column at the level of the affected nerve, the "posterior perforating point," upon the sternum, or the rectus abdominis, close to the middle line.

f. Mastodynia

Neuralgia of the mammary glands or mastodynia, is a special variety of intercostal neuralgia which is limited to the definite branches of the second to sixth intercostal nerves innervating the breast. This extremely obstinate complaint, which is identical with *Astley Cooper's* "irritable breast," affects almost solely women, often appearing for the first time during pregnancy or the puerperium. The attacks of pain sometimes coincide with menstruation. The skin over the mamma, especially about the nipple, is extremely hyperesthetic during the attack: sometimes also, red and swollen. *Erb* has described the spontaneous discharge of a milky fluid at the height of the attack of pain. Painful points are the vertebral points of the above mentioned five intercostal nerves.

g. Lumbar Neuralgia

This very rare form of neuralgia, according to its location, is divided into lumbo-abdominal, crural, and obturator neuralgia. One should be on his guard in these cases quite as much as in brachial neuralgias against overlooking local causes (lumbar vertebræ, tumors in the pelvis).

h. Sciatica

Sciatica neuralgia, or sciatica, also called *Cotugno's* disease, after the classical description of this author (1764) is the most frequent of all the forms of neuralgia. It occurs in men somewhat oftener than in women, by preference in the fourth and fifth decades of life. Its causes are manifold, particularly often, exposure to cold, as by sitting on cold stones, camping on wet ground, etc., are accused. Also trauma plays no small rôle. This sometimes is not directly to the pelvis but may affect the periphery of the lower extremity (I saw a particularly obstinate case arise in immediate connection with stumbling, the foot being caught, through which the limb suffered a torsion). On account of causing compression of the nerve within the pelvis, retrouterine hematocele, chronic constipation with fecal impaction in the rectum, tumors of the sacrum (carcinoma metastases) also tuberculous osteitis, etc., act as causative factors of sciatica. *Quénu* has also called attention to "varicose sciatica" in consequence of dilatation of the veins accompanying the nerves to the hip. All these eventualities must be considered in taking the history as well as in estimating the condition (examination by rectum or vagina). General toxic causes which according to experience often form a substratum for sciatica, are particularly gout, diabetes (in which the otherwise very rare—and always suggestive of vertebral disease—bilateral sciatica sometimes occurs), syphilis, tuberculosis, malaria, typhoid fever, influenza, gonorrhœa. Finally those cases which are found in patients with flat foot or with diseases of the generative organs or of the rectum are to be classed as reflex neuralgias.

Sciatica can begin quite suddenly or the disease picture unfolds itself little by little to its full height. The paroxysms of pain are either separated by intervals of entire freedom, or (this applies to the majority of cases), there persists also between the attacks proper, a dull feeling of pain along the course of the affected nerve (in the region of the loins and the buttocks, the posterior surface of the thigh, in the popliteal space, on the outside of the leg, and foot). The pains of sciatica are increased by cold, by sitting and by walking, while lying and standing are in general most tolerable. However, very frequently at night, without any apparent cause, the severest paroxysms occur.

Valleix's points are sometimes absent in otherwise typical cases of sciatica but in the very great majority of cases one or other of *these* points is to be recognized as sensitive: 1. "Lumbar point" over the spinous process of the fifth lumbar vertebra; 2, "Ileosacral point" near the posterior superior spine of the ilium; 3, "Gluteal points" over the great sciatic foramen and in the fold between the trochanter and the tuber ischii; 4, "Popliteal point" in the middle of the popliteal space; 5, "Peroneal point" behind the head of the fibula; 6, "Malleolar point" behind the external malleolus; 7, "Metatarsal point" in the first intermetatarsal space of the dorsum of the foot. The gluteal points are most frequently recognizable.

Of great diagnostic importance is "*Lasègue's* sciatic phenomenon." While the patient rests in a horizontal position, the leg extended at the knee, is slowly raised by the examiner; when the foot becomes separated about 35 to 45 cm. above the bed, there is sudden and severe pain on the posterior surface of the

thigh which, however, passes away as soon as the knee is bent and the sciatic is so relaxed again. "*Bonnet's* phenomenon" depends upon the same principle; passive simultaneous flexion of the hip and knee joints calls forth no painful reaction; this occurs, however, when the leg is also brought into the position of adduction.

Finally, in only part of the severe cases of sciatica "*Bechterew*'s sciatic phenomenon" is found; if the patient extends the sound leg he can scarcely stretch the affected one, and vice versa.

In walking a patient with sciatica instinctively avoids those movements which lead to stretching the affected nerve, and on this account generally keeps the hip and knee of the side involved in a position of light flexion. Besides this he attempts to spare the affected leg as much as possible by supporting himself mainly on the sound one. Also, he often turns the foot outward. In standing also an anomaly of position is often observed; in order to relieve the affected leg, by letting the vertebral column fall over toward the healthy side, the center of gravity is shifted toward that side. There is, however, also a form of sciatica (the "spasmodic sciatica" of *Brissaud*), in which, by a contracture of the erector spinæ on the affected side, occurring reflexly, a homologous scoliosis is produced instead of the cross scoliosis. An "alternating sciatic scoliosis," in which the crossed and the homologous scoliosis alternate with one another has also been described. With cure of the sciatica the anomalies of position of the vertebral column, as a rule, entirely disappear. In sitting, the sciatic patient rests his weight exclusively on the sound side, he "sits on the edge of the chair"; in rising he avoids bending the body forward, as is done normally.

The objective testing of sensibility shows either no anomalies, or hypesthesia for the different qualities of sensation in the distribution of the nerve (most frequently only in one branch, *e.g.*, the peroneal), less frequently hyperesthesia. On stroking the sole, the sensation of tickling and the resulting jerking away of the foot is often absent on the affected side, while, on the other side both are present. Motility is only so far affected, in that by a long duration of the disease, there is some atrophy of the affected muscles, and that during the paroxysms of pain sometimes muscular spasms are observed. Herpes is rare, also vasomotor phenomena, *e.g.*, local reduction of temperature. The Achilles tendon reflex, which occurs through the sciatic, is either reduced or absent, or (especially in *Brissaud's* spasmodic sciatica already mentioned), on the contrary exaggerated. The same remarks apply, as a rule, to the patella reflexes both of the affected and of the sound side. Finally, I will relate two more interesting objective symptoms which are not at all infrequent in severe sciatic neuralgias; the Achilles tendon on the affected side feels more relaxed than upon the sound side (*Oppenheim*); the gluteal fold hangs lower upon the affected side and shows its greater flaccidity sometimes by a peculiar folding (*Bonnet*).

From a differential diagnostic point of view, sciatic neuritis comes specially into consideration. This is, as compared to sciatic neuralgia characterized especially by continuity of the pain, greater intensity of the objective disturbances of sensibility (sometimes there is in places complete anesthesia!), more marked muscular atrophy, finally, reaction of degeneration, more prominent

vasomotor and trophic anomalies. In addition there is increase in the size of the nerve which can sometimes be felt as a firm cord behind the trochanter.

Very frequently chronic sciatica is confused with malum coxæ senilis. This confusion can be avoided, however, if it is remembered that *Lasègue's* phenomenon is absent in the latter disease, while, on the contrary, movements at the hip joint are more difficult and painful, that pain is already very severe on beginning locomotion, but grows less in the course of walking; that further, in contradistinction to sciatica, it is dependent upon the nature of the road, is increased by unevenness of the ground, and that finally, not the nerve trunk but the hip joint shows itself sensitive to pressure. Not adduction, but abduction is hindered, thrusting the head of the femur into the joint cavity (by a blow upon the trochanter) calls forth a sharp pain reaction.

I. Neuralgias of the Feet

The foot is a situation of predilection for certain pains limited to a narrow region and of plainly neuralgic character whose nature is often obscure, at whose basis, however, there is usually some anomaly, though perhaps a trifling one, of the neighboring structures (skeleton, tendons, etc.). There is metatarsalgia or *Morton's* disease, which is located in the periarticular nerve branches of the fourth metatarso-phalangeal joint, and presumably is caused by an abnormally movable fifth metatarsal bone being pushed under the fourth by a too narrow shoe and there exerting pressure. We may also mention talagia or tarsalgia (pain in the heel), for which, sometimes, exostoses on the calcaneum, or diseases of the serous membranes are underlying causes, which, however, can be observed without any objective findings; the same is true of Achillodynia, which originates from the point of insertion of the Achilles tendon.

J. Spermatic Neuralgia

This form of neuralgia is characterized by exceedingly severe paroxysms of pain which radiates either from the testicle along the spermatic cord to the inguinal canal, or in the opposite direction and is almost always unilateral. At the same time there is great hyperesthesia of the scrotum and testicles, so that even a slight touch is very painful ("irritable testis" of the English). In making this diagnosis one must not forget to think of local diseases of the testicle as well as renal colic, in which the pain usually radiates into the testicles. We will learn further, on considering tabes dorsalis, that in this disease of the spinal cord also, pains in the testicles occurring in paroxysms and of very severe character, may be observed.

K. Coccygodynia

In coccygodynia, or neuralgia of the coccyx, attacks of pain either occur spontaneously or are set up on sitting down, by defecation, or by walking. Pressure points are found over the coccyx. As a chief causal factor for this disease, which attacks females by preference, difficult labors are mentioned.

Traumatic or inflammatory lesions of the coccyx or of the muscles inserted into it, usually form the basis of the disease, however.

D. Treatment of Diseases of the Peripheral Nerves

1. THE THERAPY OF PERIPHERAL PARESES AND PARALYSES

We will refrain from considering at all, at length, things like primary nerve suture in the severing of peripheral nerves, the removal of tumors which compress nerves, the separation of nerves from scars or cicatrices and masses of callus, all of which you will find described in surgical works. All the more important will it be for us to undertake a careful description of the relations in those cases of interruption of conduction which present no indication for the procedures indicated (and they represent the impressive majority).

In the early stage, indeed, too active therapy should be avoided, the affected extremity should be put at rest—in a middle position for the upper, in a suitable one for the lower extremities. For example, if in a peroneal paralysis we do not want to furnish the opportunity for development of a pes equinovarus, the pressure of the bedclothes must be kept off the feet; supporting the covering upon a frame, however, often causes in patients an unpleasant sensation of cold in the legs unless a thin layer of felt is spread over them. In order that the feet be kept as near as possible in a position at right angles to the legs we furnish them with a suitable support which may be improvised with the aid of a footstool, a cigar box, or a pasteboard box. Absolute immobilization of limbs with neuritis (in a padded wire splint, for instance), I would, in general, advise against, and would prefer, rather, comparative immobilization by placing them between sandbags; this method permits greater individualization in the matter of this important postulate, to relieve tension in the affected nerve as much as possible and to protect it from pressure. In musculo-spiral paralysis, to avoid overextension of the extensors of the hand, the latter must not be allowed to remain permanently in a position of "wrist-drop"; here the application of a small, well-padded dorsal splint will make possible more rapid return of contractility in the muscles supplied by this nerve.

In the second place it is important to keep in mind the general condition of nutrition of the patient, since the better this is, the more reason have we to hope for conditions favoring the regeneration of the nerve and the return of its ability to conduct. This indication is frequently a "causal indication," in so far as we can find decided anemia, marantic conditions, glycosuria, etc. In alcoholic neuritis and polyneuritis absolute prohibition of alcohol is in place. An exception is only to be made when the condition of the heart gives reason to fear sudden collapse upon complete withdrawal, or if the psychical condition is such as to arouse suspicion of threatening delirium. In such cases the dose of alcohol is to be reduced as far as is considered safe and complete withdrawal is to be gradually accomplished.

After about fourteen days, if we feel sure that the motor symptoms no longer

show a progressive tendency (and if the continuance of severe pains does not suggest further abstention—see below, page 57 et seq.), the aim of our therapy should be energetic support of the process of repair.

Electrotherapy must here be mentioned as our most valuable agent; when possible applied daily. The indifferent, large electrode is placed upon the chest or back of the patient, the "different" or "stimulating" electrode, about 10 cm square, successively upon each of the paralyzed or paretic muscles, in which at first 5 to 10, later 20 to 40 contractions should be produced. Rule: The weakest current necessary to produce a decided effect should be used, and as different electrode that pole to which the muscle reacts best (also usually, on account of the reaction of degeneration, first the anode, later, as there is improvement, the cathode). Faradization of the muscles is usually useless in this stage, since they do not generally react to the induced current; in later stages of convalescence it may (in the form of faradic massage with the roller electrode) be in place. Used too early, strong faradization in neuritis, and especially in polyneuritis, may be directly injurious by reawakening pain and by unsuitable application (stimulating the antagonists) may favor contractures. That in spite of this, unfortunately, in peripheral paralysis faradism is far too much used, is due to the fact that many physicians are obsessed by the opinion that electrization consists in planlessly running over the parts with the faradic electrode, and hence neglect to provide themselves with galvanic apparatus or to learn its use.

Mild massage (with avoidance of too firm petrissage) will also be of advantage in the stage of repair of peripheral interruptions of conduction. Particularly important, however, is the intelligent and conscientious application of passive movement as prophylactic against contractures and faulty positions of the joints. They may be advantageously applied in or after a warm bath, which finally, with a gradually increasing content of sea salt, brine, pine needle extract, or other coniferous preparations is usually found strengthening and agreeable. It is also encouraging for the patient to note the increased ease with which he can move the paretic limbs in the bath ("kinetotherapeutic baths"); in this factor, which depends on the relative reduction in weight through the buoyant effect of the water, there is a welcome reëducatory influence. In exercise therapy, which is best undertaken in connection with electrotherapeutic applications, the greatest stress should be laid upon movements against graduated resistance. In these, the patient is encouraged to exert contractions of the muscles against moderate resistance made by the physician, which, separating the origin and the insertion of the muscle, stretches this, and so facilitates its voluntary contraction. If apparatus for half active (that is, with the aid of machinery), gymnastics is available, it can be used with advantage.

We can, and should also, support the reparative processes with drugs. Strychnin, by increasing the reflex irritability in the cells of the anterior horn of the spinal cord, favors "path formation" by the motor impulses. Besides this, it, as well as certain phosphorus derivatives (glycero phosphate of calcium, phytin, etc.), works as a tonic, increasing the appetite, perhaps specially favors the nutrition of the nervous system. It can be given as tablets con-

taining grm. 0.001 (grain 1/60) of strychnin nitrate, one or two twice daily, or as tinc. nuc. vom., 10 drops t. i. d., or, finally, as injections of strychnin nitrate 0.001—0.004 (gr. 1/60—gr. 1/15) once a day. Finally, arsenical medication may come into question (Pil. Asiatic, Sol. Fowler, Sol. Pearson, injections of Cacodylate of Sodium, etc.).

Finally, if after peripheral paralyses, power is so far regained that the question of a sojourn at a health resort can be considered (in which case besides the baths, other curative factors must be available to the patient), we have the choice between the different kinds of resorts, among which the selection should rest more upon the financial resources of the patient, the time of year, and the agreeability of the resort, than upon a specific difference of character of the springs. There come into consideration: 1. Saline baths (for example, Rheinfelden, Kreuznach, and in the United States Saratoga and Mt. Clemens, Mich.). 2. Carbonic acid salt baths (Nauheim, Byron Hot Springs, California). 3. Thermal waters of indifferent composition (Teplitz, Ragaz, the Hot Springs of Virginia and of Arkansas). 4. Thermal waters containing common salt (Baden-Baden, Wiesbaden). 5. Hot Sulphur baths (Aix-les-Bains, and in the United States, Clifton or Richfield Springs, New York, Glenwood Hot Springs, Colorado, and Gilroy Hot Springs, California). 6. Mud, peat and fango baths (Franzenbad, Postyen, Las Vegas Hot Springs, N. M., Paso Robles, California).*

The symptoms which remain in spite of the different therapeutic measures which we have described are to be treated by the orthopedic surgeon with apparatus for extension, or for support, by tendon and nerve transplantation, etc.

2. THE TREATMENT OF ANESTHESIA

Where there is decided reduction of sensibility there is always the danger that the patient, on account of his insensibility, will not protect himself from injuries which can cause deep wounds tending to heal very slowly, hence care must be taken that hot-water bottles, poultices, the thermaphore, etc., are not applied too hot. The so-called "neuro-paralytic keratitis" which is sometimes observed in anesthesia of the upper branch of the trigeminus nerve, is only in part to be considered as a trophic disturbance and the fact that the eyeball, on account of its loss of sensation is much exposed to trauma, erosions, etc., plays in this case the chief rôle. If the action of foreign bodies from the outside is prevented by suitable protecting glasses (which also prevent the drying of the epithelium, caused by interference with the tear secretion), this serious complication can usually be prevented. For anesthetic skin areas, under some circumstances "dry faradization" with the wire brush electrode is to be recommended.

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* Names of American resorts added by the Translator.

3. THE TREATMENT OF NEURITIC AND POLYNEURITIC PAIN

In patients with fresh acute neuritis or polyneuritis, our first duty is to help the patient over the days of the worst pain. The different "antineuralgics" exercise, it is true, an individually variable, but in general a quite satisfactory effect. As a rule, pyramidon, 0.3 to 0.5 (grs. 4 to 7, t. i. d.), works most efficaciously, some patients, however, respond better to a mixture of the other analgesics. Since a change of remedies may be necessary from time to time, I will give a few combinations which have proved useful to me:

℞	Acetanilidi	0.2	(gr. 3)
	Phenacetini	0.1	(gr. 1½)
	Quinine valer.	0.05	(gr. ¾)
℞	Exalgini	0.1	(gr. 1½)
	Caffeini citric.	0.05	(gr. ¾)
	Phenacetini	0.2	(gr. 3)
	Antipyrini	0.4	(gr. 6)
℞	Phenacetini	0.6	(gr. 8)
	Acetanilidi	0.3	(gr. 5)
	Codeini	0.04	(gr. ½)
℞	Aspirini,		
	Lactophenini āā	0.5	(gr. 7½)
	Caffeini citrici	0.1	(gr. 1½)

It will be well to prescribe at night an analgesic hypnotic combination, for example, Pyramidon, 0.3 (grs. 4½); Veronal, 0.5 (grs. 7½), or Lactophenin, Trional, āā 0.5 (grs. 7½). With ordinary hypnotics, if the pain is severe, almost nothing is accomplished. The following mixture will give good service:

℞	Potass. bromide	10.0	(℥iiss.)
	Chlorali hydrati	5.0	(gr. lxxv)
	Antipyrini	3.0	(gr. xlv)
	Codeini phosph.	0.4	(gr. vi)
	Aq. menth. pip ad	150.0	(℥v)

M.S. Tablespoonful at night.

The injection of morphine or of one of its modern succedanea in neuritis and polyneuritis should never, or only very exceptionally, be resorted to.

When the period of severe spontaneous pains has been overcome so that these, in spite of intense sensitiveness to pressure which persists, occur with less intensity, that is, after about four or five days, the application of heat may be begun; particularly applicable are apparatus for hot air, for example, *Bier's* boxes.

By the application of the so-called "Phœnix" ("Phenix a l'air chaud"),

which permits an intensive thermotherapy to be carried on in bed without disturbing the patient, we may obtain at the same time a conveniently regulated diaphoresis. Encouragement of the sweat secretion is often considered as of itself a curative agent which acts upon the cause in toxic polyneuritis, and in any event contributes to the alleviation of polyneuritic pains. The duration of this sort of a sweat bath (to be given once a day) should be from 10 to 15 minutes; careful control of the pulse is absolutely necessary where the heart is not entirely intact. When the treatment with hot air is inapplicable on account of weakness of the heart, or for some other reason, the application of moist heat (two to three times a day from one-half hour to one hour) is proper, in spite of its inconveniences; nevertheless the application of a compress which is best soaked with warm mildly stimulating decoctions (camomile, matricaria) as well as the envelopment of the part with waterproof tissue, is the more disagreeable for the patient the larger the skin surface which must be treated. Linseed meal poultices are often unpleasant on account of their weight, the same is true of moist compresses kept warm by the thermaphore; the dry thermaphore is often found disagreeable.

During this time we should endeavor to dispense with the analgesics prescribed at the start, or at least, only to order them at night. I often substitute for these drugs aconitin or colchicin, which are withdrawn as soon as the curative effect of the warm applications is plainly evident. Aconitin is best given once or twice a day in the form of 1/10 mg (gr. 1/600), taking care to use an active preparation (*Merck's* or *Clin's* preparation), colchicin in the form of tinctura colchici, 5 drops t. i. d. (eventually tinct. aconiti, tinct. colchici, āā 10 drops t. i. d.).

The withdrawal of the internal analgesics is facilitated by some external applications, which are to be recommended upon psychological grounds, since they occupy and divert the patient. Massage (also the rubbing in of ointment) is to be avoided as long as the acute painful stage is not certainly over; also any stimulating electric application. On the other hand the stabile treatment of the nerve trunks with the anode (application of the electrode upon the place of greatest sensitiveness to pressure for from 3 to 5 minutes) with weak current (3 to 5 milliamperes an electrode of the size of a dollar), with the cautious turning on and off the current is to be unqualifiedly recommended. For rubbing on, fluid liniments applicable without pressure are suitable. Of these the following are examples:

1. Ol. juniperi, 2.0; ol. terebinthini, spts. camphori, spts. saponis, liq. ammon. caust., āā., 12.0. (Shake well.)
2. Camphor, chloral hydrat, āā., 20.0.
3. Chloroform, 10.0; liq. ammon., 40.0.
4. Veratrin., 1.0; chloroform, alcohol, āā., 24.5 (cave oculos).

Upon areas which are the seats of annoying paresthesia, pieces of flannel soaked in the following solution can be laid on: Menthol, guaiacol, āā., 1.0; alcohol abs., 20.0 (cave oculos).

4. THE TREATMENT OF NEURALGIAS

In every fresh neuralgia of any considerable intensity, bodily rest is an urgent requisite; in trigeminal or intercostal neuralgia, as a rule, keeping to one's room (in which case visits, etc., are to be avoided) is sufficient. In sciatica, rest in bed should be enforced. In this the painful limb should be carefully placed in such a position as relaxes the affected nerve as much as possible. This is best accomplished by placing a roll of felt under the bend of the knee and keeping the extremity in a position of moderate abduction with sand bags. Sometimes (in varicose sciatica, for example) it is desirable to raise the foot of the bed. A patient with sciatica should not leave his bed until the neuralgic paroxysms have ceased, or at least have plainly abated. He should be told in advance that it will be necessary to remain in bed two or three weeks, perhaps longer; at the same time antineuralgic remedies should be given and in sufficient doses—a point on which I lay considerable stress, since many failures are due to too timid medication. In sciatica, salicylate of sodium should always be tried first. This has a decidedly more energetic effect than aspirin. This last is to be preferred only in people with sensitive stomachs.

℞ Sodii salicylat, 20.0 (℥v); syr. aurantii cort., 40.0 (℥x); aq. menth. pip., ad 300.0 (℥x). M.S. A tablespoonful 4 times a day in half a glass of water after meals. To avoid disturbance of the stomach a solution of about 30 grains of sodium bicarbonate can be taken after the salicylate.

The daily dose can soon be reduced to from two to three grammes (30 to 45 grs.) a day. As to the other antineuralgics, I would refer to the prescriptions recommended for neuritic and polyneuritic pains. Particularly in trigeminal and occipital neuralgia some other drugs are applicable, sometimes with quite happy effects: Migranin (citrate of antipyrin and caffein), in single doses of grm. 1.00 (15 grs.), butylchloral or its combination with pyramidon, trigemin (which must only be used when it is of a clear, white color), in single doses 0.5 (7½ grs.), atropin or methylatropin, the first in doses of 0.0005 (gr. 1/120), of the last 0.002 (gr. 1/30); finally tinct. gelsemii, 10 to 15 drops. Aconitin, already mentioned in the treatment of neuritis, is also frequently a very efficacious remedy in trigeminal neuralgia, especially when it is used for a long time with a daily dose of saline laxative.

The thermotherapy of neuralgia is the same as that already indicated for neuritis and polyneuritis. In the treatment of sciatica, especially in subchronic and chronic cases, besides this, the following procedures are suitable: Electric light baths, hot air douches, sun baths, steam douches, "Fango-packs," hot baths. Cold is in general only found agreeable in fresh cases of facial neuralgia, and in these may even work curatively; however, on this point there are so great individual differences that it is necessary to try it out from case to case. Freezing the skin over the *Valleix* points with the chloride of ethyl spray used for local anesthesia deserves special mention. It can be tried in all forms of neuralgia with plainly localized sensitiveness to pressure (in the neighborhood of the eyes great caution is necessary; the eyes must be carefully covered with cotton). Naturally the freezing of the skin areas, which is made evident by

their white color, can only last a few seconds; otherwise there is danger of circumscribed necrosis. After the freezing the affected part must be carefully rubbed with some simple ointment. This application can only be repeated after the reddening of the affected part (which sometimes persists long after the application of the chloride of ethyl) has disappeared and the parts look normal again. Other "revulsives," whose application is naturally restricted for the most part to the trunk and extremities, are blistering with cantharides, painting with iodine, dry cups, mustard plasters, capsicum plasters, faradization with the wire brush, ignipuncture, etc., also local bleeding (with wet cups or leeches) is occasionally applied. In all these applications care should be taken that the integument should not be injured to an extent which would prevent the later application of the electric current.

In galvano-therapy the stabile application of the anode to the pressure points is to be preferred. In persistent neuralgias the stabile application to the diseased nerve, the two electrodes, of about the size of a dollar, being placed, one upon the most proximal and the other upon the most distal point in the course of the nerve, which is accessible, a current of about five milliamperes gradually introduced and allowed to pass for from five to ten minutes (whether its direction is ascending or descending is a matter of indifference), can be used. In fresh cases of neuralgia, massage, apart from light vibration of the painful points is to be avoided; in old cases, on the other hand, particularly in sciatica, petrissage of the affected region is an important curative measure, particularly when properly carried out vibration of the nerve trunk is added to it. In sciatica also, "bloodless stretching" of the nerve is a mechanical therapeutic procedure much to be recommended; it is especially easy to carry out. The leg, extended at the knee, is raised from the bed as in *Lasègue's* test (see above, page 51), but only so far as it will go without provoking any considerable pain, and is held for several minutes in this position; at the end of this time it is stretched somewhat more strongly, until severe but still tolerable pain is produced, and then carefully laid down again. It will often be noticed that from one treatment to another, the distance through which the leg can be bent without pain increases. "Bloody nerve stretching" is now quite abandoned.

A useful procedure is the injection treatment of neuralgia. We distinguish:

1. *Lange's* method of perineural infiltration. This is a modification of *Schleich's* infiltration anesthesia. In the trigeminus a few cubic centimeters of the following solution: Beta-eucaine, 0.1; normal salt solution, 100.0—are injected over *Valleix's* points in the immediate neighborhood of the affected nerve branch. A stovain-adrenalin solution according to the following formula is also used: ℞ Sol. adrenalin (1%), gtts. v-x; stovain, 0.1-0.2; sol. sodii chloridi (0.8%), ad 100.00. In the sciatic large quantities (70-100 cc.) are injected. A *Schleich* infiltration is made over the point of exit of the nerve (in the middle of a line joining the trochanter and tuber ischii) and a 10 cm. long cannula is passed carefully through this down into the nerve (which is here 1½ centimeters in diameter) when a sensation of pain (and often a contraction of the muscles of the leg) announces that the cannula has entered the nerve. The injection is then made by means of a syringe or an irrigator. Whether the use of ice cold solutions gives better results is questionable, how-

ever, the action of simple physiological salt solution without the addition of an anesthetic sometimes is very satisfactory. The therapeutic effect of this procedure is perhaps purely mechanical; a swelling, perhaps a stretching of the nerve which can then produce a cure by reactive inflammation is its result, according to *Lange*.

2. Neurolytic injections. In these methods we endeavor to injure the nerve by the injection of different solutions, also in a way, to resect it chemically. We reserve them on this account for severe cases, since they have as a result anesthesia of the skin, and avoid them in mixed nerves (for example, the sciatic) in order not to risk a motor paralysis. Further, they should not be made into the supraorbital canal, since this, in many individuals, communicates with the orbit, the penetration into which of neurolytic solutions might severely injure the optic nerve. We will not discuss all the substances which have been applied as neurolytics (ether, carbolic acid, silver nitrate, chloroform, etc.), but will only indicate two solutions from which we have sometimes seen good results: (a) 1% osmic acid solution; (b) 80% alcohol, with the addition of an anesthetic ("*Schlösser's* injections"). Formulæ to be recommended are: Alcohol (80%), 20 cc.; menthol, 0.4; novocain, 0.2; or alcohol (80%), 20 cc.; stovain, 0.2. In the branches of the trigeminus, one to one and a half cc. are injected into the nerve, if possible, or at least into its immediate neighborhood; *Ostwalt*, indeed, seeks these branches directly upon the base of the brain (in the foramen ovale or the foramen rotundum), for which he uses a specially bent (bayonet formed) cannula, passing it from the mouth; *Levy* and *Baudoin* pass the needle in front of the coronoid process of the lower jaw, or between this and the articular process through the cheek. These methods, however, have not been universally accepted. In every injection of such solutions, the penetration of a vessel must be carefully avoided by first inserting the empty needle and drawing out its contents. The injection of osmic acid, a drop at a time, into the nerve branch laid bare by operation, and indeed, from the exposed base of the skull into the *Gasserian* ganglion, has been carried out. This brings us to the surgical therapy proper of prosopalgias. The extra cranial methods are easiest; simple section (neurotomy) has been given up, since rapid reunion occurs, and for it has been substituted nerve evulsion after *Thiersch-Witzel* (neurexaresis) which permits a tearing out of the affected nerve from the base of the skull to its terminal ramifications. In spite of this, many prosopalgias which have been subjected to neurexaresis recur, as do many cases treated with neurolytic injections. While, however, the last can be repeated a number of times, after evulsion of all three branches of the trigeminus, the trouble must be attacked at its root, in the fullest sense of the word and extirpation of the *Gasserian* ganglion must be performed. *Fedor Krause*, the originator of this operation, states that in sixteen years he has performed it sixty-four times and has never observed a recurrence. In two cases, which I know from my own observation, recovery also occurred. However, this severe operation remains a last resource, in the fortunately rare, specially obstinate cases of prosopalgia.

In severe intercostal neuralgias the corresponding posterior roots have been divided with good effect. Neurectomy has been done not so infrequently in

occipital and spermatic neuralgias. In the X-ray treatment of neuralgias I have had no personal experience.

Finally, it must be remarked that a change of diet to a purely egg, milk and vegetable régime, can be of the greatest use in neuralgia, and that of course the treatment of any recognizable underlying disease must never be neglected (iron, quinine, mercury, iodide of potassium, etc.), that in all senile forms of neuralgia an arsenic cure should not be left untried, and finally, that the balneotherapy of neuralgias is entirely similar to that recommended for neuritis.

LECTURE IV

The Dyskinesias

GENTLEMEN: In neurology, abnormal motor processes which occur in part as symptoms accompanying different diseases of the nervous system, in part are to be considered as autonomous pictures "sui generis," play an important and interesting rôle. In this lecture and in the next one we will occupy ourselves with these "*Dyskinesias.*"

A. Tremor

As tremor we designate involuntary rhythmic oscillatory movements, which affect now all the muscles, again only single groups. According to the rapidity of the oscillations, we speak of rapid or slow tremor (the extremes are about between 4 and 10 oscillations per second), according to the amplitude of the excursions, of coarse, medium and fine tremor. The coarsest form is denominated tottering, the finest as vibrating tremor or thrill. If the tremor is absent during rest, appearing only upon carrying out movements, it is called intention tremor. A particular kind of tremor of the fingers is the so-called *Quinquaud's* phenomenon, which was formerly considered a pathognomonic symptom of chronic alcoholism, but can occur in all disease conditions of which tremor is a symptom. If the examiner presses his hands against the tips of the spread fingers of the patient, he will perceive in these a peculiar unrest in the articulations, in which, presumably on account of a tremor of the interossei, the joint ends of the phalanges are pulled from side to side. Within normal limits, tremor, as is well known, occurs on shivering, in great fatigue, and in the emotion of apprehension; also the tremor of old age, as long as it remains within moderate bounds, may be considered as a physiological phenomenon; with pathological tremor of the most varied origin, on the other hand, we will meet in the further course of these lectures, quite frequently as a symptom of many functional and organic nervous diseases. At present, however, we will only consider that form of tremor which in itself constitutes a disease.

ESSENTIAL TREMOR

In this disease, which occurs mainly as a hereditary family affection, the single symptom is a rhythmical tremor of small amplitude, but without characteristic tempo. Nevertheless, it is in general more rapid in young persons, slower in old ones. It ceases during sleep, sometimes also while the patient is awake, but in absolute physical and mental rest; in any event, it is much less marked under these last conditions. Many patients can control the tremor

by an effort of the will, while in others such an attempt, on the contrary, increases the tremor. The carrying out of voluntary movements can arrest the tremor; one of my patients, for instance, was able, by strongly clenching the right fist, to arrest the tremor in both hands; on the other hand, however, an intentional character of this essential tremor is occasionally observed. All the voluntary muscles can be affected. The symptom is often exclusively observed in the hands. Tremor of the legs can sometimes interfere with walking, that of the tongue with speech. Frequent complications are malformations, epilepsy, psychoses, on which account the French speak of "tremor of the degenerate." Here and there alcoholism of the parents has been accused of being an etiological factor; nevertheless, there are families with tremor in which the ascendants have been characterized by great abstemiousness. As exciting causes, overexertion, strong emotion, infectious diseases, have been mentioned. The disease is now congenital, again it begins during childhood or puberty, or even later. The fortieth year of life is about the farther boundary.

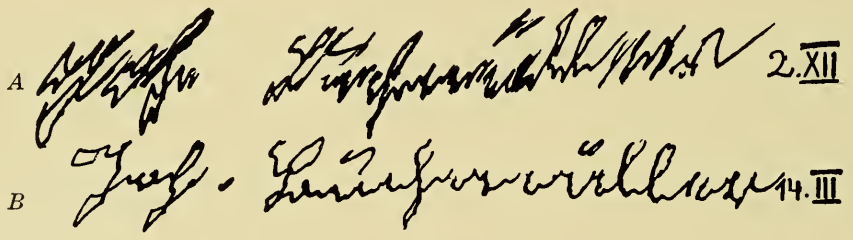

Fig. 27.

Hereditary Family Essential Tremor. Handwriting.
A. Before treatment. B. During treatment.

The tremor may be stationary, may progress, or, rarely, may grow less, but never ceases entirely. Therapy can accomplish but little. *Regnault* claims to have obtained improvement by limiting the amount of alcohol used; on the other hand, in a family with tremor, described by *Nágy*, the persons who drank most had the least tremor. In the particularly severe case under my own observation, whose handwriting is shown in Fig. 27, after the taking of wine the tremor much decreased, while after coffee it decidedly increased. Bromides, veronal, adalin, scopolamin, were without influence, while, on the other hand, the patient reacted to the "pilulæ hyoscyami compositæ" (extr. hyosc. zinc. oxid., āā 5.0 (gr. lxxv); extract. valer. 10.0 (℥iiss). M. Fiat pil. No. 100. S. 3-5 per day) with decided improvement of the tremor, so that the formerly indecipherable handwriting became legible again (see Fig. 27).

B. Fibrillary Contractions

We give this name to a symptom of motor irritation which manifests itself in isolated and successive contractions of the single fiber-bundles of a muscle. To motor manifestations proper, these contractions, which are usually of lightning-like rapidity and plainly perceptible both by inspection and pal-

pation, do not, as a rule, lead, though occasionally when the phenomenon is very marked in the thenar muscles, a slight twitching of the thumb may result. Fibrillary contractions occur in functional neuroses and can, indeed, be provoked in normal people (for example, by refrigeration); but they are of most importance clinically when they inform us of a pathological process in the anterior horns of the spinal cord (see, farther, Lectures VI and VII). A variety of fibrillary contractions is myokymia.

C. Muscular Spasms

In the large group of symptoms of motor irritation (hyperkinesias) which we denominate "cramps" or "spasms," two categories are to be separated from each other: tonic and clonic spasms.

Contractions of single muscles, or of muscle groups, of short duration repeating themselves in fits and starts are characteristic of clonic, protracted contractions of tonic spasm. By a succession of clonic and tonic spasms the complicated irritative phenomena denominated as convulsions arise. These we will have to study more closely under epilepsy. They constitute the most important symptom of motor irritation of cerebral origin. Also by the action of pathological irritants upon the spinal motor tracts in the lateral columns, there occur sometimes spasmodic phenomena; for example, the so-called *Brown-Séquard's* spinal epilepsy, a spontaneous clonic twitching occurring in attacks, a more or less severe shaking of the affected extremities on account of alternating contractions of their extensors and flexors. The spasm-producing properties of many animal and vegetable poisons are known to you; for instance, those of the toxine of tetanus and of strychnine. We will not, however, enter into these things, but will turn our attention to the "idiopathic" forms of spasm.

1. LOCAL SPASMS

As to the etiology of these conditions we are very poorly informed; we can only say that neuropathic individuals and those afflicted with neuroses and psychoses are predisposed to localized muscle spasms, and that in a good part of the cases irritants, working reflexly, appear to be the starting-point for the affection. So, for example, carious teeth, arthritis of the jaw-joint, suppuration of the jaw cavities, eye diseases, have been held responsible for spasms of the jaw and facial muscles. Only seldom can a disease process, working as a direct irritant upon the nerves of the muscles affected with spasm, be discovered: for example, in facial spasm, aneurism of the vertebral artery, gumma at the base of the brain, or caries of the petrous bone, etc.

The most important types of localized muscle spasm are:

1. *Masticatory Spasm.*—While tonic spasms of the masticatory muscles (the so-called lock-jaw or trismus) play a notable clinical rôle as a symptom of tetanus, of meningitis, and of other general diseases, as a purely local disease manifestation they are rare, occurring most frequently in connection with inflammatory lesions of the jaws. Clonic spasm is somewhat more fre-

quent, a gnashing of the teeth occurring in the form of attacks at more or less short intervals. The prognosis is in general favorable. *Oppenheim* recommends in the treatment of fresh cases in which a rheumatic influence may be in action, diaphoresis; further, some derivative procedure (cantharidal plaster over the temples or upon the mastoid process, eventually the actual cautery to the back of the neck), sedative drugs, and the galvanic current.

2. *Mimic cramp*, or facial spasm, one of the most frequent locations of peripheral spasm, is usually clonic; a tonic facial spasm is the cause of the peculiar facial expression in tetanus, which has been called "*Risus* sardonicus" (see Fig. 28). It seldom affects all the facial muscles, but is usually restricted, either to the muscles of the mouth and chin (Orbicularis oris, Levator anguli

Fig. 28.

"Risus Sardonicus" in Traumatic Tetanus.

oris, Mentalis, Zygomaticus), or to the Orbicularis palpebrarum, in which latter case, when the spasm is tonic, we speak of blepharospasm. The expressions nictitation, spasmus nictitans, and blepharoclonus denote specially the clonic forms of lid spasm. With the exception of spasms of the lids, which in many cases are relatively easy to treat and to cure permanently, the prognosis of facial spasm is in general not good, since it is an obstinate complaint and even after it is relieved recurrence is to be feared. Further, there are unfortunately many cases which cannot be cured and the patient must then choose between the facial spasm and an incurable facial paralysis; that is, he must permit a neurectomy or neurexaresis. In these cases, also, in patients hereditarily neuropathic, one is never sure that the spasm may not later attack some other nerve. As regards the therapy of facial spasms, apart from the last resort above mentioned, pathological conditions which

might set up the trouble by reflex influences should be removed (diseases of the eye, nose, accessory sinuses, and ears, caries of the teeth, retained wisdom teeth, etc.). In the way of medication, a trial of bromide of potassium in daily doses of at least 4 grms (3j), or of scopolamin, twice daily, 0.0005 (gr. 1/120) is justified. Unfortunately, these medicines only exceptionally help. *Edinger* recommends antipyrin (in combination with bromide of potassium). I have never seen any result from this in facial spasm. From electrical treatment stabile applications the anode over the trunk of the facial nerve, or the whole pes anserinus (under the general rules mentioned for the treatment of neuritis, on page 55), I have seen—with sufficient patience and persistence on the part of patient and doctor—occasionally the best results in spasm of the orbicularis oris, only exceptionally, however, any favorable influence worth mentioning in the other localizations of facial spasm. Still another electro-therapeutic procedure comes into consideration in blepharospasm, the stabile application of the anode to a recognizable "pressure point" (that is, in some place from which an attack of spasm can be cut short by firm compression, acting reflexly); this is usually one of the already mentioned *Valleix* points in the region of the ophthalmic division. When the above-mentioned methods fail one must turn to the neurolytic injections (after *Schlösser*), which have been described at length under the treatment of neuralgia. Eighty per cent. alcohol with a local anesthetic (see formulæ on page 61) is injected through a cannula inserted between the external auditory canal and the mastoid process in the neighborhood of the stylomastoid foramen indicated by the styloid process; that is, into the facial nerve at its point of exit. If the injection is successful a facial paralysis which lasts for several months is produced; if this is not to be obtained, this result is reached by exposing the nerve and stretching it vigorously (pulling it up with a "squint hook"). In favorable cases after the facial paralysis has recovered, spasm does not return; unfortunately, there are cases enough which have recurred both after alcohol injections and after nerve stretching.

To be distinguished from facial spasm is the psychogenic, isolated "tic facial" of the French writers (*Brissaud*, *Meige*, *Feindel*), which is a twitching or grimacing which has become a habit, which has the character of voluntary movement, and is to be treated and cured by exercises. The object of these exercises is gradually to carry out definite movements with the facial muscles affected with tic upon command, later to the beat of a metronome; between these, however, to keep the face still. Later the patient is allowed to practice complicated and more continued series of movements of head, neck, and face muscles until these follow quietly without being interrupted by spasms. All these exercises are to be carried out before a mirror. When there is an emotional basis for facial tic, *Dubois* has accomplished cures by pure psychotherapy.

3. *Spasm of the Tongue* (*Glossospasm*).—Isolated spasms of the tongue are excessively infrequent. They can be tonic and clonic. As reflex causes setting them off, inflammatory affections of the buccal cavity have been especially mentioned. The prognosis is usually not bad. Therapeutically stabile galvanization, the anode on the hypoglossus, is particularly recommended;

in a case of *Lange's* this nerve also was stretched and later resected, but only division of the genioglossal muscles effected a definite cure.

4. *Spasm of the Pharyngeal Muscles* (*Pharyngism*).—When this form of spasm does not arise upon the basis of a general disease of the nervous system, for example, hydrophobia or tetanus, or is not an expression of a local organic disease (retro-pharyngeal abscess, carcinoma of the œsophagus, a foreign body, *e.g.*, a fish-bone in the mucous membrane), it is usually a symptom of hysteria.

5. *Spasm of the Œsophagus* (*Œsophagism*).—To this phenomenon the remarks made regarding pharyngism equally apply. After tonic spasms of the lower parts of the œsophagus or of the cardiac orifice of the stomach, dilatations of the upper part of the œsophagus have been observed.

6. *Spasm of the Larynx* (*Laryngism*).—This form of spasm is not at all infrequent in children, particularly in rachitic infants. We will later have the opportunity of referring to its relation to the tetany of infants. It occurs in the form of tonic contraction of the muscles closing the glottis (thyroarytenoids and interarytenoids) coming on in attacks which may lead to severe dyspnœa and cyanosis, indeed, to asphyxia. Well-known popular remedies like douching the child with cold water, the production of vomiting by tickling the back of the throat, etc., sometimes suffice to cut short the attack; on the other hand, tracheotomy may be necessary. Along with the treatment of rickets, which is often present, the effect of the different internal sedatives is usually very satisfactory, so that the prognosis in general may be said to be favorable. To be recommended is a prescription of *Babinski:* ℞ Potassii bromid, 1.5-3.0 (gr. 20-45); tinct. moschi, 1.0-2.0 (gr. 15-30); syr. simp., 15.0 (½ oz.); aq. dest., q.s., ad 100.0 (3 ozs.). M.S., a dessert-spoonful every two hours. Reflex spasm of the larynx as a result of laryngitis occurs both in children and adults.

7. Spasms of the muscles of the throat, neck and shoulders are not very frequent, but are usually characterized by great persistence. They occur nearly always in neuropathic individuals; exciting causes are, in the psychical sphere, fright; depressive emotions, etc.; in the somatic, local trauma, diseases of the vertebral column or of its ligaments, etc. Not infrequently its etiology is obscure. We distinguish different forms of spasm which are quite characteristic in their manner of occurrence.

Tonic unilateral spasm of the sternocleidomastoid is called spasmodic torticolis (Caput obstipum spasticum); in this the patient turns the head and raises the chin toward the sound side, while, on the affected side, ear and shoulder are approached to one another. The sternocleidomastoid, on the side on which the head is drawn down, stands up under the skin like a firm cord (see Fig. 29). This affection is not to be confused with the harmless, temporary stiff neck due to a rheumatic affection of the sternocleidomastoid; characteristic of this latter is the decided painfulness of the muscle upon pressure and in passive movement.* Clonic unilateral spasm of the sterno-

* I once saw, however, in a patient with hysterical predisposition, a spasm of the sternocleidomastoid develop as a sequel to a rheumatic stiff neck.

cleidomastoid proceeds in the form of attacks of twitching through which the head is pulled by fits and starts into the position described above. Spasm of the sternocleidomastoid is usually combined with one of the trapezius, which is also supplied by the spinal accessory nerve; the head then is either clonically thrown backward or there is a tonic "retrocollis." Rotary spasm of the head depends upon clonic contractions in the obliquus capitis inferior; nodding

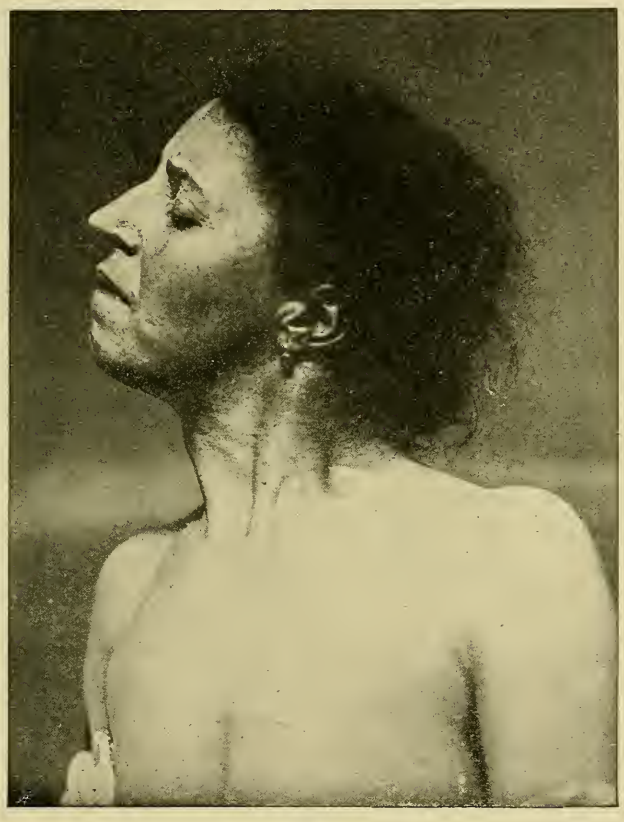

Fig. 29.

Spasmodic Torticollis.

spasm (spasmus nutans) (called also "salaam spasm" after the method of salutation of the Orientals) upon those of the deep muscles of the neck (rectus capitis, longus colli, etc.), usually with involvement also of the sternocleidomastoids. This last-mentioned form of spasm is observed especially in children, and, according to *Kassowitz*, is always due to rickets, which, however, other pediatrists deny. The following mainly tonic spasms should be mentioned: spasm of the splenius, in which the head is drawn backward toward the affected side and at the same time is somewhat rotated, while laterally from the cervical portion of the trapezius the contracted muscle stands plainly out; spasm of the rhomboid, which draws the scapula into an oblique position so that its median border runs oblique from below and within to above and

without; finally, the rare spasms of the Levator anguli scapulæ, Platysma myoides, and Omohyoid.

In connection with treatment I would recommend, after correction of whatever causal factor can be found, next to take care that the patient is isolated for several weeks in order to reduce external irritants, particularly psychical ones, to a minimum. He should be allowed to see no one except those with whom he is entirely familiar, and about whom he does not have to trouble himself, besides the doctor. Psychotherapeutically, one should untiringly endeavor to keep the patient placid and confident. In addition, bromide of potassium should be given in daily doses of 3 or 4 grammes (grs. 45-3j) (best in one dose in the morning), eventually combined with grm. 1.0 (grs. 15) of antipyrin. Also scopolamin hydrobrom., 0.0005 (gr. 1/120) twice a day is worth trying. At the same time the stabile application of galvanism with the anode and a current gradually raised to 3-5 milleamperes should be used. Points of application: 1. Pressure points. 2. The motor points of the affected muscles (see Fig. 3, page 25). 3. Those of the nerve trunk in question. In connection with this, I am accustomed to faradize the antagonists. Spring-supporting apparatus, with a head holder which opposes the pathological anomaly of position in tonic spasm of the neck muscles, sometimes aids our therapy decidedly; one of my patients, a machinist, very intelligently constructed such a thing for himself. I would decidedly warn you against rigid apparatus (plaster, etc.), in spite of its recommendation by *Strümpell*. *Oppenheim* recommends, further, the application of derivatives at the back of the neck (blistering plaster, a hair seton, and particularly the actual cautery); I am, however, no great friend of these procedures. If isolation, psychotherapy, drugs, galvanization, etc., supported by careful stretching massage and prolonged tepid baths, are without effect, neurolytic injections are to be considered, which, however, in order not to injure the important neighboring structures should only be made into the exposed nerves of the muscles affected with spasm; however, it is more rational to undertake the mechanical rather than the chemical injury of the nerve; that is, stretching it until paresis begins; neurectomy is the next most radical procedure. It, like myotomy of the muscles affected with spasm, has found more partisans among the surgeons than among the neurologists, who have often after the operation seen the spasm pass over to neighboring, up to this time sound, muscles.

Even the *Kocher-de Quervain* operation, in which successively the following muscles are cut: Trapezius, Splenius, Sternocleidomastoid, Complexus and Obliquus colli—with subsequent curative gymnastics, does not produce a cure in every case; in the case shown in Fig. 29, which did not react to other methods and in spite of being easily hypnotizable, not even to hypnosis, myotomy indeed, effected a cure.

8. *Localized Muscular Spasms in the Extremities.*—These are, on the whole, quite rare. Tonic forms of spasm, sometimes unilateral, sometimes bilateral, have been observed among others in the following muscles: Pectoralis major, Latissimus dorsi, Deltoid, Biceps, Supinator Longus, the flexors of the fingers, the adductors, Gastrocnemius, and Tibialis anticus, clonic forms in the Ileopsoas and the Peronei.

Not to be confused with these true, local spasmodic conditions of the muscles of the extremities, are the painful cramps which, as is well known, occur after fatiguing marches, long swimming, mountain climbing, etc.; also in healthy people, in the calf muscles (less frequently in the Abductor hallucis, Quadriceps, Tibialis anticus) and for which certain individuals have an exaggerated predisposition so that relatively slight exertion suffices to set up cramp. In contradistinction to true clonic spasms, cramp is relieved by massage and passive stretching.

9. Spasm of the diaphragm in tonic form is very rare and is almost never a disease in itself; it is most frequently observed in tetanus. All the more familiar is clonic spasm (singultus) which occurs in many healthy people, now reflexly (as from drinking strong spirits), again without apparent ground as an occasional temporary and entirely harmless phenomenon. Obstinate pathological forms of singultus occur in diseases of the cervical region of the cord (in the neighborhood of the diaphragm center) further in peripheral irritation of the phrenic nerve (aortic aneurism, mediastinitis, pericarditis, pleurisy, etc.), occasionally, also, as an apparently idiopathic and localized spasm. Familiar popular means of cutting short singultus (holding the breath, sipping cold water, etc.) fail in severe forms. Energetic faradization of the region of the stomach with the wire brush often succeeds here. Other derivatives to be applied in this last location are sinapisms, blisters, ignipuncture, chloride of ethyl spray. Of internal sedatives, besides bromide of potassium, opium is recommended. *Laborde* has introduced rhythmical tractions of the tongue, which sometimes succeed. Finally, the galvanization (anodal) of the phrenic nerve comes into consideration.

Complicated respiratory spasms which, indeed, only occur in the hysterical, are yawning spasm, sneezing spasm, screaming spasm, snoring spasm. These are of psychic origin, "tics," like voluntary movements, not true spasms, and their treatment, *mutatis mutandis*, has to be carried out from the points of view considered under facial tic. We must still mention that there is a diseased condition characterized by the most manifold motor automatisms which has been described as "Myospasia convulsiva," "Maladie des tics," or "general tic." This occurs chiefly in individuals of neuropathic constitution beginning in childhood, and is often accompanied with psychic disturbances; for example, the compulsory ejaculation of certain, sometimes senseless, often obscene or blasphemous, words (Koprolalia). Often there is also the compulsion to repeat words heard or gestures seen (Echolalia, Echopraxia). There are very severe incurable cases of this disease. In one mild case which I saw, cure resulted from a "mast cure," combined with the most rigid isolation in a dark room, to which, later, a course of re-education of movements was added; in other cases considerable improvement; these were in children. The older the individual the worse, in general, the prognosis. Drugs and physical therapeutics are useless, so is hypnosis. Similar to myospasia convulsiva, but of favorable prognosis, usually recovering spontaneously after weeks or months, is the "saltatory reflex spasm" described by *Bamberger*, in which the patients on standing break into jumping movements, while on sitting and lying they hold themselves quite normally.

2. "OCCUPATION SPASMS"

From the disease pictures summed up under the name of "Local Muscular Spasms" we separate a pathogenetically unique nosological group which is distinguished by its great practical importance. Its members have been called "occupational spasms," also (after *Benedikt*) "co-ordinatory occupation neuroses," and had already been recognized and their clinical peculiarities noted by *Bell* and *Duchenne*, namely, as spasms which appear only in connection with a definite activity acquired by practice. They usually appear not only as spasms (which practically always are of tonic character), but not at all infrequently as tremor, and occasionally as a refusal to act on the part of the affected muscles, which last it has been attempted to separate off as the "paralytic" variety of the occupation neuroses. As a diagnostic criterion the limitation of these different dyskinetic disturbances, not only to definite motor acts, but also to the muscle groups acting together in this function, is of decisive importance. For the occurrence of this affection two factors are necessary: 1. A neuropathic disposition (usually hereditary). 2. The frequent repetition of the act in question (usually through the carrying out of one's calling). That along with these, alcoholism, nephritis, chronic lead poisoning, and a number of other general injurious influences contribute to the production of occupation spasms, is made probable by a number of striking observations. Of local predisposing factors, diseases of the tendon sheaths (as ganglion), farther, exostoses on the bones, muscular and articular rheumatism, have been mentioned. As exciting causes, somatic and psychic traumata have been especially frequently determined.

The best-known example of occupation spasm is writer's cramp ("crampe des écrivains," graphospasm). It occurs usually either on beginning writing, or only after some lines or pages have been written, as a tonic contraction of the flexors of the fingers; much less frequently there is spasm of the extensors or abductors; occasionally the hand begins to tremble or the fingers lose power and let fall the pen; cramplike pains can occur also.* Other occupation spasms of the upper limbs are: Milker's cramp, described in 1851 by *Basedow;* farther, piano player's, telegrapher's, shoemaker's, drummer's, tailor's, cigarmaker's, violinist's, 'cellist's, flutist's, seamstresses', sawyer's, newspaper folder's, smith's, zither player's, and watchmaker's cramps. In the legs there occur ballet-dancer's cramp; in the muscles of the mouth and tongue, trumpeter's and clarinetist's cramps. *Oppenheim* has described a "shaving cramp" ("keirospasm" or "xyrospasm"). The most "modern" spasms are "daktylographer's" and "automobilist's" cramps.

These occupation neuroses are not to be confused with the professional pareses—which are mainly of neuritic nature—already mentioned. I have, however, seen develop in a cigarmaker, a year after recovery from a typical cigar-roller's cramp, a not less typical cigar-roller's paresis, with atrophy of the small muscles of the hand, which then, after rest and later change of occupation, entirely disappeared. This last, on account of the great obstinacy

* For the vasomotor form of "writer's cramp" see Lecture XXV.

of many occupation spasms, is not very infrequently the end result of vain therapeutic endeavors. A considerable number of the cases, however, are fortunately not refractory to the means of treatment at our disposal.

First, the specific harmful occupation must be entirely given up for a period of weeks, during which time complete therapeutic repose is to be sought for in the first place, since anxiety about the spasm and fear as to the eventuality of having to give up one's calling, usually greatly depress such patients and serve to keep up the neurasthenia which furnishes the basis of the occupational neuroses. Prolonged tepid baths, Swedish gymnastics, stabile anodal galvanization of the nerves and muscles in question (with avoidance of all sudden variations of current strength), a sojourn in the mountains or at the seaside, a course of arsenic or iron, are to be prescribed, according to circumstances. Later, the patient is allowed to carefully take up again the "critical" occupation, in which case this is to be undertaken in the most rational and easy manner possible; writing, for instance, with a soft, not too sharp, steel pen in a thick cork penholder, or, even better, special "writers' cramp" penholders, as, for example, that of *Zabludowski*, or "*Nussbaum's* bracelet," which is held by the spread fingers; also by finding out the easiest position for the hand. I let the patient at first go over for a few minutes at a time a very coarse copy placed on tracing paper, later writing it smaller. Little by little he is allowed to practice longer, and finally to write without a copy.

For patients with pianist's cramp, *Zabludowski* recommends practice on a juvenile piano, which has a considerably smaller scale than the regular instrument. Telegraphists should, when possible, exchange the *Morse* apparatus for that of *Hughes*.

3. THE MYOKLONIAS

Under this name we include some rare, pathogenetically still obscure, disease conditions whose common chief symptom consists in clonic contractions of the individual muscles which usually produce but little movement, or none at all. Three chief forms may be distinguished.

1. Paramyoklonus multiplex (*Friedreich*) occurs usually about the fiftieth year of life, sometimes set up by a psychic or somatic trauma, infectious diseases, excesses, and sometimes without any recognizable cause. It shows itself in attacks lasting a minute or so (of irregular rhythm and a frequency of 10 to 15 per minute) of lightning-like contractions of different, usually symmetrical muscles of the trunk and extremities; for example, the latissimus dorsi, gastrocnemius, quadriceps, pectoralis, rectus abdominis, biceps brachii, triceps, etc. The contractions are most marked during rest (in sleep, indeed, they usually stop, or at least grow less); on movement they are less. In one case I described a coincidence with congenital defect of the pectoralis, which is, of course, to be considered as a stigma of congenital deficiency of the muscles.

2. *Myoklonus Epilepsy* (*Unverricht*).—Symptomatologically particularly characterized by the myoklonic contractions affecting the tongue, the pharyngeal, and diaphragmatic muscles, this affection appears almost always in

several children of a family, not rarely in several successive generations. Noteworthy also is its association with occasional epileptiform attacks occurring particularly at night, which sometimes show themselves as precursors of the myoklonic manifestations (the last usually begin about the tenth year of life). The myoklonic phenomena increase on movement and in psychical excitement. The tendon reflexes, as well as the mechanical irritability of the muscles and nerves, are increased. Life is seldom directly threatened (for example, by aspiration pneumonia as a sequel to myoklonic spasms of the pharynx). The patients can live to be as old as seventy years, in which case there is usually gradual mental failure and termination in "Dementia myoclonica" and marasmus.

3. *Nystagmus-myoclonia* (*Lenoble-Aubineau*).—A rare affection, almost exclusively observed in the Celtic families of Brittany and Great Britain, in whom as a congenital and stationary condition contractions of the external eye muscles, as well as of the extremities, which are increased by cold and by tapping the muscles, but can to a certain extent be voluntarily controlled, have been observed. The reflexes are usually much exaggerated, different trophic and vasomotor disturbances (for example, deformities of the teeth, asymmetry of the face and the body, local sweating, circumscribed œdema, lividity of the skin) are also found.

Therapeutically, stabile galvanization of the muscles with the anode, warm baths, bromide of potassium, chloral hydrate, can be used in the myoclonias as fulfilling to some extent the symptomatic indications.

4. TETANY

As characteristic of this disease, given its name by *Corvisart*, we can consider tonic spasms, which specially affect the peripheral portions of the limbs, occur in attacks, and are usually combined with more or less decided pains.

It is usual to separate a "primary" tetany from the different "secondary" or symptomatic tetanies. The first, whose etiology is still obscure, attacks, as a rule, previously healthy and strong males between fifteen and twenty-five years old, and by preference those carrying on some definite calling, particularly shoemakers and tailors, more rarely joiners and locksmiths. Cold and damp weather favors the onset of the disease; the majority of cases occur in the months from January to April. The geographical distribution of the disease is striking; it is most frequent in the neighborhood of Vienna and Budapest, while it is quite infrequent in France, and has decreased in frequency from decade to decade; in Basle and its neighborhood tetany of workers is practically unknown. I sought in vain for years for a case and have had no word of any definite observation by any one else, a fact all the more remarkable since, in the not very distant Heidelberg, primary tetany is comparatively frequent. The "secondary" tetanies occur in the course of different affections, and hence are grouped as follows: 1. Gastrointestinal tetany, which is occasionally observed in profuse diarrhea or, on the other hand, in obstinate constipation; also in dilatation of the stomach and stenosis of the pylorus, in carcinoma of the stomach, appendicitis, intestinal helminthiasis and chole-

cystitis. 2. Intoxication tetany, which can occur, for example, in ergotine, lead, opium, atropin, and alcohol poisoning. 3. Tetany in acute infectious diseases; this is occasionally observed, sometimes in the beginning, sometimes in the course of typhoid fever, dysentery, measles, cholera, scarlatina, influenza, diphtheria, malaria, acute rheumatism, etc. 4. Maternal tetany, which develops in pregnant, puerperal, and nursing women, and was first described by *Trousseau* under the name of "contracture rhumatismale des nourrices." 5. Parathyroid tetany, the result of destruction of the parathyroid glands. It is not long since tetany occurring after extensive or total thyreoidectomy was regarded as due to the loss of this gland. To-day, however, clinical and experimental proofs have been furnished by *Vassale, Generali, Moussu, Pineles, Erdheim, de Quervain, Hagenbach, Iselin*, and others, that usually the loss of the accessory glands to the thyroid provokes the onset of tetanic phenomena. 6. The tetany of children, which, as a rule, occurs in rachitic children and in those suffering from gastrointestinal disturbances. The theory that all varieties of tetany depend upon an insufficiency of the parathyroid glands is not proved, but has much to recommend it; their function is plainly an antitoxic one; it can be conceived that tetany occurs not only when the parathyroids are destroyed, but also if their ability to neutralize exogenic or endogenic poisons is not sufficient. In the tetany of workers and maternal tetany, hypothetical toxines entering the organism or produced in it have long been spoken of. According to *Yanase*, however, a number of cases of the tetany of children depend upon hemorrhages into the parathyroids (probably at birth).

The symptomatology of tetany is characterized by a number of exceedingly typical phenomena. Sometimes (namely in primary tetany) there is complaint of certain prodromal symptoms, namely, paresthesias in the hands and feet, general malaise, muscular unrest; from these prodromes patients who have already had tetany are able to recognize the oncoming new attack. These last are entirely dominated in their clinical aspect by the attacks of spasm which are usually preceded by a feeling of tingling in the affected limbs, the so-called "sensory aura." The upper extremities are most frequently and severely affected, particularly the hands, whose characteristic attitude *Trousseau* has described in a classical manner. The thumb is brought into a forced position of adduction, the closely approximated remaining fingers are bent against it predominantly at the metacarpophalangeal joints; also the palm of the hand forms a hollow, on account of the approximation of its radial and ulnar borders, and the whole hand takes the wedge shape assumed by the obstetrician for vaginal examination ("accoucheur's hand"). The wrist is tonically flexed, and in extensive spasms, the elbow also, while by firm contraction of the pectoralis, the upper arm is pressed against the thorax; only exceptionally does the arm take a position of extension. Extension at the hip and knee joints, on the contrary, is typical in tetanic spasm of the legs; the feet, however, imitate the "obstetric hand" by approximation of the maximally flexed toes, under which the overadducted big toe is forced, hollowing the sole. In severe tetany, tonic spasms may affect the face muscles; *Escherich* has called attention to a trunklike protrusion of the lips occurring

in this case. In involvement of the masticatory muscles trismus occurs; in that of the external eye muscles, squinting; sometimes the long muscles of the back and the sphincters are also affected. Particularly important are spasms of the glottis which for a long time were regarded as the privilege of infantile tetany, which, however, *Pineles* has shown are not so infrequent in that of adults. The spasms of tetany are usually symmetrical, but one side can be

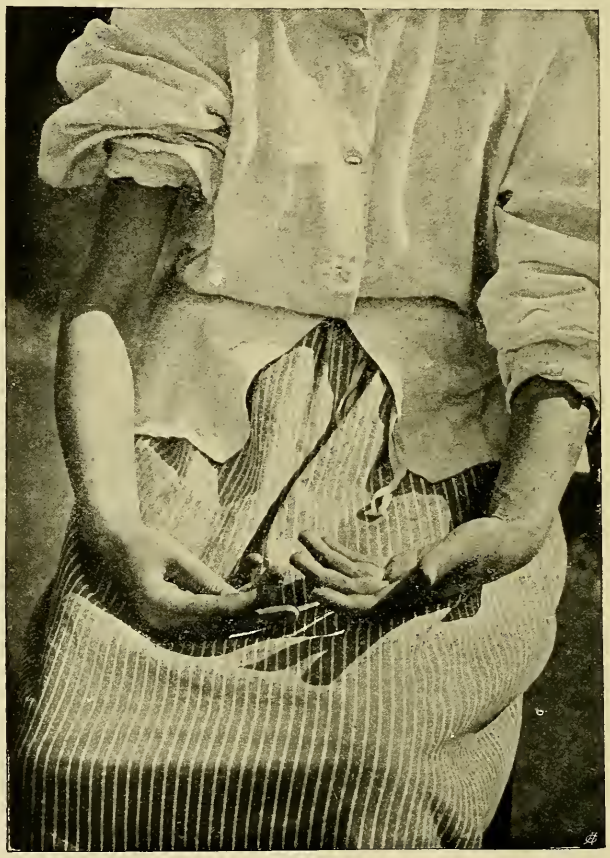

Fig. 30.

Parathyroid Tetany.—Position of the hands in the attack (instantaneous photograph).

affected before the other, or to a greater extent. As to the length of the attacks, they vary from a few minutes to several days; in cases of the last sort, ulcers have been known to be produced by the unintermitting pressing together of the fingers. As already said, more or less manifest painful sensations usually accompany the tetanic spasms.

Besides the tonic spasms (which when they are set up by voluntary movement are called "intention spasms"), tetany presents a number of exceedingly interesting motor phenomena, which occasionally, in rudimentary or "latent" tetany, can occur without an attack of spasm; on the other hand, they need not all be present in typical tetany. These are *Trousseau's, Chvostek's, Erb's,* and the *Pool-Schlesinger* phenomena.

1. *Trousseau's* sign: If with the fingers energetic pressure is exerted upon the nerves of an extremity (especially upon the nerves of the upper arm in the internal bicipital sulcus), or if a limb is firmly bound with a rubber tube, after a few seconds or minutes a typical attack of spasm occurs in it.

2. *Chvostek's* sign: Tapping the trunk of the facial nerve in front of the ear, or even stroking this region, sets up contractions in the muscles of the face.

3. *Erb's* sign: There is galvanic overexcitability of the muscles and nerves. The cathodal closing contraction appears with a very weak current (in the case of the girl shown in Fig. 30 with one-eighth milleampere on the ulnar). It is further characteristic that the anodal contractions (AnOC and AnClC) can be obtained with a comparatively weak current. Finally, according to *Mann* and *Thiemich* in infantile tetany, and according to *Pineles*, sometimes in that of adults also, it is typical that contraction on cathodal opening occurs below milleamperes and that AnOC is greater than AnClC. Faradic overirritability is rarer.

4. The *Pool-Schlesinger* sign ("leg phenomenon"): If in the time between attacks the leg extended at the knee is flexed at the hip (or if the patient lying on his back is ordered to sit up, his knees being kept pressed together), after a few minutes, or often after a few seconds, there occurs a spasm of extension at the knee and a tonic spasm at the ankle, with severe pain. The spasm, as a rule, is limited to the extremity under examination, and usually ceases when flexion at the hip is relaxed.

Galvanic hyperexcitability of the sensory nerves, occasionally observed in tetany, is known as *Hoffman's* phenomenon; the supraorbital nerve, for instance, reacts to 0.2 instead of to 1.0 milleampere. According to *Chvostek, Jr.*, and *v. Frankl-Hochwart*, galvanic overexcitability of the nerves of hearing and taste can also occur.

It remains for us to enumerate a number of rare and inconstant symptoms of tetany. First, along with the tonic spasms there occur more or less severe attacks of clonic contractions which sometimes are accompanied by alteration of consciousness, are "epileptiform"; this is most frequent in the tetany of infants, since in general infancy is predisposed to eclampsia.

Moderate rise of temperature, or, on the contrary, hypothermia is sometimes observed. Other trophic and vasomotor accompanying phenomena are a certain puffiness of the face which in primary tetany has even been described as the "tetany countenance"; further, there is sometimes a redness or cyanosis of the face or of the extremities, œdema of the hands and feet, also of the joints, falling out of the nails and hair, and finally cataract. Trophic symptoms are in general characteristic of severe chronic relapsing tetany. Of psychic phenomena, hallucinatory conditions in sequence to the attacks of spasm are to be mentioned. The tendon reflexes are, as a rule, exaggerated.

The course and prognosis of tetany vary within comparatively wide limits. Maternal tetany always runs an acute and favorable course; also cases occurring in infectious diseases and intoxications always recover, the cause ceasing to act.

In parathyroid tetany, all depends upon the amount of uninjured parathyroid substance left; total ablation of these bodies causes chronic tetany.

Gastrointestinal tetany is a severe disease, with a tendency to relapse and to pass over into chronicity. The prognosis depends above everything else upon whether the stomach or intestinal affection can be removed. In infantile tetany, attacks of eclampsia, or spasm of the glottis, may cause death. As to primary tetany, *Pineles* writes that the prognosis in a certain portion of the cases is quite favorable, since the disease is recovered from in a longer or shorter time, that however, in a not inconsiderable remainder of them, it takes a chronic course through many years, relapses occurring chiefly in the "tetany season." In this form, trophic disturbances are comparatively frequent, nevertheless its prognosis as to life is favorable, since no case of death from idiopathic, uncomplicated tetany has so far been observed. From a diagnostic point of view, with regard to the "spasmophilia" of infancy as well as to the tetanoid conditions of adults, reference must be made to the fact that galvanic overirritability is the single constant and obligatory symptom of tetany, around which then the remaining symptoms can group themselves in different combinations. In a child in a condition of latent tetany, an intercurrent digestive disturbance or taking cold can at any time lead to an outbreak of spasm in the extremities or to spasm of the glottis.

Arthrogryposis, an affection of the first years of life, which is characterized by tonic spasms and positions of contracture of one or several limbs, lasting from days to weeks, seems to have relationships with infantile tetany; the legs are fixed now in extension, again in flexion, the arms spasmodically flexed. Recovery usually occurs; *Strümpell*, however, has seen two cases end fatally (with negative post-mortem findings). Important for its distinction from true tetany, is the absence of mechanical and galvanic irritability of the nerves.

The therapy of tetany must, of course, be a causal one in all secondary forms; so, for example, in lactation tetany, nursing must be forbidden; in gastric tetany, according to conditions, lavage is to be practiced or—and not too late—pyloroplasty or gastroenterostomy is to be recommended, etc. In the tetany of workers, in order to prevent recurrence, too great crowding together of these people is to be prevented, and they are to be protected from the influence of cold; further, alcohol is to be forbidden them; eventually change of residence and calling should be insisted upon. Parathyroid tetany ought to be dying out on account of the increasing care on the part of surgeons to avoid injuring the parathyroids in goiter operations.

For all varieties of tetany rest in bed and tepid baths of long duration are to be recommended. The diet should be purely ovolacto-vegetarian (withdrawal of meat, in animals experimentally made subject to tetany, reduces the frequency and intensity af the spasms). Of drugs, the alkaline bromides (in daily doses of from 3 to 6 grms—gr. xlv-5jss.) come specially under consideration in adults; as does scopolamin, 0.0005 (gr. 1/120), twice a day; this last acting specially on the pains, for which also chloral hydrate, salicylic preparations, antipyrin, pyramidon, lactophenin, etc., may be tried; occasionally morphine is necessary. *Loeb* and *McCallum* recommend calcium lactate, which reduces the overirritability of the muscles in animals without parathyroids. A teaspoonful of a ten-per-cent. solution is given three or four

times a day. The great hopes which had been placed upon organotherapy with parathyroid preparations have not been realized. The transplantation of parathyroid tissues also has given no result worth speaking of, since these do not remain capable of functioning in their new location, but speedily perish, although the transplanted structures, so long as they are not reabsorbed, act favorably. In the treatment of infantile tetany, on account of its frequent relation to rickets, along with bromide of potassium (see formula, page 68) and chloral (0.15 to 0.5—grs. 2 to 7—by clysma), phosphorus, best given in the form of *Kassowitz's* emulsion, plays, with right, a considerable rôle.

℞ Phosphori ..... 0.1 (gr. 1½)
Ol. amygdal. dulc. ..... 10.0 (℥ijss.)
Sacch. alb.,
Gummi mimosæ .....āā 5.0 (gr. lxxv)
Aq. dest., q. s.; ad emuls.....100.0 (℥iij)
M. S. A teaspoonful once or twice a day.

LECTURE V

The Dyskinesias

D. The Choreiform Diseases

Involuntary, quick movements, co-ordinated, indeed, but nevertheless of aimless and contrary character, which are not rhythmic but rather give the impression of a jerking, continuous unrest of the affected extremities, we designate chorea; the name comes from the Greek (χορός = dance), and is in so far suitable, since when the lower extremities are affected, the gait of the patient can degenerate into dancing and hopping. The most frequent, and practically by far the most important, form of chorea is the disease known as Chorea minor, Chorea sancti viti ("Veitstanz," "Danse de Saint Guy"), or *Sydenham's* chorea; much more rare is *Huntington*'s degenerative or hereditary chorea, which is also an autonomous disease. Usually of symptomatic indication, on the other hand, are hysterical chorea (also "chorea major"), which occurs auto-suggestively by imitation of chorea minor, and not infrequently leads to school or class epidemics, chorea in lesions of the cerebellar peduncles (see Lecture XX), as well as pre- and post-hemiplegic chorea, which sometimes is observed as the precursor or result of an apoplectic attack from cerebral hemorrhage.

This last variety of chorea, affecting only the extremities of one side, is also a hemichorea. Its presence justifies the conclusion that there is a hemorrhage into the optic thalamus, the lenticular nucleus, or into the most posterior part of the internal capsule of the opposite side of the brain. The affected extremities show lively jerking or shaking movements ("hemiballism"), which are absent during sleep, but cannot be controlled voluntarily, even may be exaggerated upon attempting this. The designation of certain clinical pictures described as great rarities by the name "Electric Chorea" is incorrect. In these cases the involuntary movements follow one another with lightning-like rapidity, in the form of violent jerking. *Henoch's* form of electric chorea is probably an atypical myoclonia, that of *Bergeron*, a hysterical phenomenon. On the other hand, *Dubini's* electric chorea, a disease observed in Lombardy, in which besides muscular twitchings there is fever, pain in the neck and back, epileptiform attacks, loss of faradic muscular irritability, pareses, etc., and which usually terminates fatally, is probably an infectious disease of obscure etiology.

1. CHOREA MINOR, SYDENHAM'S CHOREA

This disease, in the great majority of cases, attacks children, chiefly girls, particularly between the sixth year of life and puberty. Younger children are only exceptionally attacked, also the disease is very rare between fifteen

and twenty-five years (these "juvenile forms" nearly always occur in females). Cases occurring at a later age are great rarities, though even "senile forms" have been known.

In the etiology neuropathic predisposition probably plays a considerable rôle (heredity is here and there recognized). In the majority of cases, however, the action of some infectious influence is decisive. In the first place must be mentioned acute rheumatism and endocarditis, which often precede the attack of chorea minor, accompany or follow it. Less frequently a connection with the acute exanthemata, typhoid fever, pneumonia or erysipelas has been determined. Exposure to cold has often been accused; chorea minor, indeed, is more frequent in cold, moist localities and in the winter months. In the juvenile forms pregnancy always plays an important rôle. It is seen in young primiparæ, who are attacked in the early months of pregnancy. The disease but rarely begins suddenly. As a rule, the family notices first a general unrest with psychical disturbance (anxiety, ill humor, unsociableness), as well as the appearance of illness and poor appetite; in this stage also many patients complain of weakness and vague painful sensations in the limbs. Next, movements become clumsy and finally plainly jerky; there is also involuntary grimacing. Once the disease picture has reached full development, the choreic character of the movements is unmistakable; we notice a continual gesticulating, there is no longer any quiet sitting or standing, short, motiveless movements occur, now in the hands and fingers, again in the feet, now an arm, now the head, and again a leg is jerked hither or thither; a wrinkling of the forehead, a smacking with the tongue, winking, protruding the lips, shows involvement of the muscles innervated by the cranial nerves. The French speak very appropriately of a "Folie musculaire" ("muscular madness"). In walking, the patient sometimes gives the impression of a "jumping jack," which moves when the string is pulled. In severe cases, however, walking is impossible, and the patient, condemned to keep her bed, throws herself about in alarming fashion. The muscles of the diaphragm, the œsophagus and the larynx may be involved (irregular breathing, disturbances in swallowing, broken speech) even in the pupils, alternating contraction of sphincter and dilator, a so-called "hippus," independent of the influence of light, accommodation or convergence, has been observed.

Chorea almost always begins in the upper limbs and in the face, affecting the legs later. Not infrequently the limbs of one side are attacked first; as a rule, however, this original "hemichorea" sooner or later passes over to the other side also.

Psychical excitement, even the consciousness of being observed, increases the choreic phenomena, which, however, cease during sleep—only in the very severe, fatal cases the intensity of the movements does not permit sleep, and the patient perishes from exhaustion ("Status choreicus").

In chorea minor the psyche scarcely ever remains entirely intact; however, its anomalies are usually inconsiderable; irritability, a high degree of distractibility, an ill-humored deportment, lability of mood. In the above-mentioned malignant cases, fortunately rare, on the contrary there may be hallucinatory delirium, and even maniacal attacks.

The rare symptoms of chorea are: slight rise of temperature (only in status choreicus and in complications with polyarthritis, endocarditis acuta, etc., is there high fever), motor weakness ("Chorea mollis," incorrectly "Chorea paralytica"), pain along the spinal column. On account of the anemia often developing with chorea minor, sometimes anemic heart murmurs occur, which should not be confused with those of complicating endocarditis. On tapping the patellar tendon, it is sometimes noticed that the foot, which is drawn up by the contraction of the quadriceps, does not sink down again immediately, but remains raised a few seconds before falling ("tonic reflex," *Gordon's* symptom).

In general the prognosis is favorable, and recovery occurs with gradual lessening of the symptoms from one to three months after the beginning of the disease. However, there is often a tendency to relapses (these occur in about 25 per cent. of the cases). Less favorable in prognosis are the choreas occurring after the acute exanthemata and typhoid and those in adults in which the danger of endocarditis is very great. Most dangerous, however, is undoubtedly chorea gravidarum with a mortality of 30 per cent.

As to the pathological anatomy of *Sydenham's* chorea, the lesions found in the corpus striatum and in the optic thalamus by some investigators are of interest. Occasionally multiple emboli have been found in these parts, to which our attention had been already directed on account of symptomatic hemichorea, in cerebral hemorrhages (*Broadhurst*); more frequently the so-called "chorea bodies" (*Elischer, Jakowenko*), concentric stratified, highly refractile bodies are seen in the vascular sheaths. However, all these findings are still the subject of controversy.

The therapy of chorea minor, when carefully carried out, usually gives very satisfactory results, and hence will be considered at length. On account of the great preponderance of the disease in late childhood and about puberty, I will give the doses of medicine for children of from ten to twelve years; from these, those suitable for younger and older patients can easily be calculated.

Every case, even the mildest chorea minor, is during the whole duration of the disease to be prevented from going to school, to public nurseries, etc., also at home or in the hospital as strict isolation as possible should be enforced. The patient should sleep in a single room, should not eat or play with the other children, and in the most severe cases, apart from the physician, should see only one and the same nurse. As a rule, the children can be allowed to amuse themselves with picture books, quiet games, dolls, etc.; also by unexciting reading (best read for them). Everything, however, in small "portions" only, with the interposition of long periods of complete rest. A full measure of rest in bed is to be prescribed, even in very mild cases; for example, from 8 P.M. until 10 A.M., and from 2 till 4 P.M., namely, sixteen hours in bed. In severe cases we order strict rest in bed until there is decided improvement; in very severe ones, besides this, the sick-room is kept dark, and by suitable padding care is taken that the patient does not bruise or injure herself. The diet should be purely ovolacto-vegetarian; alcohol, coffee, tea, spices, and very salt food should be forbidden. To prevent constipation we give corn bread

and plenty of fruit; further, taking as much milk as possible is of use; for example, a half cup every hour, eventually yoghurt, kefir, curds, etc. Where pure milk proves unpleasantly laxative, the addition of about a teaspoonful of the purest powdered gum arabic to the quart is to be recommended (mix first with some cold milk, add the rest of the milk and boil the whole). Daily tepid baths of about twenty minutes' duration, followed by rubbing down with water at from 22° to 25° C., act favorably also. On the other hand, you are expressly warned against cold douches and similar applications. From among the large number of drugs which have been introduced in the therapy of *Sydenham's* chorea, only three appear to me to have a specific effect: arsenic, antipyrin and cannabis indica. Arsenic is incontestably the most efficient of the pharmacological agents, and should hence be preferred. In using it, it is desirable to begin with small doses after well-known principles, and gradually to increase them to large and very large doses. Chorea patients are especially tolerant of arsenic, and it almost never has to be withdrawn on account of digestive disturbances, herpes, or conjunctivitis. To antipyrin also great activity is attributed, especially by French pediatrists. Nevertheless, when it is the only drug given it is necessary to exhibit it in such large doses (3.0-6.0 grm. a day in children from 6 to 10 years old, 5.0-6.0 grms. in those from 10 to 15 years), that we must reject this method as too dangerous. The matter is otherwise in its combination with arsenic preparations in which small and safe doses of antipyrin may prove a good adjuvant to the chief drug. A combination of arsenic acid and extract of cannabis indica (provided a good and fresh extract is used) gives even better results. The "Pilulæ cannabinæ compositæ," which I have recommended, have proved very useful.

℞ Extract. cannabis indicæ ..... 0.3 (gr. 4½)
Acidi arsenios ..... 0.04-0.12 (0.15) (gr. ⅗ to 1⅘)
Quinin sulphat ..... 1.0 (gr. xv)
Extract. valerian ..... q. s.

Ut. f. pil. No. XXX.

M. S.—One pill three times a day after meals (daily dose of extract cannabis ind., 0.03, and acid. arsenic, 0.004-0.012, or eventually 0.015).

Other prescriptions are:

℞ Sol. Fowleri ..... gtts. xxx-l
Aq. menth. pip. 80.0
Sr. simpl. 20.0

M. S.—A dessertspoonful in water three times a day after meals.

℞ Sol. Fowleri 10.0
Spts. melissæ co. 30.0

M. S.—Ten drops in milk, from one to three times a day.

℞ Sodii arseniatis ..... 0.01-0.03 (gr. 1/6-1/2)
Antipyrin ..... 5.0 (gr. lxxv)
Syr. aurantii cort. ..... 50.0 (℥jss.)
Aq. dest. .....q. s. ad 150.0 (℥v)

M. S.

A teaspoonful after meals. Daily dose, gr. 1/60 to gr. 1/20 sodium arseniate, and 7½ grs. antipyrin.

Besides these drugs, it may be necessary to prescribe hypnotics for a longer or shorter time, as it is of great importance that the patients sleep long and deeply; it should naturally be endeavored to get along with the most harmless hypnotics possible and in order to prevent habituation, to change them often, using, for example, adalin, bromural, veronal and trional. Occasionally an evening dose of 2.0 grms. of bromide of potassium will be sufficient; salicylic preparations are only indicated in choreas with rheumatic and cardiac complications. I recommend to you further (but only after the irritative symptoms of chorea are plainly on the decline) to let the patient practice slow, rhythmical movements of the extremities upon command, best while lying in bed. Even in the early stages as well as at the height of the disease, however, the undertaking of rational breathing exercises is to be recommended, since many chorea patients breathe badly and superficially. To this end I place a pillow under the sacral region of the patient lying on his back, and have him practice, for five to seven minutes at a time, deep, rhythmical inspiration and expiration, this once or twice a day. Whenever it is possible after the cure, the patient should spend from four to six weeks at some quiet resort. In anemic or delicate children a combination of this with salt baths or a course of iron is advantageous.

Chorea gravidarum can furnish a cause for artificial abortion; *Sarvay* gives the following indications for this: Prevention of the taking of nourishment by the violence of the irritative phenomena, loss of strength, decided alteration of the psychical functions.

2. HEREDITARY CHOREA, HUNTINGTON'S CHOREA

In 1882 the American physician, *Huntington*, on Long Island, called attention to a rare affection, which characterized in the main by progressive choreiform muscle unrest and dementia, attacks chiefly individuals from 35 to 40 years old, and is usually marked by decided heredity. Sporadic cases, indeed, sometimes occur, also it may begin between 25 and 30 or between 40 and 55 years of age, while only very exceptionally do the initial symptoms commence about puberty or even earlier. The disease has the tendency to begin at an earlier average age in each succeeding generation ("anticipatory heredity"). Still less frequently than in America, the disease occurs in the Old World, particularly in England and Germany. As exciting causes, psychic traumata, exposure to cold, the puerperium, excesses, have been brought forward. Syphilis and the acute infectious diseases, on the other hand, appear to play no rôle at all. Like chorea minor, the affection usually begins in the arms and in the

face, later to attack the legs; also it is sometimes at the start a hemichorea. In contradistinction to *Sydenham's* chorea, however, patients with *Huntington's* disease when in a state of complete quiet and abstraction can suppress the shaking in carrying out intentional movements so that they can pursue their occupations. In sleep there is usually complete quiet of the muscles; psychically a depression is evident which cannot be said to be motiveless, since the patients are only too well aware of the incurable nature of the family scourge which is coming upon them and of the mental failure which is approaching; in this stage suicide or attempts at this are common. Later the patient becomes apathetic with episodal conditions of excitement, the memory fails, sometimes there are delusions of grandeur or of persecution, also periods of hallucinatory confusion, mental failure continually advances, and there can be finally complete dementia. There are, however, cases with relatively little disturbance of intelligence, and there is even a variety of the disease beginning at puberty, which usually becomes stationary after some time, and does not alter the mental personality. Apart from this last form, the prognosis is very unfavorable; the disease can, indeed, last for twenty or more years, so that the patients not infrequently live to be sixty or seventy years old; death ensues in dementia and marasmus, or from intercurrent diseases.

In its pathologico-anatomical relations we have a number of heterogeneous findings whose interpretation is not yet possible. The newest investigations are those of *Alzheimer*. He finds very grave changes in the cerebral cortex, as well as in the corpus striatum and in the nucleus ruber, namely, a surrounding and in part a destruction of the ganglion cells by the "amœboid" elements of the glia.

The therapy is purely symptomatic, and, as such, gives little prospect of results. Only in very early stages arsenic and scopolamin (½-2 mg. per dose once a day) appear to be able to procure alleviation. It may be attempted to modify somewhat the choreiform unrest by sedative hydriatic procedures, curative exercises, etc. It is best to bring these patients as soon as possible into the seclusion of an institution where, apart from the elimination of all irritants, intelligent care of the episodal states of excitement (baths, opium, veronal, rest in bed, etc.) can best be carried out. In the latest stages the care of the disturbed and demented patient is particularly difficult.

E. The Athetoses

In the year 1871 attention was called by the American neurologist *Hammond* to a peculiar phenomenon of motor irritation which he designated by the name Athetosis (ἄθετος = restless). The hands and feet, that is, the individual fingers and toes of the patients, make constant, slow excursions, beginning with hyperextension (analogous to the movements of the tentacles of an octopus), which do not cease during sleep, but up to a certain point can be controlled by an effort of the will. Fig. 31 shows one phase of athetoid movements of the hand after an instantaneous photograph. It refers to a child in whom the athetosis developed as a result of an acute encephalitis. In fact, focal lesions which are situated in the parts of the thalamus, some-

times also in those of the lenticular nucleus, bordering on the internal capsule (more rarely also in the internal capsule itself, that is, in the posterior third of its posterior limb), can produce not only hemichorea, as already mentioned, but also hemiathetosis, on the opposite side. The last, indeed, is much more frequent than cerebral hemichorea. Children are particularly predisposed. There are indeed transition forms between hemichorea and hemiathetosis; also in lesions of the basal ganglia on both sides there is a bilateral athetosis.

Besides this symptomatic athetosis there is also an idiopathic form, the so-called "double athetosis," a disease in itself without a certainly determined anatomical substratum, in which the motor phenomena described present the only pathological manifestation, which usually is present from birth or develops in earliest childhood. Here, besides the extremities, the muscles of the face and neck are also affected; the patients continually distort the mouth, make faces, constantly turn and twist the head, and, on account of athetosis of the tongue muscles, are much embarrassed in speaking. The condition is a stationary one, although remissions occur. Whether their occurrence is to any extent favored by drugs, as arsenic, bromides, cannabis, or scopolamin, or by hydriatic procedures, is, however, a question.

Fig. 31.

Athetosis (instantaneous photograph).

Related to double athetosis, probably, is a rare progressive and incurable disease, that *Oppenheim* has described as "Dysbasia lordotica progressiva," *Ziehen* as "torsion neurosis." It affects children between eight and fourteen years old (mainly Russian Jews). The affection is characterized by a lordoscoliosis of the lower portion of the vertebral column, evident only on walking and standing, usually disappearing when the patient is lying down, along with flexion and external or internal rotation of the legs. In the rotators of the thigh, also, in the tibialis anticus and in the biceps, along with tonic tension, clonic contractions may occur. The electrical irritability is unaltered.

F. Paralysis Agitans

By this name *James Parkinson*, in 1817, denominated a disease characterized by muscular rigidity and peculiar shaking movements, which in honor of its discoverer has also been called *Parkinson's* disease. Paralysis agitans appears in the vast majority of cases between the fortieth and the sixtieth years of life, and in general affects by preference the male sex; many authors assert that the poorer classes of the population are more frequently attacked. Unexplained conditions of local nature appear here to exert an influence; since in our neighborhood, where the disease is relatively frequent, there is no evidence of a less predisposition to it in the higher classes of society. Its

etiology is quite obscure; mental stress, physical overexertion, exposure to cold and trauma, have been accused; it is probable that these factors may occasionally play the rôle of exciting causes (namely, when the first symptoms of the disease appear directly in an overstrained or traumatized limb), but in a majority of cases there is nothing of the sort, and we must confess our lack of knowledge with regard to the real causal connection. The hereditary occurrence of the disease has been only noted as a great rarity.

Parkinson's disease usually begins gradually and unremarked; the patient notices a slight general fatiguability, little by little a temporary tremor, which is at first limited to one hand (oftener the right), but gradually becomes a permanent symptom, and finally attacks the remaining extremities one after another. At the same time a continual tension of the muscles which slows all movements is added to the tremor; in a minority of the cases the last phenomenon precedes the appearance of the tremor.

In the fully developed disease the tremor presents the following charac-

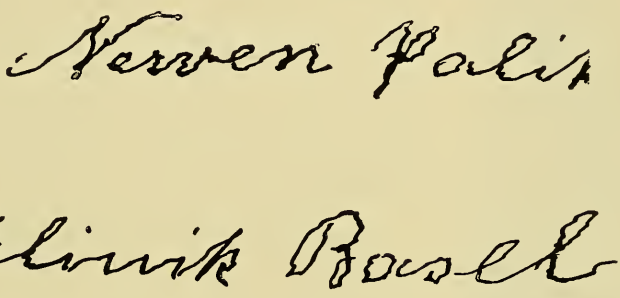

Fig. 32.
Paralysis Agitans. Handwriting.

ters: it is rhythmical, slow (two to five oscillations a second), persists during complete rest, to cease during sleep, and in the hands imitates certain complicated movements, namely, "pill rolling" and "coin counting." On carrying out a movement the tremor usually lessens, and indeed, when the disease is not far advanced, may cease, so that, for instance, a hunter, on aiming, can keep still and make the shot. Less frequently the tremor increases somewhat upon movement. Excitement always increases the shaking. While at the start, writing, eating, dressing, etc., are still possible, in the advanced stages (the disease can last twenty years or more) these functions are impossible. The tremor of the legs makes itself evident by the patient, when seated, "beating time" with his foot; usually the oscillations in the upper and lower extremities are synchronous.

The attitude of *Parkinson* patients is, as a rule, uncommonly typical (see Fig. 33). The head and trunk are bent forward, arms and legs adducted, knees and elbows flexed, in the hands the metacarpo-phalangeal joints flexed, the middle and end phalanges extended, the finger-tips of the thumb and index finger held together. This attitude is maintained so firmly that all the joints might be thought to be contractured. (Only exceptionally do the pa-

tients stand straight upright in statuesque stiffness—*Charcot's* "extension type"). The features have the immobility of an antique mask, only the eyes move in a lively manner; in looking, turning of the head is avoided, the patient turns himself rather, as a whole, about his axis. This occurs slowly with the aid of a number of small steps; especially all locomotion is usually begun with very slow, deliberate steps, so that the gait, indeed, has something solemn about it; little by little, however, the steps often become more rapid, "tripping," the bent forward patient "runs after his center of gravity," in order not to fall, and finally, in order to stop his forward movement, has to bring up against the wall or some other object. This we call "propulsion." Analogous phenomena are "retropulsion" and "latero-pulsion," which we make evident by giving the patient a slight push backward or sideways. *Parkinson* patients, on account of the hypertonus of all their muscles, as well as on account of the fact that they can only carry out an intentional movement with more or less delay, no longer have the power of regaining the balance of their bodies which have been put out of equilibrium.

Fig. 33.

Typical Attitude in Paralysis Agitans.

Disturbances manifest themselves also in the speech muscles; the voice is weak and plaintive, the speech slow, sometimes chopped off, reminding one of that of a rider upon a horse which is trotting fast. In two of my patients I observed a continual chattering of the teeth.

Sensibility is for objective tests practically always normal; exceptionally there is slight reduction of sensation in the hands and feet. On the other hand many patients complain of drawing sensations in the extremities, of cramplike pains in the calves, of paresthesia in the face ("as if something was crawling on it"), principally, however, of a burning heat of the surface of the body, particularly upon the abdomen and upon the back. On this account patients with paralysis agitans gladly uncover themselves while in bed.

The reflexes are usually exaggerated, less frequently normal. The electric muscular irritability is unaltered. The "paradoxal foot phenomenon" of *Westphal* is often found; if one brings the foot of the patient into a position of extension (dorsal flexion) the tibialis anticus, extensor digitorum, and peronei contract, since their origin and their insertion are brought nearer together, so that the foot remains for a long time in the position given it.

In most cases there is often a very distressing salivation. Hyperidrosis is rarer. A not very infrequent variety of paralysis agitans is *Parkinson's* disease without agitation. In this, with otherwise typical symptomatology, the characteristic tremor is absent.

The prognosis of the disease is not unfavorable as to life, but very unfavorable as to recovery; remissions occur, but unfortunately they are not usually of long duration. In the late stages the fate of the unhappy invalid is a wretched one, since with fully retained intelligence he is condemned to absolute helplessness, and must depend upon outside aid for the most elementary needs (for example, for turning himself in bed, wiping off the saliva that flows from his mouth). Death follows either in marasmus or from intercurrent diseases, among which cerebral hemorrhage is relatively frequent.

What is the anatomical basis of this terrible disease? As to this question it was formerly classed among the "functional neuroses." To-day, however, it is known that in the spinal cord in *Parkinson's* disease, proliferation of the glia and abnormal pigmentation of the cells of the anterior horn, together with changes in the vessels, are to be found (*Redlich, Sander, Dubief, Ballet*), also the muscles show changes according to some authors (*Blocq, Gauthier*). Nevertheless, it is not far-fetched to consider these not very characteristic lesions, which may be interpreted as an accentuation of senile changes, as only secondary, and to think of another "primum movens," perhaps in the glandular apparatus of our bodies. So *Roussy* and *Clunet* would make the parathyroid glands responsible, claiming to have found in them changes which they designate by the expression "Hyperparathyroidosis." According to *Haberfeld*, indeed, the changes found are due only to senility. The question is still open.

With regard to the treatment of *Parkinson's* disease, *Oppenheim's* remark is recommended for your consideration: "The physician can in this disease do much harm and little good." In order to avoid the first, one should refrain from trying cold water treatment, exercises, sun-baths, vigorous massage, faradization, while—indeed, seldom enough—carrying out careful passive movements, giving tepid baths (or the cooler, indifferent warm baths, and even electric baths) and a mild vibration massage can alleviate the symptoms. More useful as palliatives are certain drugs, above everything hydrobromide of scopolamin, either in the form of injections or as tablets. Of this, from 0.0002 to 0.0004 (1/300 to 1/150 gr.) once or twice a day; also duboisin sulphate in the same dose, usually moderates the tremor and muscular tension considerably. I have given both medicines, sometimes alone and sometimes combined, for years, without having observed any symptoms of intoxication. One must not decide upon this continuous method of administration too soon, since it is a last refuge. As long as the disease is not at a too advanced stage it is well to give this medicine from time to time for a period of twenty days; in the interim an arsenic cure is to be recommended; for example, Sol. Fowleri, in doses of two drops, increasing from twice to seven times a day, and then slowly returning to twice a day; also the drinking of arsenic containing waters (*Durkheimer-Maxquelle, Val Sinestra, Levico*) can be carried out after the manner recommended at these resorts. Of other drugs recommended instead of scopolamin and duboisin, from my own experience I can mention as occasionally efficient, tinct. veratri viridis (two or three times a day, 3-4 drops in thin mucilage). To be strictly avoided are all drugs increasing sweating, namely, the salicylic preparations. Most patients who have been given parathyroid substance experimentally have reacted with an increase of their trouble

(I could convince myself of this in one case), which would support the views of *Roussy* and *Clunet*. Placing the patient among as quiet surroundings as possible, preventing all exciting visits, pleasures, etc., are important; still more important in the later stages is the choice of a nurse of inexhaustible patience and great skill in carrying out all the technique of the difficult and tiresome nursing.

G. The Myotonias

A peculiar disease condition was made known by the Schleswig physician *Thomsen* in 1876. His material was furnished by his own family, which in five generations had presented over twenty cases. This "*Thomsen's* disease" later received the name of Myotonia congenita. In the vast majority of cases it is observed as a hereditary family complaint; sometimes consanguinity of the parents is to be recognized; very frequently in the relatives a heaping up of the most varied psychoses and neuropathies are found. As exciting causes acting upon the latent predisposition, physical excesses play the greatest rôle; rarely fright or trauma, very rarely infectious diseases, are accused.

The disease is either first noticed in early childhood, or it begins about the time of puberty, or somewhat later (for example, at the time of entering the army). Ninety per cent. of the cases are in males. In general the symptoms of the disease increase slowly and continually during a number of years, then become stationary; later, indeed, the intensity of the symptoms may again decrease, though such remissions are not very frequent; a recovery, on the other hand, is excluded. The disturbance pathognomonic of *Thomsen's* disease is characterized by the persistence of a condition of muscular contraction opposed to the intended end on the carrying out of a voluntary movement. If the patient, after long periods of rest, attempts to carry out any movement, this follows in the promptest manner; the groups of muscles brought into contraction together can, in spite of every effort of the will, be relaxed again only after from 5 to 30 seconds. For example, if we ask the patient to shake hands, he cannot let go for some time. If, however, the patient undertakes the movement a second, third, or fourth time, we notice that the inhibition of relaxation ceases each time after a shorter interval, so that finally the movement can take its normal course. If these patients march, at first they stand as if rooted to the ground, then follow the first most difficult steps which are interrupted by repeated tonic contractions. Little by little, however, "the machine gets going," and finally long distances are traversed without any trouble; the patient can even dance. All voluntary muscles can be the seat of the myotonic disturbances of movement. However, as a rule, the muscles of respiration are free, those of the trunk and neck less involved than those of the limbs, and of these last the upper less than the lower. The muscles of the face, of mastication, the external eye muscles, the tongue, and even the muscles of the pharynx and larynx, can be affected; while the myotonic dyskinesia makes itself apparent on grimacing and putting out the tongue in most cases; in phonation, turning the eyes, swallowing and chewing, it is

seldom observed. Psychical excitement, cold and dampness increase the myotonic symptoms.

Almost always there is increase in volume of the muscles, on account of which the patients, in spite of usually somewhat subnormal strength, present an athletic appearance.

The tendon reflexes are either normal, they exhibit tonic contractions, or they are reduced; they are seldom absent. The mechanical and electric muscular irritability are, as *Erb* has shown, very characteristically altered. Percussion of the muscles sets up the formation either of a depression or of a swelling which does not disappear for from 5 to 30 seconds. Direct faradic stimulation of muscles shows increased irritability; on stimulating with strong currents persistent myotonus appears, on continued strong faradization there is sometimes a decided "muscle wave." Direct galvanic irritation brings out the most striking anomalies: slow and persistent character of the contractions, abnormally low threshold of irritation, loss of the contractions on opening the circuit, AnC1C = or >KC1C; besides this, sometimes upon the stabile application of strong currents there are rhythmical undulations from the cathode to the anode (sometimes only appearing after repeated application of the current). These different anomalies *Erb* sums up as the "Myotonic reaction."

As a pathological change a uniform hypertrophy of all the fibers depending upon an increase of the undifferentiated protoplasm is found, while the contractile substance and its morphological expression the striation is defective. The sarcolemma shows increase of nuclei, the fibrilli vacuolization. Besides this there is slight increase of the interstitial connective tissue. *Knoblauch* thinks that there is an abnormal preponderance of the red, slow-acting muscle fibers as compared to the light, quick-acting ones, and that there is a hypertrophy of the first. Along with these muscle changes, opposing relations of innervation can play a rôle, as *Jaquet* has shown by myographic investigations.

From a therapeutic point of view, above everything else prevention of the factors, which are shown by experience to work unfavorably, is important (keeping the patient warm, prevention of emotional excitement, suitable occupation). Further, systematic exercises, warm baths and massage can alleviate the troubles. The drugs here and there recommended (strychnine, antipyrin, testicular and thyroid extract, atropin, iodide of potassium), since they are useless, have been entirely given up. A moderate amount of alcohol, on the contrary, acts favorably. The formerly practiced nerve stretching, which has as its object causing decrease of the muscular hypertrophy by injuring the nerves, is to be warned against.

Varieties of myotonia are: 1. Myotonia atrophica, a combination of *Thomsen's* disease with progressive muscular atrophy, to be described in the next lecture: this is not at all rare.

2. Myotonia acquisita (*Talma, Jolly*), an incurable condition characterized by the myotonic reaction which follows infectious diseases and traumata, in which, however, there is usually a certain muscular rigidity even when at rest, while long-continued movement favors the occurrence of myotonus.

3. Paramyotonia congenita (*Eulenberg*), very rare. This is a condition of permanent muscular contraction which usually, under the influence of cold,

occurs in certain muscle groups (of the neck, face, muscles of deglutition, and those of the extremities) and relaxes in from a quarter of an hour to several hours. The myotonic reaction is absent, the histo-pathological picture, however, much resembles that in *Thomsen's* disease. The disease is, as a rule, decidedly a hereditary family one, and is observed immediately after birth.

H. Congenital Muscular Atony

Oppenheim, in the year 1900, first called attention to a congenital pathological condition which he denominated "Myatonia congenita." This name, however, has the disadvantage that it furnishes an opportunity for confusion with the myotonia congenita described above. On this account I consider the expression "Congenital muscular atony" more correct. In such children immediately after birth a striking lack of movement makes itself evident. This is chiefly limited to the legs; in severe cases, however, it can also affect the arms, the thorax, the neck, so that the patient gives the impression of complete paralysis. Nevertheless, it is only pseudo-paralysis; by painful stimuli weak movements of protection can be set up. The muscles are doughy, but not atrophied. The muscular flaccidity is so extreme that one, for example, can raise the shoulders up above the ears, can bring the feet up behind the neck and cross them there, etc. In severe cases the limbs hang like loose appendages to the body. The tendon reflexes are absent, the skin reflexes usually retained. Electric irritability of the muscles to both varieties of current is, as a rule, reduced, seldom lost; reaction of degeneration is never present; other anomalies cannot be made out. The disease is never progressive; on the other hand, it shows a tendency very slowly to improve. Recovery is probably the most frequent termination; we endeavor to accelerate it by strong, deep-acting faradization. After an electro-therapeutic treatment a temporary reappearance of the patellar reflex is sometimes to be noted, more frequently the appearance of a few voluntary movements, which then become more and more a permanent acquisition. Further, irritants favoring movement, and later exercises, come into consideration; finally, carefully managed strychnine and arsenic cures. The prognosis is impaired by susceptibility to fatal broncho-pneumonias in case the respiratory muscles are attacked. The disease is not hereditary, almost never a family one. A unique anatomical substratum does not appear to be present. In one case examined by me, a piece of muscle, removed for diagnosis, showed only abnormal richness in nuclei. Other observers have, however, seen loss of the cross striation and striking irregularities in the contour and caliber of the fibers. Also anomalies have been found in the spinal cord, for example, abnormal smallness of the motor cells of the anterior horn.

A clinical likeness to congenital muscular atony is presented by the "myopathia rachitica," isolated by *Hagenbach* and myself between 1904 and 1907, a muscular disease never congenital, but developing in rachitic children, in which "snake man like" positions, crossing the feet behind the neck, etc., are found exactly as in the former. Histological alterations are found in this disease in relatively few and severe cases, and are then characterized, as I

have shown, by uniform disturbance of growth of the muscular tissue in the direction of a regression to a comparatively undifferentiated stage (see Fig. 34).

I. Periodic or Paroxysmal Paralysis

This disease, whose first description (1882) comes from the Russian physician *Schachnowicz*, very often, but not always, occurs as a family disease, and usually in early life The affected persons present at intervals of different length, transitory paralyses of extensive muscular groups. These rarely affect only the legs after the paraplegic type, usually the arms are also attacked, often the muscles of the trunk, too, while the muscles of the face and the eyes almost always remain unaffected. The paralysis, usually most intense in the legs and starting here, as a rule, can be complete, or only moderate paresis. On electric examination direct and indirect irritability is reduced at the beginning of the attack and while it lasts. The paralysis is usually flaccid; here and there, however, increased tension of individual muscles has been observed; the tendon reflexes in the paralyzed parts are reduced or lost, only occasionally exaggerated; the skin reflexes, on the other hand, are normal. The paralysis is little by little established in the course of about an hour; movement of the body delays its appearance; on this account nocturnal attacks are especially frequent. Return to normal is accomplished by degrees after a few hours; the paroxysms, however, repeat themselves at intervals of different lengths (days, weeks, or even months). Between the individual attacks the patients are quite healthy. It is probably a disease due to autointoxication; the abnormally constituted muscles appear to react to the formation of a poison which is accumulated during rest, by failure of function; the urine voided during the attack shows increased toxicity, often contains acetone. Therapeutically, massage and strong faradization are recommended; further, atropin in daily doses of 0.002 until mild intoxication is beginning; bromine and iodine salts, strychnine and eserine have been tried without any results worth speaking of.

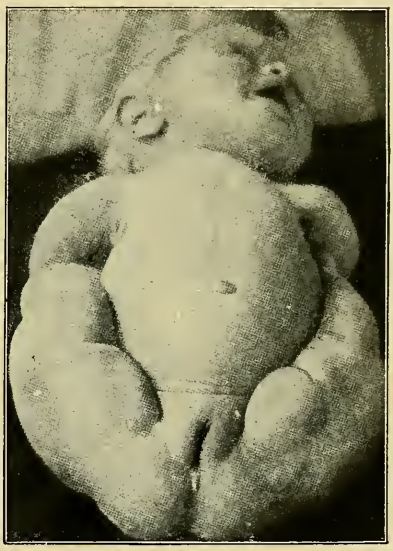

Fig. 34.
Rachitic Myopathy.

In this connection "paralyzing vertigo" (*Gerlier*)—"Kubisagari" (*Miura*) may be mentioned. It is a disease occurring in stable-men in the canton of Geneva, and endemic in Japan, probably of miasmatic origin, and recovering upon removal from the atmosphere of the stable. It consists also in flaccid periodically occurring paralyses, with intervals of health, but it begins with very severe vertigo, besides the extremities it attacks also the muscles of the neck and the levatores palpebrarum, and can, indeed, avoid the muscles of the

limbs. Difficulty in swallowing, paralysis of the masseters, diplopia, and amblyopia are not infrequent. Not rest, but work, seems to favor the outbreak of the attacks.

J. Myasthenia

Myasthenia gravis pseudoparalytica, or the *Erb-Goldflam* disease, is a disease which is characterized by excessive fatigability of the muscles, leading to paralytic weakness, and for which the muscles supplied by the motor cranial nerves show special predilection.

This comparatively rare disease usually develops gradually (commonly between the ages of twenty and forty years, and in women somewhat more frequently than in men). As to its etiology we know nothing certain; if in the anamnesis we hear very frequently of exhausting diseases having been passed through or of severe overexertion, nevertheless on account of the rarity of the disease we must assume that these factors exert their pathogenic action only upon the ground of an already present predisposition. There is probably a congenital inferiority of the muscular system; in this sense speaks its relatively frequent accompaniment by malformations; for example, of the toes and fingers, of the mamma, of the genitals, and of the lungs, or by progressive muscular dystrophy, which likewise develops upon an endogenic basis. (See the next lecture.)

A more and more rapidly appearing fatigability which leads to more and more complete paralysis (which disappears on rest) makes itself felt, first in the external eye muscles, the mimic muscles, those of the pharynx, the masticatory muscles, and those of the tongue; later the phenomenon usually extends also to the territory innervated from the spinal cord—neck, trunk and extremities. All muscular action in the region of the affected structures begins at first without delay; after a few movements, however, the contraction is less extensive, always feebler in its results, and finally the muscle is entirely paralyzed, or better, pseudo-paralyzed, since after a more or less extended rest, its function is regained. Least manifest is this anomaly in the morning after awakening, increasing during the day and reaching its maximum by evening. Not all muscles are equally fatigable; most severely affected, we find almost always, those which are naturally used a great deal, so, above all, the eye muscles. Hence double vision as well as failure of the levator palpebræ (ptosis) belong to the most invariable symptoms of myasthenia. Besides this, it leads to facial diplegia, to disturbances of speech, chewing and swallowing, to falling of the head upon the breast, to exhaustion paralysis in the arms and legs (namely at the root of the extremities); occasionally, however, the muscles of the trunk and limbs remain unaffected while their involvement alone is of the greatest rarity.

The disease has a great tendency to intermissions, in which the patients for weeks, months and years can be entirely free from trouble; termination in recovery is, on the other hand, rare, and the prognosis is on this account unfavorable, since the harmless initial stage of the disease can stretch over exceedingly long periods of time characterized by long intermissions and short

exacerbations. (In one case of *Curshmann's*, twenty-two years.) Once the myasthenia has reached its full development, however, the situation is exceedingly dangerous. The pseudo-paralysis is constantly more permanent, the muscles recover almost not at all. Again the symptoms are more severe in the external eye muscles and give the picture of external ophthalmoplegia. The voice is aphonic on account of failure of the larynx muscles; the face resembles a death mask; the pseudo-paralysis of the extremities and of the muscles of the back and neck compels the patient to keep his bed; on taking food he chokes, and fluid nourishment regurgitates through the nose if it does not enter the trachea. Conditions favoring the occurrence of hypostatic and aspiration-pneumonia are hence furnished in the highest degree; if paralysis of the respiratory muscles (perhaps also of the heart) does not sooner put an end to life.

Clinical examination of the diseased muscles shows with entire absence of atrophy, fibrillary contraction and reaction of degeneration, almost always, the so-called "myasthenic reaction" which can be defined shortly as an overfatigability for the faradic current: if a muscle is faradized continually for half a minute to one minute, a uniform reduction in the contraction is observed, which according to the strength of the stimulus is lost entirely, sooner or later. A pause of scarcely a minute suffices, however, to allow the stimulus to become again active as before. The faradic tetanization can be carried out, not continuously but at intervals of from two to three seconds; the contraction is then observed to become weaker at each stimulation until finally the muscle no longer responds, to react again, however, when it has been permitted some rest. The myasthenic reaction is obtainable from the nerve as well as from the muscle; it is not absolutely pathognomonic of the *Erb-Goldflam* disease, as it has been found in different organic nervous diseases and in a slight form even in healthy people. Overfatigability of the tendon reflexes by repeatedly eliciting them is occasionally to be found; they usually remain normal, however.

As far as the pathological anatomy of the disease is concerned it is certain that the brain, spinal cord and peripheral nerves remain normal in myasthenia. In the muscles themselves, however, changes have been repeatedly found, of which the most uniform is the increase of lymphoid cells figured by *Weigert*. *Knoblauch* has made an abnormal preponderance of the light muscle fibers over the dark ones responsible, hence, according to his view, the *Erb-Goldflam* disease is the opposite of *Thomsen's* disease (see page 90). The fact that here and there persistence of the thymus, or tumors of this body have been found at autopsy in myasthenics, attracts attention to its possible connection with internal secretions. *Lundborg*, on theoretical considerations, has conceived myasthenia as being a hyperparathyroidosis, as an expression of the overfunction of the parathyroid glands. On the other hand I have not been able to find the least trace of myasthenia or the myasthenic reaction in rats which had been made artificially hyperparathyroiditic by *Iselin* by gland transplantation. In its pathogenetic relation *Marinesco* attributes an important rôle to an insufficiency in formation of the oxydase necessary for the recuperation of the working muscle.

The treatment of myasthenia gravis pseudo-paralytica must above everything be a therapy of avoidance. No electro-therapy! Even electro-diagnostic testing is not to be undertaken oftener than is absolutely necessary. During the exacerbations of the disease rest in bed; during the intermissions, long periods of entire rest introduced into the régime of the day. Rest cures in the open air, at climatic stations where possible, should be carried out every year. On the other hand all athletic exercises are to be strictly forbidden. Naturally the disease causes unfitness for military service. Hydrotherapy is to be avoided. In the end stage the use of the œsophageal tube is to be avoided as much as possible, since *Oppenheim* has observed sudden death from asphyxia while it was being passed. Long periods of rest after meals should be enforced. Nourishment must be selected according to the viewpoint of the most nutrition in the smallest volume. Of drugs, the combination of arsenic and strychnine is to be recommended; for example, ℞ Sol. Fowleri, 5.0; tinct. nuc. vom., 10.0. M. S.—6 to 12 drops t. i. d.

LECTURE VI

The Progressive Muscular Atrophies

GENTLEMEN: When we speak to-day of "Progressive Muscular Atrophies," we use this term in a certain sense as a proper name for quite definite chronic degenerative processes in the motor apparatus. In no way, however, for other conditions setting in with progressive degeneration of the muscles. Such degeneration, indeed, can occur upon the basis of a great number of organic nervous diseases, of which, for example, we have already become acquainted with the polyneuritides, and in later lectures will study amyotrophic lateral sclerosis, syringomyelia, and other conditions. We differentiate a myopathic, a neural and a spinal form of "progressive muscular atrophy" whose different anatomical and clinical peculiarities we will proceed to study. At the start, however, we must understand that many facts warn us against drawing the line between these three subvarieties too schematically. On the one hand there are numerous clinical transition forms, on the other, autopsies have shown that in the neural form almost always, in the myopathic not rarely, changes are found in the spinal cord.

Common to the whole group is its endogenic basis, which in the myopathic and neural forms, shows itself through its predominantly hereditary family occurrence, while in the spinal form a more frequent nonfamily type stands on the one hand, a rarer family type on the other. The congenital inferiority, upon the basis of which the progressive muscular atrophies develop, shows itself in an occasional combination with other heredo-degenerative diseases or with malformations and congenital defects. (For example, with myotonia congenita (see page 90); further, with hereditary ataxia, with family spastic paraplegia, with congenital muscular anomalies, with funnel chest, skull and jaw deformities, atrophy of the bones, symmetrical exostoses, hemihyperplasia of the skeleton, shortening of the thumb, idiocy and deafmutism. etc.)

A. The Myopathic Form

This is also called primary progressive muscular atrophy; much used, however, in German-speaking countries, is the name proposed by *Erb*, "Dystrophia Musculorum Progressiva" (Progressive Muscular Dystrophy), while in France *Charcot* procured the adoption of the term "Myopathie primitive progressive."

As already said, this is a disease of chiefly hereditary-family occurrence, which nearly always begins in early life within the two first decades, and is unnoted at the start. The first symptoms of muscular wasting make themselves evident at the roots of the extremities and upon the trunk, seldom about the head, almost never in the peripheral segments of the limbs, which, as a rule, remain intact even in the later course of the disease. In the muscles under-

going atrophy, reaction of degeneration is almost never observed, but only simple quantitative reduction of irritability. Fibrillary contractions are also absent. The usually symmetrically affected muscles show a particularly characteristic peculiarity, namely that, along with the atrophic processes there occur changes which cause an increase in volume of the affected parts and striking abnormalities in the form of the body; the combination of this "pseudohypertrophy" of certain muscles with atrophy of others is comparatively regular; sensibility, the function of the sense organs, the sphincters of the bladder and of the rectum remain normal.

Along with cases impressing themselves by typical heredo-family occurrence in which transmission occurs chiefly through the mother, who herself, however, can remain free from the disease—I designate this last as "maternal metrapectic inheritance,"*—come also sporadic cases, namely, in the "juvenile scapulohumeral type" (*Erb*) which we will consider more at length under the discussion of the special forms of dystrophy. Not infrequently infectious diseases, trauma, or overexertion give the signal for the outbreak of the disease; the same factors also may cause new progress after temporary arrest.

Pathological Anatomy

The disease process which lies at the basis of this trouble has been thoroughly studied by *Erb* and *Marinesco*. Macroscopically the "fish flesh" ap-

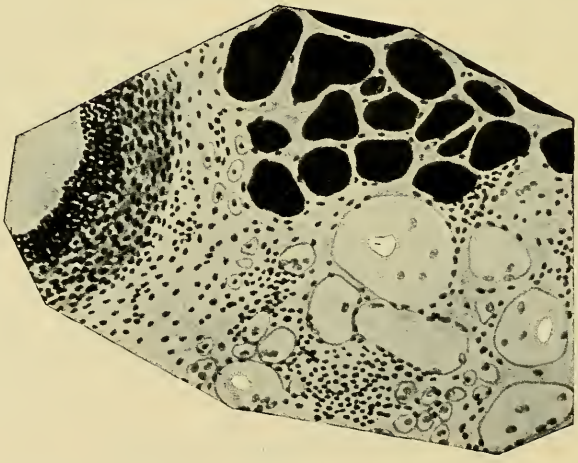

Fig. 35.

Alteration of the Muscle in Progressive Muscular Atrophy. Hematoxylin-eosin-osmic acid stain. Gastrocnemius. Zeiss Oc. 2, Obj. E.

pearance of the affected muscles, which are plainly distinguishable from the healthy brown-red muscles, but hardly at all from the surrounding fatty tissue in color, is particularly striking. In sections (see Fig. 35) the striking inequality in size of the fibers is first noticed. While, according to *Erb*, the extremes in the normal cross sections of the fibers vary between 20 and 80

* Απεχειν = To avoid. τη μητηρ = The mother.

microns and ninety per cent. of the fibers measure between 20 and 60 microns, among a larger or smaller contingent of such normal sized fibers are found numerous atrophic ones measuring from 7 to 15 microns, also hypertrophic ones of 100, 125 even 200 microns and more. The hypertrophy of the fibers seems to represent the first stage of the process, the atrophy the latter one; it finally passes over to entire loss of fibers when only empty sarcolemma sheaths are left. One need not think that in pseudo-hypertrophic muscles the thick, and in macroscopically atrophic muscles, the thin, fibers especially predominate, since nothing is more like the parenchyma of an atrophic muscle in dystrophy, under the microscope, than the tissue of a pseudo-hypertrophic one. Determining the total volume, indeed, is the smaller or greater degree of proliferation of fat and connective tissue. Both can remain within moderate bounds or may reach an excessive degree.

The muscle fibrilli almost all show a considerable increase of both the border and of the interior nuclei. Many are fragmented, split, or show in the central part, round vacuoles that look as if punched out. However great the atrophy of the fibers, the striation is plainly preserved everywhere. In the areas most diseased the longitudinal striation usually seen is increased to definite fibrillation; upon longitudinal sections the muscles sometimes appear bordered with rosary-like swellings. The intramuscular vessels undergo to a great extent endoarteritic and mesoarteritic changes which can cause great thickening of their walls; in the periarterial connective tissue, and also in the perimysum, considerable increase of nuclei is often found; this can increase to definite areas of infiltration. In contradistinction to these constant changes in the muscular tissue, the lesions in the anterior horn cells of the spinal cord, found in only exceptional cases, fall entirely into the background. Since, however, the trophic centers for the muscles lie in these last, it is conceivable that functional alterations of these cells are responsible for the perverse method of development and disturbances of nutrition in the muscles, even in those cases in which the anterior horn cells appear morphologically unaltered.

CLINICAL BEHAVIOR OF THE DYSTROPHIC MUSCLES

Upon investigating the muscles of dystrophic patients we will find naturally different conditions depending upon whether we are observing an atrophic or a pseudo-hypertrophic region.

In the first case the muscles are either uniformly thin as a whole, which process can go so far that the affected portions of the extremities look like those of a skeleton, or in the middle of the muscle among the atrophic parts, round, sometimes globular prominences, are found. This phenomenon, which sometimes is first seen when the muscle contracts, is entirely pathognomonic of muscular dystrophy and appears as "partial pseudo-hypertrophy" to present a transition to the second manner of appearance, dystrophic myopathy. In this last form the affected muscles increase in mass to such an extent that athletic contours are produced (Fig. 36). On palpation one experiences in them either a firm, often peculiar fluctuating rubber-like resistance, or (in advanced stages) on the other hand, a peculiar dead feeling. Here and there I have seen

linear atrophies of the skin like the well known striæ of pregnancy in women, over such pseudo-hypertrophic muscles. It can happen that a muscle whose ability to functionate is almost suspended by the dystrophy, presents a normal outline; in it the atrophic and the pseudo-hypertrophic process balance one another. That atrophic or pseudo-hypertrophic muscles shorten and produce contractures like that in Fig. 38, as well as that visible in the left leg of the boy in Fig. 36, is not a frequent occurrence.

With the morphological anomalies related, disturbances of function go hand in hand; the strength and the extent to which the affected muscles are able to

Fig. 36.

Progressive Muscular Dystrophy. Pseudo-hypertrophic form.

contract, in the course of the disease continually grow less; at the same time the electrical and mechanical irritability continually decrease, to be finally lost. The exceedingly rare cases, in which, besides this, indications of reaction of degeneration are found, are usually otherwise atypical and probably should be considered as transition types to the neural and spinal forms of progressive muscular atrophy.

The tendon reflexes in the affected region decrease and are finally lost.

Progressive muscular dystrophy has its "favorité muscles." Other muscles it attacks only exceptionally, still others it regularly avoids. As to this peculiar

electivity, you may be guided by the following table in which I have also indicated, by italicizing, the muscles which are particularly predisposed to pseudo-hypertrophy.

Usually Affected.	Less Frequently Affected.	Exceptionally Affected.
Pectoralis major (pars sternalis)	Sternocleidomastoid	Diaphragm
Pectoralis minor	*Infraspinatus*	Masticatory muscles
Latissimus dorsi	Supraspinatus	Eye muscles
Serratus anticus major	Levator scapulæ	Heart muscle
Rhomboideus	Coracobrachialis	Tongue *
Trapezius (especially pars inferior)	Triceps	
Erectores spinæ	Teres major	
Deltoid	Teres minor	
Biceps brachii	Abdominal muscles	
Brachialis anticus	*Sartorius*	
Supinator longus	Tensor fasciæ latæ	
Glutei		
Quadriceps		
Adductores femoris		
Peronei		
Tibialis anticus		
Gastrocnemius		
Orbicularis oris		
Orbicularis palpebrarum		

TYPICAL ANOMALIES OF CONFIGURATIONS AND ATTITUDE

Looking at the nude bodies of dystrophic patients we are impressed by the fact that the deep depressions of the atrophic parts and the great swellings of the pseudo-hypertrophic ones, join immediately on to the well-formed portions of the limbs. On this account the whole figure takes on an inharmonious outline. Besides this, however, we come face to face with a number of exceedingly characteristic local disfigurements, for example, the picture of scapulæ alatæ which occupied us already in the first lecture; this occurs on account of elimination of the action of the serrati (see Fig. 37). If to this is added, however, that of the pectorales, the trapezii and the latissimus dorsi, the shoulder-blades with the attached arms become loose appendages of the thorax, and we speak of "loose shoulder." The atrophy of the sternal portion of the pectoralis causes a flattening of the thorax; under certain circumstances, indeed, the sternum and ribs form a sort of a fissure. Decided lordosis occurs on account of the fact that extension at the hip-joint is impaired and the center of gravity must be moved forward; the weakening of the contractions of the abdominal muscles can increase this disfigurement to the highest degree; between the approximated shoulder-blades and the hips, a number of cross folds produced by the forcing together of the soft parts are observed. The "gnomes calves" occur on account of the pseudo-hypertrophy of the gastrocnemii, the "tapir snout" through the trunk-like increase of volume of the upper lip. Through imperfections of the orbicular muscles of the eyes and the mouth the "myopathic facies" is produced, the gloomy rigid "Sphinx countenance." If

* Our patient of Fig. 36 showed great enlargement of the tongue. Besides this an enlargement of the heart was demonstrable by percussion and by orthodiagraphy.

such a patient laughs, however, on account of the imperfect function of the orbicularis oris, his mouth is drawn into an ugly fissure ("transverse laughing") while the two nasolabial folds impose themselves as deep vertical furrows, the "coups de hache" of the French; the atrophy of the trunk muscles can lead to the so-called "wasp waist."

INDIVIDUAL FORMS OF PROGRESSIVE MUSCULAR DYSTROPHY

We have passed before us the characteristic anomalies which fully developed cases of dystrophy present. We must now, however, go into the

Fig. 37.

Progressive Muscular Dystrophy. (Type *"Erb."*) Winged Scapula.

definite grouping of symptoms which, characterized also by peculiarities as to the time of beginning and course, impose themselves as special types of dystrophy. We distinguish:

1. Forms with initial and preponderant involvement of the pelvic girdle and thigh muscles.

a. Atrophic variety (type, *Leyden-Mœbius*).

b. Pseudo-hypertrophic variety (type, *Duchenne-Griesinger*).

2. Forms with initial and preponderant involvement of the shoulder-girdle and upper arm muscles.

a. Juvenile, scapulo-humeral variety (type, *Erb*).

b. Infantile, facio-scapulo-humeral variety (type, *Landouzy-Dejerine*).

The pelvic-femoral forms (1a and 1b) begin, as a rule, in childhood and affect by preference boys. The atrophic variety usually begins later than the pseudo-hypertrophic; the first usually in the 8th to 10th year of life, the latter about the 3d to 5th year. The first symptom which usually strikes the

parents is a waddling gait caused by weakening of the glutæi. The children fall easily and experience great difficulty in climbing stairs. Often, on walking, they let the feet fall stamping, a result of the quadriceps paralysis. A lordosis is usually early apparent also. If we lay such a child upon the floor and then let him rise, on account of the weakness of his back, thigh, and leg extensors, he is compelled first to assume the position of a quadruped and then, by aid of his hands, to climb up his legs, as it were. In the pseudo-hypertrophic *Duchenne-Griesinger* type, the great increase in volume of the calves and buttocks makes itself early evident (Fig. 36); while in time the disease also involves the shoulder-girdle and upper arm muscles, pseudo-hypertrophy

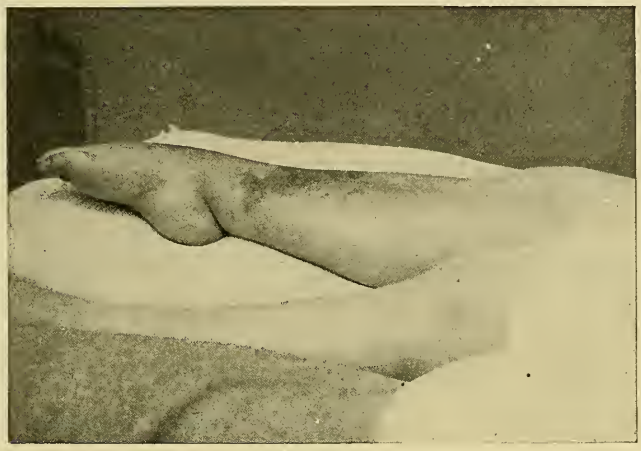

Fig. 38.

Excessive Clubbing of the Foot on Account of Contracture of the Gastrocnemius in Progressive Muscular Atrophy.

usually does not occur in these; later, indeed, it falls off in the glutei and gastrocnemii to give place to atrophy. The muscles of the face are later and less regularly affected in the hypertrophic variety than in the atrophic.

Of the two shoulder-girdle upper arm types, that of *Erb*, the "juvenile" (2, a, see Fig. 37) begins in youth (or even during the third decade of life) and usually develops very slowly, while the *Landouzy-Dejerine*, or "infantile" variety, commences in early childhood and spreads somewhat more rapidly. In the muscles of the shoulder-girdle and upper arm mentioned in the table on page 101, in the infantile type at the same time in those of the face, an increasing weakness and atrophy become apparent. In *Erb's* type pseudo-hypertrophy is common (in the deltoid, triceps and infraspinatus), in the *Landouzy-Dejerine* type it is almost always absent. Only when the last has later attacked the legs also, it may cause increase in volume of the calves, like *Erb's* type.

After we have exposed the four chief forms of progressive muscular dystrophy in their chief differential points, we will not neglect to point out how very frequently atypical cases come under observation; whether it is, that the time and the topography of the commencement of the diseases do not

agree with one another (for example, when a pelvic and thigh form does not begin until the end of the twentieth year or even later), or that the grouping of the affected muscles, or the order of their involvent presents unusual relations.

Differential Diagnosis

All in all, one can say that progressive muscular dystrophy in its fully developed stage can hardly be confused with any other disease; as to its special differences from the "spinal" and the "neural" forms of progressive muscular atrophy, I will refer in this connection to a description of these last. In the early stages, on the other hand, one must be on his guard against diagnostic errors. The abnormal manner of rising occurs in beginning spondylitis, in circumscribed acute lumbar polyomyelitis, in abnormally localized post diphtheritic paralysis, in rickets; the waddling gait in rickets and osteomalacia; the "gnome-like" calves in spastic spinal paralysis and in hydrocephalus; myotonia congenita runs its course with increase in volume of the muscles, this can occur locally, also, as a congestive phenomenon in thrombosis of veins; finally, there is a genuine congenital muscular hypertrophy. The opposite to the last condition is furnished by congenital muscle defects which, indeed, attack by preference the "favorite muscles" of dystrophy (*Erb, Bing,* etc.), but are almost always asymmetrical.

Course and Prognosis

Though pauses for ten, fifteen, and even thirty years may interrupt the progress of muscular dystrophy, nevertheless its course is almost always steadily, though slowly, progressive. In general it is the rule that in the average the cases beginning early and the pelvic-femoral forms run the most rapid course. Such patients, as a rule, die comparatively young, since the earlier they become permanently bedridden, the more threateningly hangs over them the Damocles' sword of tuberculosis and broncho-pneumonia, to which most of them fall victims. Patients with the juvenile type of the disease, however, may reach the age of seventy years or more. That progressive muscular dystrophy is the direct cause of death is exceedingly rare; in such cases the result depends upon the involvement of the diaphragm and respiratory muscles; still rarer, however, are recoveries of typically dystrophic children, as they have been observed by *Erb* and *Marina.*

Treatment

What can, and shall we do, not to put a stop to this fatal disease (since this does not lie within our power), but at least to oppose its progress as much as possible? In the first place it is necessary to improve the general condition as much as possible by good air and by nourishing food, not too rich in fats and carbohydrates. Care should be taken that daily such an amount of exercise and movement as his condition permits, without the production of a feeling of fatigue, should be carried out by the patient. "Ne quid nimis"

should also be the motto in treatment by massage and electricity; the last I apply only in the form of the "electric hand" with very weak faradism, or as galvanization, and I would warn you directly against producing faradic contractions of the muscles. I have here and there seen good results from salt baths beginning with very weak concentrations and increasing gradually to moderately strong ones, also from prolonged tepid baths and corresponding indifferent thermal ones, that is, a decided improvement in the function of the muscles. As regards attempts at medication, however, I can only report failures from the much used strychnine and from thyroid and hypophysis preparations; also the results of injections of muscle juice vaunted by certain enthusiasts have, as was to be expected, remained unconfirmed. Not infrequently we are in a position to call in the aid of the orthopedist; in contractions of the dystrophic calf muscles tenotomy of the Achilles tendon may be of use; in very slowly progressive cases with suitable topographic distribution of the still healthy and of the diseased muscles, tendon transplantation, in "loose shoulders" fixation of the scapulæ to the thorax (scapulo-pexia) or approximation of the scapulæ to one another, further, the correction of scoliosis by supportive apparatus, etc. From a prophylactic point of view I would advise you to take to heart the idea that every intercurrent disease in dystrophic patients, even the slightest one, and especially bronchitides, should be treated from the start with special care.

B. The Neural Form

This variety of progressive muscular atrophy is characteristically an heredo-family disease. "Erratic" cases are quite rare, while, on the other hand, up to thirty cases have been counted in the same family. The male members of these families are attacked by preference, if not exclusively. As exciting causes, infectious diseases and overexertion are accused.

This form is also called "The *Charcot-Marie* type" of progressive muscular atrophy. Besides these two authors, *Tooth* and *Hoffmann* have rendered service in establishing the clinical picture of this disease. The term "neurotic" muscular atrophy, introduced by the last, should naturally be corrected into "neural," since the disease has not the least thing to do with the neuroses.

Symptomatology, Course and Prognosis

The disease begins, as a rule, between the sixth year of life and puberty; however, cases have been known to begin at the end of the first year of life, or in the third and even in the fourth decade.

The disease consists in a slowly progressive muscular wasting which begins symmetrically at the extremities of the limbs, to attack later the leg and forearm. Usually the feet are first affected, less frequently the hands, or the hands and feet simultaneously. The following symptoms are produced: atrophy of the smaller muscles of the feet, the peronei, extensor communis digitorum, club-foot and claw-toes, atrophy of the calf muscles; wasting of the small muscles of the hand, with claw-hand, later, atrophy of the muscles of the fore-

arm with eventually loss of the claw position of the fingers. Usually the disease halts abruptly at the level of the knees and elbows; through this the picture of "stork legs" can occur. In advanced stages the thigh muscles may be also attacked, as a rule, however, only the vastus internus. One can always plainly perceive that the atrophy is the primary phenomenon, the paretic symptoms only its consequence; these are, however, not rarely very slight, also the extremely slow progression of the disease admits of the possibility that the still retained parts of the muscles may adapt themselves functionally to the abnormal condition. One of my patients in his forty-fifth year could still cut tin.

The tendon reflexes decrease and finally are lost, the skin reflexes are nearly always normal. Fibrillary tremor, whose importance in the picture of spinal progressive muscular atrophy we will point out later, is also frequent here, but in no way constant. The mechanical and electric irritability of the affected muscles decrease and are finally lost; reaction of degeneration, on the other hand, is almost always incomplete, since for a long time a large contingent of normal muscular fibers are mixed with the degenerated ones. Pseudo-hypertrophies never occur, contractures of the calf muscles are very rare.

In contradistinction to the other forms of muscular atrophy it must be observed that sensibility is disturbed in many cases, both subjectively and objectively. One or more of the following symptoms are observed: Paresthesias, lancinating or cramplike pains, sensitiveness to pressure of the nerve trunks, hypesthesia of the integuments (usually slight) of the peripheral portions of the extremities, reduction or loss of vibratory sensation in the same territory, disturbances of sense of position, ataxic phenomena. Vasomotor disturbances also occur, as hyperidrosis of the feet, hypothermia, cyanosis, and marbling of the legs, and even perforating ulcer of the foot! If cure is excluded, nevertheless, the disease cannot only halt definitely at the elbows and the knees, but may be interrupted in its evolution by year long pauses. Although in general there is great loss of resistance to intercurrent diseases, especially to those of the lungs, nevertheless, many of these patients live to be old people.

Pathological Anatomy

According to the anatomical substratum of "neural progressive atrophy" this nosological characterization must appear entirely insufficient in almost all cases, since, as a fact, along with the neural lesions (degeneration of the peripheral nerves, usually also of the anterior and posterior roots as well as of the spinal ganglia) and the degenerative muscular wasting dependent upon them, there are in almost all cases which come to autopsy spinal cord changes in addition, as I said at the beginning of this lecture. These are degenerations in the posterior, and often also in the lateral columns of the cord. Since the degenerative processes reach their maximum at the periphery and grow less toward the center, it is possible, as some authors affirm, that the spinal alterations are only secondary.

Treatment

The treatment to be recommended in neural progressive muscular atrophy agrees in the main with that of the spinal form which will be discussed at the end of this lecture. In the neural type, however, on account of its relative mildness and its exceedingly slow development a better result may be promised from surgical treatment (arthrodesis, tendon transplantation, tenotomy) and on this account it can find more frequent use.

G. The Spinal Form

In considering this third chief form of the progressive muscular atrophies I will confine myself to the type described about the middle of the last century by *Aran* and *Duchenne*, not considering the exceedingly rare cases which have been denominated the *Werdnig-Hoffmann* type. This last is distinguished from the *Aran-Duchenne* type by beginning in early childhood, terminating in from one to six years through broncho-pneumonia, or by extension of the process to the medulla, attacking the muscles of the pelvis, the trunk and the thigh, and corresponding to the rapid extension of the atrophy presenting a complete reaction of degeneration. Further the *Werdnig-Hoffmann* type is almost always characterized by hereditary family occurrence, while the classical *Aran-Duchenne* type usually appears as an isolated phenomenon. On this account it takes a special position in the circle of the progressive muscular atrophies, since in it the rôle of the endogenic factors appears to be less preponderant. Also other facts give the impression that here exogenic factors act to a somewhat more definite degree.

Etiology

As a fact it is striking that people of the working classes are to a so preponderant extent victims of this disease, much more so, indeed, than would be accounted for by the numerical excess of this class of society. The thought that the evil influence of physical excesses is much rather to be held responsible, gains probability from the finding that the atrophy begins usually on the right, in right-handed people, on the left, in left-handed ones. Smiths and weavers are specially predisposed. As atypical localization of the initial symptoms they have been known to begin in the calves in a ballet-dancer, in the right deltoid in a ribbon-weaver who had to hold his right arm up continually, in the muscles of the radial side of the forearm in a grave-digger, etc. Very important is the circumstance that an acute poliomyelitis anterior in childhood seems to furnish a locus minoris resistentiæ which can predispose to the later outbreak of a spinal progressive muscular atrophy. Finally, the not rare history of previous traumatic influences and acute infectious diseases is to be mentioned, while the cases impressing themselves clinically as progressive muscular atrophy, but which depend upon syphilis, form an affection by themselves which can usually be brought to a standstill by antisyphilitic treatment.

Course and Symptomatology

The beginning of the disease, which usually occurs between the twenty-fifth and forty-fifth year and affects especially men, is gradual and little marked. A thinning of the small muscles of the hand, namely those of the ball of the thumb which is already plain, usually first makes the patient aware of his disease. The hand attacked first (as already said, usually the right) even later remains ahead of the other in the intensity of the muscular wasting. After the abductor pollicis brevis, opponens, flexor pollicis brevis and abductor pollicis one by one are affected by the atrophy ("individual atrophy") the first metacarpal bone is pulled backward through the preponderant action of the extensor pollicis longus, so that the thumb comes to lie on the same plane with the other fingers. This is the so-called "Ape-hand," which by the atrophy of the interossei later becomes a "claw-hand" and, still later, when the deep and superficial flexors and the hypothenar muscles have become atrophied, a "skeleton hand." It is now typical of the further course of the affection that the atrophy proceeds farther "by leaps." After the forearm muscles or indeed, before them, the muscles of the shoulder-girdle are affected, not those of the arms. The shoulders become "loose," the arms dangle like flails from their insertion. Little by little, however, the arms, as well as the trunk muscles, are affected, when in order to maintain the equilibrium the physiological curves of the spinal column become exaggerated. Finally, the entire upper half of the body may look like that of a skeleton. On the other hand the lower limbs are either attacked not at all, or only in the last stages of the disease (with very rare exceptions). The clavicular part of the trapezius as "ultimum moriens" usually remains longest intact of the shoulder muscles, while the muscles of the neck are usually not attacked at all.

As opposed to the above sketched, comparatively stereotyped course, commencement in the shoulder muscles marks the most frequent atypical form.

As terminal symptom-complexes, which, however, only develop in a small number of cases, I would mention 1, affection of the respiratory muscles, especially of the diaphragm; 2, the occurrence of bulbar paralysis, which is to be described in the next lecture.

The clinical characters of the muscular affection are in the *Aran-Duchenne* disease on the whole quite homogeneous. Above everything it is to be emphasized that the atrophy is the primary condition, the paralysis only its result; the earliest functional symptom is an increased fatigability. With wasting of the muscles, sometimes preceding this, the phenomenon of "fibrillary contractions" (see page 64) becomes evident, in the thenar muscles sometimes so marked that a motor effect (twitching of the thumb) occurs. In the atrophic region the tendon reflexes are diminished, finally lost. Electric irritability is reduced, but the reaction of degeneration is usually only incomplete (slow contraction to galvanism, retained faradic irritability). Also it is rather hard to find places in which it can be obtained, since there is almost always a very considerable number of normal fibers mixed with the degenerated ones, even to the advanced stage, and these mask the anomalies of contraction. The reaction is found most frequently in the ball of the thumb. After the changes in the

upper extremity first affected, which have preponderated for some time, are overtaken by those in the other arm, the disease usually progresses symmetrically, and indeed, so gradually, that by practice (the development of new synergies, etc.), the patients are long able to use their skeleton arms for some acts astonishingly well.

Apart from occasional sensitiveness to pressure in the wasting muscles and from the paresthesias which the dead weight of the hanging extremity dragging upon the brachial plexus produces, sensibility remains intact. Vasomotor disturbances in the form of coldness, marbling, lividity of the integuments occur quite frequently, more rarely there is firm œdema of the hands. There is often a tendency to profuse sweating. Of trophic phenomena loss of subcutaneous fat, thinning of the skin over the atrophic parts, as well as the very infrequent atrophies of bone, and arthropathies are to be mentioned. The sphincters and the innervation of the genitals always remain normal.

Pathological Anatomy

There is a progressive sclerotic atrophy of the cells of the anterior horns of the spinal cord with pigment deposit in them, in the segments corresponding to the affected muscles; with this there is of course atrophy of the peripheral motor neurone as well as the picture of secondary degeneration of muscles (reduction of the size of the fibrilli, granular or fatty degeneration of the sarcoplasm, empty sarcolemma sheaths with increase of nuclei, proliferation of the connective tissue of the septa, finally moderate fat infiltration and a few hypertrophic fibers).

Differential Diagnosis

Progressive dystrophy begins at a much earlier age than spinal progressive muscular atrophy, is usually a family disease, leaves the hands uninvolved, is often accompanied by pseudo-hypertrophies, never with fibrillary contractions, etc.; neural progressive muscular atrophy begins in childhood at the peripheral ends of the upper as well as of the lower extremities. does not extend to the roots of the extremities, is a family disease.

The professional pareses (see above, page 3), drummer's paralysis, cigarmaker's paralysis, etc., can lead to an "individual atrophy" of the small hand muscles, which, however, the cause ceasing to act, soon present not a progressive, but rather a regressive character. On account of the frequent unilateral commencement of spinal muscular atrophy, these conditions must always be kept in mind in differential diagnosis. The same remarks apply also to certain rare forms of lead paralysis, in which the small hand muscles are affected with paralysis and atrophy before the radial muscles. The antecedents of the patient (occupation, lead colic) and the objective signs of saturnism (blue line on the gums, tremor, hard pulse, anemia) are to be looked for. Still other intoxications (arsenic, alcohol, bisulphide of carbon) produce similar clinical pictures.

The differential points between this disease and amyotrophic lateral sclerosis, syringomyelia and spinal gliosis, spondylitis and hypertrophic cervical

pachymeningitis are furnished by the difference in the reflexes and in the sensibility. They will be described in connection with these diseases.

At this point we might also consider the so-called "chronic anterior poliomyelitis," a disease whose separation from spinal progressive muscular atrophy is still sub judice. It is a rare disease of mature age, which is possibly of autotoxic origin—for example, a number of cases in diabetics have come under observation. The disease begins usually in the legs, more rarely in the muscles of the shoulder-girdle, as weakness which, little by little, passes over into paralysis and progresses by no means so slowly as the *Aran-Duchenne* disease. Extensive segments of the extremities are attacked simultaneously, reaction of degeneration and paresis or paralysis precede the atrophy, and its extension is not by leaps. The disease can come to a standstill, indeed recoveries have been observed. On the other hand a relatively rapid fatal termination (in from 1 to 3 years) as a result of asphyxia, aspiration pneumonia, etc., is to be feared.

Prognosis and Treatment

The prognosis of progressive spinal muscular atropny as regards recovery or even as to definite standstill, is altogether bad. Nevertheless the progression of the disease is almost always a very slow one extending over decades, and a direct menace to life through bulbar paralysis or respiratory failure is comparatively rare. The treatment can only be inspired by the hope of slowing and stopping the process for a time and of supporting functional compensation. Moderate and careful exercise and massage of the muscles not yet affected is the most important thing; also labile electrical treatment with weak currents, the cathode on the affected muscles, can act favorably. I would warn you, however, against all energetic measures, also against faradization, and advise you above everything to leave the already affected muscles entirely at rest. Further, galvanization of the spinal cord, the technique of which will be considered in the next lecture will come into consideration. As to drugs, strychnine injections (0.0015 (gr. 1/40) once a day) may be tried according to *Gowers'* recommendation; the formerly much-vaunted silver nitrate and ergot cures are certainly entirely useless. Bath cures and hydrotherapy also accomplish nothing; when the patient demands them one should limit himself according to the motto "non nocere," in that care is taken to avoid excesses, and above everything, cold water treatment.

LECTURE VII

While in the last lecture we have become acquainted with clinical pictures whose anatomical substratum has its location partly in the peripheral (spino-muscular) motor neurones, partly in their end apparatus the muscles, we will turn our attention to-day to degenerative affections in which the central (cortico-spinal) motor neurones are affected by the disease process. Since this occurs with avoidance of neighboring but physiologically different fiber systems, that is in elective manner, we speak of "system diseases" in contrast to the "diffuse diseases" of the nerve centers (as, for example, inflammatory affections, multiple sclerosis, syringomyelia, etc.).

We will begin with the study of the so-called spastic spinal paralysis, although this clinically very typical disease picture, as I will remark in advance, is not to be considered as the expression of a unique and definite disease of the spinal cord, but rather presents a syndrome which occurs in the course of a heterogeneous, but without exception, organic set of spinal affections, either to establish itself definitely or to pass over into other symptom complexes.

Spastic Spinal Paralysis

In 1875 and 1876 *Erb* and *Charcot*, independent of one another, under the respective designations of "Spastische Spinalparalyse" and "Tabès dorsal spasmodique," described for the first time, a disease condition which they correctly suspected, and which we to-day know certainly, to depend upon more or less symmetrical lesions of the spinal pyramidal tracts, in whose territory cortico-spinal conduction is broken. Although indeed it was originally assumed that this must be always an elective degeneration, a system-disease, of that neurone, later experience has proved the untenableness of this view; so, for example, compression of the spinal cord (by a tumor an exotosis of the vertebral column, etc.) can manifest itself for a long time only by symptoms of involvement of the pyramidal tract, in other words by the phenomena of spastic spinal paralysis. The same thing occurs in "multiple sclerosis," which is characterized by the dissemination of areas of proliferated glia throughout all the nerve centers, also in many brain diseases, for example in internal hydrocephalus. It hence seems as if the pyramidal tracts, mainly in their distal portions, are easily disturbed in their integrity by the most varied pathological conditions.

On this account one must always be aware that in making what is, as we will see, the exceedingly easy diagnosis of "spastic spinal paralysis" he is mentioning only a syndrome, not a disease, and should investigate industriously

and carefully further, if new symptoms do not present the situation in another light. The clinical picture usually unfolds itself so slowly and gradually that only after years does it unmask itself as an expression of one of the disease conditions mentioned above. There remain finally, however, as *Minkowsky, Bernhardt, Dejerine, Schultze,* and others have shown, still cases which prove to be due to a purely systemic degeneration of the cortical-spinal tract in its spinal portion, a degeneration with which a decided reactive or compensatory glia proliferation (sclerosis) goes hand in hand. Indeed, we have learned through *Strümpell* that true spastic spinal paralysis can occur as a hereditary family disease, a proof that it may depend upon endogenic factors. A reduced power of resistance of the pyramidal tracts from the start, perhaps also plays a rôle in those forms which are set up by exogenic factors; for example, anemia, carcinosis, syphilis, the puerperium, lead poisoning, lathyrism (chronic intoxication by the use of spoiled chick peas—lathyrus cicera and L. sativa). All these factors, however, frequently lead, instead of to isolated pyramidal degeneration to simultaneous involvement of the lateral and posterior columns, the so-called system diseases in which motor and sensory phenomena are united. This last is also the case in pellagra, an intoxication from spoiled corn-flour endemic in Lombardy, in Roumania [and of recent years observed in the United States—*Translator*].

Symptomatology

In full development of "spastic spinal paralysis" there are in both legs, only in a small number of cases also in the arms, the anomalies which we denominate the spastic symptom complex, and which pathologically are to be referred to the elimination of the function of the pyramidal tracts in a longer or shorter portion of their spinal course.

Physiology of the Pyramidal Tracts

The pyramidal tracts or cortico-spinal tracts have, as is known, their origin in the motor region of the cerebral cortex. They pass through the internal capsule, the crus cerebri and the pons to the medulla. There a separation takes place into the lateral pyramidal tract which passes, by way of the "decussation of the pyramids," into the opposite lateral column of the spinal cord, and the anterior pyramidal tract which descends on the same side in the anterior column. Both categories of fibers end about the cells of the anterior horn at different levels of the spinal cord, those of the anterior pyramidal tract, however, only after they have passed through the anterior commissure of the spinal cord, that is, have crossed to the opposite side.

Through these pyramidal tracts, now, the psycho-motor impulse is transmitted from the cortical motor centers of the cerebrum to the spinal centers of the different muscles of the body. But not exclusively by this path. Rather are these chief motor tracts assisted in their task by a number of accessory motor tracts, by fiber tracts which I have included under the name "sub-cortico-spinal tracts"; their points of origin (the red nucleus, the optic thalamus, the

roof of the mid brain, *Deiters'* nucleus) are connected with the motor region of the cerebral cortex, so that, thanks to these last, even on elimination of the pyramidal tracts, part of the psychomotor impulses can be transmitted in a roundabout way to the cells of the anterior horn and hence to the muscles.

Now, however, the pyramidal tracts also exercise an influence upon the important mechanism of the regulation of the tonus already mentioned and also upon the production of the tendon and bone reflexes. They exercise an inhibition, in that they take care that reflex action and the degree of tonus does not go beyond a definite and useful degree. Without this inhibitory action of the pyramids, tonus and reflexes would not present such constant relations as is actually the case. If, however, the pyramids are destroyed, hypertonia and hyperreflexia take the upper hand to a degree hindering motion, so that the muscles cannot be sufficiently relaxed and even the mere jarring of the body in locomotion suffices to set up reflex contractions.

After this short physiological introduction, it is understandable that in consequence of the vicarious action of the subcortico spinal tracts* the affected extremities are not entirely paralyzed, but only more or less paretic; this paresis, however, affects nearly all the muscles of the limbs in question, to the same extent. In the paretic muscles a spastic rigidity is present, as we can easily convince ourselves by palpation, by testing resistance and by passive movements. On the other hand there is no atrophy of the affected muscles—apart from an atrophy from disuse which occurs late and is never very great—as the muscles remain in spite of the pyramidal affection, in uninterrupted connection with their trophic centers, the cells of the anterior horn. For the same reason, along with anatomical integrity of their structure, the muscles preserve their electric irritability.

The gait of such patients, in consequence of the paresis and rigidity of the legs, is altered, in very characteristic fashion. Since the rigidity is especially marked in the glutei, the quadriceps, the adductors, and the gastrocnemius, the leg is extended rigidly at the knee and hip, the thigh is held in adduction, the heels are raised from the floor so that the patient sometimes walks only on the balls of his feet, and in order to prevent falling backward, must shift his center of gravity forward, which forces him to make use of a walking-cane. Locomotion is carried out with short, audibly dragging and careful steps; since the joints are held by the muscular rigidity in a condition of extensive "muscular ankylosis" (*Strümpell*) the feet can scarcely be raised from the floor, but must alternately be swung forward in a lateral arc one around the other ("helikopodia" "circumduction"). Sometimes a "see-sawing" of the body at every step is noticeable. In long duration of the disease, the anomaly of position of the leg is more and more fixed, on account of structural shortening of the hypertonic muscles so that a true pes equinovarus can develop. Only exceptionally are there contractures in flexion at the knee and at the hip, an occurrence that puts an end to the patient's ability to walk.

In the rare "ascending" cases of spastic spinal paralysis, in which after the

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* Besides this, usually only the lateral pyramidal tracts, not the anterior pyramidal tracts, are affected.

legs, the upper extremities are also affected, the arms, on account of the unequal distribution of rigidity and paresis, usually take the following position: adduction, flexion at the elbow, wrist, and finger joints, pronation.

The "spastic symptom complex" is completed by anomalies of reflexes and associated movements. The exaggeration of the tendon and bone reflexes in the region of the paretic muscles (which is, pathogenetically, the analogue of hypertonia) is usually very great. At the same time as this hyperreflexia in the affected musculo-tendinous apparatus, there can be demonstrated the phenomenon known as "clonus" ("trépidation épileptoïde" of the French). For example, if the tendo Achillis is suddenly stretched by pushing up the anterior portion of the foot, rhythmical oscillations of the foot, which last as long as the tension of the tendon is kept up, are produced. This is by far the most frequent clonus—"ankle clonus." "Patellar clonus" cannot infrequently be

Fig. 39.

The *Babinski* Reflex (instantaneous photograph).

produced by suddenly drawing down the patella and attempting to hold it in this position. On the other hand "wrist," "finger" and "elbow clonus" are rarely demonstrable. The behavior of the skin reflexes is much less constant than that of the tendon reflexes; the plantar reflex is found exaggerated with a certain degree of regularity, but the degree of reflex reply to skin irritation is very difficult to estimate as to its pathological indications. All the more valuable to us are certain abnormal skin reflexes which occur when the pyramidal tracts are interrupted. Of the first importance is the *Babinski* phenomenon, in which irritation of the sole of the foot causes a rather slow, tonic hyperextension of the great toe (see Fig. 39), sometimes with accompanying plantar flexion or fanlike spreading of the other toes. In the second place stands the *Oppenheim* reflex, in which the same reaction is produced by firmly stroking the skin on the inner surface of the leg. A pathological tendon reflex is the *Mendel-Bechterew* phenomenon ("dorsal foot reflex"); in lesions of the pyramidal tracts

percussion on the lateral part of the dorsum of the foot (over the 4th and 5th metatarsal bones) may cause plantar flexion of the toes; normally there is either no reflex or extension of the toes.

That the "pathological reflexes" occur normally in children in the early months of life, depends most probably upon the fact that the pyramidal tracts develop their full function only after their axis-cylinders have taken on their medullary sheaths in the course of the early part of the post-embryonal period. A satisfactory pathological explanation of these phenomena is wanting. We can at most conceive that the reflex irritation, if it is not inhibited from above (that is, from the cerebrum) breaks into paths otherwise closed to it. The same may be the case with the voluntary impulse intended to produce a definite movement of the paretic muscles so that it comes to set up unintended "associated movements." When, for example, the leg cannot be drawn up on the body, without decided dorsal flexion of the foot occurring at the same time, we speak of the tibial phenomenon or "*Strümpell's* sign"; compulsory hyperextension of the big toe under the same conditions furnishes the toe phenomenon; in the hand, the radial phenomenon occurs (compulsory dorsal flexion of the hand with palmar flexion of the fingers), further the pronation phenomenon (compulsory pronation when the forearm is bent), etc.

Sensory, vasomotor and trophic disturbances, also anomalies in the emptying of the bladder and rectum, as well as those of the sexual functions, do not belong to the picture of spastic spinal paralysis.

Course and Prognosis

The disease begins, as a rule, gradually and little marked, at first with only subjective, often but temporary difficulties; after long walking the patient feels weakness and sense of tension in the legs, symptoms which can increase to a "spinal intermittent limping" (*Dejerine*) which, however, for a long time disappear again when the patient is rested. Only later do they become permanent and then the objective symptoms of the disease grow more and more plainly apparent. If the clinical picture does not, as is usually the case, unfold itself as the initial stage of some other disease more rich in symptoms, but definitely restricts itself to the spastic symptom-complex, the progression is usually a very slow one, so that the patients may remain capable of movement for ten or twenty years or more, indeed, it may come to a stop when the disease has made relatively but little progress. The prognosis as to life is hence not unfavorable in the true spastic spinal paralyses, which usually commence between the ages of 20 and 40 years; only when the patients (generally on account of contractures in flexion of the legs) have become bedridden, it is impaired by the danger of hypostatic pneumonia. As to the prognosis of the symptomatic form, no general statement can be made. It depends naturally upon the nature of the disease causing the syndrome "spastic spinal paralysis"; for example, where it depends upon compression of the cord by a tumor which can be removed by operation, cure is possible.

Treatment

In the very first place it is necessary to warn these patients against exhausting gymnastic exercises in which many of them are inclined to seek cure, and to make clear to them the necessity of prolonged rest and self-care. The rigidity can very often be alleviated by careful massage and passive movements; protracted tepid baths work in the same way. The practice of active movements is best carried out in the bath, since the influence upon their weight which the limbs experience on account of the buoyant effect of the water (this can be increased by the addition of salt) aids greatly in the work to be accomplished, while on the other hand the diminution of the stimuli from the skin acts to some extent in reducing the tonus. On account of the long duration of the disease drugs should be used but sparingly; though occasional courses of bromides or scopolamin (for dosage see *Parkinson's* disease, page 89) can be recommended without hesitation. Faradization is strongly contraindicated; if stabile galvanization of the rigid muscles or their nerves or the "galvanization of the spinal cord" (two large electrodes, one on the neck and one in the sacral region, gradual introduction of a current of a few milleamperes, let it pass for 5 minutes, slowly reduce it, change the poles and repeat the procedure) is of any use is questionable; in any case it does no harm. In contractures, orthopedic surgery is to be considered (tenotomy, supportive apparatus). The most modern surgical procedure recommended for spastic paresis is "Rhizotomia posterior" proposed by *Förster*. Based upon the idea that the production of tonus depends upon peripheral stimuli which are transmitted through the posterior roots of the spinal cord to the cells of the anterior horn (see page 8) several pairs of these roots are resected in order to make movements more easy by getting rid of the rigidity. We will consider this operation again later in describing *Little's* disease, where it may have a good result (provided it is followed by thorough and conscientious gymnastic after-treatment). In spastic spinal paralysis, on the other hand, it seems to promise less. In the only case with which I am acquainted from my own observation, it remained without effect.

As to the causal treatment of the forms dependent upon exogenic factors, we need not consider it further at this point; the therapy of nervous syphilis will find a connected description in a later lecture.

Amyotrophic Lateral Sclerosis and Progressive Bulbar Paralysis

These names have become attached to two symptom-complexes which do not represent different diseases at all, but only describe a different localization of one and the same affection; in the one case, in the spinal, in the other, in the bulbar nerve centers. This affection, however, presents itself anatomically as a primary, progressive degeneration of the whole motor tract due to endogenic factors (though occurring as a hereditary family disease only in a continually decreasing number of cases).

Parenchymatous degeneration of the neurones with reactive proliferation of the supportive tissues lies at the base of both diseases. In amyotrophic

lateral sclerosis these changes affect the cells of the anterior horn of the spinal cord and their processes, in progressive bulbar paralysis the motor nuclei of the medulla (the homologues of the anterior horn cells). Besides this, the fibers of the pyramidal tracts going to the diseased spinal and bulbar nuclei (cortico-spinal and cortico-bulbar) degenerate throughout their whole extent.

Indeed, the motor intersegmental fibers of the antero-lateral tract in the spinal cord and the cortical fibræ propriæ in the cerebrum have been found altered.

Both disease forms are rather rare, and in the great majority of cases occur between the 30th and the 55th years of life, only very exceptionally in youth or in childhood; amyotrophic lateral sclerosis on an average, earlier than bulbar paralysis, the first beginning mainly in the 4th, the last in the 5th or 6th decades. As exciting causes exposure to cold, exhausting diseases and overexertion, have been noted here and there. It is often found in amyotrophic lateral sclerosis that these have affected particularly the hand in which the symptoms of the disease begins. Here and there progressive bulbar paralysis also has occurred in connection with severe fatiguing of the muscles of the lips, cheeks, and tongue (in players of wind instruments and glass blowers). Any special preference of one sex over the other is not apparent. The unity of the two disease forms is indicated by the fact that every amyotrophic lateral sclerosis sooner or later is combined with progressive bulbar paralysis, provided that the patient is not earlier removed by some intercurrent disease. That this statement is not reversed, that is, that progressive bulbar paralysis may occur without spinal symptoms, is explainable from the fact that it puts an end to life, and hence to the further extent of the degenerative process, relatively early.

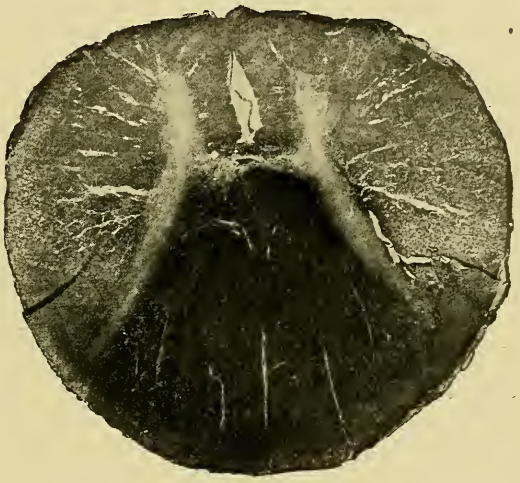


Fig. 40.

Alteration of the Spinal Cord in Amyotrophic Lateral Sclerosis. *Weigert-Pal* Stain.

Symptomatology

In sketching the symptomatology we will take as an example a typical case which, beginning with spinal symptoms, ends with bulbar involvement, and which has been denominated as *Charcot's* disease, after the great neurologist who described it for the first time in 1865.

As to the name amyotrophic lateral sclerosis, proposed by *Charcot* himself, it characterizes in a happy manner the clinical picture which represents the combination of two syndromes already studied by us, namely, 1, that of spinal

progressive muscular atrophy (the expression of degeneration of the peripheral motor neurone), and 2, that of spastic spinal paralysis (that is, pyramidal tract—or lateral sclerosis, an expression of the degeneration of the central motor neurone).

The disease begins in the upper extremities and almost never in both at the same time. The right hand usually shows the first anomaly and in the progress of the disease the symptoms generally predominate for a considerable time on the one side. The patient first notices a rapid tiring and a feeling of weakness, awkwardness and stiffness, often also of tension, particularly in the finer movements; soon objective anomalies appear, before anything else a progressive wasting of the small muscles of the hand. The abductor pollicis brevis usually atrophies first, then little by little the other muscles of the ball of the thumb, the hypothenar muscles and the interossei. The different deformities which we met in the last lecture in spinal muscular atrophy occur here in an analogous manner, namely the "ape-hand," "claw-hand," and "skeleton-hand." On the other hand, the paretic symptoms are much more marked than in the *Aran-Duchenne* disease, and the paresis is accompanied with such manifest hypertonia that in the early stages of the disease there may be active contractures which fix the finger and wrist joints in flexion. Also spastic adduction of the arm, flexion and pronation of the forearm occur. However, the spasticity in the upper extremities diminishes again with the progressive atrophy (this soon affects also the extensor side of the forearm and shoulders).

As long as the muscles needed for their production are present, exaggeration of the tendon and periosteal reflexes are to be found; now and then wrist clonus also can be elicited. In the atrophic muscles fibrillary contractions are very manifest. As in progressive spinal muscular atrophy and on account of the same conditions (namely, the gradual progress of the degeneration from fiber to fiber in the individual muscles) reaction of degeneration is found only in places (particularly in the thenar muscles) and only in incomplete development; in general, faradic and galvanic irritability are reduced in proportion to the degree of muscular wasting.

For a comparatively long time, the lower limbs remain normal except for a marked increase of reflexes. Little by little, however, the patient experiences a weakness and tension in the legs; also the whole spastic symptom-complex, as we have already described it (see page 112), develops. (Only the *Babinski* and *Oppenheim* reflexes are comparatively frequently absent.) Here, in contradistinction to the arms, the amyotrophic component is entirely subordinate to the spastic one and sometimes atrophy of the muscles of the leg and foot occurs only late in the disease.

Trophic, sensory, and sphincter disturbances are absent during the whole of the disease. On the other hand, the last stage of the inexorably progressive affection is a period of grave increase in the symptoms on account of the addition to them of progressive bulbar paralysis (glosso-labio-pharyngeal paralysis). The first description of this last we owe to *Duchenne* (1860).

Its prodromal symptoms are: Slight difficulty in speaking and in the other movements of the tongue, occasional difficulty in swallowing, inability to whistle, etc. Little by little the disturbances of function in the muscles of the tongue

and lips, of mastication, of swallowing and of the respiratory muscles become more marked. The speech becomes continually more inarticulate. The pronunciation of the linguals, particularly of r and l, is first affected; soon after that of the labials (f, w, p, b). Among the vowels, o and u, whose production is favored by the contraction of the orbicularis oris, are worst articulated. On account of involvement of the muscles of the velum palati there is difficulty in the formation of the gutturals (g and k); this also gives the voice a nasal sound, since the nasal cavities can no longer be shut off from the pharynx and act as resonance chambers. Besides this, the speech becomes monotonous on account of weakness of the larynx muscles. The paralysis of the velum palati, as well as that of the other pharynx muscles, causes appreciable disturbances in swallowing; food, particularly that of a liquid character, easily gets into the larynx and produces attacks of choking, or it regurgitates through the nose. Chewing, on account of the progressive paralysis of the masseter and temporal muscles, becomes more and more difficult; the involvement of the internal pterygoid makes itself evident, in inability to draw the jaw laterally in order to grind the food. The taking of nourishment is rendered difficult by still other factors; the tongue can no longer shove the bolus between the teeth and throw it back again, on which account it remains caught between the gums and the cheek; also, it cannot form a trough to direct the fluids into the gullet, hence the patient when drinking must throw the head backward. The saliva also is imperfectly swallowed and flows out of the corners of the mouth.

Objective examination shows that the functionally so seriously affected muscles are also attacked by degenerative atrophy, although, corresponding to the condition of the small muscles of the hand, reaction of degeneration is to be found, as a rule, only in a few muscles (lips and tongue) and scarcely ever in complete development. The tongue is flattened, markedly wrinkled, as if its coating of mucous membrane had become too large for it, and is the seat of marked fibrillary contractions. These last are usually very evident in the lips also, while here the atrophy is usually concealed on account of fatty deposit. The velum palati hangs down like a thin, sagging curtain. The temporal fossa presents a depression, and occasionally fibrillary contractions are to be observed in the masseters.

The masseter reflex is usually exaggerated. The palate and pharyngeal reflexes are almost always diminished or lost. The face of the victim of bulbar paralysis presents a characteristic change. The lower part in contradistinction to the expressive eyes and perfectly movable forehead, is of mask-like immobility; the mouth, on account of the paresis of the orbicularis and the contracture of its antagonists, is half open, and extended laterally ("rire transversal") or its angles hang down deeply, which gives to the physiognomy a lacrimose expression. The skin over the emaciated muscles of the cheeks and chin shows exaggerated folding, only very exceptionally is the upper facial region affected.

This condition, which the mental clearness renders specially shocking, finally terminates either by vagus paralysis (permanent severe tachycardia, heart weakness) or paralysis of the diaphragm. Every bronchitis, besides this, contains in itself the greatest danger to life, since coughing and expectoration

are impossible. Also, the danger of aspiration of particles of food into the lungs cannot indefinitely be avoided in any of these patients.

Course and Prognosis

By far most frequently we see *Charcot's* disease begin in the small muscles of the hand and develop further in the manner sketched above; only very exceptionally are the muscles of the shoulders attacked before those of the hand (scapulo-humeral form). Another rare method of beginning is that the spastic paralysis of the lower limbs precedes the spastic atrophic manifestations in the upper ones. This is also a further proof that spastic spinal paralysis in its later course presents itself as only a partial symptom of another disease process.

Finally, we have as a third variety, the cases which begin at once with bulbar symptoms. In only a very small number of these cases an extension of the motor disturbances to the muscles of the neck, the shoulders and the upper extremities occurs; most of them die too soon.

The prognosis both as to life and as to recovery is absolutely unfavorable. Death occurs in from six months to ten years. Relatively the best prognosis is given by the form beginning as paraplegia; by far the most unfavorable, by primary progressive bulbar paralysis, of which no case lives beyond the fifth year of the disease, only few beyond the second year.

Differential Diagnosis

In amyotrophic lateral sclerosis, the resemblance at the start to progressive spinal muscular atrophy is great; the hyperreflexia as well as the tendency to contracture, however, enable us to exclude this as well as the other varieties of progressive muscular atrophy; also the progress of the atrophy is much more rapid in *Charcot's* disease than in that of *Aran-Duchenne*. We have already alluded to the liability to confusion of the paraplegic form of amyotrophic lateral sclerosis with spastic spinal paralysis, an error from which reservation of opinion until the disease has developed further, will protect us. Finally, it must be mentioned that an affection of the spinal cord which we will consider in the next lecture, namely, syringomyelia, may begin with atrophy of the small muscles of the hand and spastic phenomena; however, there are always disturbances of sensibility peculiar to it, which are without exception absent in *Charcot's* disease.

As to the glosso-labio-pharyngeal paralysis, an acquaintance with the clinical peculiarities of myasthenia (see page 94) will permit distinguishing the two diseases without difficulty. Similar pictures are given also by supranuclear pseudo-bulbar paralysis (see Lecture XV) which occurs through symmetrical interruption of the neurones passing from the cortex to the motor nuclei of the bulb, from foci of disease of arteriosclerotic origin. These occur, however, in connection with repeated minor apoplectiform attacks and there is neither muscular atrophy, reaction of degeneration, nor fibrillary contractions, but usually disturbances of intelligence as well as those in the function of the bladder.

Further, disease pictures resembling those of bulbar paralysis occur in

neuritis of the motor nerves arising from the medulla (for example, after diphtheria), also from acute inflammatory febrile processes (polioencephalitis inferior), from hemorrhages and softening of the medulla (embolism and thrombosis of the basilar artery) beginning acutely, and finally from compression of the medulla (through tumors, caries of bone, etc.). In these last, however, there is never complete symmetry on the two sides; besides, there is local sensitiveness to pain, often headaches and vomiting. Also further symptoms may be found, as, for example, involvement of the sensory portions of the trigeminus, of the acusticus, etc.

We must consider here "superior bulbar paralysis" or the ophthalmoplegia chronica progressiva of *Graefe*, which presents an analogue of progressive glosso-labio-pharyngeal paralysis and may be combined with this. This is characterized by a constantly progressive, gradual atrophic paralysis of the different eye muscles which, so long as it is not combined with glosso-labio-pharyngeal symptoms, naturally does not threaten life. Much more frequently, indeed, progressive ophthalmoplegia is not a definite primary degenerative disease like progressive bulbar paralysis, but a part of different organic diseases of the brain and spinal cord, as, for example, lues cerebrospinalis, tabes dorsalis, dementia paralytica, sclerosis multiplex, etc. There are also neuritic ophthalmoplegias; further, acute or subacute attacks arising upon an inflammatory basis (polioencephalitis superior) as well as those which are caused by local processes in the brain and at the base of the skull (for example, meningitides, tumors, etc.).

Treatment

The treatment of amyotrophic lateral sclerosis comprises that of its components, spinal progressive muscular atrophy and spastic spinal paralysis. In progressive bulbar paralysis, arsenic, iodide of potassium and strychnine are given, although a favorable, or even a palliative effect of these medicaments has not been recognized with certainty. Of electro-therapeutic procedures galvanization transversely through the medulla oblongata (electrodes the size of a dollar upon both mastoid processes, two milleamperes current strength, slowly introduced and reduced again), as well as the production of swallowing movements, are recommended. (Anode on the back of the neck, cathode moved up and down on the side of the neck.) We can counteract the flow of saliva by atropin, scopolamin, or duboisin in doses of $\frac{1}{2}$ mg. (gr. $\frac{1}{120}$). As long as swallowing is appreciably impaired the nourishment should be half liquid and should contain as many calories in as small a volume as possible (nutritive preparations, meat-juice, etc.). In advanced stages tube feeding must be resorted to.

In order to aid you in obtaining a comprehensive view of the different forms of primary degeneration of the motor tracts as well as of the muscles as we have learned them in the last two lectures, I have prepared for you a synoptic table showing these affections and their most important differential characteristics.

Seat of the Lesion	Author	Beginning	Age of Onset	Heredo-famil.	Atrophy	Reflexes	R. D.	Fibrill. Contract
A. Central Neuron								
1. Spastic Spinalparalysis — Lateral-sclerosis — "Tabes dorsal spasmodique"	Erb-Charcot	Lower Extremities	Adult age, rarely in childhood	Rarely	0	Increased	0	0
B. Peripheral Neuron (Including the Muscles)								
2. Progressive Muscular Atrophies								
a) Myopathic ("Dystrophy," "Primitive Myopathy")								
α) Pelvifemoral Forms								
1. Atrophic variety	Leyden-Mobius	Pelvic Girdle, Thigh	Childhood	Usually	+	− or 0	0	0
2. Pseudohypertrophic Variety	Duchenne-Griesinger							
β) Scapulohumeral, Juvenile Form	Erb	Shoulder Girdle, Arm	Puberty (Rarely later)		+	− or 0	0	0
γ) Facioscapulohumeral, Infant. Form	Landouzy-Dejerine	Face, Shoulder Girdle, Arm	Childhood		+	− or 0	0	0
b) Neural ("Peroneal")	Charcot-Marie, Tooth	Ends of the Extremities	Childhood (Rarely later)		+	− or 0	Incomplete	Often
c) Spinal								
α) Classic Form	Aran-Duchenne	Small Hand Muscles (rarely shoulder)	Adult Age	Rarely	+	− or 0	Incomplete	
β) Infantile Variety	Werdnig-Hoffman	Trunk, Thigh	Childhood	Usually	+	− or 0	+	
C. Central and Peripheral Neuron								
3. Amyotrophic Lateral Sclerosis	Charcot	Small Hand Muscles (Rarely lower extremities)	Mature Age (Rarely earlier)	Rarely	+	Increased. Later lost.	Incomplete	+
4. Progressive Bulbar Paralysis	Duchenne	"Glossolabial-pharyngeal"				Usually increased		

LECTURE VIII

The Hereditary Family Ataxias

Gentlemen: Since we have in the preceding lectures become acquainted with the dyskinesias, the progressive muscular atrophies, spastic spinal paralysis, amyotrophic lateral sclerosis and progressive bulbar paralysis, a series of disease conditions which characterize themselves by preponderant hereditary-family occurrence, we will consider to-day some specially interesting representatives of the heredo-degenerative nervous diseases. These are the hereditary family ataxias which since their clinical groundwork was laid by the great Heidelberg clinician *Friedreich* (1861-1863), have always held medical interest; of late, however, have gained in actuality in that alongside of the typical clinically and anatomically well-founded picture of "*Friedreich's* disease" certain other varieties have become known. We will first make the acquaintance of the classical type of the disease.

I. Friedreich's Disease (Spinal Heredo-ataxia)

Symptomatology

The fundamental symptom of this disease, which begins, as a rule, in childhood, is static and locomotor incoördination which, from a scarcely noticeable beginning, progressively increases to the highest degrees of intensity, and almost without exception affects first the legs, only later the arms. The parents of such a child, which usually has learned to walk early and well, but as time progresses begins to complain of weariness in the lower extremities, notice that little by little its gait becomes uncertain, spraddling and jerky. Soon a "wabbling in the loins" is added to this and the gait comes more and more to resemble that of a drunken man. Finally, the excessive degree of this ataxia no longer permits locomotion and the patient becomes permanently bedridden. At the same time as the locomotor, static incoördination has also developed: even when sitting quietly, the body sways from side to side, each limb when held free, and also the head, oscillates irregularly.

This ataxia is, as we may assume from the description of the pathological anatomy of the disease, the clinical corollary of degeneration in the posterior columns and in the spino-cerebellar tracts of the spinal cord. Both these systems conduct impulses, which belong in the category of deep sensibility (see page 7), and which have come by way of the peripheral nerves through the posterior roots into the spinal cord, toward the brain. While now one portion of these impulses passing up into the posterior columns and proceeding by way

of the optic thalamus to the cerebrum exerts a quite general movement regulating action, the second portion which passes by way of the direct cerebellar tract and column of *Gowers*, to the cerebellum, enters into the special function of maintaining the equilibrium on standing and walking. The incoördination in *Freidreich's* disease is now a mixed form of these two types of ataxia. The one component, which is to be considered as cerebellar in nature, manifests itself by severe disturbances of equilibrium which we have just mentioned, through staggering and wabbling on locomotion, through swaying of the body also while sitting quietly. We will have an opportunity to further consider these symptoms when discussing diseases of the cerebellum.

Exceedingly frequent further, are choreiform movements, which, however, are of much less intensity than those in true chorea. Now it is a constant playing of the fingers, again an unrest of the muscles of the neck and face. I have occasionally noted it only in the upper lid. Athetoid movements have been described as a rare occurrence.

In isolated cases, muscular paralyses have developed in the extremities of patients with *Friedreich's* disease. It appears to me quite doubtful that these are uncomplicated cases of the disease, since in one such case of my own I was able to recognize anatomically the combination with progressive muscular atrophy, while, on the other hand, neuritic processes have been found. Again, speech disturbances are among the most regular phenomena in spinal heredoataxia; in advanced cases they are almost never absent. The words are brought out slowly, with difficulty, and often in a slightly scanning and explosive manner (hesitating speech). A frequent changing into falsetto (bitonality) has been pointed out by *Dejerine*, *Thomas* and myself. *Soca* has noted disturbances of articulation in the pronunciation of certain letters, namely l, k, v and i.

Among disturbances of the eye muscles, nystagmus takes the first place on account of its great frequency. In the early stages of the disease, however, it is generally absent. Usually it appears after from 3 to 5 years' duration of the affection, sometimes only later; in but very few cases is it absent entirely. As a rule, it is a dynamic horizontal nystagmus, that is, made up of rhythmical contractions of both eyes which occur in the horizontal plane and are set up by looking to one side. Almost never, on the other hand, do we find a static nystagmus, that is, one present during rest. Paralyses of the eye muscles are exceedingly rare in *Friedreich's* disease.

The tendon and bone reflexes are diminished in the early stages, entirely lost later. The reflexes in the lower extremities disappear first, in the upper extremities only some years later. In almost all cases *Babinski's* foot phenomenon is present in typical form, occasionally a slow hyperextension of the big toe can be produced also by stroking the inner side of the leg (*Oppenheim's* reflex). In general the skin reflexes are undisturbed, as well as those of the mucous membranes and of the pupil.

Particularly interesting are certain deformities which are typical accompaniments of *Friedreich's* disease. A peculiar change in the form of the foot in the great majority of cases early develops; the dorsum curves upward so that the foot assumes a short, compressed and concave form (pes cavus); the toes, particularly the great toe, become permanently held in dorsi-flexion at their

metatarsophalangeal joints, the extensor tendons of the great toe give the impression of being considerably shortened and stand out on the back of the foot; finally, the foot assumes a more or less pronounced equinus position (see Fig. 41). Along with this typical deformity, there are other less marked varieties of the "*Friedreich's* foot." In the production of this deformity the "balancing" action, the continual contraction on the one hand of the tibialis anticus and posticus muscles, and on the other of the muscles of the calf and sole, due to the ataxia in gait, plays a great part; besides this—as the dorsal contraction of the great toe, which may be considered as a permanent *Babinski* reflex, indicates—the continuous irritation of the sole on standing and walking contributes to this too. I have seen this hyperextension disappear entirely again after the patient had become bedridden. Besides this, a more or less marked curvature of the vertebral column occurs in time in most cases. This is mainly a scoliosis, though sometimes a kyphosis, more rarely a lordosis. A satisfactory explanation of this symptom has not yet been given. The same remarks apply to a deformity of the hand described by *Friedreich* as a rarity, which was indicated in one of our cases (permanent hyperextension of the thumb).

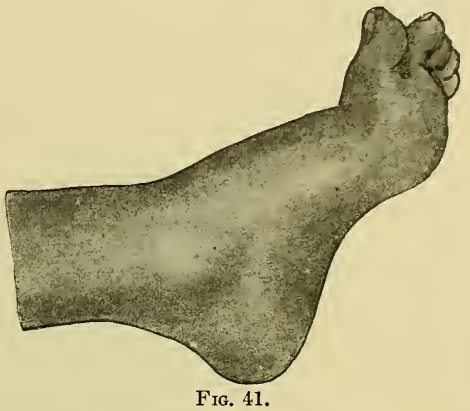


Fig. 41.

Friedreich's Disease. Typical Foot Deformity.

I will mention also some unusual symptoms of *Friedreich's* disease. These are mainly certain sensory disturbances. Diminution of the sensibility of the skin occurs only in the latest stages, for example, in one case I could trace its first beginning only after the disease had existed for 37 years. It occurs chiefly on the feet. The muscular sense and the vibration sense can be affected early, but in most cases remain long intact. In the patient mentioned above a reduction of stereo-esthesia in the right hand occurred first, with superficial hypesthesia in the feet. Among sensory irritative symptoms, *Bramwell* and I have described painful cramps of the calves. Many patients complain of attacks of rotary vertigo or even of a permanent state of dizziness of little intensity. Further, the disturbances of vegetative function which have been occasionally observed in *Friedreich's* disease should be considered. These are profuse salivation, polyuria, hyperidrosis, œdema, diabetes, dyspnœa, jerky respiration, disturbances in swallowing, late beginning of the menses. The sphincters of the bladder and the rectum practically always functionate correctly, at most, slight degrees of urinary incontinence occur. Impotence is never present. In conclusion the rare psychical disturbances must still be considered. One patient studied by me was imbecile to a high degree, at the same time vicious, impulsive, and given to coprolalia. Other authors have seen a combination with idiocy. Slight defect in intellectual development occurs somewhat frequently; usually, however, the thoroughly normal intelligence of such patients is in marked con-

trast to the weakminded appearance which they make in consequence of their stumbling speech and their often dull expression of countenance.

Pathological Anatomy

The spinal cord in *Friedreich's* disease is thin and small to a degree observed in no other disease; also, since the duration of the disease makes no difference in this, it is evidently not an atrophy but a hypoplasia of the organ.

We consider the columnar degenerations which we find in the spinal cord in *Friedreich's* disease as consequences of an arrest of development; these regularly affect the posterior columns, the direct cerebellar tracts, and the lateral pyramidal tracts, sometimes also *Gower's* columns and the anterior pyramidal tracts. Fig. 42 shows you the characteristic picture with the *Weigert-Pal* stain for the

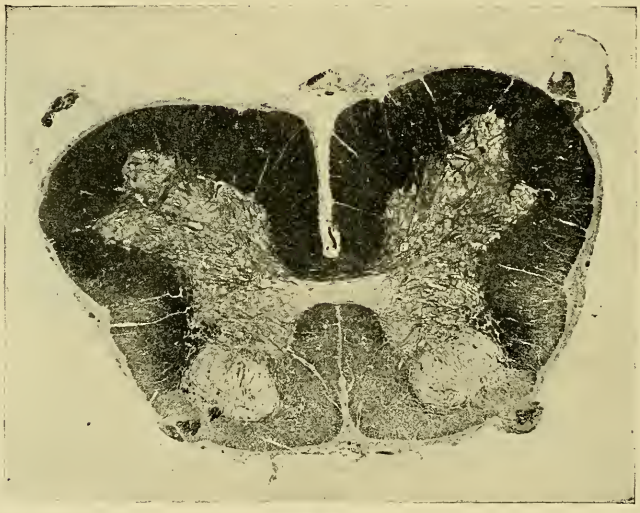


Fig. 42.

Alteration of the Spinal Cord in *Friedreich's* Disease. *Weigert-Pal* Stain.

medullary sheath. Only very exceptionally have these typical combined columnar scleroses been seen to develop in a well-formed and in no way hypoplastic spinal cord (*Ed. Müller*). The connection of the ataxia with the degeneration of the posterior columns and the spino-cerebellar tracts we have already brought out. Upon comparing the anatomical and the clinical pictures, however, the fact that in spite of extensive alteration of the posterior columns so little sensory defects are to be found appears paradoxical. It seems as if, in system diseases which affect the youthful organism the nervous conduction paths so organize themselves, that by a roundabout way through the gray substance the loss of the posterior columns is partially compensated. The substratum for nystagmus, choreic unrest and disturbances of speech is anatomically not yet made clear. Hypo- or areflexia appears to correspond to the loss of function of the fibers of the posterior roots. The degeneration of the pyramidal tracts usually manifests itself only by *Babinski's*, though sometimes also by *Oppenheim's*, reflex.

Course and Prognosis

The beginning of the disease, in the great majority of cases, falls between the 6th and the 15th years of life. An earlier commencement (3 to 4 years) is, however, not exactly rare, neither are cases which present their first symptoms at from 15 to 20 years. Unusual, however, are the "late forms" of the disease in which the initial symptoms appear in the third decade of life. As to the sequence of disease symptoms, they usually begin with ataxia of the legs and hyper-extension of the great toe; next the patellar reflexes disappear. The speech disturbance generally is not long delayed, neither is involvement of the arms, while nystagmus is postponed somewhat longer. Nevertheless, within 5 or 6 years after its beginning, the disease picture is usually present in a typical manner. Further progression occurs then, as a rule, much more slowly, so that the patients can live 3 or 4 decades longer, until some intercurrent disease (usually broncho-pneumonia) causes death. Remissions are rare, more frequent are sudden exacerbations in connection with intercurrent acute infections.

Etiology and Pathogenesis

In spite of the "erratic" cases which not so rarely come under observation, *Friedreich's* disease is in general characterized by the following criteria, as an essentially heredo-degenerative disease, corresponding to a pathological variety of the species. It attacks, as a rule, several members of the same generation (homologous heredity) and occurs in these mainly at about the same age (homochronous heredity); it is not to be referred to external factors during intra- or extra-uterine life (traumata, intoxications, infections); in such factors an exciting cause can at most be recognized. The injury has already affected the germ (endogenic basis); and finally from the moment of its beginning it is marked by unceasing progression. That *Friedreich's* disease not only as a disease of the single individual, but also as that of a line of descendants, manifests a progressive and severe degenerative character, is shown from the confirmed fact that from generation to generation the age at which it begins, becomes lower and lower. Also it can happen that the disease skips several generations to appear again (by atavistic reversion), which is denominated latent heredity. The most convincing example of latent heredity is furnished by a genealogical tree prepared by *Konrad Frey*, which refers to a large group of cases of *Friedreich's* disease in a village of the Swiss Jura. The author was able, by referring to the church records, to recognize the common descent of these patients from an ancestor of the sixteenth century. In the descendants of this man, distributed among six collateral lines, the disease, however, only appeared in the 11th or 12th generation, although in the intervening generations consanguine marriages with the loss of ancestors and a history of heavy drinking was often noted.

As signs of congenital defect of certain parts of the central nervous system may be mentioned the frequent accompaniment of *Friedreich's* disease by the most varied congenital defective conditions and malformations, for example, by hypospadias, facial asymmetry, "mongoloid" conformation of the face, anoma-

lies of the central canal of the spinal cord, ectopy of the gray matter of the spinal cord, etc. Or the combination with other heredo-degenerative diseases also occurs; so, for example, I have described a patient with hereditary ataxia and muscular dystrophy at the same time, and *Kollorits* has seen the combination of *Friedreich's* disease and *Huntington's* chorea. Now how this congenital defective condition ("abiotrophy" according to *Gowers*) leads to the gradual degenerative destruction of certain tracts, there may be different conceptions. *Jendrássik* writes: "In many families striking peculiarities occur. Some lose their hair prematurely, in others the pyramidal tracts degenerate." *Raymond* has spoken of "premature senescence," *Edinger* of the using up, I of the wear and tear of the congenitally defective systems. As causes for the first appearance of a heredo-degenerative disease in a definite family line, certain "germ injuring" or "blastophthoric" factors cannot infrequently be made responsible. In *Friedreich's* disease it is very often alcoholism in the parents (in many cases there is a spontaneous statement that the affected children were begotten while the father was in a drunken condition!) Further, consanguinity of the parents, advanced age, or great disproportion in their ages. As exciting causes of the disease we find exceedingly frequently the history of having passed through some exhausting disease (variola, typhoid fever, scarlatina, measles, pneumonia, pertussis, meningitis, influenza).

Treatment

Unfortunately we can in no way influence the disease process in *Friedreich's* disease. In order to delay its course as much as we can, it is indicated that the patients be placed as soon as possible under the best hygienic conditions and under continued medical care, for example, best in an institution. In every case care should be taken that they get plenty of bodily rest, best lying out in the fresh air, while the amount of daily exercise should be very carefully regulated so that overexertion, which is under all circumstances injurious, is avoided. By careful massage it should be endeavored to strengthen the muscles still capable of function and to effect some correcture of the foot deformity. In order to relieve the ataxia the *Frenkel-Leyden* compensation therapy which we will consider at length under the discussion of tabes in the next lectures, should be tried in every case; however, I have only once obtained a decided effect which was lasting. From time to time strychnine cures (for adults daily 0.002 to 0.005 (gr. 1/30 to gr. 1/12) subcutaneously, or 4 times a day 5 to 10 drops of tinct. nucis vomicæ), are also to be recommended. In order to prevent bed-sores, in bedridden patients there should be the most thorough cleansing and care of the whole back, as well as proper attention to position in bed.

II. Cerebellar Heredo Ataxia

In the year 1893 *Senator* called attention to some cases of hereditary ataxia in which, along with the spinal lesion which we have described (see page 125) there was also a striking diminution in size of the cerebellum, and in the same year *P. Marie* attempted to construct from these atypical cases a disease picture to which he gave the name "hérédo-ataxie cérébelleuse," and

which was to be separated both clinically and anatomically from *Friedreich's* disease.

According to *Marie's* description, the marked cases present the picture of a disease which begins after the twentieth year of life with a slowly and progressively increasing unsteadiness in walking and standing; sometimes, too, with pains in the loins or in the lower extremities. After from one to three years the ataxia involves the arms also, while at about the same time disturbances of speech and of vision become evident. Optic atrophy and narrowing of the visual field, often also a loss of the pupillary reaction to light, are found. The patellar reflexes are exaggerated, or at least of normal activity. Often there are also other spastic phenomena; for example, ankle clonus. Frequently a certain mental weakness is observed. Nystagmus and disturbance of speech develop as in spinal hereditary ataxia. On the other hand, the deformities of foot and spine so characteristic of the latter are absent. Quite often there are disturbances of sensibility, less frequently those of swallowing and of the bladder; sometimes there is hypertonia of the muscles, again choreiform movements. This presents a symptom complex differing in many important points from *Friedreich's* disease, which seems to justify definite separation from affections resembling it in the disturbances of coördination and occurrence in families; but it has been shown that the boundary line cannot be sharply drawn either from an anatomical or from a clinical point of view. As to the post-mortem findings, it suffices to point out that *Thomas* and *Roux* in one case which corresponded particularly well with the above description of *Marie*, have reported a negative finding as regards the cerebellum, with hypoplasia and systematic degeneration of the spinal cord, and that I found in a patient who presented clinically not a single one of the symptoms characteristic for *Marie's* type, the most profound alterations of the cerebellum which have ever been observed in the hereditarily ataxic. The weight of the cerebellum was reduced from the normal figure of 145.2 grms. to 43.2 grms. (see Figs. 43 and 44). From a clinical point of view, we know, too, that *Marie's* type of hereditary ataxia can occur just as frequently in childhood as that of *Friedreich* in adult age; that in it there may be intact sensibility and an intact optic nerve; that scoliosis and pes cavus with dorsal contracture of the great toe occur now and then, that loss of the pupillary light reflex is usually absent, etc. Exaggeration of the reflexes has best preserved itself as a clinical criterion; but with observing that the patellar reflex, while abnormally lively at the start, can be lost during the course of the affection, this last distinction must fail. Also hereditarily ataxic brothers and sisters who, while presenting an otherwise identical picture, show a different condition of the tendon reflexes, are not at all rare. Hence, we can recognize cerebellar-heredo-ataxia only as a relatively characteristic and frequent variety of the true *Friedreich's* disease, and at the bedside must refrain from making a diagnosis as to the anatomical distribution of the lesion with too great positiveness (especially as to the involvement or not of the cerebellum). Indeed, I have been able to recognize experimentally that in the dog the same ataxic symptom complex can be produced, according to choice, either by a lesion of the cerebellar tracts and spinal cord or by destruction of their endings in the vermis cerebelli.

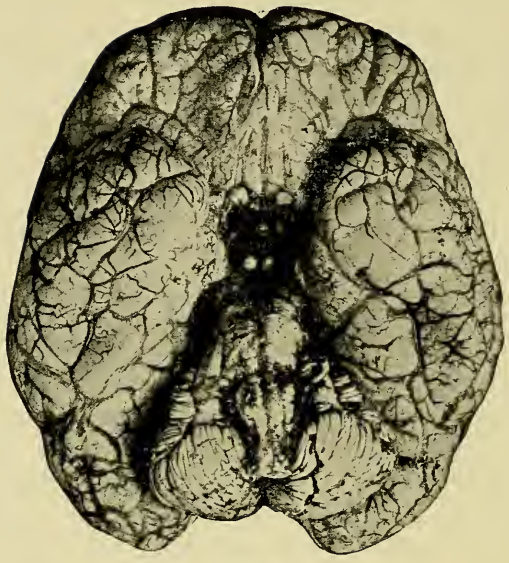


Fig. 43.

Atrophic Cerebellum of a Patient Aged Forty Years, with Hereditary Ataxia.

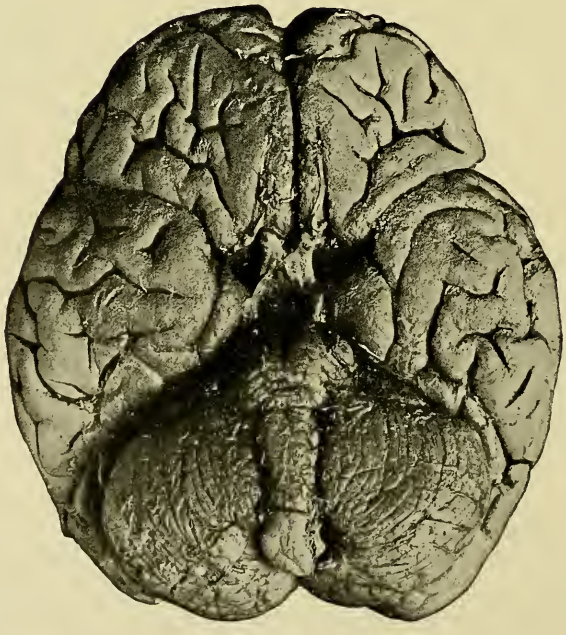


Fig. 44.

Normal Cerebellum of a Man Aged Forty Years.

This discovery bridges over physiologically also the clinically as well as anatomically already obliterated boundaries between the *Freidreich* and the *Marie* types. As regards etiology and therapy, there is nothing to add to what has already been stated in connection with the discussion of *Friedreich's* disease.

III. Infantile Progressive Hypertrophic Neuritis

In the year 1890 *Dejerine* described a "special form of *Friedreich's* disease with muscular atrophy and disturbances of sensibility" which since then has been recognized as a clinically and anatomically quite special disease. The affection begins in childhood, occurs almost always in children of the same family, and has the following symptoms: Marked ataxia of all four limbs; pes equinovarus excavatus; kyphoscoliosis; nystagmus—so far criteria of *Friedreich's* disease. To these are added, however, loss of the pupillary light reflex, psychical abnormalities, marked disturbances of sensibility, and marked muscular atrophy in the peripheral parts of the extremities, neuralgiform pains. Further, a very great hypertrophy of the peripheral nerve trunks, recognizable on palpation, is found in these patients. This depends, as post mortems have shown, upon a very great increase of their fine and coarse connective tissue, which proliferated in great circles, surrounds the partly preserved, partly degenerated, nerve fibers. The nerve trunks, on account of this, are thickened to double their normal size, which particularly gives to the cauda equina a characteristic appearance. In the spinal cord there is a systematic affection of the posterior columns.

LECTURE IX

Multiple Sclerosis

GENTLEMEN: The disease with which we must now occupy ourselves and which was first recognized as a disease picture *sui generis* and studied by *Vulpian* and *Charcot* (in the sixties of the last century) must, since of late our attention has been directed to its rudimentary and atypical forms, be considered as a comparatively frequent nervous disease. I see many of these cases among a country population and this experience agrees with that of other neurologists.

Multiple or disseminated sclerosis of the brain and spinal cord, the "sclérose en plaques" of *Charcot*, takes, in the circle of nervous diseases, a special position, in consequence of a remarkable antithesis which exists between its clinical and its pathologico-anatomical habitus. Namely, while from autopsy to autopsy the topographical distribution of the disease foci varies so much that the preparations from the brain and spinal cord in two cases never represent the same picture, clinically a number of types can easily be separated in which the individual cases may present a remarkable similarity.

Nevertheless, while no region of the cerebro-spinal apparatus is avoided by the lesions of multiple sclerosis, on comparing the different sections it is clear that the sclerotic foci plainly show a predilection for certain regions; in the pons they affect chiefly the basal regions; in the cerebrum, the walls of the ventricles and the corpus callosum; in the cerebellum, the medullary substance; in the spinal cord their preference for the white matter is so evident that *Ribbert* in his time affirmed that the borders of the gray matter set an impassable limit to the extension of the sclerotic foci—a view which to-day is indeed disproven.

Pathological Anatomy

Fig. 45 shows you with how little reference to the structural arrangements of the nervous elements the sclerotic foci are bounded. Macroscopically these plaques, whose number in the brain and spinal cord can vary from a few to many hundred, present a reddish or yellowish gray appearance; on palpation they give a feeling of increased resistance. Microscopically the sclerotic tissue presents itself as a thick felt-work of proliferated glia through which many nerve fibers still pass. These last have, indeed, mostly lost their medullary sheaths, but the naked axis cylinders are preserved; indeed, a new formation of axis cylinders appears to have occurred. This persistence of the conducting elements makes the absence of secondary ascending or descending degenerations in multiple sclerosis explainable; it speaks for the fact that the glia

proliferation is the primary, while the destruction of the medullary sheath is usually a secondary phenomenon (to which, indeed, the phagocytic action of the glia cells contributes). Also the ganglion cells in the foci are preserved; on the other hand, the blood vessels are usually markedly affected; they are thickened, infiltrated, and the lymph channels of the adventitia are filled with granular cells.

Etiology

From an etiological point of view, multiple sclerosis presents much that is puzzling. The most widespread opinion, that firmly held by *P. Marie*, attributes to it an infectious basis. Previous typhoid, variola, measles, scarlatina, have frequently been brought into causal connection with it. The occurrence of great epidemics of influenza naturally must also be considered among possible etiological factors. Less frequently puerperal infections, diphtheria, erysipelas, cholera, dysentery, and malaria have been held responsible. Also previous syphilis we find occasionally noted; indeed, one of the cases reported by *Thomas* and *Long* is remarkable on account of the anatomically recognized combination of luetic meningo-myelitis and true multiple sclerosis.

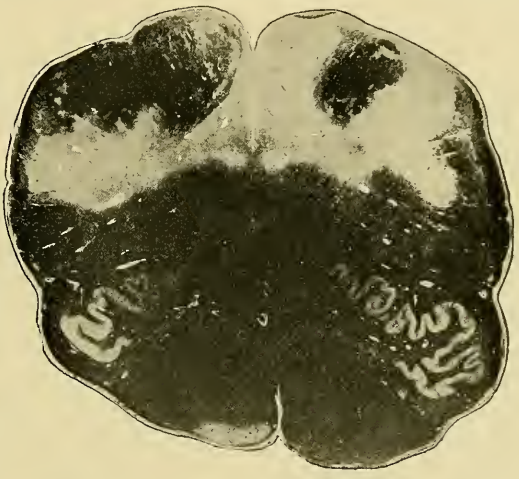


Fig. 45.

Multiple Sclerosis. Section through the Medulla. *Weigert-Pal* Stain.

In the second place, poisonings by metals have been brought forward as causal factors, chiefly by *Oppenheim*. In these cases the active agent has usually been lead, but some cases have also been attributed to intoxication by mercury, zinc and manganese. Among non-metallic poisons, carbonic oxide must be mentioned, since in a number of observations there has been a history of poisoning by emanations from coal.

Finally, in a few publications the disease has been referred to a trauma or to a violent emotional disturbance.

Where so varied a list of factors has been brought into etiological relation with one and the same disease, we are quite justified in seeing in these factors rather "provoking agents" than direct causes, and in looking deeper for the actual causes. This is also the view of *Strümpell*, who, among 24 cases observed in Erlangen, was never able to prove the connection with previous infections or intoxications. Apparently multiple sclerosis is not an exogenic, but an endogenic affection dependent upon the congenital makeup of the nervous system, like syringomyelia to be described later, since a combination of the two diseases occurs. This agrees with the experience that multiple sclerosis, as a rule, is a disease of early life; its beginning is usually at the

end of the second or the beginning of the third decade. In children, indeed, the disease is quite rare; here one cannot be too cautious about the diagnosis if one does not wish to incur the danger of being contradicted by the autopsy or by the further course of the disease. The diagnosis "infantile multiple sclerosis," however, was confirmed, for example, in one case of *Eichhorst*, which was also remarkable in that the mother of the child died of the same disease: however, hereditary occurrence of the disease is extremely rare. Also, the histo-pathological picture seems to me to speak decidedly against an inflammatory basis for the disease as assumed by different authors; much rather, it appears to me as the correlate of a congenital tendency to perverse growths somewhat resembling tumor formations. The Russian neurologist *Rossolymo* goes so far as to consider multiple sclerosis, gliosis and glioma of the central nervous system as three stages of intensity of the same process.

Symptomatology

When now we come to sketch the clinical symptoms we will confine ourselves at first to the typical, fully developed cases, upon which *Charcot* has based his classical description.

In the foreground of the picture stand disturbances of motility, above everything, tremor. This presents such sharp criteria in multiple sclerosis that in spite of the appearance of tremor in all possible nervous diseases it can be considered the most characteristic symptom of this disease. This is the so-called "intention tremor" which is absent during rest and only shows itself on voluntary movements (much more rarely on automatic, reflex, or associated movements). It disappears during sleep; under the influence of emotional disturbances and exertion it increases and its oscillations are proportional in their range to the extent of the movements carried out; the further the intended movement goes, the more rapid the oscillations become, but in spite of this they preserve a certain rhythm.

In a classical experiment the patient is solicited to carry a glass of water to his mouth. The moment the hand begins to move the oscillations commence; they increase in extent and violence the nearer the glass gets to the lips; it strikes clinking against the teeth, the contents are spilled, and the tremor ceases only after the glass has been taken away from the patient. If the hand of the patient is laid upon his bed and he is made to bend one single finger, this isolated intended movement suffices to set the whole hand, even the entire arm, into violent oscillation. The tremor of multiple sclerosis is a massive one, affecting the extremities as a whole, and increases in intensity toward their roots. In like manner intention tremor can be provoked in the lower extremities; the head and neck also present it; as long as the patient keeps his head on the pillow they are quiet, but upon raising it up or turning it sideways the most violent nodding and rotating oscillations commence. The handwriting, even in the early stages of the disease, assumes a characteristic appearance (as in Fig. 46).

Not only the impulse necessary to carry out a movement, but also that needed to keep a certain position, sets up the tremor. So the body of the

patient, as soon as he attempts to hold it upright, is shaken violently up and down, or forward and backward.

In the arms, as in the legs, the tremor may be first unilateral, or at least predominating upon one side, only later to spread to the opposite side.

As to the material basis of this symptom, there are various hypotheses which can be as little proved as disproved. *Charcot* considered it the expression of the leaping over of the stimulus from one neuron to another (on account of the loss of insulation by destruction of the medullary sheath). Later authors explain the intention tremor as a focal symptom produced by sclerotic plaques in the pons, in the corpora quadrigemina, in the thalamus, and finally in the cerebellum.

Less characteristic is the disturbance of gait in patients with disseminated sclerosis. Its study is considerably interfered with by the tremor which is at the same time set up on walking. In general, however, three sorts of gait abnormalities can be distinguished: the spastic, the spastic cerebellar, and the purely cerebellar gait.

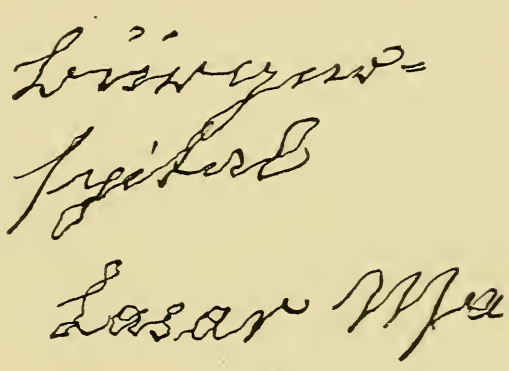


Fig. 46.

Multiple Sclerosis. Handwriting.

In the first, with the beginning of locomotion there is tonic rigidity of both lower limbs; they are extended at all joints, and besides this held in adduction. The patient balances himself on the tips of his toes, as we have described under spastic paralysis. This form is not rarely observed; still more frequently, however, the spastic element is less prominent and is combined with cerebellar-atactic symptoms, widening the supporting surface by spreading the legs, swinging the feet, and deviating from side to side from the straight line. Much more rare, on the other hand, is the third, the pure cerebellar type of locomotion, in which the gait is staggering and uncertain; the feet seem to stick to the floor and forward movement is carried out in a more or less zigzag manner. To the true staggering gait of the drunken man, however, it does not usually come, because sooner or later spasticity occurs. Through this, finally, walking becomes impossible, and the patient is condemned to bed on account of true spastic paraplegia.

Among the eye muscle disturbances of multiple sclerosis, nystagmus takes the first place. In the early stages of the disease it must be sought for by making the patient look far to the side and up and down. It is a dynamic nystagmus, an intention nystagmus, whose intensity increases with the extent of the movement. Almost always lateral nystagmus is observed, more rarely vertical nystagmus. As a rarity, rotary nystagmus, turning the eyes like a wheel, has been described. Do not forget that a slight grade of nystagmus sometimes occurs in healthy people by strained looking to one side; only a definite oscillation of the eyes, not the so-called nystagmiform contraction, is to be relied upon for the diagnosis of multiple sclerosis.

Other eye-muscle disturbances are less constant. According to *Dejerine* and *Thomas*, they only occur in a sixth of the cases. Usually it is an isolated paralysis of an external eye muscle. Most frequently affected is the abducens, more rarely the oculo-motorius. *Oppenheim* has seen three cases of total ophthalmoplegia. All these paralyses come on slowly and by degrees, and can disappear later. They are accompanied by diplopia and strabismus. According to *Kunn*, however, the strabismus of multiple sclerosis is not always the expression of eye-muscle paralysis, but the result of sclerotic foci which interrupt the association tracts between the individual eye muscle nuclei. Myosis, inequality of the pupils, weakness of accommodation, occasionally occur. More frequently hippus, unrest of the pupils, abnormally lively pupillary reflexes, are observed, which, according to *Parinaud*, are to be considered as the homologues of intention tremor and increase of reflexes in the muscles of the body.

These last affect the tendon and bone reflexes in all the limbs, particularly in the lower extremities, however, and are generally very marked. Usually the whole "spastic symptom-complex," as we have studied it in Lecture VIII, is found together with the abnormal reflexes (*Babinski, Oppenheim, Mendel-Bechterew*); sometimes, however, the ankle clonus cannot be obtained on account of the foot being held in too strong contraction. The skin reflexes, namely those from the abdominal wall, are, however, usually diminished or lost, as *Ed. Müller* has specially pointed out.*

The tongue, also, and the speech apparatus are the seats of marked disturbances of motility. When the patient puts out his tongue it is noticed to be in a state of rhythmical unrest; it is alternately protruded and drawn back, in rather rapid succession.

The speech shows marked disturbances. Only slowed at the beginning, it takes on later a scanning character. Each syllable is chopped off and brought out in an explosive manner. This speech has very appropriately been compared with that of a child spelling. The individual syllables are uttered explosively with oscillations of the head, of the lower jaw, and lips. A few authors claim to have observed a tremor or a paresis of the vocal cords in phonation. *Oppenheim* has seen this sometimes in respiration, which becomes jerky on this account. In spite of recent attempts, particularly by laryngologists (for instance, *Rethi*, in Vienna), to analyze the disturbances of speech in multiple sclerosis, a satisfactory explanation of its method of production is still wanting.

We must conclude the enumeration of motor disturbances with description of the pareses and contractures as well as of the apoplectiform attacks. At the start the gross strength in arms and legs is well preserved; soon, however, the muscular energy diminishes so that finally there may be actual paralysis. Along with this, even in the early stages and particularly in the lower extremities, there is usually a condition of latent contracture which is expressed in

* However, abdominal hyporeflexia and areflexia are only to be considered as a pathological finding in young persons with firm abdominal walls which contain little fat and in the absence of disease of the abdominal organs! Also repeated testing with stimuli of graduated intensity is necessary before the abdominal reflexes are put down as weakened or lost.

abnormal resistance to passive movements. The paretic disturbances are often more marked on one side, also according to whether they affect only one or both limbs of the affected side, they present the monoplegic or the hemiplegic type. An uncommon symptom, on the other hand, is degenerative muscular atrophy, which manifests itself by qualitative alterations of electric irritability. Complete reaction of degeneration, so far as I know, has never yet been recognized. Nevertheless, such cases have been mistaken for amyotrophic lateral sclerosis by no less a person than *Dejerine*. Actual attacks of hemiplegia occur in only a small part of the cases. These are accompanied by loss of consciousness and leave behind a one-sided paralysis, which, however, usually disappears very quickly, in a few hours or days. I have frequently seen them accompanied by elevation of temperature. Also epileptiform attacks equally accompanied by a rise of temperature have been described in multiple sclerosis.

Bladder troubles are not at all infrequent, though in my experience they occur more frequently in the atypical forms than in cases with the classical combination of symptoms; they are, however, usually of temporary duration. They are generally pollakiuria, or ischuria paradoxa, in which, when the bladder has become filled to a certain point, the urine begins to leak away in drops. More rare is true incontinence with permanent dribbling of urine; still less frequent, retention of urine on account of spasm of the sphincters. Disturbances of the rectal and of the sexual functions are entirely inconstant and episodic. When impotence or priapism are complained of, they are usually temporary conditions. "Dissociated disturbance of potence," in which erections still occur but ejaculation and orgasm are suspended, deserve special mention.

Disturbances of sensibility were formerly regarded as rarities; they are, however, not so exceptional. *Freund* first directed attention to them. They are chiefly paresthesias, tickling, formication, "going to sleep" of the peripheral parts of the extremities. More rarely actual pains, stabbing, boring, girdle or constriction sensations. Even lancinating pains, like those in tabes, have been observed in cases certainly multiple sclerosis. Neuralgias also occur. As cause of such pains in the trigeminus distribution, in one case a focus of sclerosis situated just at the exit of the nerve has been described. I would particularly point out the arthralgias which are not at all rare, and which may lead to confusion with articular rheumatism in case they occur at the start.

Upon objective examination, diminution of the pain and temperature sense, more rarely of that of touch and vibration, as well as that of position sense, is found reduced in some patients. The topography of these disturbances is extremely inconstant, also they can totally disappear again. Total anesthesia scarcely ever occurs as an actual symptom of multiple sclerosis. If this is present and, still more, if it is definitely unilateral, one should think of hysteria which is a relatively frequent complication of this disease. Rarely, and usually as an episode, hyperesthesia is complained of.

Upon the part of the organs of sense we may next mention changes in the optic nerve, which are to be found, indeed, in the majority of cases. They affect now only one, again both eyes, and produce either amaurosis or amblyopia, or more or less circumscribed defects in the visual field. These last

can be centrally or peripherally located, but have usually a quite irregular outline. Further, the visual disturbances are exceedingly variable in their intensity and can, indeed, entirely disappear again. The alterations of the eye ground can present themselves under very varied aspects which have been studied, among others, by *Unthoff*, *Parinaud*, *Bruns* and *Stölting*. Only exceptionally has the papilla the appearance of optic neuritis (hyperemic, somewhat prominent, cloudy with dilated vessels); usually it is, on the contrary, flat and gray-white, or porcelain-white, in color. The circumstance is remarkable that the ophthalmoscopic change almost never affects the optic nerve *in toto*, but is usually, even in the late stages, limited to one-half, generally the temporal.* These different anomalies contribute to give the patient the peculiar dull look with which *Charcot* was struck when he expressed himself, "These patients have a vacant look."

While hearing, as a rule, remains intact, vertigo of vestibular origin belongs to the most frequent symptoms of multiple sclerosis. It is true rotary vertigo, sometimes of great intensity, so that the patient has the feeling of actually being whirled around, and on this account falls to the ground. As a rule, this vertigo occurs in attacks and suddenly, set up either by change of position of the head (raising up in bed, looking upward, locomotion), or without recognizable cause. Only very exceptionally it comes on, like *Menière's* disease, with ringing or whistling in the ears; also it does not, as a rule, lead to vomiting. In some cases, specially marked by decided vertigo, it has been assumed that there were sclerotic foci in the course of the vestibular fibers, but in only a very few have there been any positive findings. Though *Charcot* asserted that vertigo was an initial symptom in three-fourths of all the cases of multiple sclerosis, from my personal material I would say that these figures are too high; of vertiginous initial symptoms I could find a history in scarcely half the cases. Taste and smell are usually uninfluenced in disseminated sclerosis.

Of other cerebral disease manifestations, we must mention headache and psychical alterations. The first is very frequently complained of; it can be like migraine. The latter often occur first after a long period of normal mental condition; finally, however, in the majority of cases. The patients are either silly and strikingly euphoric, or, on the other hand, morose and uninterested. Also, an abnormal lability of mood which manifests itself by sudden change between laughter and tears is not rare; along with this there occurs also actual compulsory laughing and crying. When multiple sclerosis begins in early life, it usually has particularly serious results psychically, and puts a complete stop to further mental development. On the other hand, where hallucinations and delirium have been described, there was certainly a combination with psychoses.

Finally, trophic alterations must be considered. These only exceptionally occur; epidermolysis, dropsy of joints, localized œdema and sweating, falling

* Since normally the temporal half of the papilla appears paler than the nasal half (the thinner layer of nerve fibers allows the white lamina cribrosa to be seen more plainly there), in doubtful cases it is always advisable to get the opinion of an experienced ophthalmologist.

out of the hair, brittleness of the finger nails, have been described. Only in advanced stages of the disease, when the patients are bedridden, and particularly in cases of very rapid course, is there a marked tendency to bedsores.

Atypical Forms of Multiple Sclerosis

The classical or *Charcot's* form of disseminated sclerosis, as we have now become acquainted with it, is marked by so great a number of definite symptoms that its diagnosis is to be considered a very easy one—at least, as easy as that of tabes. As particularly characteristic, since the time of *Charcot*, the "symptomatological triad"—intention tremor, nystagmus and scanning speech—have been brought forward. But the difficulty in recognizing the disease is given by its atypical forms, which include a striking majority of the cases, according to my experience, at least 90 per cent., and knowledge of which is on this account extremely necessary. Along with the more or less rudimentary forms of the disease in which different cardinal symptoms are absent (besides *Charcot's* triad, I would add to these last also spasticity of the legs and loss of the abdominal skin reflexes *), along with these "formes frustes" there are still some atypical forms distinguished by characteristic clinical pictures of which I will only describe to you the four most important.

(*a*) *The Bulbar Form.*—In this difficulty in swallowing and chewing, disturbances of articulation and phonation, stand in the foreground, glycosuria, rapidity of the pulse, and asphyxic conditions occur. When one considers how often upon autopsy large sclerotic areas are found in the medulla, it appears paradoxical that these clinical disturbances are really great rarities (see Fig. 45, page 132).

(*b*) *The Hemiparetic Form.*—In this form spastic paresis and loss of the abdominal reflexes are present only on one side; with these intention tremor may occur on one side only. Here and there disturbances of sensibility are found upon the opposite side in such cases, also the *Brown-Séquard* symptom-complex of half-sided spinal cord lesions. In conditions of this kind a positive diagnosis, indeed, can scarcely be made before the autopsy, but with very slow and intermittent progress of that symptom one should not forget to think of multiple sclerosis. In one case of *Oppenheim* this diagnosis could be confirmed.

(*c*) *Paraplegic Form.*—In contrast to the two already related, this atypical form is extremely frequently observed. In the initial stages the picture of spastic spinal paralysis may be present; in characteristic form, however, usually loss of the abdominal skin reflexes, changes in the optic nerve, or slight intention tremor in the upper extremities may direct attention in the right direction. Slight disturbances of sensibility and of the bladder function occur in an appreciable number of these cases. In the further course very severe spastic conditions may develop; either only the legs, or all four extremities become contractured little by little, the arms in flexion, the legs usually in extension, but often likewise in flexion. The tendon reflexes are exaggerated in the highest degree, and on testing them marked clonus is produced. Later

* With the exceptions mentioned in note on page 135.

permanent organic contractions of the tendons and muscles can occur (I once saw the knees gradually drawn up to the chin on this account), when the reflexes can no longer be obtained.

(*d*) *The Amyotrophic Form.*—I have already cursorily mentioned this, and indicated that it can imitate amyotrophic lateral sclerosis. There is atrophic paralysis of the muscles of the trunk and limbs, usually with complete reaction of degeneration and permanent contractures in the paralyzed region, great increase of the reflexes and bulbar symptoms.

Differential Diagnosis

The special peculiarity of multiple sclerosis, in beginning often with rudimentary and atypical symptoms not infrequently leads to its confusion with other nervous diseases. In the early stages, when the skin reflexes are normal and there is yet no spasticity of definitely organic character (that is, no *Babinski* or *Oppenheim* reflex or ankle clonus, etc.), a wrong diagnosis of hysteria is sometimes made even by experienced observers, a mistake which is all the more frequent since the combination of both diseases is, as I have already remarked, not at all infrequent. In such cases it is well to be very cautious and to put off making a diagnosis and prognosis until the further course of the disease has been observed. In those rudimentary cases where tremor is the only symptom (they are exceedingly rare) a consideration of the typical character of this tremor will make a confusion with chorea or paralysis agitans impossible. A differential diagnosis between this and mercurial tremor may be more difficult; this is often an intention tremor, which, however, does not cease entirely during rest. In the course, or as a consequence, of infectious diseases, disseminated myeloencephalitis, whose symptomatology presents a great similarity to that of multiple sclerosis, may occur. This runs a course with elevation of temperature, and is usually characterized by some focal symptoms which point to large inflammatory foci in the brain axis; for example, an alternating hemiplegia with paralysis of the face on one side and of the extremities on the other. Acute onset does not suffice to exclude multiple sclerosis; this also, as we will soon see, may commence acutely. In many cases only the further course will clear up the diagnosis. Cerebro-spinal syphilis can also simulate multiple sclerosis. Lumbar puncture is very valuable here when syphilitic infection has not been definitely shown. According to *Long* and others, in multiple sclerosis there is no lymphocytosis, or only a minimal one is found.

In general, also, the *Wassermann* complement-fixation reaction is a good differential guide. However, I once saw a positive result from this in a case which proved on autopsy to be certainly multiple sclerosis, although there was neither a history nor any objective signs signifying the assumption of previous syphilis. (It was in a virgin.) In the picture of progressive paralysis, also, spastic paresis can figure, and this also shows tremor, apoplectiform attacks, and speech disturbances. Here, however, the tremor is rapid and fine and affects chiefly the ends of the extremities; the speech, too, is not scanning and chopped off, but hesitating and drawling. Finally, also, the

psychical disturbances are of quite definite character. It is to be noted that in a number of cases the combination of progressive paralysis with multiple sclerosis has been observed (for example, by *Schultz*). Those rare cases of disseminated sclerosis which begin in an apoplectiform manner can present difficulty, since post hemiplegic tremor after apoplexy can exceptionally have a typical intentional character. In such cases one must often await further developments before coming to a conclusion. Nevertheless, in suddenly or gradually occurring hemiplegias in young persons in whom alcoholism, brain tumor, syphilis, heart disease and hysteria can be excluded, the possibility of multiple sclerosis should not be lost sight of. Many points in common are shown by the symptomatology of multiple sclerosis on the one hand, and of the hereditary ataxias on the other, namely, nystagmus, scanning speech and cerebellar atactic phenomena. However, there can be no confusion with the spinal variety of *Friedreich's* disease, since it is characterized by loss of tendon reflexes, by scoliosis, and by the foot deformity; besides the tremor combined with ataxia peculiar to it has entirely a different character from that of multiple sclerosis (see the previous lecture). Rather is there a possibility of confusing multiple sclerosis with *Marie's* cerebellar heredo-ataxia, since in this the reflexes are exaggerated, scoliosis and foot deformities are absent, and the tremor may have a definitely intentional character; further, changes in the optic nerve occur. Here family character would speak for heredo-ataxia; besides, in this last there is usually beginning with purely cerebellar symptoms. Brain tumors sometimes produce clinical pictures similar to multiple sclerosis, as *Bruns* and *Nonne* have shown. The differential diagnosis can be greatly involved, when on account of complicating hydrops ventriculorum the multiple sclerosis is accompanied by symptoms of brain pressure (continued headache, œdema of the papilla, etc.). In general, however, these last are not so intense, and particularly not so progressive, as in intracranial tumors. Besides, these are exceptionally rare cases. Again, the differentiation between tumor of the spinal cord and multiple sclerosis is sometimes not easy. I intend to consider this practically important point in a later lecture when describing tumors of the spinal cord. Finally, *Westphal's* "pseudo-sclerosis," a rare and not yet sufficiently explained disease, in which, anatomically, the findings have been negative or only a very inconsiderable degree of diffuse glia proliferation, must be mentioned. The clinical picture has great analogies with that of multiple sclerosis; still, on the one hand nystagmus and optic nerve changes are regularly wanting, and on the other, in pseudo-sclerosis, along with the nearly constant occurrence of epileptiform attacks, there are other symptoms which do not belong to true sclerosis; namely, deep apathy and dementia, delirium, outbreaks of violence and a peculiar slowing of the movements of the face and eye muscles. Here, also, upon a proper estimation of the symptoms, mistakes can be avoided all the more, since pseudo-sclerosis is a disease of childhood, while multiple sclerosis, as has already been indicated, almost never occurs in children.

Course and Prognosis

We can distinguish three methods of beginning of multiple sclerosis. It can begin insidiously, and this is most frequent, and usually then announces itself by "outpost symptoms," as *Oppenheim's* appropriate expression has it, for example, by disturbances of the eye muscles with double vision or by changes in the optic nerve (often wrongly considered as retro-bulbar neuritis), or by bladder troubles; to these are usually added spastic symptoms in the legs, which little by little increase in intensity; less frequent is a rapid (acute or subacute) beginning, usually introduced by severe vertigo, still more rarely sudden brutal onset, with an apoplectiform or epileptiform attack. The disease itself runs a more or less chronic course, either progressively or in exacerbations. In the first case the disease usually leads to death in from 5 to 20 years. Proximal causes of death are: intercurrent diseases or the occurrence of bulbar disturbances, cachexia with bedsores, or an apoplectic attack. There are cases described as acute multiple sclerosis, which in from 3 months to 2 years develop rapidly the most severe symptoms and end fatally. Probably these, or at least part of them, are instances of another disease, namely the post infectious disseminated myeloencephalitis mentioned in the discussion of differential diagnosis. When remissions occur they can last for years; in one of my cases affecting a 50-year-old lady, the history mentioned that at the beginning of her third decade of life she had suffered from a temporary spastic paraplegia with tremor; a nearly thirty-year remission must hence be assumed in this case. The possibility of recovery, or at least of a definite standstill, of the disease is affirmed by a few authors, but these reports must unfortunately be received with great reserve, as the above case with a recurrence after 27 years shows. Important from the point of view of prophylaxis is the condition that new exacerbations are often started up by overexertion, getting chilled, infectious diseases, the puerperium, etc.

Treatment

After entering upon hospital treatment considerable improvement (which is mainly to be attributed to the physical and psychical rest which the patient gets) is often observed. Also in patients cared for at home, the chief weight is to be laid upon the keeping away of causes for overexertion and emotions. In this connection it is to be remembered that these patients often have a strong libido, whose gratification or even excitation is to be suppressed as far as possible. This point must be considered also in the choice of nurses; an endeavor must be made to strengthen and build up the patient. Warm baths are just as injurious as cold water treatment. The same remarks apply to strong electric applications. Of all drugs, only arsenic seems to have any active effect in the way of favoring remissions. I give it either in the form of *Fowler's* solution or as the Asiatic pill (raising it as rapidly as possible to considerable doses, 7 drops of *Fowler's* solution or 0.005 (gr. $\frac{1}{15}$) of acid. arsenios, t.i.d.), eventually also in combination with quinine or iron, or, however, if there is no effect after 3 weeks from this medication, subcutaneously as sodium cacodylate, 1 ampoule containing 0.05 (gr. $\frac{3}{4}$), later 0.1 to 0.15 (gr. $1\frac{1}{2}$ to gr. 2).

LECTURE X

A. Spinal Gliosis and Syringomyelia

The two disease conditions of which we will now give a common description cannot be separated from one another either symptomatologically or upon a basis of their pathologico-anatomical substratum, and their pathogenesis; since spinal gliosis not only is a condition regularly antecedent to the occurrence of the cavity formation which we call "syringomyelia," but both conditions are usually found alongside of one another in the spinal cord of one and the same patient.

Pathological Anatomy

A typical syringomyelic spinal cord can macroscopically closely resemble a tube, justifying the characteristic appellation introduced by *Ollivier d'Angers*.

The former view that it was the product of an ectasy and a hydrops of the central canal must be given up, since the entire independence of the cavity formation from the spinal canal can often be made out. From other cases there is evidence that the confluence of the ependymal canal and the syringomyelic cavity have occurred secondarily only. This last usually begins at the posterior gray commissure and extends more or less irregularly in the cross section into the gray substance of the posterior and anterior horns. In a longitudinal direction it extends from the cervical region (at which level the cavity formation usually begins) in the majority of cases, getting smaller continually, into the middle or lower thoracic region. There are, however, instances in which it has extended deep into the lumbar region, indeed, into the conus terminalis. On the other hand the process can ascend until the cavity opens into the 4th ventricle of the brain axis and involves an extensive part of the medulla, when "syringo-bulbia" can properly be spoken of.

Microscopical examination now furnishes information as to why these cavities occur. Their walls consist mainly of a thick zone of firm, greatly proliferated, neuroglia rich in cells—that is of the same tissue as the neoplasms which develop from the supporting substance of the nerve centers, the gliomata. While glioma, however, spreads from its point of origin diffusely in every direction without its growth following any definite path determined by anatomical relations so far as can be recognized, the process upon a basis of which syringomyelia occurs—"central gliosis of the spinal cord"—is characterized by more or less cylindrical or prismatic outline, by excessively slow growth and tendency to quiescence for long periods, and by exceedingly sharp separation from the healthy tissues. Peculiar to it, further, is its tendency to break down and to

form cavities; still, cases occur in which the last are absent or only rudimentary, and in which disease-pictures, not to be separated clinically from syringomyelia, depend upon a substitution of solid masses of glia for central portions of the spinal cord. Above and below, too, a portion of solid gliosis is often joined to the glia tube of syringomyelia. Hence, when we in the following, for the sake of simplicity, speak always of syringomyelia, it must be understood in advance that our remarks apply throughout to both conditions which can be separated neither clinically nor anatomically.

The syringomyelic cavities have a lining of ciliated cylindrical cells only in front where they have become united with the central canal, some of the ependy-

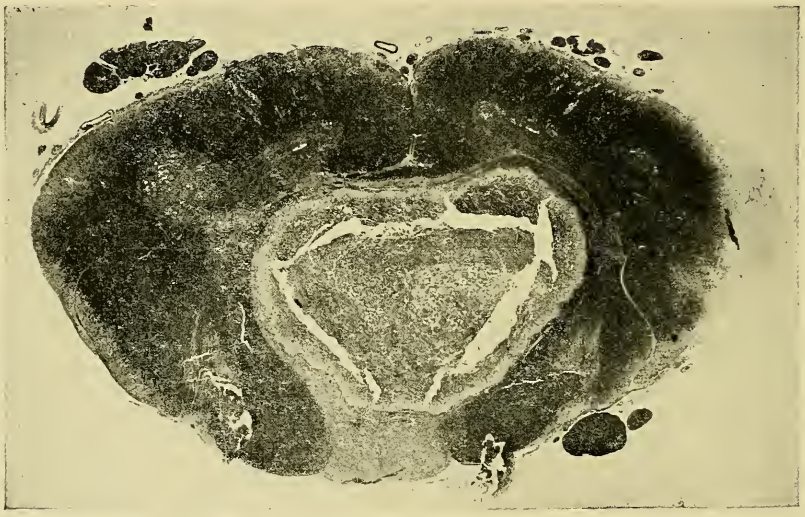


Fig. 47.

Central Gliosis Passing Over Into Syringomyelia. *Weigert-Pal* Stain.

mal lining of which has remained. Finally, where the gliomatous masses have overstepped the boundaries of the gray substance into the white columns, ascending or descending degeneration of the long fiber tracts, for example, of the posterior columns or of the pyramidal tracts results.

Etiology

Although a varied line of exciting causes have been held responsible for the outbreak of syringomyelia—that is, for its first clinically determined symptoms—there can be no doubt that it is at the base a congenital process, namely that the affected individual, on account of a congenitally defective structure of the spinal cord, is condemned to defective growth and degeneration of the central glia leading to disturbances, first unnoticeable, but gradually becoming more marked. The disease affects men about three times as frequently as women; it shows peculiar variations in its geographical distribution; for example, while quite frequent in Vienna, in our neighborhood (northwest Switzerland, southern Baden and upper Alsace) it is excessively rare.

Symptomatology

The most characteristic symptom of syringomyelia is the so-called "dissociated anesthesia," that is, loss of pain and temperature sense with intact touch sense and deep sensibility. This peculiar disturbance, first thoroughly studied by *Kahler* and *Schultze*, has been denominated the "posterior horn type of sensory anomaly." As to its anatomico-physiological basis we have quite accurate knowledge. Certain definite short fibers from the posterior roots end in the posterior horn of the spinal cord on the same side. From this point the impulses transmitted centripetally by these are conducted farther through the gray substance to the lateral columns of the opposite side and through them to the optic thalamus and the cerebrum (by way of the "tractus spino-thalamicus").

The senses of pain and temperature are conducted exclusively by these paths, hence in destruction of the posterior horns or of the central gray matter (through which the "tractus spino-thalamicus" passes) the perception of these sensations is lost, while to touch sense and deep sensibility, still other paths remain open (the posterior columns, etc., see Fig. 50).

In typical cases of syringomyelia corresponding to the predominant distribution of the pathologico-anatomical process in the upper segments of the spinal cord, the dissociated loss of sensation is found in the upper limbs and on the chest and trunk. The patients themselves often become aware of these anomalies, since they are struck by the absolute painlessness of accidental burns or cuts and the absence of feeling of cold in winter. It is remarkable also that in taking the history of individuals in whom syringomyelia has only been diagnosed in the third or fourth decade of life through the onset of other symptoms, we sometimes meet with the statement that even as children they had been remarkable through experiencing no pain from wounds and burns.

If the dissociated loss of sensation presents nothing alarming to the patient or his surrounders, on which account the physician is generally not called into counsel, the two other so-called "cardinal symptoms" of syringomyelia present the more striking and disturbing anomalies; namely, the progressive muscular wasting usually limited to the upper extremities and the shoulders and the manifold tropho-vasomotor disturbances equally affecting by preference these parts.

The muscular atrophy is usually for a long time unilateral or, at least, much more marked on one side; only in the later course of the disease does it become more or less symmetrical. It begins almost always in the small muscles of the hand, especially in those of the first intermetacarpal space and of the thenar and hypothenar regions. By degrees deformities which we have fully described in Lecture VI under Spinal Muscular Atrophy, of the type *Aran-Duchenne*, occur. The "leaping over" of the atrophy from the hand to the shoulder muscles occurs less frequently in syringomyelia than in the *Aran-Duchenne* disease; somewhat more frequently regular ascent, in which first the forearm, and then the arm muscles are attacked, is observed. In the forearm it is remarkable that the extensor muscles long remain intact; the preponderance that they gain over the atrophied muscles of the median and ulnar distribution sometimes results in a permanent contracture of the hand in a position of hyperextension which it

has been attempted to characterize, not very happily, as the "preacher's hand." Much less frequently we meet syringomyelic muscular atrophy in the muscles of the trunk and leg; these have a somewhat variable manner of appearance and can give rise to different anomalies of position (for example, pes equinovarus, pes calcaneus, clawfoot, etc.). In the neck the sternocleidomastoid, the scaleni and the trapezii are affected by the muscular wasting. The affected muscles almost always atrophy, not as a whole, but after the "fascicular" type, that is, one bundle after another. The proximity of degenerating and intact muscular fibers brings it about that, like in spinal progressive muscular atrophy, reaction of degeneration is often very difficult to recognize, and then is only partial. Fibrillary contractions are usually plainly manifest; they are found most easily in the deltoid.

The vasomotor trophic anomalies affect, above everything else, the skin and its derivatives—hair, nails, etc. A firm œdema of the back of the hand, coldness and livid coloration of the integuments (*Marinesco's* "succulent hand") is frequent. A similar thing is sometimes observed in the foot. Further, abnormal dryness, scaling, and fissuring of the skin, tendency to herpetic eruptions, separation of the epidermis and superficial ulcerations are observed. In the origin of these last phenomena, a considerable rôle should be attributed to the fact that the patients injure their analgesic and thermo-anesthetic hands by striking them against things, rubbing their wounds, handling hot or very cold objects. There is also a more or less extended atrophy of the skin with loss of its normal superficial outline, the so-called "glossy skin." I would mention further hyperkeratosis of the palms and soles, brittleness or falling out of the finger nails, loss of the skin hairs. The trophic disturbances affecting the subcutaneous connective tissue and the bones of the phalanges are particularly severe. Their points of origin are usually areas of suppuration which, starting in the fissures of the cracked skin, whose circulation also is much impaired, do not tend to heal normally, but continually to grow deeper. Their entirely painless course justifies the expression "panaris analgésique" used by *Morvan* for such cases. This can go on to severe mutilations as in leprosy, to the loss of the ends of, or of whole, fingers. Finally, in syringomyelia "osteoarthropathies" in the extremities, also in the vertebral column, occur, and indeed, quite frequently (according to *Schlesinger* in 30 per cent. of the cases). These affect, usually, the shoulder or the elbow. Atrophic and hypertrophic changes in the head of the bone, erosion of the socket, loosening of the ligaments, great intracapsular effusions, etc., are found.

We leave now the so-called "cardinal symptoms" to turn our attention to the less frequent manifestations of syringomyelia. Most important among these last are the oculo-pupillary symptoms which have been united under the name of *Horner's* symptom-complex. The relative frequency of this syndrome depends upon the fact that gliosis and syringomyelia have a strong predilection for the level of the lower cervical cord. At the level of the 8th cervical segment in the gray substance, however, is located the so-called "cilio-spinal center" from which fibers proceed by way of the anterior roots of the 8th cervical and two upper dorsal nerves into the lower ganglion of the cervical sympathetic, from which by another set of neurones there is further conduction to the superior

tarsal, the orbital and the dilator pupillæ muscles. The first mentioned is made up of the involuntary muscular fibers of the levator palpebræ; the second bridges over the lower portion of the orbital fissure and so prevents the contents of the orbit from sinking downward and backward; the third is the antagonist of the sphincter pupillæ, innervated from the motor oculi. The cutting off of influences from the cilio-spinal center is manifested, 1, by a paralytic myosis in which the pupil narrowed on account of paralysis of the dilator fibers, does not enlarge when the eye is shaded; 2, by a narrowing of the palpebral fissure, the so-called "sympathetic ptosis"; 3, by a sinking of the eyeball back into the orbit, the so-called "enophthalmus." This last component of *Horner's* symptom-complex is, indeed, not always definitely to be found.

Obviously this symptom-complex can also occur in lesions of the cervical sympathetic, of the anterior roots of the lowest cervical and two upper thoracic nerves and their rami communicantes; we have met it as an accompaniment of the "lower armplexus paralysis" ("*Klumpke's* paralysis") already in Lecture I. (See also Lecture XXV.)

Further, spastic symptoms which owe their origin to the involvement of the lateral columns by the pathological process, are not at all infrequent, particularly in the lower extremities, as in the arms, atrophy goes hand in hand with loss of reflexes; exaggeration of the patellar and Achilles reflexes are found; often, also, *Babinski's* phenomenon and ankle clonus; the hypertonia of the muscles only seldom reaches such a degree that the characteristic gait of spastic spinal paralysis occurs.

Both the oculo-pupillary and the spastic phenomena usually are limited to one side.

Of the atypical forms of syringomyelia we will give a short clinical outline of "syringobulbia" already mentioned; in this, there are found (often only on one side) dissociated sensory paralysis in the distribution of the trigeminus, atrophy of the tongue or masticatory muscles, recurrens paralysis, formation of ulcers on the posterior wall of the pharynx, paralysis of the eye muscles, ageusia, etc.

Differential Diagnosis

The differentiation of syringomyelia from progressive spinal muscular atrophy, since as far as the muscle symptoms go, they can greatly resemble one another, depends upon the demonstration of the typical dissociation of sensation. Also the trophic disturbances found eventually in the *Aran-Duchenne* disease are of much less intensity than those of syringomyelia. In spastic syringomyelia the determination of the condition of sensibility likewise furnishes the means of deciding between it and amyotrophic lateral sclerosis. The differential diagnosis between syringomyelia and lepra anesthetica can sometimes be very difficult, a question which in our neighborhood, fortunately, scarcely ever comes up. [It does in the United States.—*Translator.*] Decisive as to lepra is the recognition of nodular thickenings of the peripheral nerve trunks, particularly, however, the finding of the *Armauer-Hansen* bacillus.

Trophic disturbances about the face occur frequently in lepra, very rarely in syringomyelia; for lepra speaks further the finding of skin areas with in-

creased or decreased pigment, in which sensibility is diminished (insular anesthesia or hypesthesia) as well as the occasional occurrence of febrile paroxysms.

A rare disease isolated by *Charcot* and *Joffroy*, hypertrophic cervical pachymeningitis can produce clinical pictures very similar to syringomyelia. Pathologico-anatomically it consists in a great proliferation of the meninges in the cervical region which finally surround the cord like a thick cicatricial band which cannot be separated from the cord proper. Contractions within this great connective tissue envelope lead to solutions of continuity within the gray matter of the spinal cord, which evidence themselves in dissociated anesthesia, and to lesions of the pyramids recognizable by spastic-paretic phenomena. Almost regularly the "preacher's hand" already mentioned, which may depend partly upon secondary alterations in the anterior horn and partly upon changes in the motor roots, is found. The regular involvement of the sensory roots in the meningeal thickening leads without exception to a neuralgiform initial stage of the disease, upon which in its differential diagnosis from syringomyelia, most importance is to be placed. In the late stages of the disease paralysis of sphincters, formation of bed-sores and marked spastic paraplegia of the legs appear and the picture loses its resemblance to syringomyelia, to pass over more and more into that of compression of the spinal cord.

Course and Prognosis

As a rule, the first beginning of the disease is, as already said, so little marked, that the patients only late—that is, when they become frightened by the occurrence of muscular atrophy or trophic skin and bone disturbances—come to a physician, usually between the ages of 20 and 25 years. Nevertheless, the presence of the disease in childhood can be affirmed, not only retrospectively from a history of analgesia, etc., but is sometimes directly determined by a physician, the last indeed seldom enough. The further development of the disease proceeds extremely slowly; exacerbations of rapid progress and long stationary periods alternate with one another. As to life, the prognosis is in general good; the patients can live to be old, in any case they only rarely succumb to the syringomyelic paralysis of the muscles of deglutition or of those of respiration, sepsis, etc., but die usually of some intercurrent disease. As to recovery, the outlook is naturally poor; nevertheless, trophic lesions can heal up not to return again.

Treatment

In the treatment of syringomyelics, prophylaxis on the one hand against overexertion, injurious climatic effects, etc., on the other against wounds and burns, plays the chief rôle. Against the disease itself occasional arsenic cures or galvanization of the spinal cord can be tried. For the technique of this last, see Lecture VII, page 116. The results which have been described from the subjection of the cervical cord to the X-ray are not very convincing, on account of the well-known tendency of the disease to come spontaneously to a standstill; nevertheless, further experiences must be awaited before forming a definite opinion as to this new method.

B. Hematomyelia

Hematomyelia, or hemorrhage into the spinal cord occurs rarely spontaneously, relatively frequently as the result of trauma. In the first case it is either the result of rupture of a vessel on account of pathological brittleness of the arterial wall (arteriosclerosis, syphilis, pernicious anemia) in which usually we find given as causes factors which raise the blood pressure, like sneezing, straining, coitus, etc., or the hemorrhage follows secondarily in an already diseased tissue.

I saw one case (published by *Gerhardt*) in which the hemorrhage occurred in an intramedullary glioma, which up to this time had remained entirely latent. Also in acute and chronic myelitis secondary spontaneous hemorrhages have been described.

Traumatic hematomyelia occurs in severe contusions of the spinal column, injuries by lightning or electrocution by powerful currents; also, as a result of certain operative measures in pathological labors (difficult forceps extractions, the *Veit-Smellie* manipulation), or in *Schultze's* method of artificial respiration in infants born asphyxiated. Further, by forced bending forward or backward of the cervical spine (prize-fighters, "jiu-jitsu," etc.).

In typical cases the patient experiences a sudden and severe pain in the region of the vertebral column corresponding to the hemorrhagic focus and falls to the ground with more or less marked paralysis of the muscles distal to this point. Less frequently there is a somewhat slower development of the motor symptoms to which also disturbance of sensibility and of the bladder and rectum are usually added. A great part of these symptoms do not depend upon the direct destruction of the elements of the spinal cord by the extravasated blood, but usually represent remote effects of the hemorrhage which are alluded to as "shock," "inhibition," "diaschisis phenomena." These are in their nature temporary, so usually in the days, sometimes in the hours, next succeeding the hemorrhage, partial subsidence of the symptoms occurs. Since now the hemorrhage is usually in the central region the predominating syndrome of hematomyelia often presents the greatest analogies to the syringomyelic symptom-complex, particularly in the specially frequent cases in which the location of the hemorrhage is in the cervical region; then along with spastic phenomena in the lower limbs, atrophy of the small muscles of the hand, with the "ape-hand" or "claw-hand" deformity, a typical dissociated anesthesia and the *Horner* oculo-pupillary symptom-complex can be found (see page 146). Making of a prognosis is naturally only possible after the remote symptoms have cleared up. It depends mainly upon the location of the lesion; if the hemorrhage has occurred high up in the cervical region the prognosis is very gloomy, since in consequence of its nearness to the medulla, paralysis of the heart and respiration with high fever is apt to occur. Also location low in the spinal cord has very evil results, since permanent paralysis of the bladder carries with it the danger of ascending infection of the urinary apparatus. Many cases of hematomyelia high up in the cord perish rapidly in shock. On the other hand, small hemorrhages in the dorsal and lumbar regions can recover almost without residual symptoms.

Therapeutically, in the early stages absolute rest is the chief thing. Care must be taken that through the use of purgatives movement of the bowels takes place without straining, that is without any considerable elevation of the blood pressure. Bleeding, or a series of leeches or wet cups along the spine can be of use under some circumstances. The patients must not get up before the end of four weeks! The residual pareses, sphincter disturbances, etc., often need symptomatic treatment; we need not go into these things here, since they are considered elsewhere.

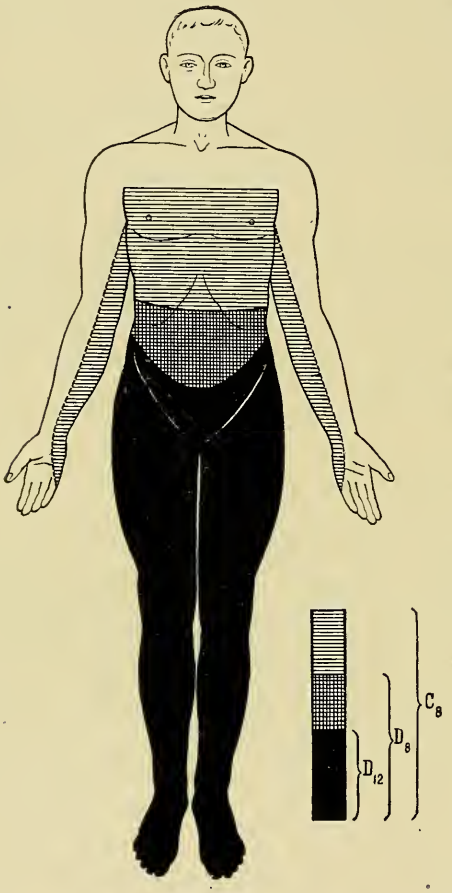


Fig. 48.

Extent of the Anesthesia in Transverse Lesions at Different Levels.

C. Transverse Lesions of the Spinal Cord

Considered from a clinical point of view, discussion of a rather heterogeneous array of disease conditions under this unique designation, is entirely justified; since disease or injury of the cross-section of the spinal cord, whether it owes its origin to a myelitis, to a traumatic solution of continuity, to compression by extra-medullary tumors or tuberculous spondylitis, or to the development of a neoplasm in the spinal cord itself, produces symptom-complexes agreeing to the fullest extent.*

On this account I intend deviating from the usual method, first to present to you the facts common to the group of disease conditions indicated, then, in order to avoid repetition, I will expose the individual pathogenetic sub-varieties according to their particular pathologico-anatomical and clinical aspects.

1. General Symptomatology of "Transverse Lesions"

A lesion of the spinal cord which so severely injures the total cross section of this cylindrical organ at a definite level, that the conduction of impulses by

* The same remarks apply to the so-called "concussion of the spinal cord," which though anatomically still little investigated, can give clinically very suggestive pictures. *A. Visscher* has communicated to me from the Balkan War some interesting observations of typical "transverse lesions" which had occurred without any direct traumatism in soldiers in whose neighborhood a grenade had burst or even had passed close by them.

all the fiber tracts is here interrupted, causes, of course, entire loss of sensibility and motility in the portions of the body distal to this.

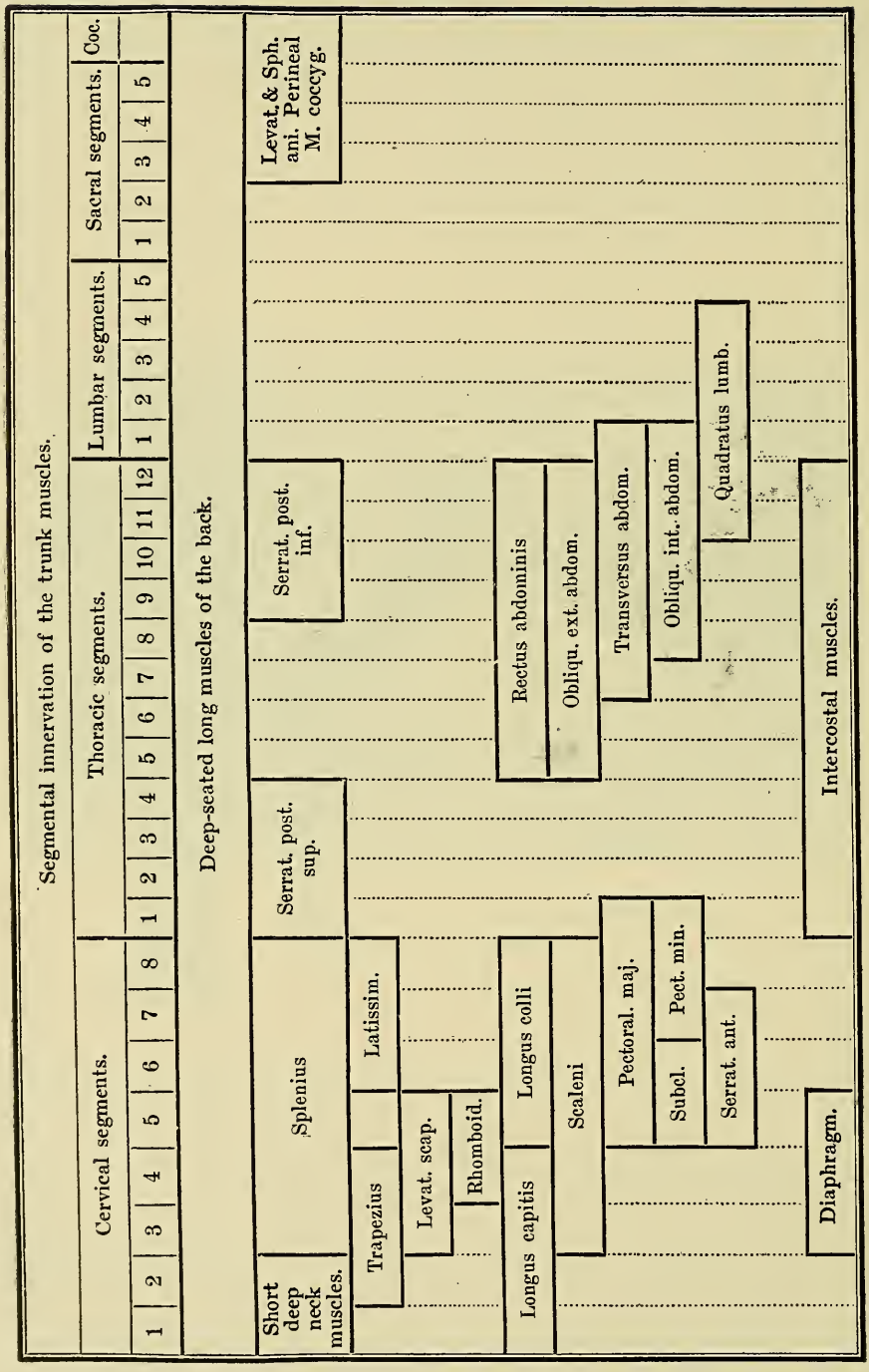
Fig. 48 shows the extent of anesthesia in total and symmetrical transverse lesions at different levels. The extent of motor paralysis is shown in the following tables after *E. Villiger*. For every assumed transverse lesion, the complex of muscles paralyzed is here exposed (those in the column representing the affected segment and all those to the right of this column). In injuries of the spinal cord (dislocation or fracture of the vertebral column) besides this paralysis, irritative symptoms from the region immediately above the lesion come into consideration in the clinical estimation of the case. So, for example, as shown in Fig. 49 in destruction of the 7th cervical segment, as a rule, the forearms are

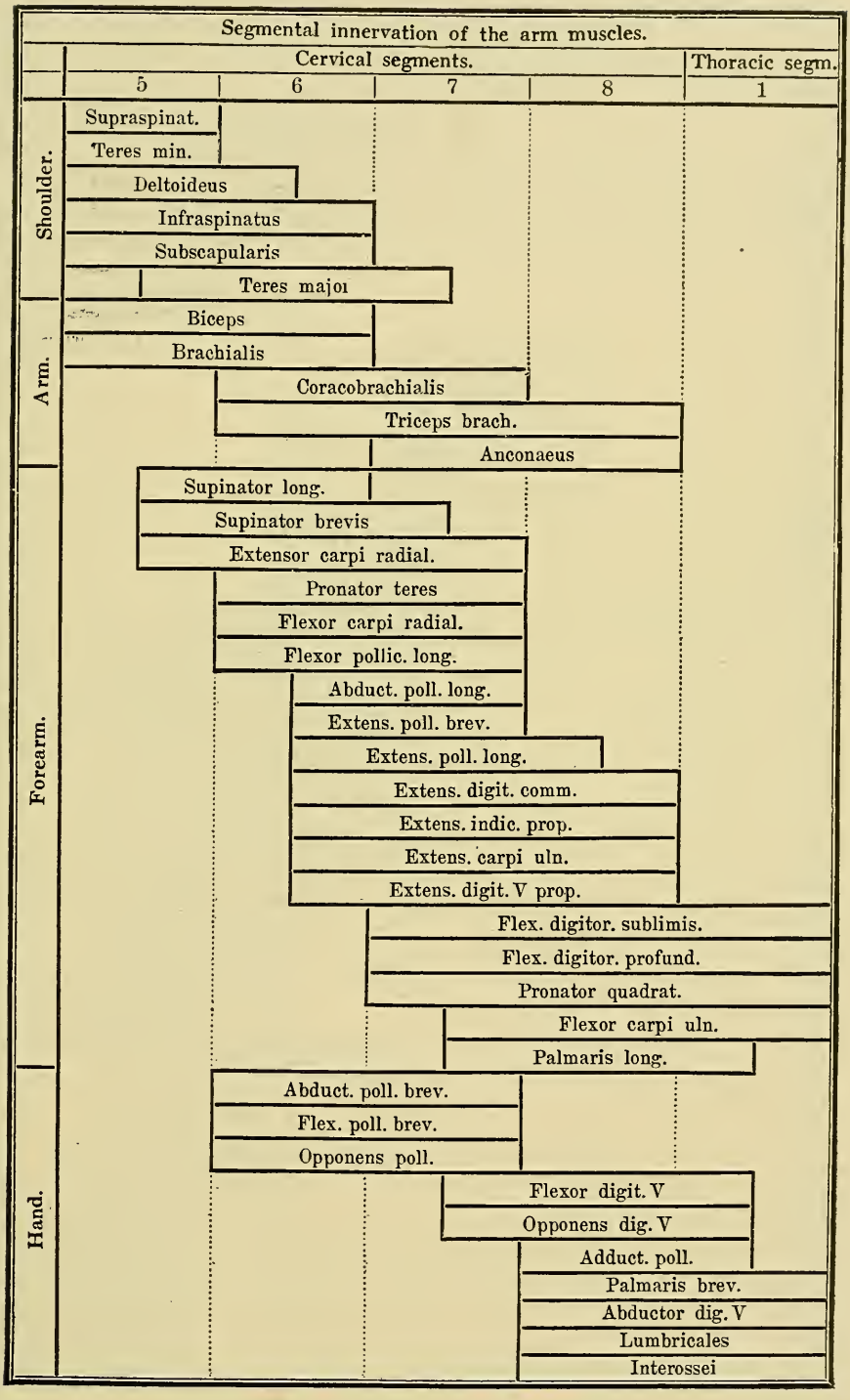
FIG. 49.

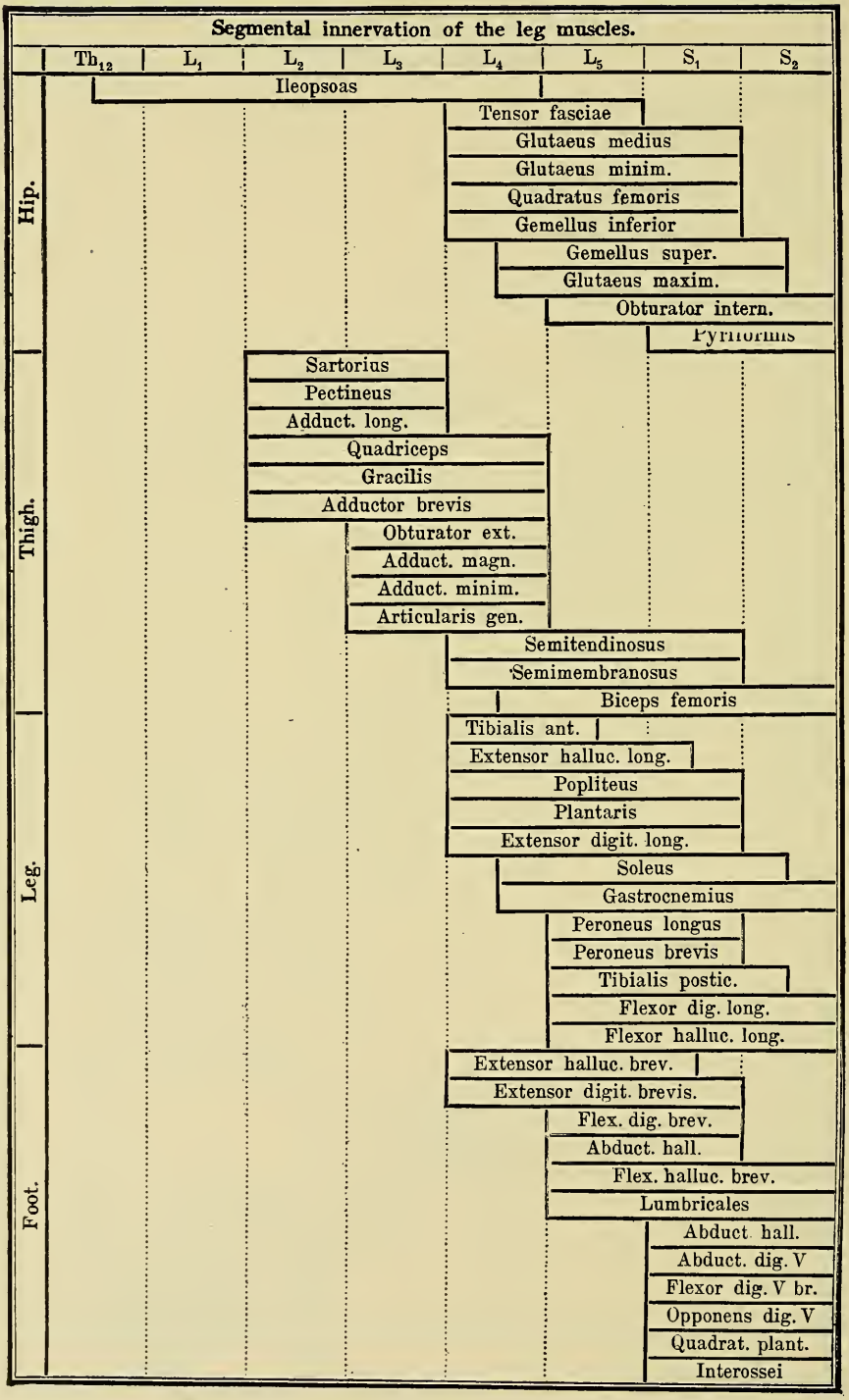
Complete Severing of the Spinal Cord at the Level of the VIIth Cervical Segment. (Vertebral Fracture.)

held in forced flexion by contracture of the biceps and the brachialis anticus muscles.

In total solutions of continuity, anesthesia extends, of course, to all qualities of sensation; indeed, all centripetal conduction is suspended; the motor paralysis, too, is absolute (no paresis, but paralysis), since there is severing not only of the chief motor tracts, that is the pyramids, but of each and every connection between the organ of the will and the spinal centers of movement. Now as to the character of the paralysis: according to what we have said in Lecture VII (page 112), a spastic paralysis would be expected, as below the level of the separation the reflex arcs are intact and freed by the lesion from inhibiting cortico-spinal influences. In complete transverse separation of continuity in the upper parts of the spinal cord, however, an absolute atonia and areflexia in the paralyzed region is found almost without exception, at any rate, in the early stages. It has been attempted to explain this as due to shock; this, however, is not satisfactory since, first, in many cases atonia and areflexia


Segmental innervation of the trunk muscles.
Cervical segments.
1 2 3 4 5 6 7 8
Thoracic segments.
1 2 3 4 5 6 7 8 9 10 11 12
Lumbar segments.
1 2 3 4 5
Sacral segments.
1 2 3 4 5
Coc.
Deep-seated long muscles of the back.
Short deep neck muscles.
Splenius
Serrat. post. sup.
Serrat. post. inf.
Levat. & Sph. ani. Perineal M. coccyg.
Trapezius
Latissim.
Levat. scap.
Rhomboid.
Longus capitis
Longus colli
Scaleni
Rectus abdominis
Obliqu. ext. abdom.
Transversus abdom.
Obliqu. int. abdom.
Quadratus lumb.
Pectoral. maj.
Subcl.
Pect. min.
Serrat. ant.
Diaphragm.
Intercostal muscles.


Segmental innervation of the arm muscles.
Cervical segments.
Thoracic segm.
5
6
7
8
1
Shoulder.
Supraspinat.
Teres min.
Deltoideus
Infraspinatus
Subscapularis
Teres major
Arm.
Biceps
Brachialis
Coracobrachialis
Triceps brach.
Anconaeus
Forearm.
Supinator long.
Supinator brevis
Extensor carpi radial.
Pronator teres
Flexor carpi radial.
Flexor pollic. long.
Abduct. poll. long.
Extens. poll. brev.
Extens. poll. long.
Extens. digit. comm.
Extens. indic. prop.
Extens. carpi uln.
Extens. digit. V prop.
Flex. digitor. sublimis.
Flex. digitor. profund.
Pronator quadrat.
Flexor carpi uln.
Palmaris long.
Hand.
Abduct. poll. brev.
Flex. poll. brev.
Opponens poll.
Flexor digit. V
Opponens dig. V
Adduct. poll.
Palmaris brev.
Abductor dig. V
Lumbricales
Interossei


Segmental innervation of the leg muscles.
Th12
L1
L2
L3
L4
L5
S1
S2
Hip.
Thigh.
Leg.
Foot.
Ileopsoas
Tensor fasciae
Glutaeus medius
Glutaeus minim.
Quadratus femoris
Gemellus inferior
Gemellus super.
Glutaeus maxim.
Obturator intern.
Pyriformis
Sartorius
Pectineus
Adduct. long.
Quadriceps
Gracilis
Adductor brevis
Obturator ext.
Adduct. magn.
Adduct. minim.
Articularis gen.
Semitendinosus
Semimembranosus
Biceps femoris
Tibialis ant.
Extensor halluc. long.
Popliteus
Plantaris
Extensor digit. long.
Soleus
Gastrocnemius
Peroneus longus
Peroneus brevis
Tibialis postic.
Flexor dig. long.
Flexor halluc. long.
Extensor halluc. brev.
Extensor digit. brevis.
Flex. dig. brev.
Abduct. hall.
Flex. halluc. brev.
Lumbricales
Abduct. hall.
Abduct. dig. V
Flexor dig. V br.
Opponens dig. V
Quadrat. plant.
Interossei

remain permanently; second, they may occur also in non-traumatic cases (for example, in transverse myelitis). Probably it is the result of the disturbances of lymph and blood circulation occurring in complete transverse affections high up in the cord which lead to serious functional injury of the posterior roots and anterior horn cells in the lower levels.

Complete transverse lesions of the spinal cord always lead to bladder and rectal disturbances; there is retention of stools; in the bladder, retention of urine is usually the most prominent symptom; when, however, the distention of the bladder has reached a certain degree, there can be a reflex involuntary dis-

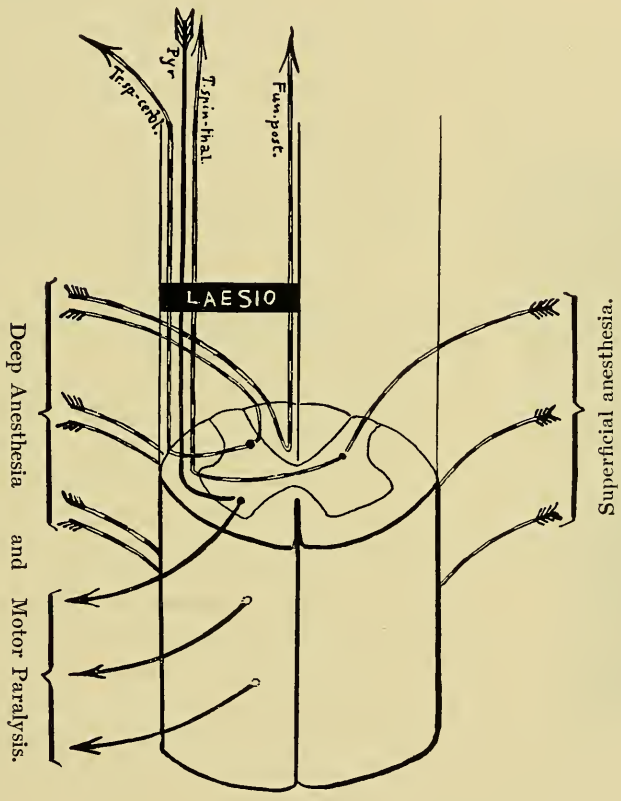


Fig. 50.

The *Brown-Séquard* Symptom-Complex.

charge of urine ("intermittent incontinence"). Only in lesions which are situated very low down and destroy the vesico-spinal and ano-spinal centers of the sacral region, other symptoms occur; either a continued dribbling of the urine ("permanent incontinence") or discharge of the urine in drops from the moment the filling of the bladder has reached a certain degree ("ischuria paradoxa"). Further, incontinence of the bowels. Of vasomotor and trophic disturbances are to be mentioned the dilatation of vessels to be observed in the paralyzed region in fresh cases, which later gives way to conditions of cold-

ness and cyanosis and the marked tendency to deep and rapidly spreading bed-sores.

Of great prognostic importance is the differentiation between partial and total transverse lesions. In making this, the following points are to be considered: In incomplete transverse lesions the paralysis is not symmetrical as in the complete ones and has, further, the tendency to partial recovery (naturally, provided the cause ceases); also when the lesion is located high, the patellar reflexes are never permanently lost, but, as a rule, exaggerated. Frequently there is a difference between them on the right and on the left. Vasomotor and sphincter disturbances are only present to a slight degree. Further, irritative symptoms may be evident distal from the lesion (pains, twitching of the muscles in injuries to the cord, also permanent erection of the penis, the so-called "priapism").

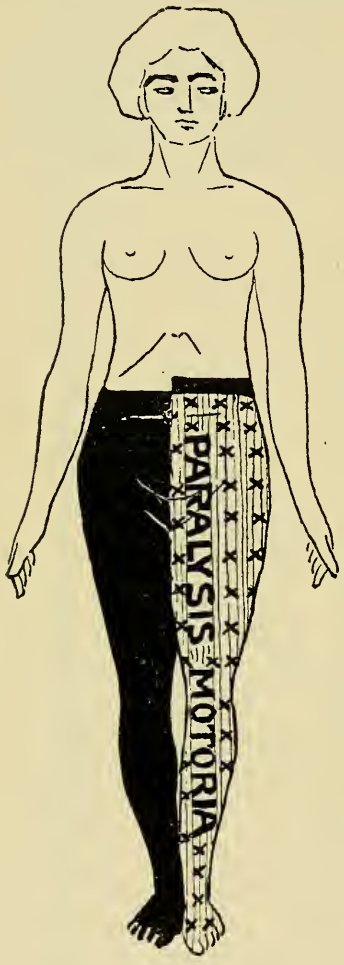


Fig. 51.

Brown-Séquard's Symptom-Complex. Lesion of the Left Half of the Spinal Cord.

Disturbances of Sensibility.

= Superficial Anesthesia.

= Deep Anesthesia.

= Superficial Hyperesthesia.

The phenomena which occur in an only half-sided transverse lesion of the spinal cord, most usually from trauma or in tumor formation, make up what has been called the "*Brown-Séquard* symptom complex." This is composed of the following symptoms which present themselves in the parts of the body distal to the seat of the lesion: On the side of the lesion motor paralysis, disturbance of the deep sensibility and hyperesthesia for touch; on the opposite side disturbance of the superficial sensibility, namely for pain and temperature stimuli.

The motor paralysis on the side of the lesion has naturally a spastic character; its method of production is readily understandable from Fig. 50. The disturbance of the deep sensibility (marked involvement of the sense of position, ataxia) on the same side, also considered in that diagram, is explained by the almost exclusively homolateral ascent of the paths for "bathyesthesia," that is, of the posterior columns and the direct cerebellar tracts. Also vibration sense is, as I have been able to recognize, disturbed or lost on the side of the lesion in *Brown-Séquard's* paralysis. The crossed superficial anesthesia affects always pain and temperature sense, since conduction of these qualities of sensation is almost exclusively carried out through fibers passing over to the other half of the spinal cord. In typical cases touch sense is altered, too, but much

less intensely. This, also, is explainable from the anatomico-physiological conditions. Tactile impressions are conducted toward the brain partly uncrossed (in the posterior columns) partly crossed (in the lateral columns). Sometimes there is neither tactile anesthesia nor hypoesthesia. The explanation of superficial hyperesthesia on the same side (which is only a temporary symptom) is difficult. Probably after a one-sided lesion, the cells of the posterior horn, which are intercalated in the crossed sensory tracts are loaded down with too much work (since now only these crossed tracts are open to tactile stimuli, part of which normally pass up in the uncrossed tracts) and this overwork manifests itself, until the organ has become accustomed to the new relations, by touch stimuli being perceived as painful.

If the unilateral lesion is located in the cervical region we have a "spinal hemiplegia," if it is in the dorsal region, a "hemiparaplegia." Fig. 51 illustrates such a case. The narrow anesthetic zone above the region of motor paralysis must be attributed to destruction of the posterior roots entering the affected region of the cord. Typical "classic" forms of the *Brown-Séquard* symptom-complex are, however, much rarer than atypical ones in which only an incomplete half-sided lesion or, on the contrary, one also affecting the other half of the spinal cord, is present.

2. DIFFUSE MYELITIS

We begin our consideration of the individual affections of the spinal cord leading to transverse lesions with Diffuse Myelitis, that is, with that inflammatory process, which in its diffusion in the diseased organ is not limited to definite anatomical elements (in contradistinction, for example, to "poliomyelitis anterior," which, as we will see, does not, as a rule, pass the boundaries of the anterior horns, and which also does not occur in diffused foci (like disseminated myelitis already mentioned in Lecture IX).

The myelitides present the following pathologico-anatomical characters: In very fresh cases, macroscopically the spinal cord in the affected regions appears swollen and somewhat red in color; in the advanced disease it may be softened to a discolored pulp. Microscopically the early stage of the disease is characterized by cloudy swelling and vacuolization of the ganglion cells, with loss of their finer structure as well as of the nerve fibers arising from them; further, by great dilatation of the vessels, minute extravasations of blood, infiltration of round cells, crowding of the perivascular lymph sheaths with granular cells, etc. In the later stages of the disease the infiltrative processes progressively decrease and "reparatory" proliferative processes in the neuroglia occur. Above and below the inflammatory foci the so-called "secondary degenerations" take place in the long fiber tracts of the spinal cord; following "*Waller's* law" (see page 10), the degeneration in the pyramidal tracts is descending, that in the posterior columns and in the spino-cerebellar tract, ascending (see Fig. 52). Purulent myelitis, in which there is breaking down of the spinal cord with pus formation and sometimes an abscess membrane, is excessively rare. Very infrequent also are the subchronic and chronic diffuse myeli-

tides in which parenchymatous destruction is combined from the start with proliferative alterations of the neuroglia and of the vessel walls.

From an etiological point of view the diffuse myelitides are quite heterogenous. The acute forms can, as a whole, be considered infectious; usually they are connected with general diseases (influenza, typhoid fever, acute rheumatism, variola, erysipelas, malaria, pneumonia, diphtheria, dysentery, gonococcemia, measles, syphilis). In the so-called "primary or idiopathic acute myelitis," for which sometimes exposure to cold, getting wet, and overexertion have been held responsible, most probably some obscure infection is always re-

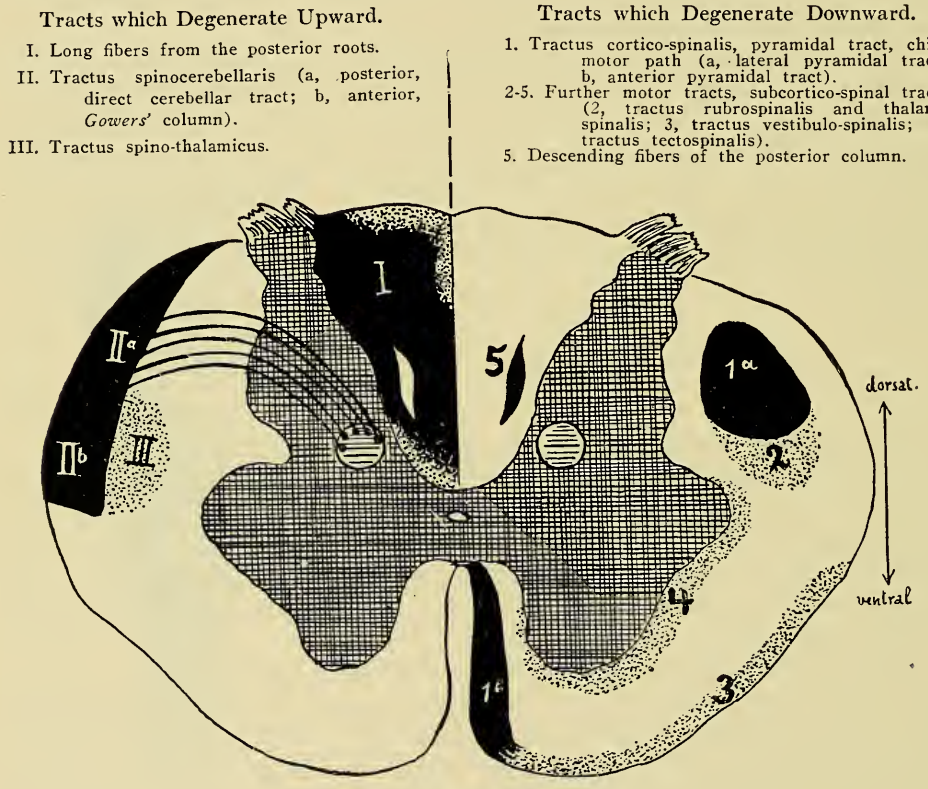


Fig. 52.

sponsible, and the factors mentioned have only acted as exciting causes. In fact, streptococci, staphylococci, and colon bacilli have occasionally been recognized in the diseased spinal cord. On the other hand, it seems certain that infectious myelitides can occur also without microbic invasion of the organ, usually from the action of toxines. For the subacute and chronic forms, syphilis comes particularly into question as a causal factor; occasionally, also, exogenic, chronic intoxications, as saturnism and alcoholism. Abscess of the spinal cord occurs almost only from metastasis, particularly after ulcerative endocarditis. A special position belongs to those forms of myelitis which occur in connection with multiple gas embolisms in the vessels of the spinal cord; these have been observed in consequence of the development of bubbles of air

in the blood upon abruptly passing from a high to a low atmospheric pressure. Workers on bridge building and in tunnels are particularly exposed to this "caisson paralysis" or "diver's paralysis" if special arrangements for a gradual "decompression" have not been provided. The development and course of transverse myelitides may be summed up as follows: usually a prodromal stage which is characterized by pain in the back, paresthesias in girdle form, sensation of drawing and formication in the extremities along with general malaise, shivering and rise of temperature, precedes the development of paraplegia. Only very rarely is the beginning abrupt, "apoplectiform." A very slow beginning is not frequent and is to be considered prognostically unfavorable as to restoration of function (provided that syphilis, against which we can act energetically, is not at fault). On the other hand the acute forms sometimes recover in astounding fashion. The longer the fever lasts the more unfavorable is the prognosis; it is further mainly dependent upon the seat of the transverse lesion. Myelitides of the cervical region, on account of their proximity to the medulla and its centers so important to life, is always very dangerous. The occurrence of broncho-pneumonia, large bed-sores, pyelo-nephritis, and the tendency of the disease process to extend further (ascending paralysis) are to be considered as prognostically unfavorable criteria. Interesting in a case of lumbo-sacral myelitis observed by me was the circumstance that the patient afterward, in spite of complete motor and sensory paraplegia, went through two labors, normal except for complete painlessness.

According to the symptomatology we can distinguish a number of varieties. First, according to the level of the spinal cord in which the transverse lesion has occurred, a lumbo-sacral, dorsal and cervical form, individual forms which in clinical importance are inferior to ascending myelitis, in which after beginning in the lumbo-sacral region continually higher levels are involved in the motor and sensory paralysis. In this, the inflammation extends by way of the perivascular lymph-sheaths upward, as numerous animal experiments conducted by *V. Salle*, under my direction, have shown. The central canal plays a very subordinate rôle. Cases in which the ascent occurs in exceedingly violent and steady manner, so that within a few days from their commencement death follows from paralysis of the muscles of deglutition and of respiration, are classed with the so-called "*Landry's* paralysis," or "paralysis ascendens acutissima." They belong properly, however, with affections which pathogenetically must be separated from transverse myelitis, namely with the ascending cases of poliomyelitis anterior which come under the head of the *Heine-Medin* disease (see Lecture XVI) and particularly, also, with those of polyneuritis acutissima. As "myelitis migrans" I have described a peculiar case in which a lumbo-sacral transverse paralysis, which had arisen acutely and was to be attributed to chilling and infection, recovered. Immediately afterward, however, paretic symptoms and disturbances of sensibility occurred in the trunk. These symptoms now ascended, in a step-like manner to a certain extent, upward, with recrudescence of the fever, while at the same time from below, a return to the normal occurred, likewise by steps. After the arms had been affected and already had begun to get better, the situation became extraordinarily dangerous on account of the occurrence of paresthesia in the region of the trigeminus and violent hic-

cough. At this critical moment the process came finally to a stop, and (in spite of the duration of the progressive period for weeks) at length recovered. This conduct can well be compared with that of "migrating erysipelas."

From a therapeutic point of view, active attack upon the cause is only possible in those forms depending upon malaria and syphilis. In a typical case of very severe syphilitic lumbo-sacral myelitis I saw a most startling effect from an intramuscular injection of 0.6 grm. of salvarsan. Six days after the injection the patient began to move the previously totally paralyzed legs again; on the thirteenth day he could stand, and in the end (after several, this time intravenous, injections of salvarsan) there was complete recovery. It may be attempted to treat the other infections by injections of colloidal silver (collargol, elektrargol). Whether these really are of value, or if in the cases which have recovered under this treatment it was merely an accidental coincidence, cannot yet be decided. For the rest, we must content ourselves with symptomatic treatment and care. Prevention of bed-sores by frequent change of position, protecting dressings, bathing with alcohol, with subsequent application of ointment of balsam of Peru, rubber rings, water-cushions, etc., prevention of cystitis by the most careful asepsis on catheterizing (which is only to be done when it cannot be avoided), giving urotropin, etc. Complications which have already arisen (bed-sores, cystitis, broncho-pneumonia) are to be treated with all care. To the treatment of the paralyses, what has already been said in the discussion of spastic spinal paralysis, syringomyelia, etc., applies; I would refer you also to my later remarks in Lectures XVI and XXII.

3. TUMORS OF THE SPINAL CORD

Under tumors of the spinal cord, in a clinical sense, we include both new formations developing in the spinal cord itself and extra medullary tumors which compress the spinal cord; the last are again divided into extradural and intradural neoplasms. Of growths within the cord itself, glioma, sarcoma, tubercle and gumma are particularly found; the intradural growths are chiefly fibroma and fibro-sarcoma (usually pedunculated), attached to a vessel. Less frequently myxoma, psammoma, teratoma, lymphangioma; while the extradural formations are chiefly lipoma and echinococcus cysts; still *Bircher* and I have described an extradural pedunculated fibrosarcoma of the cervical region, which had passed out through an intervertebral foramen and had led to the formation of a second tumor in the fossa supraclavicularis. The two tumors, joined somewhat like an hour glass, could be extirpated and the patient cured.

Etiologically, we have naturally definite information only about the infectious granulomata and the echinococcus cysts. Among exciting causes trauma must sometimes be considered; so, in my case of extramedullary sarcoma in the upper lumbar region (Fig. 53), the patient had been struck on the back with a cudgel.

The development of most cases of spinal cord tumor is marked by a prodromal stage with neuralgiform pains, lasting for months or sometimes for years, to be attributed to the pressure which the tumor (in the great majority of cases located extramedullarily and on the dorso-lateral surface of the spinal

cord and growing very slowly) exerts upon the posterior roots. Later the symptom-complex of transverse lesion usually develops and increases very slowly, the spastic paresis occurring early, the disturbances of sensibility later. These last are first manifested as hypesthesia of the feet, and then gradually ascend until an upper limit corresponding to the seat of the tumor is reached. Then, while the upper boundary remains fixed, the intensity of the sensory disturbances increases, eventually to complete anesthesia. Finally, disturbances of the sphincters are added; the legs can be drawn up in contracture as Fig. 53 shows. Almost always the *Brown-Séquard* symptom-complex is present for some time in typical, or more or less modified development. Earlier or later, there are also atrophic paralyses of certain groups of muscles; for example, of the small muscles of the hand, in tumors of the lower cervical region.

Since the treatment (except in the case of the gummata which can be cured by antisyphilitic measures) can only be surgical, exact localization of the tumor has, above everything else, great importance. Into the points of view important

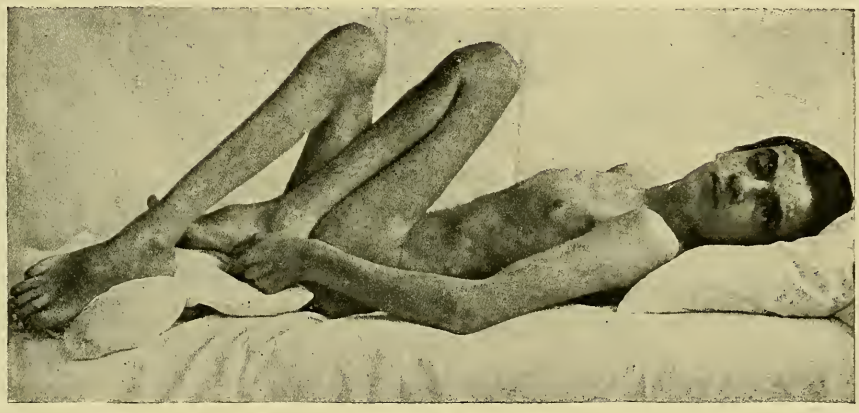


Fig. 53.

Paraplegia, with Extreme Contracture in Flexion, in Extra-medullary Fibro-sarcoma of the Lumbar Region.

to this end we cannot enter here; I would refer you to my "Compendium of Topical Brain and Spinal Cord Diagnosis," where this matter, which is too extensive for the scope of these lectures, receives a treatment sufficiently adequate for practical purposes. With very rare exceptions the intramedullary tumors present no indication for surgical intervention; not so the extramedullary, whose extirpation presents no great difficulties to modern surgical technique. It is noteworthy that from a clinical point of view all extramedullary tumors of the spinal cord, even the sarcomata, are to be considered benign, since after total extirpation no recurrence has so far been observed. The operation is to be considered as dangerous, both in the upper part of the cervical region and (on account of the danger of an infection of the hematoma) in the lower part of the spinal cord. The best outlook is furnished by tumors in the lower cervical and upper dorsal regions.

From the point of view of differential diagnosis, first "pseudo-tumor" is

to be considered, which, however, is only to be diagnosed by "autopsia in vivo," that is, by opening the vertebral canal, exploratory laminectomy. These cases are due to circumscribed cyst formations upon the basis of a localized serous meningitis which leads to local compression of the spinal cord in exactly the same manner as true tumor formations; further, must be mentioned, multiple sclerosis, a failure to recognize which, has led to many unnecessary operations. In order to protect yourselves as far as possible from such a serious error, take to heart the following rules: Multiple sclerosis develops, as a rule, slower than a neoplasm and often shows decided remissions in its course, which, in the clinical picture of tumor are either absent or are of very short duration. By closer analysis of the pain phenomena of which patients with multiple sclerosis complain, it is usually found that there is only very annoying paresthesia (a feeling of burning, of tension, of weight, of twitching). However, actual pains of severe, lancinating or pseudo-neuralgic character which are often plainly dependent upon the weather, do also occur. Pain in the joints speaks for multiple sclerosis. Disturbances of sensibility, which are sharply limited above, occur also in multiple sclerosis occasionally, but there is never complete anesthesia, as is frequently the case in the advanced stages of a tumor. The *Brown-Séquard* topography of disturbances is very unusual in multiple sclerosis.

Of the greatest prognostic importance is also the (often excessively difficult) distinction between extra- and intra-medullary tumors in which sometimes exploratory laminectomy alone can decide. In general, however, we have the following criteria (of which, observe, no one is pathognomonic): In favor of extramedullary location: 1. Slow development of the motor and sensory symptoms. 2. Tendency of the motor and sensory disturbances in spite of increasing intensity to remain stationary for a long time with regard to their extent upward. 3. Long duration of the *Brown-Séquard* syndrome. 4. The pseudo-neuralgic prodromal stage. 5. Considerable intensity of the spastic phenomena, which persists even after the occurrence of complete paraplegia. 6. The presence of motor irritative phenomena, spasms, twitching. 7. Slight development of degenerative-atrophic paralyses, striking lack of relation between the muscular atrophy and the only slight changes of electric irritability. 8. Sensitiveness to pressure over the spinal column. 9. Yellow color (xanthochromia) and increase of albumin in the cerebro-spinal fluid. On the other hand a typical and marked dissociated anesthesia speaks strongly for the assumption of an intramedullary tumor, while a simple prevalence of disturbance of the pain and temperature sense as compared to that of the other senses also is not rare in extramedullary tumors (great sensitiveness of the pain and temperature fibers to pressure).

4. LESIONS OF THE SPINAL CORD DUE TO AFFECTIONS OF THE VERTEBRAL COLUMN

We will only consider this subject quite cursorily, selecting what is important neurologically; the surgical side of the matter does not come within my province.

Luxations and fractures of the vertebral column can lead to sudden "pres-

sure paralysis" of the spinal cord or to complete compression or rupture of this latter. Luxations are more frequent in the cervical region and occur particularly between the 1st and 2d or between the 5th and 6th cervical vertebræ. Fractures, on the other hand, are strikingly more frequent in the lowest dorsal and upper lumbar region. The prognosis depends particularly upon the topography of the injury; if this has affected the upper cervical region death is usually instantaneous ("broken neck"), but even in the less fatal location of the lesion, bed-sores, paralysis of the bladder, cystitis, etc., present such great dangers that death can occur even weeks later.

Tumors of the vertebral column (above everything the dreaded cancer metastases after carcinoma of the breast, the uterus, the prostate, etc.), as well as *Potts'* disease (spondylitis tuberculosa) have, as a result, slow compression of the spinal cord. The clinical, neurological symptoms agree quite well in both processes. The picture is usually ushered in by shooting and tearing "root-pains," partly in girdle form, partly radiating into the legs, and of great intensity. Here the neuralgiform prodromal stage is, as a rule, of much shorter duration than in tumors of the spinal cord. Later, band-like hypesthetic zones usually develop, often, also (as the result of compression of the anterior spinal roots), atrophic paralyses of certain groups of muscles corresponding to the location of the pressure. An increasing exaggeration of the tendon reflexes in the leg, the appearance of the *Babinski*, *Oppenheim*, and *Mendel-Bechterew* reflexes, ankle clonus, etc., are the threatening forerunners of paraplegia, which, ushered in with increasing weakness of the legs, occurs at the start without hypertonia of the muscles, but later, however, can develop into marked spastic paresis. The tuberculous caries of the upper cervical vertebræ and of the atlanto-occipital joint which occurs both in children and in adults, presents a particularly typical picture, with which it is necessary to be acquainted, since here the "gibbus" which occurs in other locations is absent. The prodromal symptoms consist in stiffness of the neck and bilateral neuralgia of the occipital nerve, also in "*Rust's* phenomenon" in which the patient on changing his position supports his head with his hand. *Oppenheim* has also observed unilateral atrophy of the tongue and spinal accessory paralysis. Later, paralytic symptoms of motor and sensory nature appear, both reaching up to the neck.

As to the treatment of diseases and injuries of the vertebral column I would refer to the surgical text-books. As a palliative, "posterior radicotomy" (particularly dangerous here, however) in the affected region comes into question. To the treatment of the nervous symptoms remaining after cure of the causal lesions, the principles already repeatedly laid down apply.

LECTURE XI

The Syphilogenic Diseases of the Central Nervous System

As syphilogenic we denominate those lesions of the brain and spinal cord, for whose origin syphilitic disease in the affected individual is a prerequisite. Among these syphilogenic organopathies we must separate two categories from one another: 1, the syphilitic, and 2, the meta- or para-syphilitic affections. In the first, there is development of the pathologico-anatomical changes in general characteristic for syphilis, in the region of the nerve centers; in the last, however, there are degenerative lesions of definite portions of the cerebro-spinal apparatus, which histologically have nothing in common with the tertiary manifestations as we find them in other organs. Since now we will save true "lues cerebro-spinalis" for a later lecture, in the description of the meta-syphilitic nervous diseases, we will start with the so common tabes dorsalis.

The study of this will give us the opportunity of touching upon many points, which apply also to the other metasyphilitic affection of the central nervous system, to progressive paralysis, in the description of which we will hence be in a position to express ourselves somewhat more briefly.

A. Tabes Dorsalis

By this name, after *Romberg's* example, is quite generally denominated a disease for which *Duchenne*, of Boulogne, to whom we owe the first satisfactory description and nosological separation of the disease picture in question, in 1858, had proposed the name "progressive locomotor ataxia"; only in France is this last expression still used occasionally.

The syphilogenic nature of tabes is, on the other hand, a discovery of the last two decades which we owe particularly to the investigations of *Fournier* and of *Erb*. In his latest comprehensive statistics from the year 1901, *Fournier* was able to demonstrate, positively, a former syphilitic infection in 93 per cent. of tabetics; since then we have learned that also in tabetics who neither give the history of lues nor upon the most exact physical examination present residua of such a disease (scars, leucoplakia, glandular enlargements, etc.), serological recognition of syphilis is not rarely to be obtained (through the "*Wassermann* reaction" to be mentioned later). Even where this reaction is not positive, the possibility of a latent or hereditary syphilis is in no way to be excluded. *Mendel* and *Tobias*, in their cases of tabes in virgins, have been able with all certainty to demonstrate either a direct tabetic heredity or an acquired extragenital infection. To the very rare cases in which a tabetic has

recognizably acquired syphilis, any value as disproving the statement formulated by *Möbius*, "Nulla tabes sine syphili," is to be denied. Hence I would not hesitate to denominate a case presenting the clinical picture of tabes dorsalis, in which, however, acquired or hereditary syphilis is with certainty to be excluded as a "pseudo-tabes" (such a case, in fact, I never yet observed). To the conception of tabes belongs, according to our present view, along with the symptomatological criteria, the causal connection with syphilitic disease.

Now, however, arises the following question: "Why is tabes such a rare result of syphilis"? (*Reumont* has, for example, shown that among 3,600 syphilitics only 40, that is, 1.1 per cent., later became tabetic.) May not other factors be determining for the development of the disease (usually 5 to 15 years after the infection)?

In fact, the opinion is wide-spread that here neuropathic heredity plays a part. *Erb* has found in 28 per cent. of his tabetics hereditary neuropathic factors; in 42 per cent. individual nervous disturbances—not belonging to tabes. If tabetic patients are systematically investigated as to the so-called "stigmata of degeneration" (malformations of the skull, of the palate, of the ears "fissural angiomata," etc.), these are found more frequently and in the average in richer combination than in healthy persons. For the view that an originally defective structure of the spinal cord can predispose this organ to the development of tabes in case of syphilitic infection, the circumstance that it is in tabetics that most of the cases of heterotopia and abnormal course of the fibers have been described, is of some importance.

Along with reduced power of resistance (which in the origin of progressive paralysis appears to play the same rôle as in that of tabes) differences in the luetic virus also are probably determining factors in the later involvement of the central nervous system. There are apparently definite strains of spirochætæ whose toxines possess particularly strong neutropic properties—a "Lues nervosa," a "Syphilis a virus nerveux." We are forced to take this point of view by the numerous observations, in which, after the luetic infection of groups of people from a recognizable common source, syphilogenic diseases have later occurred in all or in a large number of those infected. *Erb*, for example, showed that 4 men infected from the same prostitute later all became tabetic or paralytic. *Brosius* saw, 12 years later, 5 of 7 victims of an epidemic of syphilis among glass-blowers; 4 of these were tabetic or paralytic. In an entirely analogous manner must be interpreted the considerable number of recognized cases of family tabes or general paresis, in which father or mother, or both parents, along with one or several children, are afflicted with tabes. Family predisposition cannot always be invoked here, since, as a rule, the parents are not consanguine. As group diseases in a small way, the not at all infrequent cases of conjugal tabes or paresis can be subjected to the same method of consideration.

An important question is whether the insufficiently treated syphilitic runs a greater risk of later metaluetic disease than one subjected to adequate treatment. Many authors, particularly syphilologists, have decided this question in the affirmative, while, on the other hand, there are those who would make mercurial intoxication in part responsible for the origin of tabes and

paresis. According to my idea, both views are incorrect. What could be more conclusive than conjugal cases of tabes in which we hear that one of the pair has had extended anti-syphilitic treatment, the other none at all, and where still both have equally acquired tabes? We will occupy ourselves more extensively with this question in the next lecture.

Edinger has taken a totally different standpoint, in that he attributes to functional use (that is, to the measure of work required from the metaluetic spinal cord) a considerable rôle in the origin of tabes. In many cases this view has much that is illuminating, namely, in those in which tabes begins quite acutely after severe bodily excesses* or the first symptoms are noted in an extremity continually used. Not rarely, however, *Edinger's* "overuse theory" fails us, and we know tabetics enough whose disease remains particularly mild and stationary, although they, in spite of all medical advice, continually overexert themselves (for example, as hunters and riders).

Probably race predisposition comes also into question in the tendency of syphilitics to develop metaluetic nervous diseases. It has been scientifically demonstrated that in many races saturated with syphilis, tabes either does not occur or is extraordinarily rare (for example, among the Kurghise of Central Asia, the negroes and mulattoes of North America, the Arabs of Algeria, etc.† Finally, as occasional accessory factors, getting chilled or wet and trauma should be mentioned, which, along with overexertion, explains the fact that after campaigns an increase in the number of cases of tabes has been repeatedly observed among the combatants.

Pathological Anatomy

Tabes is pathologico-anatomically a systematic degeneration in the region of the posterior roots of the spinal cord.

We call systematic, as already indicated (page 111), those diseases of the spinal cord whose lesions are limited to definite fiber systems (as, for example, amyotrophic lateral sclerosis and tabes), while "asystematic" or "diffuse" affections are those whose anatomical substratum is not limited to definite neurons (as multiple sclerosis and tumors of the spinal cord).

A knowledge of the structure of the posterior spinal roots and of the further course of their fibers is of great importance for the understanding of the pathological anatomy of tabes dorsalis.

The fibers of the posterior roots have their cells of origin in the spinal ganglia; these cells of origin, through one of their processes which runs in the course of a nerve trunk, are in relation with the different peripheral apparatus, which we find partly as free nerve endings, partly as terminal bodies (touch cells and bulbs, the *Vater-Pacini* corpuscles, etc.) in the integument, the mucous membranes, the mesentery, the joint surfaces, etc. The other or central process of the spinal ganglion cell, however, enters the spinal cord through a posterior root.

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* Corresponding to a certain extent to the generally recognized predisposing rôle which mental overexertion, excitement, etc., play in the causation of progressive paralysis.

† This statement is incorrect in so far as it relates to the American negro.—*Translator.*

The posterior root fibers, however, we separate into different categories, according to their further course. The "short fibers" pass directly through the rim zone of the posterior horn into the gray matter and break up either about the cells of the anterior horn, or about those of the posterior horn, on the same side of the cord. In the first instance they serve to convey reflex

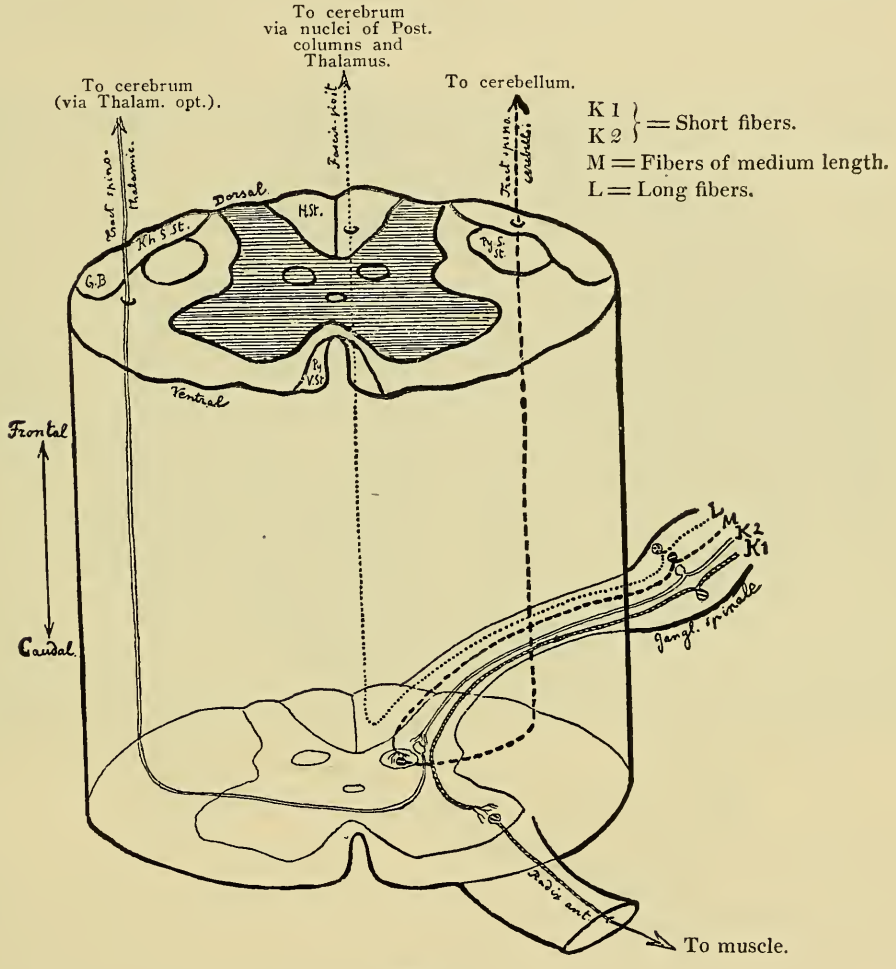

Fig. 54.

The Different Categories of Fibers in the Posterior Roots and Their Continuation in the Spinal Cord.

G. B. = *Gower's* tract.
H. St. = Posterior column.
Py. V. St. = Anterior pyramidal tract.
Kh. S. St. = Direct cerebellar tract.
Py. S. St. = Lateral pyramidal tract.

stimuli to the anterior roots, in the second to conduct pain, temperature and touch stimuli to the so-called "Tractus spino thalamicus," which, after crossing to the other side of the cord, conducts these sensations toward the brain. The "middle fibers" of the posterior roots pass through the posterior roots on into the posterior columns, and from there into the base of the posterior

horn on the same side, and end about the cells of the column of *Clarke*, from which the stimuli transmitted to the cord through this set of neurones are carried in the direct cerebellar tract to the cerebellum. The third variety, the "long fibers" of the posterior roots also enter the posterior columns through

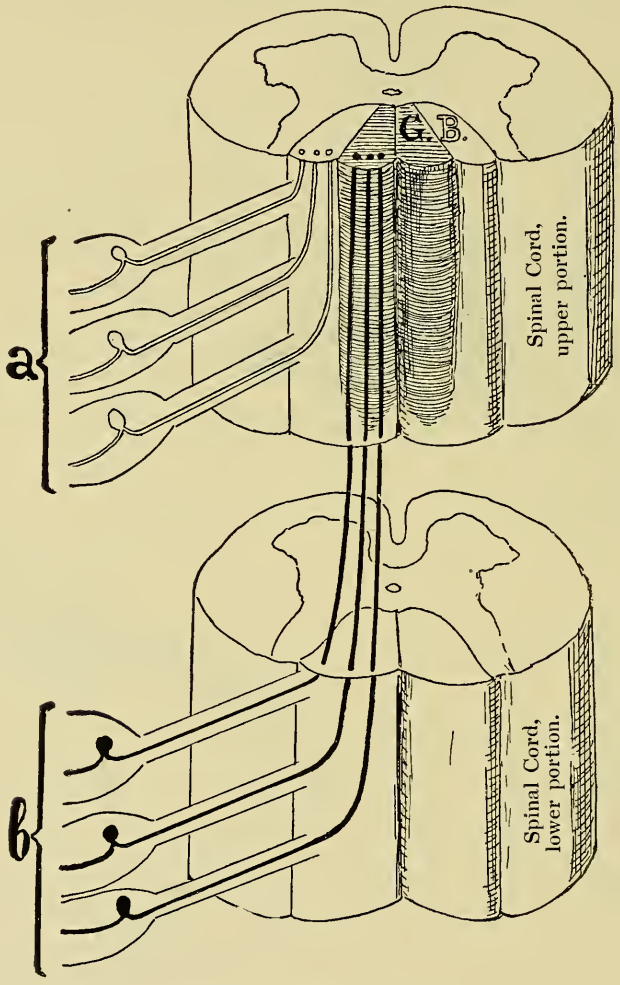

Fig. 55.

Structure of the Posterior Columns.

G—Column of Goll (Funiculus gracilis).
B—Column of Burdach (Funiculus cuneatus).
a—Long fibers of the posterior roots from the upper half of the body.
b—Long fibers of the posterior roots from the lower half of the body.

the posterior root zone, then run in these columns upward, and end in the "nuclei of the posterior columns" in the medulla, from whence the conduction of stimuli is transmitted via the optic thalamus to the opposite side of the cerebrum.

The fibers of the posterior columns as well as those of the direct cerebellar

tracts conduct "deep sensibility," with the difference, however, that the spino-cerebellar tracts serve to transmit subconscious perceptions (of tactile and coördinatory nature), while the posterior column, along with the same function, also serves to carry the conscious perceptions of position sense and movement sense to the cerebrum.

The following anatomical peculiarities of the fibers of the posterior columns should also be noted: 1. While the long fibers entering the posterior column

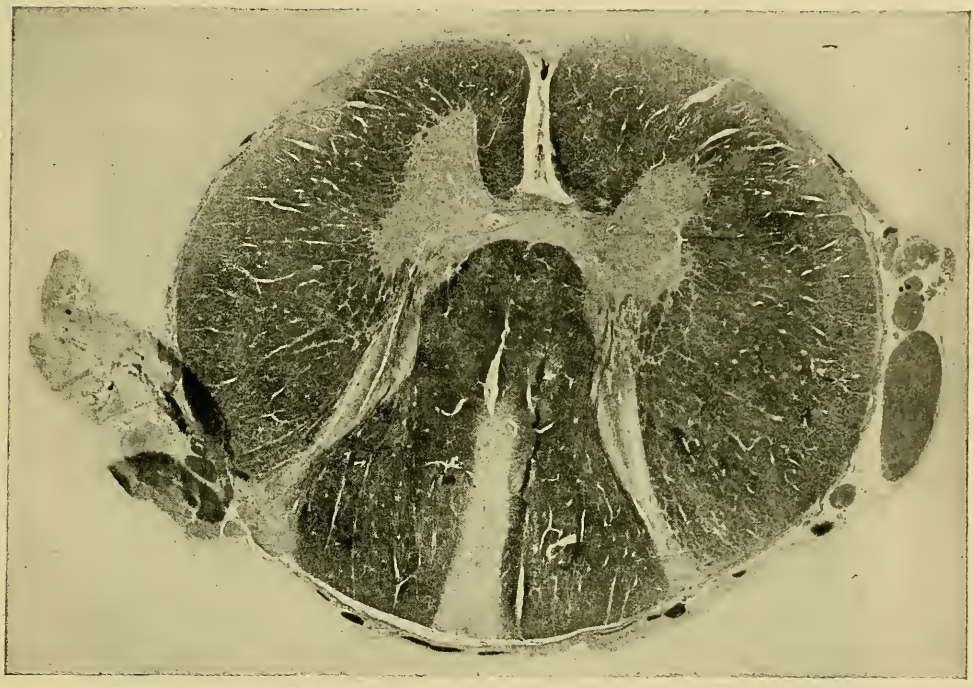

Fig. 56.

Lumbar Tabes. (Cervical Region.) *Weigert-Pal* Stain.

through a definite posterior root run upward, they are continually forced more and more toward the median line by fibers entering higher segments through the posterior root zone. On account of this, for example, in a cross section through the cervical region, the fibers of sacral origin lie next to the septum, those from the lumbar region farther out, then the dorsal fibers, and finally, next to the posterior horn, those of the cervical neurones.

In the cervical cord, however, a macroscopical separation has occurred in this way between the long fibers of the posterior roots which come from the lower half of the body (that is, from the 4th thoracic segment downward), which are united in the columns of *Goll*, and those from the upper half of the body (that is, from the 4th thoracic segment upward) which make up the column of *Burdach* (see Fig. 55).

2. Each one of the long fibers of the posterior roots, before it turns upward in the posterior columns, gives off a branch (a collateral) which descends some distance, also in the posterior column. (In Figs. 54 and 55 the

collaterals are omitted for the sake of simplicity.) These descending elements of the posterior root system lie together in a definite bundle, the so-called "*Schultze's* comma" (see above Fig. 52, page 158); this last, for example, in the upper lumbar segments contains no lumbar fibers, but those which have come in at a higher level of the spinal cord, that is, in the instance selected, belong to the dorsal segments.

3. The long fibers of the posterior root system are not the only nervous structures of the posterior column area. Rather are there mixed among them the so-called "endogenic" fibers of the posterior column, the so-called intersegmentary or association tracts, which connect with one another different levels of the posterior horns. At certain places in the posterior column area the endogenic fibers lie so close together that on cross sections the long fibers of the posterior roots appear quite thinned out. These "endogenic fields of the

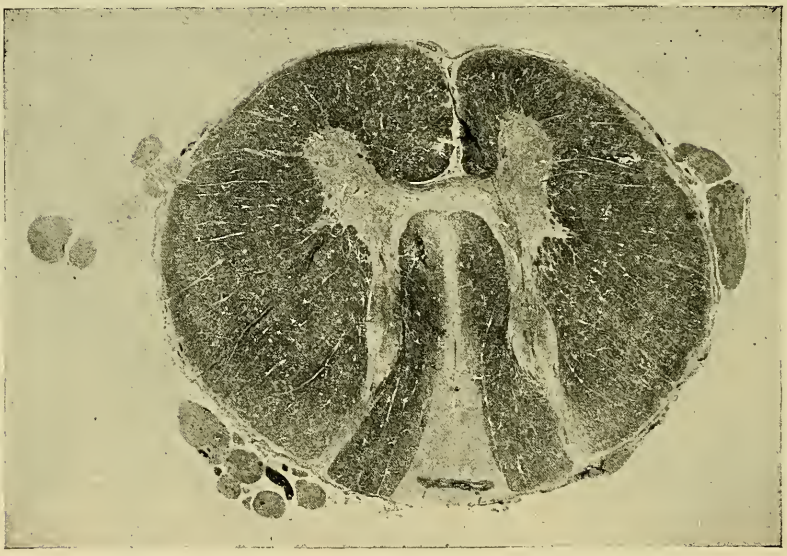

Fig. 57.

Lumbar Tabes. (Dorsal Region.) *Weigert-Pal* Stain.

posterior columns" are, in the neighborhood of the commissure, the "ventral field of the posterior column" (Zona cornu-commissuralis), at the periphery of the spinal cord, along the septum, the "median peripheral field of the posterior column" (Zona septo-marginalis).

Since now tabes, in the great majority of cases, begins as "tabes inferior," that is, as degeneration of the posterior roots in the lumbar region; after the above remarks it is understandable that in the early stages of the disease, in sections through the higher levels of the spinal cord, it is only the mesially placed long tracts of the posterior column, that is, those which have come up from the lumbar region; in other words, the columns of *Goll*, which are degenerated (see Figs. 56 and 57). In sections through the lumbar segments, however, a degeneration involving also the lateral portions of the posterior column is found, indeed, it is most marked in the region of the posterior root

zone (see Fig. 58). Upon careful microscopical examination it is found further, that "*Schultze's* comma" is not affected by the degeneration. This is not remarkable, since its fibers come from the higher roots not yet diseased. The "endogenic fields of the posterior column" are in contrast with the others by being relatively intact, since they carry a large number of fibers which have nothing to do with the posterior root system. In advanced cases of tabes on account of the involvement of higher segments of the cord in the disease process, "*Schultze's* comma" degenerates also, and finally even in the cervical region all the posterior area is involved.

The central point in tabetic disease of the spinal cord is the primary degeneration of peripheral sensory neurones. In this there remain longest

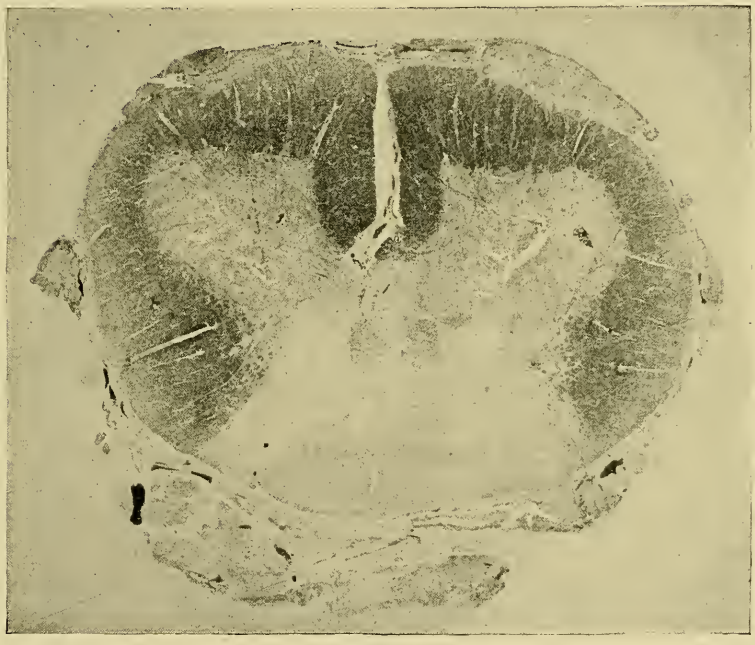

Fig. 58.

Lumbar Tabes. (Lumbar Region.) *Weigert-Pal* Stain.

intact (at least, according to external appearance) the cells themselves, that is, those of the spinal ganglia, while degeneration shows itself first in their central processes (posterior roots and posterior columns). Their peripheral processes (which are to be sought in the sensory nerves) usually are only found altered in advanced stages of the disease. That the radicular and medullary part of this neurone is more intensely affected than the peripheral part is partially explained by the following discovery of *Obersteiner* and *Redlich*. These authors showed, namely, that at the point of entrance of the posterior roots into the spinal cord there is normally a sort of strangulation of the root bundle, since at this point the pia is thickened and is covered with a particularly close layer of glia. This circumscribed part of the posterior root forms evidently a "Locus minoris resistentiæ." On this account,

even in incipient tabes, it is in the lumbar region where these constrictions are most marked that the roots are earliest and most intensely affected. The whole sensory neurone of the first order seems on account of the method of development peculiar to it (the sensory nerves grow in ontogenesis only secondarily into the spinal cord) to have become the specially vulnerable part of the central nervous system. A certain specific affinity of metasyphilitic toxines for this system of fibers (also for the others affected by tabes, *e.g.*, the optic nerve) seems probable, indeed.

Symptomatology

As we now proceed to the consideration of the manifold and interesting symptomatology of tabes dorsalis, regardless of the different clinical pictures which it can present to us, we will next pass in review all its disease symptoms in as rational grouping as possible. We will begin, of course, with those symptoms which can be directly attributed to the characteristic anatomical findings in the posterior root affection.

This posterior root syndrome is made up of the following components:

1. ATAXIA

We have already learned in the first lecture that from disturbance of such centripetal impulses as inform us at any given movement of the position of our limbs, the clinical phenomena of ataxia or disturbance of coördination results.

The ataxia of the tabetic produces an exceedingly typical disturbance of gait, hence the name "progressive locomotor ataxia" which *Duchenne*, of Boulogne, applied to this whole nosological unit. There is a "dysmetria" in the movements necessary for progression, in that the legs are swung out and shoot forward beyond the measure of a normal step; further, the steps are of unequal length. We notice also abnormal positions of the joints during locomotion, so that, for example, the swung leg remains strongly extended at the knee, the point of the foot is directed too much outward and the foot strikes the ground not with the ball, but with the heel. In the most extreme degrees of tabetic ataxia walking becomes impossible, even when the patient is supported on both sides, as every attempt to make steps degenerates into a series of contrary movements in the lower extremities.

On the other hand, in the early stages of tabes, the incoördination of the locomotor mechanisms is not to be demonstrated without special tests. Some easily applicable tests adapted to bring out an ataxia not evident in ordinary locomotion are walking on a straight line (in which each foot is alternately to be placed exactly in front of the other), quick turning about at command, walking on the tips of the toes or with the knees bent, walking backward, walking up and especially downstairs, finally walking with closed eyes.

This last test makes evident to a certain extent the compensating influence which the optical control of movements can exercise upon the coördination of these last in tabetic patients; also walking in the dark is considerably

more difficult for them. This influence, however, makes itself felt not only in relation to locomotor or dynamic, but also in static ataxia. Upon this depends the well-known *Romberg's* symptom; if a tabetic is asked to stand with his feet pressed close together (that is, his base of support is made as small as possible), and then to close his eyes, he immediately begins to stagger and would fall without support. Where in light or incipient cases *Romberg's* phenomenon is not present or is imperfect, it is well to test coördination by having the patient stand on one leg. It is then noticed that where this last is disturbed, the patient is unable to balance himself on one leg when he closes his eyes, often even with open eyes. In advanced stages of the disease static ataxia often appears spontaneously in that the body of the patient, when he is sitting upright, sways continually from side to side.

Tests for demonstrating ataxia of the legs when the patient is lying down are the following: He is asked to describe a circle with each foot; to touch an object held before him with his toe; to touch one knee with the opposite heel (first with open and then with closed eyes). For testing for ataxia in the upper extremities the finger-finger and finger-nose tests, in which the patient first with open and then with closed eyes brings his index fingers together in front of him, or touches the point of his nose, are customary.

2. HYPOTONIA

Since for the preservation of tonus a continual passage of stimuli from the posterior roots* to the motor cells of the anterior horn is necessary, it is not remarkable that the prevention of this passage by the tabetic degeneration of the posterior roots leads to an abnormal relaxation of the muscles. In the region of the pelvis and of the lower limbs the hypotonia is shown by the patient being able to spread his legs abnormally wide apart, or in that we can bring the leg, extended at the knee, into an acute angle with the trunk, and under certain conditions, indeed, we can in this manner, without trouble, "make a shoulder of the leg"—things which the healthy person can do only after long training (the "grand écart" of the ballet dancers, the "art" of the "snake-man"). In the knee, on account of the relaxation of the biceps, semitendinosus and semimembranosus a hypotonic subluxation backward, the so-called "Genu recurvatum" occurs. Fig. 59 shows this deformity, which, however, is not pathognomonic for tabes dorsalis, but can occur in muscular dystrophy, neuritis, etc. On the other hand, hypotonus of the quadriceps sometimes permits bringing the heel up against the buttock.

3. AREFLEXIA

While in the very first stages of tabes increase of the tendon reflexes is not rare (evidently as a symptom of irritation of the posterior roots), in the course of the disease these reflexes almost always gradually disappear. On

* This is by way of the short posterior root fibers which enter the anterior horn directly and break up about its motor cells.

account of the intimate relation between tonus and reflexes the pathogenesis of this hyporeflexia and areflexia corresponds to that of tabetic muscular relaxation (see also Lecture I, page 7). Most important among the reflex anomalies of tabes is the so-called *Westphal's* phenomenon, the loss of the patellar reflex. Complete loss can only be diagnosed when striking upon the patellar tendon produces no contraction in the quadriceps muscle, all tension of the thigh muscles, voluntary or involuntary, being avoided. An excellent means of attaining the best conditions for this experiment is furnished by the so-called "*Jendrássik's* maneuver"; in this the patient sits with loosely crossed legs on a comfortable chair, his head thrown back, looking at the ceiling, and the fingers of each hand hooked with those of the other; after the patient has been previously instructed to pull hard with his hands as the last number is given, 1—2—3 is counted, and immediately, while the patient's attention is directed away from his legs, a firm stroke is made with the percussion hammer upon the ligamentum patellæ. The patellar reflex can also be tested while the patient is lying down; the leg to be tested, the knee bent, is lifted from the bed by an aide, the patient being entirely passive and relaxing his muscles. [The limb is best rotated slightly inward.—*Translator.*] Testing the Achilles tendon reflex is almost as important as that of the patella tendon; it is most easily accomplished if the patient kneels upon a well-cushioned chair and turns his back to the examiner, the feet hanging over the edge of the chair. The different tendon and bone reflexes in the upper limbs (see table, page 8), usually after exaggeration at the start, are lost later than those of the lower extremities (except in the so-called tabes superior).

Fig. 59.
Genu Recurvatum.

Before reduction or loss of the tendon reflexes are interpreted as indicating tabes dorsalis, it is necessary to find out by questioning if the anomaly can be brought into connection with any former local disease; so, for example, a sciatica very often leaves behind a hypo- or areflexia of the Achilles tendon on the same side.

Loss of reflex on one side, or difference between the reflexes on the right and on the left, are frequent symptoms in the course of tabes dorsalis. In contradiction to the tendon reflexes, the skin reflexes are almost always retained; often, indeed, they are abnormally lively.

4. DISTURBANCES OF SENSIBILITY

Even in the early stages of tabes, disturbances in the power of appreciating different qualities of sensation, which in advance stages of the disease may increase to anesthesia proper, make themselves felt. Both skin and deep sensibility may be affected (see page 6).

a. Disturbances of Superficial Sensibility

As to the location of the hyperesthesias and anesthesias to be found on the skin, as a rule they plainly follow the so-called "radicular type" in that the areas of reduced or lost sensation do not agree with the distribution of peripheral nerves, but on the extremities involve more or less longitudinal areas; on the trunk, circular zones.

The difference between "peripheral" and "radicular" topography of disturbances of sensibility is explainable through the following relations. Each spinal ganglion sends its peripheral processes into different sensory nerves, each sensory nerve contains fibers which originate in different spinal ganglia. Independent of the often complicated paths which they have followed in the peripheral nerve, at the extreme periphery, that is, on the surface of the body, the sensory fibers so arrange themselves that those arising from a definite spinal root supply a definite region. This region is called a "radicular zone" or "root field." In it the sensory fibers originally united in one posterior root come together again, even when they are transmitted to the skin through different peripheral nerves. Figs. 60 and 61 expose the difference between "root fields" and peripheral nerve areas. In them the radicular zones are represented as occupying the area on each side of the line which bears their root number. The root fields of the individual posterior roots overlap one another somewhat like the tiles on a roof. The special arrangement of the radicular zones in the extremities, in which in contradistinction to the circular arrangement on the trunk a longitudinally directed distribution prevails, is explainable from their ontogenetic relations. In the embryo, the first beginnings of the limbs, growing out from the trunk, take with them the "dermatotomes" (fetal skin segments) lying in their way and corresponding to the same segments of the cord, and since the limbs grow out more or less vertically from the axis of the trunk, in them the principle of circular arrangement is not impaired by the extension of the segmental distribution in the direction of their axes (longitudinally).

The radicular hypesthetic or anesthetic zones are found particularly frequently on the inside of the leg and foot and on the inside of the upper extremities; often, also, in the form of a girdle or a half girdle (since asymmetry is not at all rare) they surround the thorax or the epigastrium (see Fig. 62). In the early stages we find sometimes instead of the more or less bandlike hypesthetic or anesthetic zones, insulated areas of hypesthesia and anesthesia, usually asymmetrically distributed. So much for the topography of the tabetic sensory disturbances in the integument. As to their quality, they can affect equally the tactile, the pain or the temperature senses. Usually one

variety of sensation is more affected than the others, and this is usually the pain sense. Apart from diminution of sense perception we often find delay in conduction in which the patient first experiences pain some seconds after a pin prick. If in such cases tactile sense is undisturbed, one prick can

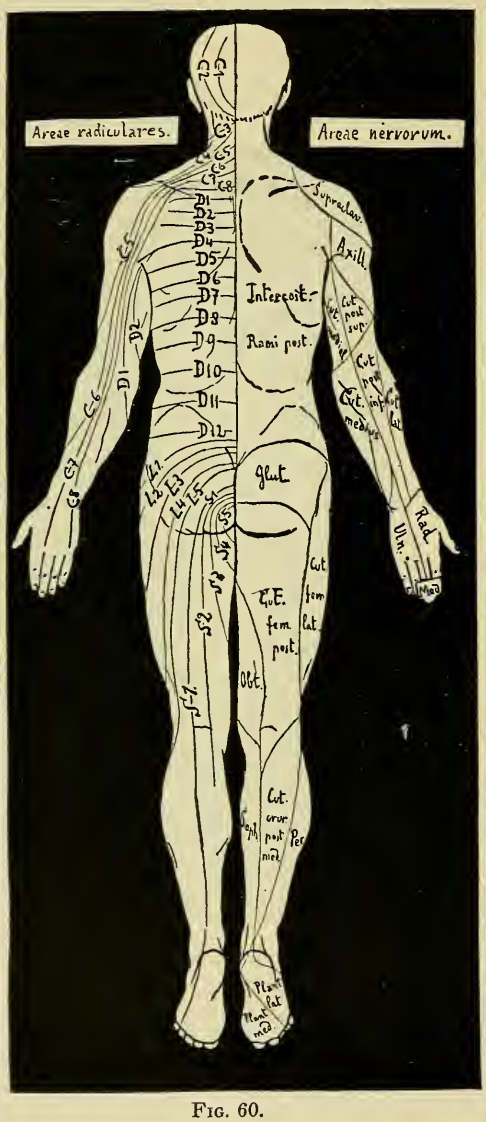

Fig. 60.

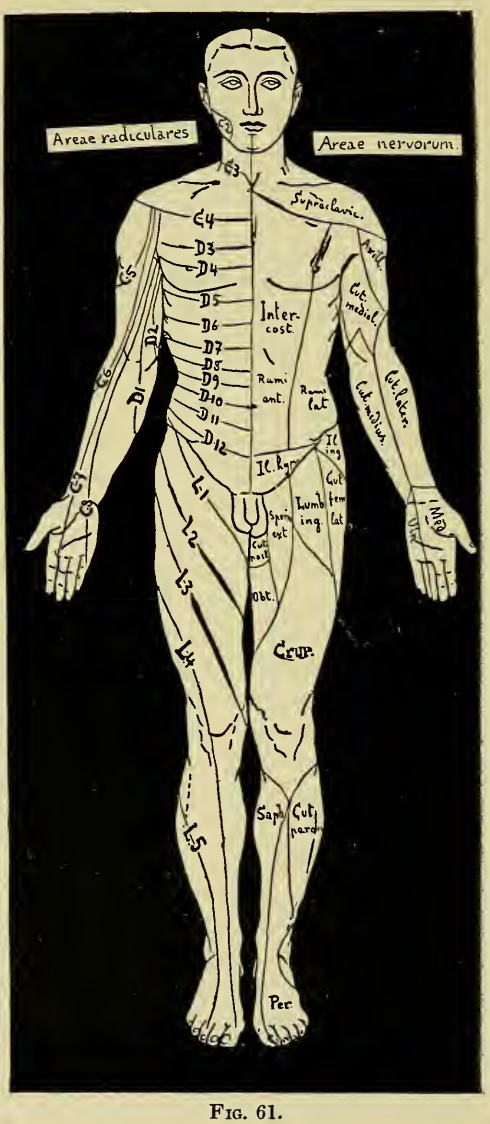

Fig. 61.

produce two sensations, in that it is experienced first as a touch, and somewhat later as pain. Further, the quite frequent faulty localization of touch, pain and temperature stimuli must be mentioned; if this is so marked that the patient, his eyes closed, when solicited to indicate the spot irritated, mistakes the side of the body, that is, locates a prick on the right hand on the left side, we speak of "Allocheiria." Hypesthesia or anesthesia of the soles

of the feet, a very common tabetic symptom, is usually experienced as very disagreeable by the patient and impairs his power of locomotion.

b. Disturbances of Deep Sensibility

Strictly considered, ataxia, already studied, belongs among the disturbances of deep sensibility, as shown in Lecture I. Besides this, interference with "bathyesthesia" is evident in most cases of tabes after they have reached a certain degree of intensity. One can convince himself of the loss of "position sense" without difficulty if, having the patient close his eyes, one extremity is put in a certain position and the subject is requested to put the opposite one in an exactly similar position. The gross errors which he makes give evidence of the fact that he has lost control over the relative attitude of his limbs without the assistance of the visual sense. In analogous manner loss of "movement sense" may be recognized. Disturbances of the position and movement sense in the fingers lead to "stereo-anesthesia," that is, inability to recognize the form of objects by handling them.

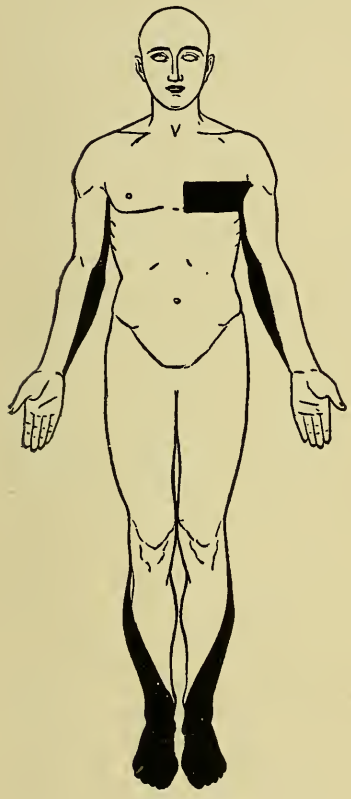

Fig. 62.

Typical "Radicular" Disturbances of Sensibility in a Case of Tabes Dorsalis.

The testing of bone sensibility with a tuning-fork can render valuable service even in the early stage of tabes. The loss of vibration sense, or its manifest reduction, is, as I have recognized, the most delicate reagent for detecting beginning lesions of the posterior columns.

To the early disturbances of deep sensibility usually belong also anesthesias of certain nerve trunks and viscera. The severe pain which firm compression of the ulnar or peroneal nerves normally produces is no longer to be elicited (*Biernacki's* symptom). The same remark applies to the exceedingly unpleasant sensation which energetic pinching of the Achilles tendon produces in healthy people; this analgesia of the Achilles tendon is called *Abadie's* tabes symptom. Of the frequent visceral analgesias to pressure I would mention those of the eyeball, the larynx, the trachea, the breast, the ovary and the testicle.

5. TROPHIC DISTURBANCES

We must assume that, normally, the trophic influence which the cells of the anterior horn of the spinal cord exert upon the bony skeleton and the integument are stimulated to a certain extent reflexly through the elements of the posterior root system. In this manner can be explained the fact that

tabetic disease of the posterior roots can sometimes lead to trophic disturbances in this apparatus.

In the skeleton the so-called "tabetic spontaneous fractures" which depend on pathological brittleness of the bones deserve special consideration. They occur from the slightest causes; for example, I saw fracture of the neck of the femur from stepping down from the pavement; fracture of the radius from striking the swinging arm against the back of a chair. Characteristic of these fractures, whose occurrence is sometimes the first symptom calling attention to the presence of tabes, is their painlessness, an important evidence of the already mentioned deep anesthesia. Repair of these fractures sometimes occurs normally; occasionally, however, the callus formation is defective, so that pseudo-arthroses occur; on the other hand, an excessive callus formation (leading to severe disturbances of function) has repeatedly been observed. The X-ray examination of the bones of tabetics, which tend to undergo spontaneous fracture, shows thinning of their compact layer; besides this, a dilatation of the *Haversian* canals and a poorness in lime salts of the bony tissue is found histologically.

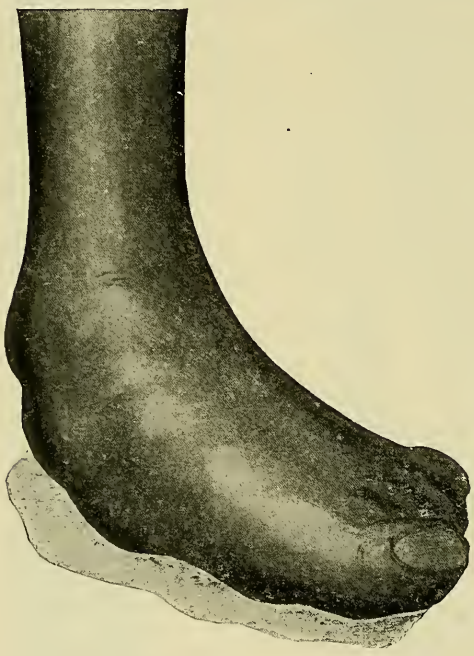

Fig. 63.
Tabetic Foot.

Still more interesting are the "tabetic arthropathies" first studied by *Charcot*. In the pathogenesis of these remarkable joint conditions, different factors combine. Small fractures in the region of the ends of the bones, and particularly tearing off of tendons, can act as a starting-point, or the hypotonic anomalies of position, for example, the genu recurvatum, lead to stretching of the capsular apparatus and to abnormal friction in the joint; to this is added, that in consequence of "deep anesthesia," the patient in such cases does not in the least spare his joint, but rather maltreats it freely; further, abnormal secretory conditions in the synovia, wearing away of the cartilage and the bone, or, on the contrary, pathological proliferation in them, etc., come into play. In this manner exceedingly striking disease pictures occur, which affect far most frequently the knee, then in diminishing frequency the ankle, hip, and shoulder joints. The greatly distended globular joint region, covered with tortuous veins, can justify the comparison with the abdomen of a dropsical child used by *Brissaud*. At other times marked flail joints, due to the relaxation of the capsular and ligamentous apparatus, occur; or the epiphysis is destroyed, in consequence of which the condyles of the femur,

the head of the femur, or of the humerus, disappear, and the shaft of the diaphysis ends free in the empty capsule of the joint. Besides these atrophic arthropathies, hypertrophic forms occur, in which the proliferative processes gain the upper hand and portions of new-formed bone grow into the joint cavities and throughout the periarticular tissues leading to rapid ankylosis. Through osteoarthropathies in the region of the root and middle bones of the feet, a particular kind of flat foot, the so-called "pied tabétique" (tabetic foot) occurs (Fig. 63).

The most important trophic disturbance of the integument is perforating ulcer ("mal perforant"), which in the great majority of cases is located on the sole of the foot (particularly on the ball of the great or of the little toe, or on the heel). This is a round, painless ulcer, which, first superficial, gradually spreads deeper and deeper, and finally may expose a joint of the foot. As "mal perforant palatin," *Letulle* has described an analogous lesion of the palate which can lead to perforation of this through into the nasal cavity. While these ulcerations are usually observed in the early stages of tabes, in the late stages there is a tendency to extended malignant bed-sores, which are located chiefly over the sacrum, but also in other places (heels, trochanters, elbows, etc.). A rare form of tabes, the "marantic tabes" of *Oppenheim*, is characterized by rapid and complete disappearance of the subcutaneous fatty tissue. As farther trophic skin symptoms of little practical importance, eruptions of herpes, hemorrhages into the skin, vitiligo, ichthytosis and circumscribed loss of hair, are observed occasionally in tabetics. Of the other epidermal structures, the nails (deformity, brittleness, falling off) and the teeth (painless and spontaneous separation) are sometimes affected.

6. RADICULAR IRRITATIVE PHENOMENA

The five previously considered categories of posterior root symptoms—ataxia, hypertonia, areflexia, sensory disturbances and trophic disturbances—are, as we have seen, to be interpreted as due to impairment of function. We must now pass on to other radicular phenomena which plainly present the character of irritative symptoms. These are tabetic hyperesthesias, paresthesias, pains and crises.

Already in the early stages of the disease many tabetics complain of prickling, formication, burning, or cold feeling, of various distribution, now located in the trunk, again in the extremities. On the trunk the circular distribution of these paresthesias sometimes bring it about, that the patients suffer continuously from a feeling of constriction, as if they were wearing a belt or a tightly laced corset around the body. Hyperesthesias also occur in spots or in girdle form, so that, for example, the rubbing of the shirt, the pressure of the shoe, etc., may become almost unbearable. Particularly frequent is a zone of marked hyperesthesia for cold, just below the ribs.

All these paresthesias and hyperesthesias are usually quite constant, and last through long periods of the disease. It is otherwise usually with tabetic pains, which almost always have a much more paroxysmal character than the attacks of neuralgic pain; we speak hence of lightning or shooting pains,

"douleurs fulgurantes et lancinantes." Their intensity is usually very great, sometimes terrible, so that the patients at every "stroke" or "stab" writhe and scream aloud. On the trunk the radicular topography of the radiations is plainest ("girdle pain"); in the extremities less so. So, many patients express themselves as having the feeling as if a knife or a red-hot iron was suddenly thrust through their extremities (by far most frequently into the legs); others have the sensation of being struck on the shin with a club; still others of the flesh being torn from their bones, etc. On the foot the type of pressure pain is particularly frequent. On this account the French, with a play upon the "Spanish boots" of the torture chamber, speak of "douleurs en brodequin." Only rarely are the pains more continuous, and of comparatively steady, often very moderate, intensity, so that such patients are for a long time considered as "rheumatics."

Radicular irritative phenomena in the region of the internal organs we call "crises." They are always accompanied with motor or secretory phenomena reflexly produced. Gastric crises are by far the most frequent. At the start they commence with severe pain in the abdomen and in the back, and intense retching and vomiting. In the X-ray picture after a bismuth or barium meal, the spasmodic contraction of the stomach to hour-glass form has been observed. Chemical examination of the gastric juice usually shows marked hyperchlorhydria; however, this sometimes is absent; indeed, there is occasionally subacidity in gastric crises. These excessively painful attacks, which occur with great disturbance of the general condition, can last hours, or even days. In particularly stubborn cases we speak of a "status criticus," after the analogy of the "status epilepticus." In them, also, in consequence of parenchymatous ecchymoses of the mucous membrane of the stomach, there can be vomiting of blood ("crises noires"). The cessation of gastric crises is just as abrupt as their beginning, and even greatly reduced patients usually recover quite promptly from them.

Other crises observed in tabes are:

Larynx crises, in which the patient is suddenly attacked by a feeling of suffocation and cough, the air is drawn into the spasmodically narrowed glottis, with difficulty and with a whistling sound, the face is cyanotic, and the patient breaks out in a sweat. In severe cases there may be syncope; the patient even may die; in rudimentary cases the attack is usually limited to a moderate "suffocating cough." As in all other crises, the return to normal is almost immediate.

Pharynx crises consist in painful swallowing spasms, œsophagus crises in painful spasms of the gullet, heart crises resemble angina pectoris. Before these last are diagnosed it is naturally necessary to exclude the (quite frequent) coincidence of tabes with luetic heart and vascular disease.

Sympathetic crises can produce fleeting pictures which resemble those of *Basedow's* disease (see Lecture XXIII). Vagus crises lead to paroxysmal alterations of the heart beat and of the breathing; indeed, to apnœa and syncope. I have interpreted as medullary crises attacks of hyperpyretic temperature with *Cheyne-Stokes* breathing.

Irritative phenomena on the part of the olfactory nerve with spasms of

sneezing and rhinorrhœa, show nasal crises, colicky pains with profuse diarrhœa, intestinal crises. The rectal and vesical crises are characterized by very severe tenesmus. We have already alluded to testicular crises in speaking of the differential diagnosis of neuralgia of the testicles (see page 53). Renal crises imitate renal colic, liver crises that due to gallstones. The clitoris crises of female tabetics are characterized by spontaneous paroxysms of sensuous feeling, which gradually pass over into painful sensations; also by hypersecretion of the vaginal mucus and emptying of the glands of *Bartholin*. Painful attacks of singultus, finally, are considered as diaphragmatic crises.

LECTURE XII

The Syphilogenic Diseases of the Central Nervous System

A. Tabes Dorsalis (*Continued*)

GENTLEMEN: In the last lecture we became acquainted with those components of the tabes symptomatology which may be classed together as "the posterior root symptom-complex" and for whose physio-pathological explanation we can point to the pathologico-anatomical considerations with which we preceded our remarks. You have noted in the description of the "crises," that tabes dorsalis passes beyond the bounds of the spinal innervation, and now the description of its important ocular symptoms will bring still more clearly before you that it is not strictly to be included among diseases of the spinal cord, but is rather to be considered as a cerebro-spinal affection. Fortunately, our clinical knowledge of this subject is more satisfactory than what we know of its pathologico-anatomical and physiological basis, hence, since so much information is still lacking, especially as to the explanation of the pupillary symptoms, I prefer not to enter into the matter in order to avoid becoming involved in a discussion of theoretical and disputed questions.

OCULAR SYMPTOMS

1. *The Pupillary Symptoms.*—Since the anomalies presented on the part of the pupillary innervation are common to tabes and progressive paralysis, our description of these will be applicable to what may be found in all the metasyphilitic diseases of the nervous system. In a great many cases of lues cerebro-spinalis we meet with the same conditions, and it is above everything a question if we should not consider them as pathognomonic of the syphilogenic nature of an existing nervous affection. Personally, I am, in common with many other neurologists, of this last opinion; indeed, I have never found the most important of the clinical manifestations now to be described, *Robertson's* symptom, in typical development in a patient, in whom a non-syphilogenic disease had to be diagnosed as was shown on autopsy. It is hence just as much a criterion of previous syphilis as a positive *Wassermann* reaction.

Testing the Pupillary Functions.—Normally, contraction of the pupils ("myosis," action of the sphincter pupillæ innervated from the oculo-motorius) occurs under the following conditions: 1. By the action of light upon the eye being examined or upon the other eye, "direct and consensual light reaction." 2. As a regular accompaniment of movements of the other eye muscles, among

which, clinically, only movements of accommodation and convergence come into consideration ("synergic pupillary reaction"). 3. In certain individuals upon the basis of the psychical conception "bright" ("ideomotor pupillary reaction"). A pupillary dilatation ("mydriasis" through the dilator pupillæ innervated from the sympathetic), on the other hand, occurs normally in many individuals when a painful stimulus is applied, particularly to the face or neck, further, in fright, anxiety, orgasm, etc.

The specific action of many alkaloids, some dilating, others contracting the pupils, is well known. Mydriatics: among others, are atropin, cocain, scopolamin, duboisin. Myotics: physostigmin, morphine, pilocarpin.

In every patient, determining the morphological relations of the pupils should precede testing their reaction, whether they are round, irregular, of medium width, large or small, if they are of the same size on both sides or if there is difference between them (anisocoria), finally, if they are centrally or excentrically located. We must also consider further whether they have been acted upon by any of the above-mentioned drugs and whether there has been, or is, any local disease (*e.g.*, iritis) which could alter the shape and function of the pupils. Next, we examine the direct reaction to light, either to daylight (before a window) or to artificial light (electric lamp, candle, match, etc.). The patient should not be frightened, however, by suddenly throwing a strong light into his eyes, since the psychical mydriasis due to fright may inhibit the light myosis and cause an imperfect reaction. The patient must further look into the distance in this test, in order to eliminate synergic convergence and accommodation reactions. Further, the eye not being tested must be covered to prevent consensual narrowing. On the other hand we test the last-mentioned reaction by observing the iris of the shaded eye. Usually on sudden illumination, the light reaction after an appreciable period of latence appears very plainly. Following the initial contraction, after several oscillations (physiological hippus) the pupil assumes a medium width. We test reaction for a convergence and accommodation, by having the patient fix his eyes on an object about 1½ meters in front of him, and then approaching it slowly toward his nose.

The isolated reflex pupillary rigidity, or the *Argyll-Robertson* symptom of tabes and general paresis, manifests itself, in that the pupil does not contract either when exposed directly to light, or when the other eye is illuminated, but does react in accommodation and convergence. The *Argyll-Robertson* symptom can occur unilaterally; if it is only partial, a slight direct and consensual light reaction can still be obtained on strong illumination, and we speak then of reflex sluggishness of the pupils. Reflex rigidity is often accompanied by myosis and irregularity of the pupil,* while abnormal width of one or both pupils is not rare. In a considerable portion of cases, anisocoria is present at some stage of the disease. Where this is variable, so that sometimes the right and sometimes the left eye presents a wide pupil, we speak of "bounding

* It is not to be forgotten that in later life, and especially in old age, the pupils are normally narrower than in young people; such narrow pupils, however, as is shown by many tabetics ("pinpoint" pupils) do not come within the physiological limit. Slow reaction to light is also proper to old age (on account of rigidity of the iris).

pupils." Absolute rigidity, that is, inability of the pupils to react either to light, to accommodation or to convergence, can occur in tabes and paresis, but here it is to be considered an atypical finding. How fundamentally important the exact examination of the pupils in tabetics is, is shown by the statistics of *Faure* and *Desvaulx* who, in 200 cases found pupillary anomalies of some sort 193 times.

2. *Tabetic Diseases of the Optic Nerve.*—Upon the basis of tabes (as well as upon that of progressive paralysis and indeed, as an independent monosymptomatic, metaluetic affection of the optic nerve) a simple noninflammatory atrophy of the optic nerve can develop. There is a primary degeneration of the optic neurones with reparatory proliferation of the supporting tissue, that is, the pathologico-anatomical analogue of the tabetic degeneration of the posterior roots and posterior columns, which is more to be dreaded in the early stages than in advanced tabes. According to *Uhthoff*, optic atrophy occurs in 10 per cent. to 15 per cent. of tabetics; according to my personal experience this percentage seems to me too high. As the study of the visual field shows, the so-called "papillomacular bundle" of the optic nerve remains longest intact, so that there is concentric narrowing of the visual field; less frequent, are irregular losses of vision, entirely atypical, central scotoma. Color perceptions (particularly for green) are usually lost before those for white. Ophthalmoscopically the general pallor of the nerve head is next observed; little by little it becomes chalk-white, on which account its outlines remain very sharp. Finally, the vessels are also affected and appear much thinned. Where, on the other hand, the atrophy develops from a neuritic process (that is, the papilla appears cloudy, washed out) there is a complicating syphilitic disease of the optic nerve and not a metaluetic, purely degenerative atrophy. This last is therapeutically little to be influenced and of very bad prognosis; it leads in a short time (1 to 3 years) to complete blindness. Only very exceptionally does the degenerative process come to a standstill.

3. *Paralyses of the Eye Muscles.*—In contradistinction to most of the tabes symptoms previously considered, which are characterized by their tendency to progress, or at least to remain constant, the tabetic eye-muscle paralyses are, in the majority of cases, of temporary, even fleeting nature. These phenomena, also, usually belong to the early stage of tabes, in which the paralysis affects by preference a single muscle ("paralysies parcellaire" of *Fournier*), most frequently the external rectus, but little more rarely the internal rectus or the levator palpebræ. Where, on the other hand, the eye muscle paralyses appear in the late stages of the disease, they are usually of greater extent. Strabismus, double vision, ptosis, are their clinical results. We are not entirely clear either as to the location or as to the nature of the pathological substratum of these paralyses, whether there are changes in the nuclei or in the nerve trunks. The circumstance that these disturbances often last only a few days, further, can vary from one day to another, seems to justify the suspicion that they are due to circulatory disturbances, probably to vascular spasm. Permanent or even progressive eye muscle paralyses in tabetics we consider rather as tertiary luetic complications.

OTHER CRANIAL NERVE SYMPTOMS

Along with the so important ocular phenomena of tabes dorsalis, its manifestations in the other cranial nerves play only a subordinate rôle. The involvement of the auditory nerve we have not rarely observed in the form of a progressive nerve deafness, with diminution of hearing through bone conduction (to be demonstrated by the *Schwabach*, *Rinne* or *Weber* tests—see Lecture II, page 34), partial loss of perception for the musical scale, tinnitus and occasionally, also, attacks of labyrinthine vertigo. *Oppenheim* and *Siemerling* among others have recognized, anatomically, the degeneration of the auditory nerve. Reduction of the olfactory and gustatory perceptions, which may increase to anosmia and ageusia, and which *Klippell* has attributed to destruction of fibers in the olfactory and glosso-pharyngeal nerves, are rare. In the trigeminus, paresthesias, hypesthesias and anesthesias occur; in the facial, unilateral or bilateral, usually temporary paralyses (exceedingly rare). In the hypoglossus, *Raymond*, *Cassirer-Schiff* and others have observed, as an excessive rarity, hemiatrophy of the tongue.

Symptoms of disturbance in the innervation of the larynx arouse great interest. They are far more frequent than the already mentioned larynx crisis with which they are classed by the French under the name "laryngisme tabétique." According to *Dejerine*, paralyses of the laryngeal muscles, or ataxia of the vocal cords, occur in nearly 45 per cent. of tabetics, the former affect by preference the muscles opening the glottis, that is, the posterior cricoarytenoids. Pareses of the posterior muscles, of slight degree can, particularly when they are unilateral, cause no clinical symptoms, and may only be discovered on laryngoscopy. In severe cases the air is drawn in with a whistling sound, and slight bodily exertion leads to severe dyspnœa; on the other hand, phonation is uninfluenced. More rare, are disturbances in the closers of the glottis which, when they are present bilaterally, show themselves through bitonality of the voice, "breaking," etc., when unilateral, however, remain latent. In ataxia of the vocal cords the voice is tremulous, sensory disturbances of the larynx (hyperesthesia or anesthesia) are also occasionally observed. All these manifestations belong to no particular stage of the disease, but occur now as early again as late symptoms. They may be inconstant, and in the same patient are sometimes present, sometimes absent.

DISTURBANCES IN THE SKELETAL MUSCLES

In general the muscles of the tabetic remain normal until the disease is very far advanced and has led to cachexia and great loss of strength, then we almost always see a wasting of the muscles without fibrillary contractions or reaction of degeneration, in which the feet of the permanently bedridden patient, partly on account of the hypotonia of the peroneal muscles which are no longer able to hold up the weight of the feet, partly from pressure of the bedclothes, etc., may take an equinovarus position. Sometimes, however, circumscribed muscular atrophies occur, even in the early stages of the disease, usually in the upper extremities (shoulder-girdle, small muscles of the hand), in which sometimes

partial reaction of degeneration, sometimes only quantitative reduction of irritability, is to be found. Corresponding to the degree of atrophy, muscular power is naturally reduced, so that finally marked pareses or even paralyses may result. These atrophic phenomena are, as far as their pathogenesis is concerned, scarcely to be considered as anything unique. In many cases they may present the analogue of the already mentioned trophic disturbances of the skeleton, integument, etc.; in others, however, which have been distinguished as "amyotrophic tabes," there may be a combination of tabes dorsalis with "syphilitic spinal amyotrophy" (*Raymond, Léri, Nonne*, etc.), which depends upon disease of the anterior horns of the spinal cord.

GENITAL AND SPHINCTER DISTURBANCES

One of the first symptoms of tabes in men is usually impotence; there are, however, cases in which potence is very long preserved; usually a period of sexual excitement precedes the development of sexual weakness; this may be responsible for the erroneous view of the older authors and which is still in existence among the laity, that sexual excesses may have tabes as a result. Anesthesia of the glans is often to be found along with disappearance of the power of erection. The genital disturbances of female tabetics present themselves apart from the clitoris crises mentioned, above everything in anaphrodisia or indeed, as total anesthesia of the genital organs; labor sometimes runs its course entirely without pain and may be accompanied by insufficient expulsive power and atonic hemorrhages.

The bladder function very frequently suffers impairment and, indeed, in various forms. Sometimes the patient, in spite of great desire to urinate, must strain a long time before he can pass water; sometimes the desire is immediately followed by emptying the bladder and the patient wets himself, or, on the other hand, the feeling of desire to urinate is lost entirely and the patient only urinates "out of consideration," as *Fournier* expresses it. All these disturbances are often already present in the early stages of tabes, but are in no way constant; rather are frequent intermissions, with entire return to the normal, as well as great variations in the intensity of the anomalies, quite frequently observed. Even severe symptoms, as, for example, complete retention which necessitates catheterism, can completely disappear after lasting for months. There occurs, further, ischuria paradoxa, in which discharge of urine (in drops) only occurs when filling of the bladder has reached a certain degree, incomplete retention, in which the bladder can never be completely emptied and a certain quantity of residual urine remains in it; finally, true incontinence. This last, characterized by continual dribbling of urine and a consequence of atony of the sphincter and of the detrusor vesicæ, belongs to the late stages of tabes. It may be only nocturnal, or can manifest itself also in the waking state. Only exceptionally, and then only in the terminal stages, incontinence of the bowels occurs in tabetics, while obstinate constipation, on account of intestinal atony, is quite frequent.

THE CONDITION OF THE CEREBRO-SPINAL FLUID

When I now pass over to the consideration of this, of late so freely discussed questions of the anomalies in the cerebro-spinal fluid in tabes and their determination—we will consider here also paresis and cerebro-spinal syphilis—I would warn you against overestimation of results obtained by these methods and the too frequent making of lumbar punctures in all patients with syphilitic nervous diseases. While this procedure, carried out with proper technique, is scarcely dangerous (see Lecture XVI), it is best reserved for a specialist who is also skilled in the necessary laboratory investigations. He, also, should only use it where an extended clinical investigation still leaves the diagnosis in doubt.

Within the limits of these lectures I cannot go into the technique of the *Wassermann* complement-fixation reaction which is to be applied in the first place to any spinal fluid removed on account of suspicion of a syphilogenic affection. (The amount of fluid necessary for making all the necessary investigations is about 5 or 6 cc.) The second test to be applied to the fluid is the globulin reaction of *Nonne-Apelt*. To the fluid, equal parts of a hot saturated and then cooled solution of ammonium sulphate is added; if in three minutes ("Phase I") there is clouding, this speaks for abnormal globulin or nucleoalbumin content of the specimen under examination. The third test consists in the cytological examination of the centrifugated sediment (*Widal, Ravaut, Nissl*); while normally only occasional lymphocytes are found, marked lymphocytosis is to be considered as a certainly pathological finding.*

The indications of these reactions to which, as a fourth, the *Wassermann* test applied to the blood serum is to be added, in the diagnosis of syphilitic diseases of the central nervous system, is best shown in the following synopsis of *Nonne:*

I. Blood Examination

Wassermann Reaction.

a. Positive.—Is characteristic for syphilis, with a few exceptions coming little or not at all into practical consideration. A positive reaction is given also by a few cases of scarlatina (only at certain very limited stages of the disease), by malaria, by frambœsia, by lepra, by the plague, etc.

A positive *Wassermann* reaction from the blood serum tells nothing further than that the individual under consideration has at some time had syphilis, hereditary or acquired, not that the disease now present must be of luetic nature.

b. Negative.—Is to be considered as speaking against paresis since with exceedingly rare exceptions the blood of paretics gives a positive *Wassermann* reaction.

II. Examination of the Spinal Fluid

a. Normal Fluid.—Pressure 90 to 130 mm water (Manometer). Phase 1 reaction negative; at most 5 to 6 cells to the cu mm (*Fuchs-Rosenthal* counting apparatus).

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* By many laboratory workers, in the United States at any rate, the *Noguchi* butyric acid test for globulin is considered more delicate than that of *Nonne-Apelt.* Also it is customary to count the cells in the fluid without centrifugating it.—*Translator.*

Wassermann reaction, applied according to the original method, using 0.2 cc of the fluid to be examined and also with larger quantities (0.3-1.0 cc fluid) negative.

b. Pathological Fluid.—

1. Discharged under increased pressure (over 150 mm water).
2. Positive phase, 1 reaction.
3. Increased cell content.

These three symptoms, in combination or alone, show that there is an organic disease of the central nervous system (specified or non-specific).

4. Whether the disease of the central nervous system is of syphilitic nature is decided by the *Wassermann* reaction applied to the spinal fluid.

If the *Wassermann* reaction, applied according to the original method (0.2 cc fluid being used), is positive there is the greatest probability that the case under investigation is paresis or tabo-paralysis, much more rarely is it cerebro-spinal syphilis, and only in exceptional cases true tabes.

In the great majority of cases of paresis the *Wassermann* reaction is positive with 0.2 cc of spinal fluid.

In a few cases of paresis, in almost all cases of cerebro-spinal syphilis and tabes, the *Wassermann reaction* is only positive when larger quantities of fluid (0.3, 0.4 to 1.0 cc) are used.

TYPICAL FINDINGS

I. Paresis or Taboparalysis.

1. *Wassermann* reaction in the blood positive (in almost 100 per cent. of cases). Lumbar pressure frequently increased.
2. Phase 1, reaction positive (in about 95 to 100 per cent. of cases).
3. Lymphocytosis (in about 95 per cent. of cases).

Wassermann on the spinal fluid:

a. Positive in about 85 to 90 per cent. when the original method is used; (0.2 cc fluid).

b. Positive in 100 per cent. when larger quantities of fluid are used.

II. Tabes without Combination with Paresis.

1. *Wassermann* reaction in the blood serum, positive in 60 to 70 per cent. of cases; lumbar pressure frequently increased.
2. Phase 1, reaction positive in about 90 to 95 per cent. of cases.
3. Lymphocytosis positive in about 90 per cent. of cases.
4. *Wassermann* on the spinal fluid:

a. Original method (0.2 cc), positive in 5 to 10 per cent.

b. Larger amounts of fluid, positive in almost 100 per cent.

III. Cerebro-spinal Syphilis.

1. *Wassermann* reaction in blood serum positive in about 80 to 90 per cent.; lumbar pressure frequently increased.
2. Phase 1, reaction negative only in exceptional cases, otherwise positive.
3. Lymphocytosis like Phase 1, almost always positive.
4. *Wassermann* on the spinal fluid:

a. Original method (0.2 cc) positive in about 10 per cent.

b. With larger quantities of fluid, almost always positive (particularly valuable in differential diagnosis from multiple sclerosis, brain tumor and spinal cord tumor).

Course and Prognosis

The average duration of tabes can be placed at from 10 to 20 years. On the one hand, however, malignant cases which lead to death in a few years occur, and on the other there are those which characterized by special mildness, slow progression, complete standstill, or even partial regression of the symptoms, permit reaching an advanced age.

For the prognostic estimation of the individual case the following principles are applicable in general, though, like all rules, they of course have their exceptions: Mildness of course is the more to be expected, the later after luetic infection the disease becomes apparent, the longer also the latent or incubation stage of tabes has lasted; with this well agrees the fact that cases of tabes in the mature period (beginning about the 5th decade) are usually characterized by their mildness. A tendency to attacks of pain persisting for a long time, although this peculiarity greatly influences the comfort of the patient, is usually associated with favorable prognosis as regards preservation of ability to walk and duration of life. On the other hand the richness in symptoms of a concrete case of tabes, is to be considered unfavorable prognostically with the single qualification that complication with paresis in typical forms of tabes with many symptoms, is scarcely to be feared. The same remark applies in greater measure to the cases of tabes characterized by location of the lesion high up (Tabes superior) in which variety the prognosis is clouded by fear of complication with bulbar symptoms. The estimation of the general condition is very important: Syphilogenic heart and vascular lesions are not at all rarely found in tabetics on close investigation and where present alter, markedly for the worse, the outlook. Finally, the striking fact is to be considered that the occurrence of blindness from tabetic optic atrophy ("amaurotic tabes") very often puts a stop to the further development of the disease, so that among French authors the expression "Tabes arrested by blindness" is in common use.

In a somewhat schematic but practically quite useful manner, three stages in the development of tabes are distinguished from one another; the preatactic, in which the gait is not yet altered; the atactic, in which it is characteristically altered on account of the disturbances of coördination; and the paralytic, in which a paralysis is combined with advanced ataxia, so that the patient is permanently bedridden. Tabes is the cause of death only in very rare cases (paralysis of the larynx, paralysis of the bladder, cystitis and uremia; bed-sores,

and sepsis; inanition and exhaustion after protracted gastric crises; status criticus, vagus crises, etc.); death usually occurs from intercurrent diseases (tuberculosis, pneumonia, influenza, etc.), against which the power of resistance is greatly reduced, not at all infrequently from syphilogenic diseases of the vessels (angina pectoris, aneurism of the aorta, cerebral hemorrhage, etc.).

We will now enumerate the courses which tabes dorsalis may take. According to its rudimentary mildness or malignancy there have been separated on the one side rudimentary or abortive tabes, in which the patients never get beyond the preatactic stage (I know of cases which have been in existence 30 years); on the other hand, tabes acutissima, "galloping consumption of the spinal cord," which in one of my cases in 9 months ran through the atactic and paralytic stages and led to death in cachexia. Through the time of its beginning the very rare juvenile tabes dorsalis is characterized, commencing in youth or even in late childhood, it arises upon a hereditary luetic basis. According to its location, special positions are taken by tabes superior or cervical tabes, in which the anomalies affect the arms, and the lower limbs present loss of patellar reflex as their sole tabetic symptom; and the opposite condition, tabes of the conus terminalis. A case of this last variety affecting a 43-year-old woman, carefully investigated by *Thomas* and myself, presented the following symptoms: Rectal crises, incontinence of urine and stools, total anesthesia of the genitals (so that coitus was not at all noticed), total peri-ano-genital anesthesia (of the riding trouser form), sometimes pain in the legs, myosis, reflex rigidity of the pupils, exaggeration of the patellar reflexes. A special topographic variety is also tabes amaurotica, of whose frequent inhibitory influence upon the further extension of the spinal lesions we have already spoken; simple metasyphilitic optic atrophy can be considered indeed, as a "tabes without spinal cord symptoms." Finally, according to the special preponderance of individual symptoms or symptom-groups, we speak of a tabes dolorosa, visceralis, amyotrophica, marantica.

Diagnosis and Differential Diagnosis

The diagnosis of fully developed tabes is one of the easiest tasks in clinical neurology and can often be made at a glance. In the early stages the disturbances are less striking, so that, unfortunately, disastrous mistakes are frequent enough; so, for example, gastric crises have been mistaken for ulcer of the stomach and laparotomized, also patients with lancinating pains are continually sent into the hospitals with diagnoses of rheumatism, neuralgia, sciatica, etc. Nevertheless, even in the early stages, tabes can be recognized nearly always with certainty, after a thorough general examination. In taking the history and estimating the present condition the following "cardinal symptoms" should be specially sought after:

1. Pupillary disturbances (reflex rigidity or slowness, anisocoria, irregularity, myosis).
2. Localized or radicular (that is on the extremities in longitudinal bands, on the trunk as zones) hypesthesias or hyperesthesias (the last particularly for cold), anesthesias of the nerve trunks or viscera.

3. Reduction or loss of tendon reflexes (their exaggeration in no way excludes tabes incipiens).

4. *Romberg's* symptom, sometimes only to be recognized when standing on one leg.

5. Lancinating pains or crises.

6. Bladder or genital disturbances.

7. Optic atrophy.

If four of these symptoms are found the diagnosis, tabes dorsalis, is absolutely assured, if three, it is almost certain. A probable diagnosis can be made from two of these symptoms and eventually confirmed by the results of blood and spinal fluid examination. The laboratory methods are a diagnostic necessity, only in the quite rare cases of beginning tabes, where most exact clinical examination shows only one cardinal symptom.

The following compilation after *Oppenheim* may give you a conception of the manifoldness of the symptom grouping in tabes incipiens:

1. Lancinating pains, *Westphal's* symptom, pupillary rigidity.

2. Bladder weakness, *Westphal's* symptom, girdle symptom.

3. Pupillary rigidity, anesthesia on the trunk.

4. Optic atrophy, *Westphal's* symptom, analgesia.

5. Optic atrophy, girdle symptom with corresponding hypesthesia, analgesia.

6. Optic atrophy, lancinating pains, impotence.

7. Attacks of vomiting, *Westphal's* symptom.

8. Attacks of vomiting, loss of the Achilles' tendon reflex.

9. Attacks of vomiting, pupillary rigidity.

10. Gastric crises, anesthesia on the trunk.

11. Joint affection, analgesia, *Westphal's* symptom.

12. Joint affection, analgesia, pupillary rigidity.

13. Paralysis of the vocal cords (with or without attacks of coughing), *Westphal's* symptom, pupillary rigidity.

14. Spontaneous falling out of the teeth, disturbances of sensation in the trigeminus distribution, *Westphal's* symptom, bladder trouble.

15. Paralysis of eye muscles, girdle symptom, analgesia.

16. Optic atrophy, anesthesia on the trunk.

Of great differential diagnostic importance is the distinction of tabes dorsalis from "peripheral pseudo-tabes" as it occurs in diphtheritic and alcoholic polyneuritis. (See above, page 42.) The *Argyll-Robertson* symptom occurs in neural tabes almost never (that is, only when the patient is an old syphilitic). Further tabes symptoms foreign to polyneuritis, are gastric crises in typical development (gastric disturbances, gastralgia, the morning vomiting of drinkers, etc., as they are not rare in patients with alcoholic neuro tabes, must not be mistaken for crises), other paroxysmal visceral disturbances, visceral anesthesias. Bladder disturbances are very rare in polyneuritis and almost never reach any considerable extent; a limitation of this remark applies only to those cases in which a "polyneuritic psychosis" develops and leads to incontinence of urine and

feces. The condition of the peripheral nerve trunks on palpation is important: In tabes usually hypo- or anesthetic to pressure, they are never so in neuritic pseudo-tabes, but are usually very sensitive to pressure and besides are often thickened. The eventual superficial anesthesias of the atactic forms of polyneuritis have not a radicular topography, but increase from the roots of the extremities toward the periphery and reach their maximum on the ends of the limbs. I have never seen girdle sensation and anesthesia in girdle form, in polyneuritis. Lancinating or boring pains, preceding by years the onset of ataxia, occur only in tabes. The early appearance of degenerative paralyses or pareses (namely in the peroneal region with "steppage," see above, page 23), speak against tabes. The affections of the optic nerve occurring eventually in both diseases, are to be differentiated without difficulty by ophthalmological examination. With the ophthalmoscope, in retro-bulbar neuritis a paling, temporally located, is found instead of the diffuse pearly or porcelain white coloration in tabes; decisive is the difference in the limitation of the visual field; in polyneuritis, central scotoma, in tabes almost always peripheral narrowing. As to the development of the disease, "acute ataxia" occurs far more frequently from "neurotabes peripherica." This increases rapidly but has a much greater tendency to regression, even to cure, of the symptoms, than tabes dorsalis.

Treatment

Although it is of course not permitted to us to cause regression of the degeneration of important nerve elements, which is the basis of tabes, and although in many cases the progress of the disease is inexorable, nevertheless, the great majority of tabetics give us plenty of opportunity for active interference in the sense of symptomatic therapy.

Before, however, we enter upon this last, we must take some stand in the much disputed question as to how far a causal treatment of tabes dorsalis is to be attempted—and who speaks here of causal therapy can readily speak of "specific" or "anti-luetic" treatment.

It is not only certain that thorough treatment of syphilis does not exclude later development of tabes or paresis, but it is indeed questionable if it to any extent reduces the risk of these diseases. This we can affirm even without the extensive and faultless statistics which to date are still wanting. Less extensive statistics, however, as, for example, those of *Spillmann*, who among 32 thoroughly treated syphilitics later saw not a single one attacked by metaluetic disease cannot be considered conclusive. *Reumont* has shown, as already mentioned, that in syphilitics only about 1 per cent. later become tabetic. If the introduction of the salvarsan treatment alters the matter we must wait and see. It is at least certain that among our tabetics and paretics, such patients as have at the time of their syphilis been thoroughly mercurialized are not few in number.* That these cases, indeed, present only a minority is not remarkable,

* I would refer to four double observations of conjugal parasyphilis, in which the husband on his part went through a specific cure; the wife, however, remained untreated, and both developed tabes or paresis about the same time, or, rather, the man earliest. See above (page 165) what was said about "Syphilis a virus nerveux."

since thorough treatment is received by only very few luetics. Also the paradoxical result of the retrospective statistics of *Eulenburg*, *Dinkler*, *Kron*, *Schuster*, and *Mendel-Tobias* cannot be disregarded; just the patients of these authors who had had the most energetic anti-syphilitic treatment showed the shortest incubation period of their tabes. It would be, of course, foolish on this account to assert that there was inadequate treatment of the syphilis, but such statistics, however, are a warning against too schematic handling of the question in this excessively difficult matter.

As to the specific treatment of already fully developed cases of tabes, in such cases in which a combination with tertiary nerve syphilis appeared to be excluded, I formerly did not use mercury and only in the last six years have I been converted to the use of mercurial and, of late, of salvarsan treatment in certain indications. The pains and crises have shown themselves most certainly to be influenced by these remedies; their action is quite frequently, though by no means always, very decided; with arsenobenzol sometimes a striking one. In incipient, or at least in pre-atactic cases, of tabes, with regard to the estimation of the influence of mercury and salvarsan, great scepticism is still justified. Nevertheless, I have the impression that (in cases with positive *Wassermann*) the course, in the sense of mildness (frequent enough otherwise, however) can be influenced. Evident tendency to rapidly progressive ataxia, as well as poor condition of nutrition, or indeed, "marantic tabes," contraindicate mercury, not, however, salvarsan, while on the other hand, this last may be dangerous in the combination with syphilogenic diseases of the heart or aorta. An unqualified contraindication against mercury cures is presented by parasyphilitic optic atrophy. I have seen several cases together with the ophthalmologist, *Paul Knapp*, which in spite of the advice of both of us, were subjected to energetic mercurial treatment and rapidly got worse. *Knapp* could indeed show four times most exactly that the degenerative process in the optic nerve progressed much more rapidly, and that upon abandonment of this treatment the progression was again slower.

Into the particularities of anti-syphilitic treatment we will not enter here. The most important points neurologically will be mentioned in the description of lues cerebro-spinalis. As to the iodide medication so much used in tabes dorsalis, it comes into consideration less as an antiluetic as on account of aiding the circulation through its reduction of the viscosity of the blood which has been shown and proven in the course of iodide cures (*Müller*, *Enada*, etc.), perhaps, also, thanks to a certain vasodilator effect, a better circulation through, and nutrition of, the affected regions may occur, which opposes the degenerative destruction. Along with the internal administration of the alkaline iodides (in doses of 0.5 (gr. 7½) 3 or 4 times a day, after 20 days, at least a 10 day pause before repetition of the cure) I used with advantage, iodipin injections (twice a week 10 cc of the 25 per cent. oil). The other organic iodine preparations (sajodin, iodostarin, lipiodin, iodglidine, iodon, iodtropin, etc.) give less favorable results; nevertheless, they can sometimes be ordered in sensitive patients or for those demanding a change of medicine.

When we look over the drugs recommended in tabes dorsalis I might mention almost only from its historical interest, silver nitrate (0.01 made into a pill with

bolus alba 3 times a day) introduced into the therapy of tabes by *Wunderlich* in his day, although it is often enough prescribed in a routine manner still. I am convinced that it does not have the slightest action and at most can come into question symptomatically in gastric crisis (as a drug limiting the secretion), here, however, it is usually not retained, but promptly vomited. According to the original instructions, pills of silver nitrate should be given for months at a time. There is in this, however, the danger of "argyria" (blue-black coloring of the skin and mucous membranes), even when the rule not to exceed a total dosage of 10 grm. has been observed. Also, protargol which can be given in 10 times the dose, can well be dispensed with, as can oxide of zinc (0.05 3 or 4 times a day) and chloride of gold and sodium (0.01 t. i. d.). As to ergotin, recommended by *Charcot*, it is certainly no specific against tabes, but sometimes opposes the tabetic bladder paralysis in consequence of the affinity for the smooth muscles of the pelvic organs also responsible for its action upon the uterus.

℞ Extract. secalis cornuti 2.5
Pulvis secalis cornuti 5.0

M. fiat Pil. No. L. S. 1 pill t. i. d.

A somewhat efficient general effect in tabetics is exerted by strychnine, however; we can administer it in pill form or subcutaneously (0.003 to 0.006 per day—gr. $\frac{1}{20}$ to gr. $\frac{1}{10}$ per day). Tinctura nucis vomicæ is also a favorite remedy:

℞ Tinct. nuc. vom. 10.0
Tinct. cinconæ co. 20.0

M. S. 30 drops t. i. d.

An excellent combination has been given by *Erb* in the form of his "tonic pills."

℞ Ferri lactat,
Extract. cinchon. aq. .....āā 5.0 (gr. lxxv)
Extract. nucis vom. ..... 0.8 (gr. xii)
Ext. gentianæ, qs. ut.
Fiat pil. No. C.

S. 2 pills t. i. d. p. c.

I have had better results from the following:

℞ Ferri lactat ..... 4.0 (gr. lx)
Quinin sulphat ..... 3.0 (gr. xlv)
Extract. nucis vom. ..... 1.0 (gr. xv)
Extract. valerian ..... 7.0 (gr. c)
Fiat pil. No. C.

S. 2 pills t. i. d.

Next to strychnine the arsenic preparations should be recommended: sometimes arsenic cures produce a manifest improvement of the nutrition and of the general condition, which occasionally has a favorable effect upon the course of the disease. In cachectic symptoms intensive subcutaneous arsenic medication is especially to be recommended. In order to avoid repetition I would remind you of what was said under the therapy of multiple sclerosis. Specially suited for the treatment of tabes appears to be the, in England officinal, iodide of arsenic (Arsenii iodidum, B. P.) in the form of *Donovan's* solution.

℞ Arsenii iodid. 0.1
Hydrarg. biniodid. 0.2
Potass. iodid. 2.0
Aq. dest. 60.0

M. S. From 5 to 100 drops gradually increased, in water 2 or 3 times a day.

While different derivatives of phosphoric acid, for example lecithin, calcium glycerophosphate, nucleinic acid or nucleinate of sodium, phytin, etc., can be occasionally tried, I can express myself as to the whole opo- and organo-therapy of tabes unfavorably only, also as to the use of spermin, cerebin, etc. From fibrolysin, which has been recommended upon theoretical grounds, in tabes, I have equally failed to see any result.

The attacks of pain in tabetics frequently call urgently for symptomatic medicinal treatment. What is to be said here agrees entirely with our previous remark as to the drug treatment of neuralgia, hence we will not again enumerate the different anti-neuralgics and their combinations, but will refer to Lecture III (page 57). As peculiar to the tabetic pains, we will only refer to their occasional susceptibility to the action of two drugs to which ordinary neuralgias scarcely ever react, namely, methylene blue and sodium nitrite. The first acts, perhaps, through its histo-chemical affinity for the axis cylinders; the last by relieving vascular spasm. Methylene blue is always to be combined with powdered nutmeg in order to prevent irritation of the bladder. Sodium nitrite only acts when used subcutaneously, and since it may act as a cardiac poison, is to be given in very carefully regulated doses.

℞ Methylene blue ..... 0.1 (gr. 1½)
Powdered nutmeg ..... 0.5 (gr. 7½)

M. Divide into 10 capsules. S. 1 t. i. d.

℞ Sodii nitritis 0.1-0.3
Aq. dest. 10.0

Should be sterilized. S. 1.0 to 1.25 (15-20 minims) daily (carefully increasing the quantity of sodium nitrite).

The too ready ordering of hypodermic injections of morphine is to be unqualifiedly condemned, since many tabetics, by carelessness of the physician in this respect, acquire the morphine habit. Further, it looks as if the use of mor-

phine in tabetics favors the occurrence of gastric crises, probably because morphine excites the secretion of gastric juice. Where morphine injections must be used as a last resort, it is best combined with atropin.

For the gastric crises *Rodari* has recommended the combination of pantopon and atropin. ℞ Pantopon, 0.2; atropin sulph., 0.01; aq. laurocerasi, 10.0; 10 to 15 drops 2 or 3 times a day. Many patients with gastric crises, however, do not tolerate the internal administration of drugs (for example, of cerium oxalate recommended in doses of grm. 0.1). Here suppositories often work well, containing, for example, codein phosph. and extract. belladonna, āā. 0.05 (gr. ¾). These last are to be used also in intestinal crises, while in laryngeal crises local application of 10 per cent. cocaine solution or inhalations of amyl nitrite are to be tried. (Pearls of nitrite of amyl containing each 5 drops are convenient.)

In disturbances of potence yohimbin can be tried. However, if in the dose of 5 to 10 drops of a 1 per cent. solution three times a day (or in tablets of 0.005 each—gr. 1/12), the effect is not satisfactory, it must be given up, as no larger doses should be used. It need not be attempted to excite the libido in such cases in which erection is no longer possible.

We will now leave the subject of the medicinal treatment of tabes and proceed to consider the exceedingly important physical methods of treatment.

For the use of electricity there is quite an extended field. Of galvanic procedures I would mention, first, the stabile application to the spine which is found beneficial by many tabetics. Two large, flat electrodes are applied, one to the neck and the other over the sacrum, and a current of 5 milleamperes gradually introduced; after 3 minutes the current is gradually reduced, the poles changed, and the procedure repeated. In lancinating pains stabile galvanization, the anode over the peripheral nerve trunks as described in Lecture III, can be tried. It helps only, however, in a very small number of cases. For this indication, more result is promised by energetic faradization with the wire brush acting as a derivative, which often is useful for the paresthesias also. The faradic current is further applied within the urethra in the treatment of weakness of the bladder; a flat electrode is placed over the lumbar region while a catheter electrode is passed one cm into the urethra; the individual applications should last 5 minutes, the current being regulated so that it is felt plainly but not painfully. Finally, the favorable effects occasionally obtained from the high frequency current in crises and lightning pains should be mentioned. As to hydro- and balneotherapy, I would warn you most decidedly against the application of the various cold water procedures. The rapid deterioration often observed in tabetics who have sought aid in "cold water institutions" speaks impressively against these measures. Tabetics are also to be warned against the other extreme, namely, hot baths, douches, etc., or mineral springs of high temperature. These remarks need to be modified only as regards a number of local methods of applying heat, which sometimes act favorably against some of the paroxysmal symptoms, for example, hot packs or application of the incandescent light to the limbs in lightning pains, application of the thermaphor or hot poultices to the epigastrium, or to the lower abdomen in gastric and intestinal crises. Of mineral springs the carbonated salt waters are to be considered principally

(Nauheim, Oeynhausen, Saratoga, New York, Ute Springs, Col., Paso Robles, Cal.), also the ordinary salt baths, since arrangements for the artificial addition of carbonic acid are available almost everywhere (Rheinfelden Ischl, etc.).

Further, the indifferent thermal waters, as those of Wildbad in Wurtemburg, Badenweiler, Teplitz, Virginia Hot Springs, Byron Hot Springs, Cal. The temperature of the baths should not exceed 35° C. (95° F.), their duration should usually be limited to half an hour. It has become the fashion of late to bring forward the radium content of these baths in order to explain their favorable action; this is quite possible, but as far as the artificial radium baths (radiogen, etc.) are concerned, they usually fail in lancinating pains, and in the rare cases in which they have been reputed to aid, the element of suggestion cannot be excluded. Good results are to be obtained also with less expensive ingredients for the baths, for example, with the slightly stimulating pine needle "ozofluin" baths, etc. Prolonged baths at 35° C. without any addition are alleviating for many tabetics. As a powerful derivative application in the lancinating pains, I can recommend applications of warm salt.

Of mechanical procedures the so-called "suspension treatment" formerly played a great rôle in tabes dorsalis. Hanging up the patient by means of *Glisson's* [or *Sayres'*—Tr.] suspension apparatus, affects an "extension of the spinal cord," which in many cases exerts a favorable effect upon the crises and attacks of pain. It has shown itself, however, very dangerous, in that softening of the spinal cord has been observed after the suspension. Also the modified proceeding in which the feet of the patient remain on the floor or the extension is carried out, the patient lying on an inclined plane, is now abandoned. Where we wish to exercise a mechanical traction upon the spinal cord or the posterior roots, we can accomplish this by having the patient lie upon his back and, draw his knees up as near to his chin as possible, then keeping them in this position for some time by a band passing around the neck and the bend of the knee. In tabetic irritative symptoms, drawing off a few cc of the cerebro-spinal fluid by lumbar puncture sometimes acts favorably.

The most important aid in the correction of tabetic ataxia is the compensatory exercise treatment developed by *Frenkel*, *Leyden*, *Goldscheider*, and others, in which the patient under the control of his eyes attempts gradually to learn again to accomplish the coördination of his movements. These exercises, however, should never be pushed to the extent of fatigue (which is best avoided by watching the pulse, since in many tabetics the sense of fatigue is absent), should be given up when the ataxia is rapidly increasing, and are best begun when we have received the impression that the tabes is in a stationary condition. It is entirely unnecessary to use the complicated apparatus often proposed; most of the requisites can be improvised; for example, the atactic is placed upon a comfortable chair and is requested to practice touching with the point of his foot some numbers which have been written on the floor in front of him until finally he can do this on command, or a small ladder is placed before him (Fig. 64), the individual rounds of which he learns to touch with his foot (practice of eumetria in vertical movements). Then exercises in walking are taken up, in which the feet of the patient are to be placed on marked areas; for this, simple lines on the floor or a strip of linoleum as in Fig. 65 and Fig. 66 can be used. A further

stage is that of making steps of different length, stepping over pieces of wood placed at regular or irregular distances apart, of like or unlike dimensions, etc.; finally, exercises in climbing stairs. For the upper extremities a chessboard, whose individual squares the patient must touch with his finger, a board with holes into which a stopper is to be placed, or an old typewriter, etc., can be used.

In treating the hypotonia and the joint troubles in tabes we may need the aid of the orthopedist to apply a supporting corset in relaxation of the trunk muscles, suitable splints in genu recurvatum and arthropathies. Massage should only be practiced by a skilled operator under the direct control of the physician, otherwise it can only increase the hypotonic and arthropathic deformities and do more harm than good.

As a surgical procedure for the relief of the gastric crises, which, however, on account of its great danger, should only be considered when everything else

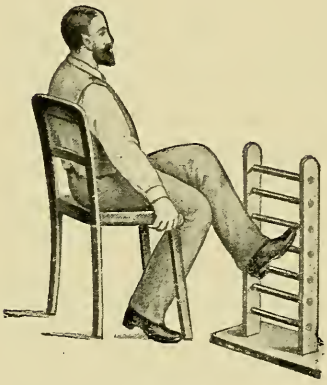

Fig. 64.

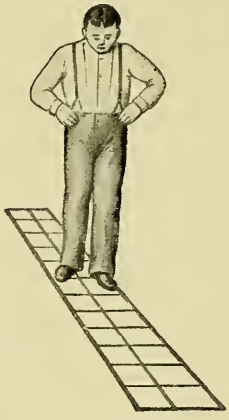

Fig. 65.

Fig. 66.

Exercise Treatment in Tabes Dorsalis.

has failed and the symptoms are excessively intense and persistent, posterior radicotomy has of late been recommended by *Mingazzini*, *Förster*, and others. This is the intradural or extradural resection of the posterior roots D. 7 to L. 1 (in several stages). *Franke* has proposed as a less dangerous substitute operation, neurexaresis of the corresponding intercostal nerves in which the posterior roots are evulsed also, *König*, the (in any case little dangerous) production of deep anesthesia by injecting the nerves in question with 0.5 per cent. novocain-suprarenin solution after *Braun*. This last method can also be tried for the lightning pains, but only temporary results can be expected. For perforating ulcer, finally, *Jaboulay* has proposed an operation which consists in dissecting out the femoral artery in *Scarpa's* triangle on the affected side and separating it for some distance from its vascular sheath. The resulting division of the vaso-constrictor nerve fibers accompanying this vessel, has as its result a peripheral vaso-dilation which has repeatedly led to the healing of ulcers which had resisted all other treatment.

In every tabetic the most minute regulation of his whole manner of life is

of extraordinary importance. The patient should be warned against the damage which every physical overexertion may do him; walks, should be broken by periods of rest, climbing stairs should be restricted as much as possible; mountain climbing, even the slightest, should be forbidden; also long standing is dangerous. Excesses in every direction, particularly in regard to alcohol, must be prevented, sexual excitement avoided. Regular, full, but unstimulating diet, long night's rest, frequent passing urine at regular intervals, combating constipation (when possible by diet); upon all these things great stress must be laid if we wish to aid in fulfilling the conditions contributing to a mild course of the disease. Where there is a tendency to cachexia, but only after gastro-intestinal crises have passed off, full-feeding cures are to be undertaken (see Lecture XXVIII). Last, but not least, the psychotherapy necessary for every tabetic should be considered. Avoidance of the tendency to depressive forebodings and to exaggerated anxious self-observation present in almost all tabetics, may in itself relieve organically caused difficulties, since psychogenic factors often reinforce disease symptoms having a material substratum and very considerably increase their intensity.

LECTURE XIII

The Syphilogenic Diseases of the Central Nervous System

B. The Progressive Paralysis of the Insane. (General Paresis)

With this important disease which *Bayle*, in 1822, isolated as a clinical entity and an intimate knowledge of which has been furnished us by the works of *Falret*, *Westphal*, *Krafft-Ebing* and others, we will occupy ourselves in this lecture only in a very condensed manner. Namely, we cannot attempt to give an accurate picture of the fully developed disease with its different clinical varieties, as nearly every psychiatrist has an opportunity to observe it in institutions, hence these matters can only be considered very cursorily. We must, however, consider quite thoroughly the particularities of the prodromal and early stages of progressive paralysis, the measures indicated in it, and the points important in differential diagnosis coming under consideration, since all this comes especially within the sphere of the neurologist, even also within that of the practical physician, and since here mistakes in diagnosis and other errors (unfortunately quite frequent) may have regrettable consequences for the patient, his family and the community.

The etiology of "paresis" (to use a designation sanctioned by custom) has already been discussed in connection with that of tabes, and we have there learned that along with the primordial syphilitic damage, different injurious factors inherent to civilization are to be considered as important contributing causes. The steady increase in morbidity since the recognition of the disease, which according to nearly all psychiatrists, has occurred, stands in close connection with these facts, though as far as I know, extended statistics proving this latter statement beyond a doubt have not so far been furnished. Also we should not forget that to-day progressive paralysis is much more surely and on this account, much more frequently recognized, than was the case some decades ago. *Arnaud* has shown that paresis occurs about 4 times more frequently in cities than in the country.

Pathological Anatomy

Progressive paralysis has nothing to do with "softening of the brain" as which it is usually designated by the laity (a peculiar persistence of the erroneous term which the psychiatrist *Parchappe* used in the year 1838 "softening of the cortical layer"). Rather is the disease characterized macroscopically by a progressive atrophy of the cerebrum, particularly of its frontal lobes; micro-

scopically by the gradual loss of the tangential fiber layer and of the ganglion cells along with compensatory glia proliferation in the cortex cerebri (particularly in the frontal regions and in the island of *Reil*); further, by alteration of the cortical vessels (ectasies with perivascular or adventitial infiltration with round cells and with the specially typical plasma cells). In many cases there is added degeneration of the pyramids, in others, the systematic degeneration of the posterior roots and the posterior columns of the spinal cord described for tabes. Only where the last mentioned lesion is already developed in the early stages, the clinical picture of "tabo-paralysis" occurs; in the terminal stage on the other hand, more or less marked changes in the posterior columns in the upper cervical region are an almost regular autopsy finding, but clinically irrelevant. Without exception also there is found a fibrous lepto-meningitis, the "arachnitis chronica," which *Bayle* considered the basis of the affection.

PRODROMAL STAGE

The beginning of the disease in the great majority of cases (over 80 per cent.) falls in the 4th and 5th decades of life. According to *Obersteiner's* observations, in 56 per cent. the disease begins between the 36th and the 45th year; the "incubation period" which has elapsed since the luetic infection, he estimates upon the average as 12½ years, the shortest period in these cases was 3, the longest 32, years. Men are about 8 to 10 times more frequently attacked than women. Infantile or juvenile paresis (that is, beginning before the 20th year) is excessively rare and occurs only upon a basis of hereditary syphilis.

The first alterations are in the sphere of the intellect and are manifested by defects of memory (particularly for recent events) further, by reduction of mental capacity and by marked distraction and forgetfulness, so that the patient, for example, loses himself on the streets of his own town, or, like one of my paretics, who repeatedly, when fishing, walked into the stream without his rubber boots. In the ethical sphere also, there is often alteration. The family complain of formerly unknown acts of neglect, of brutal egotism, of continued irritability. The patient is careless in his business, slovenly in his dress, indecorous, even obscene, in his conversation. Physical troubles also appear, however. Attacks of hemicrania not rarely in the form of the so-called "ophthalmic migraine" occur. These we will study in a later lecture (Lecture XXX). If such paroxysms of headache occur in the thirties and forties in people who have not before suffered from migraine we should be awake to the eventuality of a threatening paresis. The same remark applies to vertigo, feeling of pressure in the head, sleeplessness and other "neurasthenic" symptoms when they occur for the first time in former syphilitics, in these ominous decades, and we can find no causal factors for acquired neurasthenia (see Lecture XXVIII). Particularly alarming, however, are the following, it is true not very frequent, phenomena: Temporary paresis of one hand, temporary eye muscle paralyses with diplopia; temporary pupillary rigidity or anisocoria, fleeting nocturnal enuresis. The patient usually feels ill and has a sense of alteration of his psychical personality. The fear of threatened "softening of the brain" sometimes drives him to the physician. It can also lead to suicide or a suicidal attempt. How

valuable in confirming the diagnosis in this stage the "four reactions" of *Nonne* can be we have already mentioned.

THE INITIAL STAGE

In the initial stage there occur in different groupings a large number of objective, mainly somatic anomalies which even without the aid of biological methods and the anamnesis throw a clear light upon the nature of the affection present. These are:

1. *Pupillary Alterations.*—The extreme importance of these anomalies in the symptomatology of paresis is shown from the fact that according to the statistics of *Mignot, Schrameck* and *Parisot* only 6 per cent. of the cases present intact pupils. The anomalies of iris innervation in progressive paralysis are in the main the same as those in tabes dorsalis, and after the very careful investigation of *Bumke*, the *Argyll-Robertson* phenomenon (reflex rigidity or slowness) occupies by far the first position in frequency. It is interesting, too, that *Wolff, Gaupp, Reichardt* and *Bumke*, in altogether about 70 cases of paresis with the *Argyll-Robertson* symptom, found in each instance changes in the posterior columns in the cervical region, that is, a combination with tabes superior. *Fürstner* and *Naka* have indeed occasionally found absolutely intact posterior columns with clinically surely recognized reflex pupillary rigidity, and we are hence of the opinion that even without tabetic complications the typical *Argyll-Robertson* symptom can occur in paresis, namely in the initial stage. The cases which come to autopsy, however, have almost without exception reached the terminal stage, and we have already remarked, that in this last, the posterior columns of the upper cervical region are almost never intact. Further pupillary symptoms of paresis are absolute immobility (more frequent than in tabetics), irregularity, abnormal wideness or narrowness (the first rarer than the latter), anisocoria (very frequent), and the so-called "bounding mydriasis," that is, a rapid variation between the width of the right and of the left pupil. The last phenomenon originally considered pathognomonic for paresis and tabes, occurs also in *Basedow's* disease and even in neurasthenia and hysteria.

2. *Speech Disturbances.*—Already in 1814 the genius *Esquirol* wrote in a medical lexicon in an article on "Dementia," "Speech difficulty is a fatal sign," and it is not to be doubted that he had in mind cases which since then have been classed with dementia paralytica. Even the public knows that "softening of the brain" begins with trouble with the tongue. The first disturbances usually present themselves in the form of the so-called "syllable stumbling" which *Kussmaul* defined as follows: "A disturbance of the coördination of the whole word as a speech unit, with intact formation of sounds and syllables." The patient misplaces letters and syllables, leaves them out, or repeats them in the course of the word (perseveration), confuses them with similar sounding ones. So, for example, for artillery brigade, he says artrallery brigade; for abnormality, abnormalty; for truly rural, tooly looral, etc. If he is required to repeat these somewhat complicated words several times it is noticed that each time the disturbance, which at the start was overcome by strained attention, becomes more marked and finally the word becomes unrecognizable. One must, however, be

careful with this test since the more educated patients may so be brought to make a diagnosis of their own cases. It is better on this account to have them read a newspaper article adapted to bring out the defect under the pretext of testing their vision. In this (as well as in the course of conversation with the patient) the following ominous speech disturbances are readily noticed: First, the "drawling" speech in which it is noticeable that the more important letters are weakened, p is pronounced like b; t, like v (the form of the mouth is naturally to be noticed here), particularly that r is pronounced badly, more or less like l. Further, frequent mistakes, substitution of words, paraphasias, which the patient usually does not observe or correct (in contradistinction to neurasthenics or mentally fatigued healthy persons) and the slow and hesitating tempo, as well as the monotony and dullness of the diction, often also a certain nasal quality. In conversation, finally, the patient must strikingly often stop and think of a word or phrase, makes many grammatical errors and mistakes of syntax, stalls in the middle of a sentence, etc.

3. *Motor Irritative Symptoms.*—In the first place the exceedingly characteristic fibrillary twitching in the region of the lower facial should be mentioned; this either plays over the countenance like summer lightning, now right, now left, now about the corners of the mouth, now over the chin, or spreads as an almost continuous fluttering and trembling over the orbicularis oris, levator anguli oris and quadratus menti; indeed, occasionally (on emotion, etc.), it extends over the alæ nasi and cheeks up to the eyelids. Also the outstretched tongue waves and trembles; sometimes there are recognized in it, plainly, gross contractions of the larger muscle bundles. In the muscles of the extremities anything analogous is scarcely ever perceived. However, tremor of the fingers is very frequent. It is not very characteristic but is quick and fine, increasing upon excitement and exertion, and does not cease even upon complete rest.

4. *Alterations of the Handwriting.—Obersteiner* gives the following intelligent description of the handwriting of the paretic in the initial stage: "The individual type of the characters is altered, the handwriting is often smaller, sometimes larger, and pointed; it appears (analogue of the hesitation in speech) not as from a copy, often markedly artificial with unusual flourishes, etc. Beginning affection of the muscle sense, muscular insufficiencies manifest themselves, by the improper use of light and heavy strokes, getting off the line, unequal size of the letters, angular excursions, wavering and zigzag strokes; in addition, leaving out of words, or at least of syllables, occasionally doubling of the latter, mutilation of words, to paraphasic phenomena."

5. *The "Paralytic Facies."*—The facial expression of many paretics shows even in the initial stage so typical a picture that a diagnosis at first sight, even among the crowd in the reception-room, is not infrequently possible for the experienced physician. The patient looks like an "all nighter," sleepy, relaxed, expressionless, exhausted; his face has about it something dull and empty, the action of his muscles of expression is particularly limited. Asymmetric innervation of the facial muscles on the two sides (sometimes varying, so that to-day the right, a few days later the left naso-labial fold appears more marked), is indeed by far not so frequent as in the later stages, but is not rarely observed at this time.

The Further Clinical Picture

Along with the 5 groups of symptoms which we have now considered and which can be called the stigmata proper of beginning paresis, the clinical picture otherwise unfolds itself in this early stage in the following manner:

The psychical anomalies which impress their stamp already upon the prodromal stage appear now in much more definite manner, since the weakening of judgment and beginning disintegration of the mental personality become continually more evident.

The amnesia for recent events is to be observed in much higher degree. Along with this, there is from time to time, clouding of consciousness which leads to occurrences which can no longer be considered as the result of pathological forgetfulness and abstraction, but as true confusional conditions. In these, things may be done which bring the patient into conflict with the police and the law, for example, if he enters a strange house at night, begins to sing or to whistle in church during the sermon, urinates on the pavement of a crowded street, etc. On the other hand the defect in ethical inhibition takes the upper hand in a way which often leads to forensic consequences; particularly often there are sexual assaults upon children, exhibitionism, swindling, defalcations, aggressive behavior in public places and on the street, cheating at cards, etc. A teacher treated by me ran amuck among his pupils with a pair of compasses. The increased feeling of self-importance which, in the fully developed disease assumes the classical form of "grandiose delusions," can show itself even in the initial stage in senseless expenditure, reckless making of debts, a craze for invention, tendency to risky undertakings, etc. Inability to criticise his own acts or those of his surrounders, testifies to the growing preponderance of intellectual defects. The physical troubles enumerated for the prodromal stage either diminish in the initial stage or, on the contrary, they manifest such obstinacy and intensity (particularly vertical headache and sleeplessness) that they are markedly differentiated from the same symptoms as observed in neurasthenia. Occasionally an unconquerable desire to sleep by day is in contrast with the agrypnia by night. Consciousness of illness is now still present only for the physical troubles, not for the mental abnormality. Not rarely there appear even in the initial stage those ominous paroxysms from which only a minority of paretics are spared, but which, as a rule, do not come on until later in the disease, and which are known under the name of "paretic seizures." We differentiate apoplectiform and epileptiform attacks. In the first the patient falls unconscious as in a true attack of apoplexy, and shows a hemiplegia after he comes out of the coma. This, however, entirely disappears within a few days (occasionally even after a few hours), and the attack is accompanied by a rise in temperature which is in contrast to the subnormal temperature of cerebral hemorrhage but parallel to many of the apoplectiform paroxysms of multiple sclerosis. *Krafft-Ebing* has pointed out a further differential point in their diagnosis from cerebral hemorrhage. In the latter, the tendon reflexes in the region paralyzed are regularly lost for about 24 hours after the attack; in the paretic hemiplegias, they are exaggerated immediately after its onset. *Kraepelin* is of the opinion that many cases of death at middle age from "apo-

plexy" are really examples of a fatal ending of apoplectiform attacks in the initial stage of paresis. More frequent are epileptiform attacks, which also are often accompanied by hyperthermia. They have usually the character of the so-called "Jacksonian or cortical epilepsy" which we will study in a later lecture (XIX) more fully. At present I would only remark that they consist in tonic and clonic spasms which usually begin in one corner of the mouth and spread from there over the face and extremities of the same side and often to the other side. Sometimes, however, they exactly resemble genuine epilepsy: The patient falls with a cry, is unconscious, shows general tonic and clonic convulsions, his face is cyanotic, fæces and urine are involuntarily discharged, and he foams at the mouth. After the attack, which lasts only a few minutes, the patient is confused for a long time and regains entire consciousness only little by little. In connection with paretic seizures, both apoplectiform and epileptiform, there occasionally occur, usually very temporary, aphasic, or apractic conditions, symptom complexes, which we will subject to a thorough consideration in a special lecture (XVIII).

Changes in the optic nerve corresponding to those in tabes can also occur in paresis, but this is, on the whole, quite rare. *Joffroy*, among 227 paretics, only saw more or less developed atrophy of the papilla 27 times, and these were usually alterations which developed only in the later stages.

THE STAGE OF THE FULLY DEVELOPED PSYCHOSIS

The paroxysms just described can, in that they leave behind a considerable degree of mental deterioration which further rapidly develops into dementia, mark in rather precise manner the passing of the initial stage into dementia paralytica proper; more frequently, however, this change is imperceptible. Where, however, the disease which has fully developed manifests itself in delusional ideas and delirious conditions, the beginning of a new and more severe stage of the affection is recognizable even to the laity, the usual consequence of which is the bringing of the patient under psychiatric care.

In the usual manner this paralytic psychosis will now be subjected to a rather schematic classification in that we speak of a depressive, an expansive, an agitated, and a dement form. If it is always remembered that between these forms there are intermediate varieties, and that they even can pass over into one another, there is no objection to using didactically these divisions. We will hence, adhering closely to the descriptions of *Obersteiner* and *Kraepelin*, present the clinical criteria of these 4 chief forms.

1. *The Depressive* (*Melancholic*) *Form.*—Depressive delusional ideas which show lack of insight, loss of power to criticise and which cannot be corrected, are manifested, often in an absurd manner. These can be of hypochondriacal nature (the patient, for example, laments that his head is shrunken, that he has no stomach, that his body has turned to pus, etc.), or they can take the form of "micromania," having committed great sins, etc. (The patient complains that he has stolen a million; that he has destroyed his family; affirms that he is dead and refuses food, since a corpse cannot eat.) Into the course of this depressive form, conditions of maniacal exaltation with grand delusions

may force their way, and when the alternation is regular, we speak of the "circular form" of paresis.

2. *The Expansive* ("*Classic*") *Form.*—In this variety, to which the first descriptions of the disease applied, grandiose ideas, increasing to the immeasurable, dominate the picture. While the patient first contents himself with declaring that he is the richest, the handsomest and the healthiest man in the country; before long he takes himself for a king, an emperor, God, Supergod; he is going to marry princesses; to buy Australia; he is building a railroad to the moon, etc. One of my patients, immediately after the epileptiform attack which ushered in his psychosis, wrote to the King of the Greeks to urge him "in the name of the Swiss people gathered on the shores of the Lake of Zürich" to make a republic out of Crete.

3. *The Agitated* (*Maniacal*) *Form.*—This variety is characterized by conditions of maniacal and delirious excitement. The patient disturbs day and night, roars and sings, holds confused conversations marked by flight of ideas, sleeps hardly any, loses flesh rapidly; attacks of real frenzy and dangerous assaults upon his surrounders also occur. The most severe cases of agitated paresis have been called "Galloping Paralysis." In these the patient after a few weeks of the most extreme excitement, obstinate refusal of nourishment, complete filthiness and absolute sleeplessness, perishes from heart weakness or from dysenteric diarrhœa.

4. *The Dement Form.*—Under this name, we include all the cases in which from the start progressive dementia stands in the foreground of the psychical symptomatology. Amnesia stretches continually farther backward until finally his whole previous existence with all its impressions and experiences are obliterated from the memory of the patient. Orientation for time and for place suffers and is finally lost entirely. The power of speech is impaired more and more, not only on account of the increase of the syllable stumbling and the anomalies occurring even in the initial stage, but also through the progressive extinction of the speech memories. "Amnesic aphasia" of the most extreme degree so occurs. Since also the "conceptions of movement" are defective an "ideational apraxia" goes hand in hand with this last. The gait also is slow, clumsy, uncertain; there is incontinence of urine and fæces.

Remissions and Intermissions

The severe disease pictures just described can (sometimes even suddenly, almost from one day to another) so far disappear that the patient and his family get the impression of a cure, while we, in such cases, usually speak only of remissions, occasionally, however, of true intermissions. In the remissions a return to the psychical and somatic status of the initial stage usually occurs. Sometimes, however, there remain more or less marked speech disturbances, and regularly indeed the pupillary anomalies formerly present. During these remissions which last for weeks and months, seldom for years, the patients are often let out of institutions by psychiatrists and many times take up their callings again. It is of capital importance that they remain under permanent and careful medical observation; only too frequently, however, they are with-

drawn by their friends from such oversight until a paretic seizure or the sudden recurrence of their delusional ideas again bring to the front the great seriousness of the situation and dissipate the beautiful illusions as to cure. I have so far only seen paretics of the expansive and depressive type, who have been let out of the asylum on account of remissions, return to my care; the longest of these remissions lasted one and one-half years. Frequently I have been able from return of the agrypnia, slight speech disturbances, pathological irritability, headache, unreasoning desire to speculate, etc., to diagnose the threatened return of progression of the disease and to put the patient back in the asylum in time; in one case in which the wife of the patient could not be convinced of the necessity of this measure, there broke out on the next day a condition of extreme excitement with tendencies to violence and to suicide, in which dreadful results were only with great difficulty prevented. One should also exercise continued control over paretics during these remissions since it is not only one of the most thankless tasks, but also one of the most responsible positions in which the physician can be placed. The great frequency of remissions in the classical form of progressive paralysis is emphasized by most psychiatrists; in the agitated form they are rare, in the dement form they almost never occur.

True intermissions of year-long duration are not entirely rare. My teacher, *L. Wille*, told me about one of his paralytic patients, who, during an intermission, was able to assume again his position as minister of a small German state. A patient of *Kraepelin's* not only discharged his duties as employee in the telegraph office for 5 years, but rose to higher positions, stood examinations and married. Another one who presented grandiose delusions, speech disturbance, pupillary rigidity, *Westphal's* symptom, and attacks of vertigo, lost his delusional ideas and could again occupy his former position as janitor at a school for 6 years. A patient of *Tuczek*, who in his grandiose delusions declared himself Pope, Emperor of Germany, King of Darmstadt and father of 137 boys born at the same time, who presented the characteristic somatic stigmata of paresis, and after repeated paretic seizures, had already passed into a condition of advanced cachexia, little by little recovered completely both physically and mentally, took up again his position as conductor, and 5 years after the beginning of his intermission was promoted "on account of exceptionally good work," then 2 years later had to give up his position finally because of "tabes." Only just before his death, 20 years after the beginning of the intermission, he presented again mental anomalies, which however, *Knoblauch*, under whose care he had come, considered only as "marantic confusion." The autopsy showed in the brain none of the alterations characteristic for progressive paralysis. It was otherwise in a case of *Dobrschansky's*, in which after an intermission of 15 years, practically amounting to recovery, the autopsy showed typical paralytic alterations.

THE TERMINAL STAGE

However different the clinical pictures of florid paresis may be, the patients finally in the end stage all come into the same wretched condition. The delusions have disappeared in the predominating dementia, the patient has lost

all mental relations with the external world, even, indeed, the consciousness of his own personality, his movements are now only rudimentary, his discharges are all passed in the bed, he utters only inarticulate sounds, must be fed, the heart's action grows feeble, the skin is anesthetic, livid, œdematous, trophic disturbances, bed-sores, abscesses, hypostatic and aspiration pneumonias develop, and put an end to a vegetative existence hardly any longer to be considered as "life," if death does not occur from paralysis of the heart in a paretic seizure. In the end stage, "status epilepticus" from an unbroken series of attacks is frequent.

Prognostic

Cases like those of *Tuczek* and *Knoblauch*, are such rare exceptions that they cannot prevent us from making the general statement, that general paresis leads inexorably to death; besides, the autopsy findings in these patients also do not exclude the fact that there are particularly complete and long intermissions, during which the patient may be taken away. It is certain further, that in many of the so-called "recovered pareses" the condition was really some other symptomatologically closely related affection (alcoholic pseudo-paralysis, cerebral syphilis with psychosis, etc.). The proximate causes of death can be: in the early stages, and further, at the height of the agitated and depressive forms, suicide; in all stages, paretic seizures; in the terminal stage, aspiration or hypostatic pneumonias, ascending infection of the urinary tract, sepsis from bed-sores or injuries, asphyxia from "choking," finally, simple marantic heart failure.

Between the individual clinical varieties there is a certain difference in malignancy. Relatively more favorable is the "Classical," the expansive form; in it remissions may most frequently be hoped for, the paralytic attacks which almost always hasten the progress of the disease are comparatively rare, and finally, long stationary periods occur. So, then, this form can here and there present quite a long duration of the disease (to about 15 years); in general, however, from the beginning of the grandiose ideas to death, a period of about 3 years can be predicted. The other forms usually lead in less than 2 years to death; apart from the "galloping" variety of the agitated form, the dement type is most malignant, since in it, the paretic seizures are very frequent during the whole time of the disease and remissions are almost entirely excluded.

Differential Diagnosis

In the prodromal stage the differentiation of progressive paralysis from neurasthenia can be a matter of great difficulty when it concerns a patient who has had syphilis. Not a few former syphilitics become neurasthenic on account of fear of paresis. It has been asserted, however, often with too great positiveness, that this nosophobia speaks against paresis, since in this, prodromal autodiagnosis are not at all rare. It is notable on the other hand, that the neurasthenic occupies himself much more with his troubles and observes them much more carefully and in detail, notes them down and describes them to the physician more frequently, than the, even at the start, rather indolent paretic, who is

generally indifferent and forgetful as to the changes in his own personality. Plain alterations of character for the worse, with transgressions against propriety and custom, speaks strongly for paresis. Further, the "weakness of memory" complained of by neurasthenics on careful testing, shows itself to be only the expression of the imperfect and abnormally fatigable, power of mental concentration, while that of the paretic represents a true dys- or amnesia for recent events which, indeed, is plainer objectively than subjectively. Loss of topographic memory pictures, getting lost, never occurs in neurasthenia. Very properly, on the other hand, *Oppenheim* suggests that imperative conceptions speak decidedly against paresis. Very suggestive of the latter are temporary paresis or nocturnal enuresis. With entrance into the initial stage with its characteristic somatic anomalies and the increasingly more evident psychical disturbances, the differential diagnosis soon loses its difficulty, particularly when a paretic seizure roughly tears down the last veil. We must not forget here to allude again to the great value of the "4 reactions" of *Nonne*.

Also when the diagnosis between hysteria and beginning paresis is difficult the chemical, cytological and serological blood and spinal fluid examinations can greatly aid us. As an example I would mention to you a case described by *Jusgen* which also is important medico-legally since not rarely a head injury gives the signal for the outbreak of progressive paralysis. It was that of a workman who was struck upon the head by a piece of falling timber while in apparently good health and in whom, after about 4 weeks, mental alterations not rare in traumatic neuroses appeared; now indifference, now labile "affects" and nervous complaints. Clinical examination showed "hysterical" convulsions which partially could be relieved by suggestion; on the other hand no positive signs of progressive paralysis. Nevertheless, blood examination showed a strongly positive *Wassermann* reaction, as did also that of the spinal fluid with increased globulin and pleocytosis. The further course of the disease justified these reactions; during the first weeks of observation, indeed, the hysterical symptoms remained fully developed, namely, there was great suggestibility. Then, however, there was rapid, unmistakable change; negativism was substituted for suggestibility, the patient became restless, spoke in monotone, manifested foolish, depressive ideas and passed into a marked paralytic stupor.

The differential diagnosis from multiple sclerosis was already discussed in connection with this last (page 139). That from the so-called "pseudo-paralysis" (of luetic and arteriosclerotic nature) as well as from certain manifestations of brain tumor, will be taken up in later lectures (XIV, XV, XIX). We must not neglect to consider at this point "sleeping sickness," all the more, since this affection, as *Spielmeyer* has recognized, not only has clinical analogies with progressive paralysis, but also etiological relations (similarity between trypanosome and spirochæte infection) and a certain pathologico-anatomical agreement. Along with the somnolence, this endemic disease of the black portion of the earth's population, which is called after its most striking symptom, shows frequently disturbances of motility and of the reflexes, epileptiform attacks, speech disturbances, decrease in intelligence, weakness of memory, delusional ideas; besides this lymphocytosis of the spinal fluid. In Africa the differential diagnosis between these two conditions can sometimes be difficult,

namely when the "sleeping sickness" (which indeed is seldom the case) attacks a white person.

Treatment

On account of our inability to permanently arrest the disease process, as soon as the diagnosis is confirmed, we must endeavor to delay the development of progressive paralysis as much as possible and to favor the occurrence of remissions. In the prodromal and initial stages the most important point is immediate removal from business activity and transference to as quiet and nonirritating surroundings as possible, for example, such as are found in well managed sanitaria. You cannot be too urgently warned against sending such patients to hydrotherapeutic institutions (there are unfortunately too many) where the patients are subjected to fixed cold water procedures and incipient paretics are thereby greatly injured. If in a neurasthenic condition there is even the slightest suspicion of paresis in the prodromal stage, care should be taken that the patient is not given cold baths or douches. Great importance is to be attached to full but simple and nonconstipating diet, with avoidance of all stimulants (alcoholics, spices, tea, coffee). For sleeplessness drugs must often be resorted to (veronal, 0.5–1.0 (gr. 8–15); trional, 1.0–2.0 (gr. 15–30; scopolamin, 0.0005 (gr. $\frac{1}{120}$); paraldehyd, 3.0–5.0 (5i–5ii) (greatly diluted); amylenhydrate, 2.0–4.0 (5 ss.–5i), (greatly diluted), may be alternated as necessary). No time should be wasted on baths, sponging, or psychotherapy; if the tendency to agrypnia is reduced, the hypnotics may be gradually limited or withdrawn, although in this case they represent the less of two evils, since we must unceasingly attempt to procure rest for the diseased brain during the night.

The question as to whether antisyphilitic treatment should be instituted or not, cannot be answered in general terms. Many psychiatrists and neurologists, as *Krafft-Ebing*, *Kraepelin* and *Dejerine*, consider mercurial treatment as contraindicated when the diagnosis is certain, since it reduces the nutrition of the patients and in a metasyphilitic disease can accomplish nothing. Others, as *Leredde* and *Ziehen*, on the contrary, are of the opinion that under mercurial treatment remissions oftener occur, are more marked and last longer than without such treatment. *Ziehen* hence advises the trial in every incipient case of a course of inunctions with subsequent administration of iodides, and even in advanced cases he has an inunction made twice a week and gives 0.2 (gr. 3) iodide of sodium a day. I consider an intermediate standpoint correct, and limit the mercurial treatment to early cases which have not previously been sufficiently mercurialized, that is, when they give a decidedly positive *Wassermann* reaction; further, I avoid too energetic treatment, and content myself with a series of 10 to 12 intramuscular injections of about 1 cc of the following solution: Hydrarg. biniodid, 0.2; sodii iodid, 0.2; sodii chlorid., 0.075; aq. dest. 10 cc. (The series can be repeated later.) I have certainly never done any harm with these injections (given at intervals of from 1 to 2 days), but believe that I have occasionally aided in the establishment of remissions. As to salvarsan, with which, however, I have had but very limited experience, I can neither affirm the one thing nor the other. Since I have in two instances

gained the impression that it accelerated the disease process or provoked seizures and have had similar experiences narrated to me by colleagues, I advise on principle against this otherwise so valuable remedy, in paresis. Only in one case was there improvement, and this was only in the subjective symptoms, while speech disturbances, facial tremor, sluggishness of the pupils, intellectual weakness, etc., were not influenced in their progress by the injections.

Into the symptomatic therapy and care of the fully developed disease and of its terminal stage (sedative drugs, keeping clean those who suffer from incontinence, the prevention and treatment of bed-sores, tube feeding in refusal of food, when there is danger of choking, etc.) we need not enter; these things are mainly for the asylum psychiatrist. It may be said, however, that for getting a disturbed paretic into the asylum, a large dose of scopolamin hydrobrom. (0.001=gr. 1/60) injected subcutaneously, exerts a strikingly sedative effect ("chemical camisole").

In conclusion, a few words upon some new and interesting, although yet incomplete, therapeutic experiments. These are based upon the familiar experience that remissions in progressive paralysis occur, not very infrequently, after febrile, infectious and suppurative processes. Upon this was based the old methods of provoking suppuration by the inunction of *Autenrieth's* "Pustulating ointment" (ung. tartari-stibiati) or by vaccination. Of late the Vienna psychiatrist *Wagner v. Jauregg* has advised the systematic use of tuberculin injections, upon the favorable effects of which with regard to the occurrence of remissions and in prolongation of life in paretics, *Pilcz* and *Dobrschansky* have reported. Beginning with a dose of from 0.01 to 0.1 of tuberculin, the dose is gradually raised to 0.5; this last dose is given 7 to 12 times, each injection at the interval of about 2 days. The raising of the dose is dependent upon the degree of febrile reaction; temperatures of over 39° (103.1° F.) are avoided as far as possible. Sometimes *Wagner* precedes the tuberculin cure by antiluetic treatment. *Friedländer* has reported favorable results from dead cultures of colon and typhoid bacilli. *O. Fischer* and *Donath* produce in paretics an artificial hyperleucocytosis by injections of nucleinic acid; the latter uses the following solution: ℞ sodii nucleinat., sodii chlorid., āā 2.0; aq. dest. steril., 100. M. S. use for one or two injections. The injections, carried out at intervals of from 5 to 7 days, should produce a leucocytosis of from 16,000 to 25,000 lasting some time. Sometimes they produce decided local reaction. *Donath* found in 70 per cent. of the cases treated more or less marked improvement.

The recent discovery by *Noguchi* and *Moore* of spirochætæ in the brain substance in general paresis (confirmed by other observers) seems to settle the question of the relation between this disease and syphilis. Very recently *Noguchi* has reported finding spirochætæ in the spinal cord in one case of tabes out of twelve examined. In view of these discoveries, it has been urged by some writers that the terms "meta-" and "para-syphilitic" be abandoned. In so far, however, as they are used to imply certain pathological changes differing from those usually considered characteristic of syphilis, they may well be retained.

On account of the inaccessibility of the spirochætæ in the central nervous system to salvarsan introduced into the circulation, *Swift* and *Ellis* have recommended administering this remedy intravenously, one hour later, bleeding, centrifugating the blood, heating the serum for one hour to 56° C., letting stand on ice overnight, making a 40 per cent. dilution of this serum

with physiological salt solution, and after preliminary lumbar puncture and withdrawal of a corresponding amount of fluid, injecting into the spinal canal 15 to 330 cc of this "salvarsanized serum." It is thought that by this means the salvarsan in the blood may be brought directly into the ventricles and lymph spaces, and can attack the spirochætæ there harbored. While this method has been used to a considerable extent in the United States, it has not entirely fulfilled the hopes attached to it, and our experience with it is as yet insufficient to warrant a positive opinion as to its value. The direct injection of salvarsan into the spinal canal has been tried, but some serious results have followed this method of administration, and its technique is as yet insufficiently developed, hence it can hardly be recommended at the present time. —*Translator.*

LECTURE XIV

The Syphilogenic Diseases of the Central Nervous System

GENTLEMEN: Now that in the last three lectures we have studied the two most typical representatives of the metasyphilitic nervous diseases, we will take up to-day the consideration of syphilogenic affections, which in pathologico-anatomical and symptomatological relation present less uniformity. These are, 1, the so-called syphilogenic combined system diseases and 2, cerebro-spinal syphilis.

C. Syphilogenic Combined System Diseases

In Lecture VIII we have seen in *Friedreich's* disease a representative of the so-called combined system diseases, but put off the general consideration of these in no way frequent disease forms until later. It is now time to enter into a short discussion of this nosological conception.

While in spinal hereditary ataxia there is the combination of a purely endogenically caused degeneration in the different sets of fiber systems of the posterior and lateral columns of the spinal cord, similar lesions can occur from other causes. First from the action of poisons or toxines which are elective, but which affect at the same time several categories of long tracts and which lead to the progressive degeneration of the neurones affected. Next, as particularly *Nonne* and *Früind* have shown, disseminated focal diseases, starting from the vascular apparatus, which is situated within the areas of definite fiber systems (for example, the pyramidal tracts, or the columns of *Goll*) which in consequence of the descending or ascending degeneration of these tracts so caused, can furnish the anatomical substratum for combined system diseases. On microscopical examination this pathogenetic subvariety can usually be distinguished without great difficulty from the other varieties by the lack of sharp limitation of the sclerosed areas, as well by the marked vascular alterations; symptomatologically, however, the result can be identical. Finally, it may be emphasized that the greater susceptibility of the pyramidal tracts and the posterior columns to ischemic damage makes an apparently elective affection of just these tracts, even in more diffuse vascular lesions, comprehensible. In these different ways the combined system diseases which have been observed in the following general diseases are explainable: Chronic alcoholism, chronic lead poisoning, diabetes, carcinomatosis, pellagra (chronic intoxication by spoiled corn), lathyrism (intoxication from chickpeas), chronic nephritis, pernicious

anemia, leucemia, sepsis. In practical importance these conditions are far inferior to the syphilogenic combined system diseases with which they agree entirely from a symptomatological point of view. The description of these last may hence serve as paradigm for the whole group.

Let us add that in the production of syphilogenic combined system diseases, in part simple degenerative processes as in tabes, in part tertiary luetic endarteritic processes, are in action. The disease pictures under consideration are hence to be considered partly as metasyphilitic, partly as purely syphilitic; their description, thus, conducts us in a natural manner from tabes and paresis to cerebro-spinal syphilis.

The combined destruction of centripetal systems (affection of the posterior column) and of the most important centrifugal fiber complexes (Tractus cortico-spinalis) has as a consequence, that clinically the combination of atactic and hypesthetic phenomena predominates. Now, however, as we know from

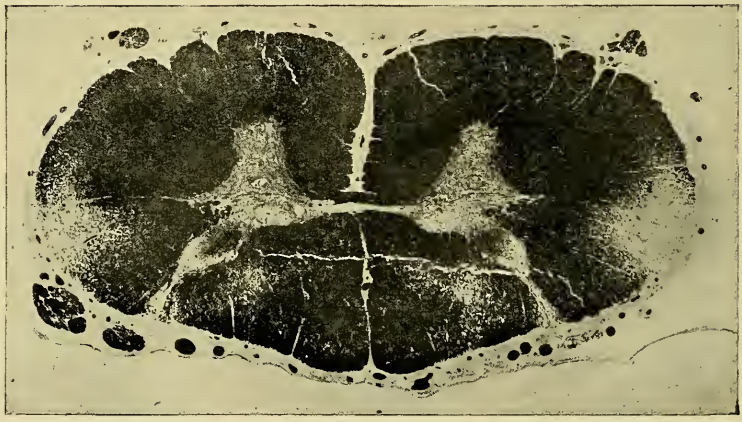

Fig. 67.

Changes in the Spinal Cord in Syphilitic Spinal Paralysis. *Weigert-Pal* Stain.

our former descriptions, elimination of the pyramidal tracts has as its consequence hypertonia and hyperreflexia, that of the posterior fasciculi, hypotonia and hyporeflexia. Hence, in combined lesions the influence of diametrically opposed tendencies make themselves felt. The result depends upon the greater or less intensity of the process in the different parts affected.

If affection of the pyramids predominates (see Fig. 67), there results the symptom-complex of spastic-atactic paraplegia with sensory disturbances and we call such cases, after *Erb's* recommendation, syphilitic spinal paralysis. The sensory disturbances are often very slight. As I have shown, they are, as a rule, most easily recognizable on testing vibration sense with a tuning-fork. Besides this, lancinating pains, bladder disturbances, reflex pupillary rigidity and optic atrophy are sometimes observed, just as in tabes dorsalis.

If, on the other hand, the pyramidal affection is less than that of the sensory tracts (see Fig. 68) a syndrome characterized by ataxia and areflexia with motor weakness, is produced, and besides this, phenomena pathognomonic for the pyramidal affection (chiefly *Babinski's* symptom, more rarely that of

Oppenheim or of *Mendel-Bechterew*) usually are present. Also, in this case, the occurrence of lancinating pains, bladder disturbances, the *Argyll-Robertson* pupil and optic atrophy are not rare, and we speak of "combined tabes."

Pure cases of syphilitic spinal paralysis and of combined tabes are rarer than their accompaniment with symptoms that belong to cerebro-spinal syphilis.

The course of both forms is slowly progressive, in combined tabes on the average more rapid than in syphilitic spinal paralysis. In it also, the prognosis is clouded by the not so rare development in addition of general paresis and also by the fact that specific treatment can accomplish much less than in syphilitic spinal paralysis. In this last, indeed, which begins often in the first years after luetic infection, the therapeutic outlook is relatively good. If

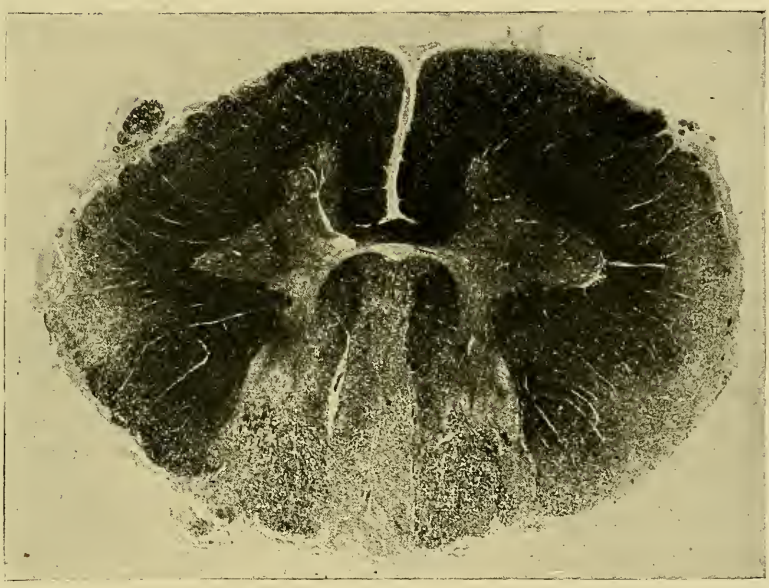

Fig. 68.

The Combined Tabes. *Weigert-Pal* Stain.

the treatment is begun very early complete recovery is possible; if later, a definite standstill can at least be obtained, and to this the disease may spontaneously tend. As an early symptom the so-called "spinal intermittent limping" is of great importance; a description of this I will take up in the next lecture when speaking of arteriosclerotic diseases of the spinal cord. We will consider treatment at the end of this lecture.

D. Cerebro-Spinal Syphilis

In the great majority of cases in which tertiary syphilitic processes develop in the central nervous system they appear simultaneously in the brain and in the spinal cord. This is explained by the facts that the syphilitic lesions of the nerve centers do not arise from the parenchyma proper, the ganglion cells and nerve fibers, but are partly of meningeal, partly of vascular origin,

and that both the meninges and the vessels of the brain and spinal cord present a unique and connected whole. Let us enumerate now the chief alterations which present the anatomical substratum of cerebro-spinal syphilis.

1. The gumma or syphilitic granulation tumor occurs but rarely as a gross and solitary structure, and then on the convexity of the cerebrum; its place of origin is then usually the dura mater and the symptomatology agrees with that of true neoplasms of corresponding size and analogous location. Much more frequent is the formation of small and multiple gummatous nodules which prefer the basal portions of the brain and usually are situated in the pia-arachnoid or on its vessels. Where they are particularly numerous we speak of "gummatous basilar meningitis." Also the gummata developing on the spinal cord are almost always multiple and originate in the meninges; they are frequently miliary, so that on microscopical examination they may be mistaken for tuberculous nodules.

2. Syphilitic endarteritis, a disease of the vessels whose chief characteristic consists in a progressive thickening of the intima by a new formed connective tissue rich in cells. It can lead to complete obliteration of the lumen of the vessel. As a rarity the deposit of minute gummata in the new formed endarteritic tissue has been described. Syphilitic endarteritis is ubiquitous in the central nervous system; it can be found as well in the meninges or on the surface of the brain and spinal cord as in their interior.

3. Meningo-encephalitis and meningo-myelitis syphilitica. At the basis of these processes lies a diffuse infiltration of the pia-arachnoid with small cells, which in later stages, leads to considerable connective tissue thickening and infiltration of this with a gelatinous deposit. Gummatous deposits, on the other hand, are absent in typical cases. In the altered meningeal region, however, the vessels without exception undergo endarteritic and periarteritic changes; their passing into the superficial portions of the central nervous system furnishes to the infiltration process a port of entry for propagation in the cortex of the brain or in the white matter of the spinal cord. In the first instance the convexity of the hemispheres, in the last, the posterior part of the periphery of the spinal cord, are seats of predilection.

Symptomatology

a. *Cerebral Symptoms.*—A particularly constant symptom is headache which usually has a paroxysmal exacerbating character and occurs with the greatest severity, particularly at night or early in the morning. At the height of its intensity it assumes, usually, a boring and hammering character; during the remissions it is usually described as a dull and continuous feeling of pressure. At the height of the paroxysm there is not very rarely nausea or vomiting, on which account on superficial diagnosis it is occasionally mistaken for migraine. The comparison of clinical observations with autopsy findings justifies us in considering this headache as an expression of basilar-meningitic processes. Only when this last assumes a high degree of intensity is syphilitic headache accompanied by stiff neck and rise of temperature. Circumscribed sensitiveness to tapping or to pressure over the skull is scarcely

ever to be found in syphilitic basal meningitis; where it is found, a suspicion that the convexity of the cerebrum is affected is justified.

An important rôle in the picture of cerebral syphilis is played, further, by the eye symptoms, which often involve the optic nerve, since the region of the chiasma presents a point of predilection for the localization of tertiary products. Ophthalmoscopic examination shows different pictures, among which papilloedema and optic neuritis depending upon increased intracranial pressure are most important. We will consider the first when speaking of brain tumors (Lecture XIX); the last—whose end result is neuritic optic atrophy—has already been considered in Lecture II among diseases of the cranial nerves. Simple, degenerative optic atrophy is sometimes found in cerebral syphilis and in a form entirely agreeing with what is observed in tabes and paresis; its occurrence always points to the (quite frequent indeed) combination of syphilitic and metasyphilitic processes. Optic neuritis can occur unilaterally, papilloedema and degenerative atrophy are practically always bilateral, indeed, often with considerable difference in intensity between right and left. Considerably more frequently than the optic nerve, however, the oculomotor is affected and indeed, now unilaterally, now bilaterally, almost never totally, but, as a rule, with avoidance of one or several of the muscles supplied by it. Particularly regularly and particularly early, occurs paralysis of the levator palpebræ muscle, so that a large number of these patients are brought to the physician, usually to an ophthalmologist, on account of ptosis as the first symptom of their brain syphilis. Rapid change in the picture of the eye muscle paralysis (it occurs also in the trochlear and the abducens, though considerably less frequently) is very characteristic of brain syphilis. As to syphilitic affection of the internal branches of the oculomotor, paralysis of the pupils, as well as its combination with paralysis of accommodation, is found (ophthalmoplegia interna), also, finally, the reflex pupillary rigidity already described (*Argyll-Robertson* pupil, see page 183). The pupillary paralysis naturally manifests itself by mydriasis and absence of reaction of the pupil for light and convergence.

Of other luetic cranial nerve symptoms the affections of the trigeminus, facial, auditory, glosso-pharyngeal, vagus, and hypoglossus should be mentioned. The trigeminus manifests its involvement in the disease process usually in the form of neuralgic pains (I would recall to you, for example, the bilateral *Seeligmüller's* neuralgia of the auriculo-temporal nerves mentioned in Lecture II (page 47), as characteristic of syphilis), rarely by sensory defect symptoms or loss of the corneal reflex, only very exceptionally in the form of trophic disturbances, as, for example, herpes zoster, corneal ulcer, etc. On the part of the facial, paralyses of peripheral type are sometimes observed and in luetic basilar meningitis, indeed, not so very rarely, as the otherwise excessively infrequent facial diplegia. The eighth pair of nerves are very frequently affected, particularly the cochlear (tinnitus aurium, nerve deafness) here and there also the vestibular branches (vertigo, *Menière's* attacks); as to the symptomatology, see page 33 et seq. The vagus and glosso-pharyngeal roots are much more rarely affected, and then almost always both together; their involvement manifests itself in attacks of cough, difficulty in swallowing, paralysis of the vocal

cord, the (also rare) basal syphilitic lesion of the hypoglossus, in hemi-atrophy of the tongue.

In the vast majority of cases the syphilitic affections of the cranial nerves are observed not isolated, but in varied combinations. The peculiarity of rapid change in the symptom picture applies not only to the syphilitic eye muscle paralyses as emphasized above, but also to the other basal phenomena.

Brain syphilis can further lead to paralyses in the extremities, both from basal meningitis and particularly from endarteritic changes. This occurs almost always as one of the varieties of hemiplegia. We will postpone the consideration of the particularities of these important syndromes to a future lecture in which an adequate presentation of the subject of half-sided paralyses of cerebral origin will be given (Lecture XVII).

It will only be remarked here that the paralytic symptoms produced by the pressure of meningitic exudates upon the crus cerebri, the pons, and the medulla are differentiated by their less intensity, and particularly by their fugacity, from the prognostically decidedly less favorable paralyses of vascular origin. Endarteritic processes in the branches of the basal and vertebral arteries can lead to bulbar paralytic disease pictures (see Lecture VII, page 120).

As to gumma and to gummatous meningitis of the cerebral convexity, with them disease processes occur which greatly resemble those which we will study when speaking of brain tumors; indeed, many gummata clinically should come under the classification of tumor. For syphilis speaks in general the slight development of papilloedema, which indeed, in gummata of considerable size, is sometimes entirely absent; further, a striking variation in the intensity of the symptoms, a rapid alternation of improvement and retrogression. In general, much depends upon the localization of the syphilitic process. For foci which are located in the motor area of the cortex, the combination of the so-called "Jacksonian" or "cortical" attacks (see Lecture XIX) with relatively limited unilateral paralyses (monoplegia facialis, faciobrachialis, etc.) is typical; left-sided foci often lead also to aphasic disturbances (see Lecture XVIII).

Psychical disturbances point nearly always to diffuse encephalitic processes, particularly to extensive disease of the brain arteries. They are not common in gummata, still more rare in syphilitic meningitis (apart from the general clouding of the sensorium in advanced cases). Upon the basis of diffuse syphilitic endarteritis, disease pictures, which may deceptively resemble dementia paralytica yet are distinguished from it by decidedly better prognosis, often occur.

As chief differential diagnostic criteria for distinguishing them from true progressive paralysis, *Kraepelin* and *Obersteiner* give the absence of general paretic symptoms, while on the other hand focal symptoms may occur very early and are much more obstinate, the absence, or much less marked development of disturbances of speech and handwriting; further, disturbances of memory and attention keep more in the background, reduction of sensitiveness to pain is not present; particularly striking is the great frequency of hallucinations of hearing. Against a true syphilitic brain disease, but only with caution, a great length of interval between infection and outbreak of the disease can be utilized. All these criteria I consider of very questionable value and I could refer to

several cases in which by psychiatric authorities, upon the basis of the psychical syndrome, a luetic psychosis was diagnosed which later, however, showed themselves to be true paresis. Wrong diagnoses can never be entirely avoided. We are most justified in assuming syphilitic pseudo-paralysis when the somatic symptoms of brain syphilis, particularly the cranial nerve paralyses, are very marked, and a decided alternation of symptoms is present, mainly, however, when antisyphilitic treatment works promptly and strikingly.

b. *Spinal Symptoms.*—Syphilitic meningomyelitis and spinal endarteritis, the syphilitic granulomata of the spinal cord and its membranes, lead to clinical pictures in which the meningeal and the medullary components of the anatomical substratum find expression in the two already well known syndromes, of radicular pains and partial transverse lesion; in the last the paralytic symptoms appear now symmetrical, that is, in paraplegic form, again unilateral in the form of the *Brown-Séquard* symptom complex. As to the rest, however, the combination of flaccid and spastic, sensory and motor, irritative and defect phenomena, varies according to the longitudinal and transverse extent of the anatomical lesions, within quite wide boundaries, and besides this, in the course of the disease very frequently a tendency to variation in intensity, decided remissions and renewed progressive advances are noticeable.

This symptomatological lability can render a differential diagnosis from multiple sclerosis considerably more difficult. In general the intensity and the frequency of bladder troubles is much greater in syphilitic processes than in multiple sclerosis. This applies even more to rectal troubles which are excessively rare in the latter. What aid we are justified in expecting from examination of the spinal fluid in cerebro-spinal syphilis has been already thoroughly considered in Lecture XII.

That the spinal symptoms of cerebro-spinal syphilis as we have just indicated them, cannot be sharply separated from those of the "pseudo-systematic" forms of "syphilitic spinal paralysis" should be clear to you without further remark; also as compared to tabes and progressive paralysis, the boundaries are frequently bridged over by the combination of tertiary and metasyphilitic processes.

Prognosis and Treatment

The prognosis of cerebro-spinal syphilis is, presuming timely and thorough specific treatment, decidedly better than that of tabes and paresis, indeed, it can be said, better than the prognosis of the other organic brain and spinal cord affections. The more, however, the cerebral symptoms occupy the foreground, the more serious the prognosis. A syphilis located principally in the brain, sometimes defies the most energetic therapy and leads in spite of all our efforts to death or to chronic invalidism.

Often we must content ourselves with incomplete recovery, or "recovery with defect." The best outlook is given by the spinal syndrome, where sometimes cures bordering on the marvelous can be recorded. I have already mentioned the surprisingly rapid cure of a complete syphilitic paraplegia as a result of treatment by salvarsan. Energetic mercurial treatment can, though less rapid, effect just as striking results in cerebro-spinal syphilis. With

all recognition of the powerful agent which we have in salvarsan, this must be expressly emphasized, as *Oppenheim*, *Nonne*, *Sänger* and others have done. Personally, my position is that in great development of cerebral symptoms I still hesitate to recommend its use instead of that of mercury, since the aggravation of the symptoms as they have been observed in such cases, after the use of salvarsan by *Mingazzini* and others (the risk of them is small, it is true) need not be feared in the employment of the less heroic but nevertheless actively efficient mercurial treatment. Where, however, salvarsan is to be given (often in response to the direct wish of the patient, in which event it is advisable after *Oppenheim's* recommendation, to insist that he assume responsibility for any untoward results before beginning the treatment), it should not be used in insufficient doses, since the "neurorezidive" observed after arsenobenzol injections (they are usually paralyses of the cranial nerves) are probably not due to direct toxic action of the remedy but are of true syphilitic nature and their occurrence is favored by the "neurotropy" of the salvarsan, but only with the provision that the spirochætæ after the injection still have sufficient power to cause organic lesions. A good rule for salvarsan treatment is presented in the scheme of *Bruno Bloch:* *a.* Intravenous salvarsan injection (0.6 grm in adults. In delicate persons, in children, etc., correspondingly less). *b.* 5 to 7 days later the first intramuscular salvarsan injection of 0.3 grm. *c.* 14 to 20 days after the intravenous injection a second intramuscular injection of 0.3 grm. *d.* 4 to 5 weeks after the first intravenous injection, finally, a second intravenous injection.

With regard to the use of mercury many neurologists still give preference to the old inunction treatment. However true it is that this latter properly carried out and sufficiently frequently repeated at graduated intervals can accomplish most excellent results, it is certain that exactly the same thing can be accomplished with the much more convenient and cleanly intramuscular injections, on which account I use these almost exclusively. Where there is danger in delay and very rapid mercurialization is necessary I usually begin with daily injections of a soluble salt of mercury, for example, the biniodide according to the formula given on page 210; instead of the daily dose of 0.02, larger doses (0.03 to 0.04) can be given, indeed, with risk of acute mercurial intoxication. This we can arrest, however, when using the soluble salts of mercury, by stopping the injections, which we cannot do with the still more active but insoluble calomel injections (in 40 per cent. emulsion) on which account this last method is best left to the experienced syphilologist. After 10 days the injections of biniodide of mercury are intermitted for several days. A series of 20 to 30 injections constitute a "cure."

In later treatment under less pressing indications, injections of salicylate or thymoloacetate of mercury (in 10 per cent. emulsion with paraffine oil) should be preferred to the soluble salts, since on account of its more protracted absorption the injections are at longer intervals, hence more convenient and less expensive. Twelve injections of 1 cc each at intervals of from 3 to 4 days constitute a normal "cure" for a strong adult; it is well, however, to test the tolerance at first by giving smaller doses. That in all mercurial injections the most careful asepsis and proper technique is necessary goes without saying.

Place of injection: External upper quadrant of the gluteal region; with avoidance of vessels and regular examination of the urine for albumin should never be neglected, particularly when large doses are used.

The erroneous idea that cerebro-spinal syphilis, since it is "tertiary," is to be cured with iodide of potassium alone, must be decidedly opposed. Iodide of potassium, however, is to be used and in not too small doses along with mercury and eventually, salvarsan. I increase it usually to 4 to 6 grms a day; many syphilologists go considerably higher (to 15 grm). After each 20 days it is intermitted for about 10 days. If iodism begins, a solution of 3 grms each of sulphanilic acid and bicarbonate of soda in 200 cc water is given. As substitutes for iodide of potassium (over which iodide of sodium has no advantages) when there are symptoms of intolerance, different organic iodine preparations come under consideration, for example, sajodin, iodgline, iodtropon, iodstarin, lipoiodin, etc. They are all, however, less active with the exception of intramuscular injections of from 10 to 20 cc of 25 per cent. sajodin, which repeated at intervals of from 3 to 4 days sometimes has striking effect.

Internal administration of mercury is at most to be made use of for later mild "intercures"; under no circumstances can it replace injection or inunction cures. I would mention in this connection mercurous tannate 0.05 (gr. ¾) in pill form t.i.d.

In conclusion a word as to the relations between the *Wassermann* reaction and the treatment of cerebro-spinal syphilis; it occurs not infrequently that in cases where this reaction is positive, even the most energetic specific treatment cannot change it into negative. This, however, in no way clouds the prognosis, since in spite of this, recovery can be obtained and preserved. In order that such patients do not become unhappy syphilophobes, we must practice a "pious fraud" and not inform them of the continued anomaly of their blood serum, lest they believe themselves continually threatened with a recurrence and doomed to life-long use of mercury.

LECTURE XV

Arteriosclerosis of the Nerve Centers

GENTLEMEN: In to-day's lecture we will consider neither the mechanical destruction which the nervous tissue suffers in consequence of the rupture of a vessel altered by arteriosclerosis, nor the extended foci of softening which the atheromatous closure of a large artery produces.

These gross and striking results of arteriosclerosis of the central nervous system will be described at length in a later lecture.

To-day I will take up those phenomena which are explainable on the ground of diffuse disturbances of nutrition in the brain and spinal cord, consequent upon alteration of their vessels, as well as certain paroxysmal or intermittent nervous disturbances, whose comparison with other arteriosclerotic symptom complexes (for example, angina pectoris or intermittent limping) imposes itself upon us. Further, I will unfold before you, different clinical pictures which, in spite of their relative frequency, are still too little considered and which stand in relationship with special peculiar modalities of arteriosclerotic tissue destruction.

Symptomatology

Of special practical importance is acquaintance with that category of disease manifestations which we can unite under the name of arteriosclerotic pseudo-neurasthenia. In the proper estimation and management of just this condition, of which *Windscheid, Erlenmeyer* and others have given us good descriptions, we will find the central point of prophylaxis against all destructive nerve lesions of arteriosclerotic origin, for example, also against the cerebral hemorrhages and foci of softening already mentioned. It is plainly the very first beginning of vascular degeneration in the central nervous system.

The patients—usually men between 40 and 55 years old—generally come under treatment on account of headache. This is usually a feeling of pressure and heaviness, particularly about the forehead which is present on rising and increases during the day to a considerable, often to a distressing, intensity. Often the headache increases particularly on mental effort, sometimes even on meditation, which the French author *Josué* characterizes as "le signe de la penseé douloureuse" (symptom of painful thinking). There are also cases in which headache is absent during complete rest but is produced by the slightest mental or bodily effort. This headache can present a great similarity to that produced by anomalies of refraction, or by moderate astigmatism in people beyond 40 years old—when the power of accommodation decreases. It will be well in such patients not to neglect the eye condition.

Further, there is frequent complaint of vertigo; sometimes this designa-

tion is applicable (namely for sensations which the patients experience in the morning upon sitting up in bed or upon rising); often, however, under this so frequently misused expression are understood only sudden feelings of weakness or momentary darkening of the visual field, or finally, feelings of oppression or anxiety. Only rarely do we encounter true rotary vertigo to the vestibular nature of which an accompanying tinnitus can point, although the otological examination may disclose no middle ear or labyrinthine affection. A noteworthy variety of arteriosclerotic vertigo which as far as I know was first described by *Homburger*, I have heard occasionally sketched; when the patient lies in bed he suffers from the painful uncertainty of not knowing exactly how he is lying—whether upon his back or upon his side—and cannot describe the location of his bed; with this a distressing anxiety and oppression may come over him and may be accompanied with a sensation of a dull humming inside of his skull. Almost always, also, there are sleep disturbances, which, however, scarcely ever lead to a severe and obstinate agrypnia, as for example, in the prodromal stage of general paresis. Usually it is only considerable difficulty in going to sleep, restless sleep and early awakening with a sense of weight in the head. Rather rare and somewhat limited to those of more advanced age among the arteriosclerotics, is the combination of ability to sleep well by day with nocturnal sleeplessness and jactitation. This paradoxical phenomenon has been called "sleep inversion." You will often note, also, that in arteriosclerotics the intensity of the sleep disturbances varies from day to day.

Ability to work suffers usually in this manner, that the patient can follow his ordinary occupation, but contrary to what was formerly the case, finds it a great exertion and is no longer able to take up new tasks. Mental concentration is particularly difficult, which is brought to the realization of the patient on reading a paper, in keeping his accounts, in hearing a lecture or sermon, etc.

The mood is usually fretful, irritable, morose, and besides this there often makes itself felt a pessimism due to the feeling of reduced capacity which first concerns the health and economic future of the patient, but soon, however, disturbs his judgment of all relationships.

Finally, many of these patients complain of abnormal sensations, usually in the form of formication, prickling, going to sleep of the extremities.

These troubles present a great similarity to neurasthenic symptom-complexes (see below, Lecture XXVII). The differential diagnosis from neurasthenia is sometimes made easier for us by going thoroughly into the history. No neurasthenia occurs without some definite etiological preceding conditions. In the constitutional form of this disease its congenital degenerative basis is clear from the fact that its first beginnings can be traced back into early youth; in the acquired, accidental form, on the other hand, we can regularly make definite exogenic causal factors responsible, among which there is usually a combination of nervous fatigue in any direction with the chronic action of disagreeable emotions. A nervosity, however, which begins apparently without cause in a person of "arteriosclerotic age" of previously healthy nervous system, imperatively demands the close investigation of the vascular apparatus. And even in genuine inveterate neurasthenics at this critical age, arteriosclerosis

should conscientiously be sought for; it is indeed, a fact long known, that neurasthenics—mainly those with vasomotor cardiac symptoms—are predisposed to arteriosclerosis. Mutual relations between both disease conditions has, a priori, much probability. I have pointed out that cases of neurasthenia with cardiac and vascular symptoms (for example, tachycardia, dermographism, etc.) are characterized very often by abnormally high blood pressure (see Lecture XXVII). Now, however, cardio-vascular hypertension, as has been experimentally recognized, plays a rôle in favoring the occurrence of atheromatous heart lesions.

I cannot refrain from mentioning at this opportunity the recognition by the American physiologist *Cannon* that in animals psychical excitement (in the cat, for example, coming into contact with a dog) produces an adrenalinema. This fact appears to me to throw a new light upon the pathogenetic rôle which in the etiology of arteriosclerosis may be attributed to the continuous increased emotivity common to all neurasthenics. Now adrenaline acts injuriously upon the vessel walls, both chemically and by raising the blood pressure. Finally, a still more direct connection between the nervous system and vascular degeneration has been experimentally rendered probable; for example, *Manouelian* has shown, that through the destruction of definite nerve twigs entering the vessel walls, sclerotic plaques may be produced in a corresponding situation.

To return to the clinical side, if there is a suspicion of arteriosclerotic nervosity in a patient, we must naturally confirm the diagnosis by recognizing the vascular affection. Here I would warn you above everything against depending upon the palpation of the peripheral arteries as is so often done. The clinic and the autopsy table in agreement, teach us that between the condition of the peripheral arteries and that of the brain and spinal cord vessels, a paradoxical incongruence often exists. I could mention many cases of cerebral hemorrhage and encephalomalacia with severe sclerotic alterations of the brain arteries, in which in the radial, the dorsalis pedis, the carotid, the brachial, and even the temporal, there was no perceptible tortuosity or hardening of the vessels. Nevertheless, I would add that (when all arteries accessible to palpation are examined) negative findings in this respect are not so very frequent.

As to the rest, in arteriosclerotic pseudo-neurasthenia we can pretty well count on registering further stigmata of vascular disease in these patients, sometimes indeed, in considerable number. So, for example, alteration of the size of the heart, especially hypertrophy of the left ventricle, accentuation of the second, impurity of the first aortic sound.

Slight albuminuria is usually to be found if the urine is repeatedly examined; and also in urine free from albumin, hyaline and granular casts are occasionally observed. Of importance, also, is the recognition of slight œdema over the ankles in the evening, provided that it is not explainable through heart weakness, nephritis, or varicose veins. Sometimes there is a tendency to nose bleed. As to stenocardiac attacks proper, or cardiac asthma, it is, as a rule, not necessary to inquire, as these conditions are usually spontaneously mentioned by the patient; still they occur sometimes in slight or rudimentary form,

scarcely observed by him; this applies, namely, to the "respiratory fatigue" of the French, which occurs on moderate bodily exertion. It is well, also, to measure the blood pressure, although its increase which according to *Broedel* occurs in about 65 per cent. of cases of arteriosclerosis, according to *Romberg* and others, is no symptom of the vascular disease itself, but of the interstitial nephritis which so often accompanies this last.

Of late it has been endeavored by *Kurt Mendel*, in disease pictures which occur without any external cause, in previously healthy men between 45 and 50 years of age and which closely correspond to the description which I have given you of arteriosclerotic pseudo-neurasthenia, in which, however (if we neglect congestions of the head and paroxysmal palpitation), cardiac vascular anomalies are absent, to refer these cases as "molimina climacteria virilia" to alterations of the inner secretion of the genital glands and to separate them from incipient arteriosclerosis of the nerve centers.

We will turn now to a group of disturbances of considerable intensity but of fleeting character which we sometimes see appear upon the basis of arteriosclerotic pseudo-neurasthenia and which demand our close attention. They inform us, namely, of an already high degree of intensity of the vascular disease and represent only too often (as the further course of such cases teaches us) the forerunners of irreparable organic lesions of the nervous system.

I would mention first, severe pains which occur in attacks, are characterized by great intensity, and are located both on the trunk, in the limbs and in the region of the trigeminus. They are mainly considered as neuralgias, although I believe, incorrectly very often. The absence of pressure points along the nerves in question, the boring, lancinating, constricting character of these attacks, recalling tabes dorsalis, makes it appear probable to me that they are due to vascular spasms in the central nervous organs or in their posterior roots. *Nothnagel* and *Huchard* long ago emphasized the rôle in the clinical picture of arteriosclerosis, which vascular spasm occurring in attacks, played alongside of the permanent vascular lesions, and the similarity of these painful crises of arteriosclerotics to angina pectoris and to arteriosclerotic colic, decidedly impresses itself. Still further analogies, however, exist between certain intermittent disturbances in the motor functions, which are occasionally found in cerebro-spinal arteriosclerosis and the interesting phenomenon which is known as "intermittent limping."

This complaint ("Claudicatio intermittens," "Dysbasia angiosclerotic intermittens") often presents a prodromal stage of the so-called spontaneous gangrene of the lower extremities. It is caused by arteriosclerosis of the leg arteries, though functional causes also play a rôle. While namely, on walking, the movements of such patients are first normal, painless and unhindered, after a certain time—this can be in one case, a few minutes, in another a half or even three-quarters of an hour—there occurs an extreme weakness of the lower extremities with cramplike sensation, which soon makes further progression impossible for the patient. At the same time it can be remarked how single toes, single portions of the skin of the feet, alter their color under the eyes of the observer, now deathly pale, now livid, again cyanotic —a plain evidence that it is not exclusively the expression of blood supply

insufficient for the locomotor requirements, but that a vasomotor spasm is also in action—after a short rest, however, all the trouble has disappeared again and the patient can move in a normal manner; indeed, only after about the same time as formerly, to be again attacked by the trouble and to be compelled to rest. And so the same thing is repeated upon all attempts at walking; with the total duration of the locomotor effort, the time of unhindered walking usually becoming continually shorter. Dysbasia angiosclerotica intermittens, occurs very rarely with us; in Poland and Russia, however, it is strikingly frequent, which may have a connection with the very cold climate. There is in that region an apparent predisposition among the Jewish race which is not the case in our neighborhood (of 5 cases which I have seen, not one was a Jew). Excessive smoking certainly plays an important etiological rôle. The treatment—apart from the general treatment of arteriosclerosis—falls in with that of the vasomotor neurotic forms of intermittent limping (see Lecture XXV).

There occurs now in arteriosclerosis of the central nervous system a disturbance which appears so like the classical picture of intermittent claudication introduced into the nosology by *Charcot* and *Erb*, that *Dejerine* denominates it as the spinal variety of this disease, an "intermittent claudication of the spinal cord." I have seen two exceedingly instructive cases of this. In distinction from the peripheral form, all vasomotor disturbance in the lower extremities is absent and both arteries on the feet (dorsalis pedis and tibialis posticus) are to be felt normal. With the beginning of the limping, there occur the following symptoms characteristic of it: an increase in the patellar and Achilles reflexes, often a decided ankle clonus, and sometimes also, the *Babinsky* reflex, as clinical corollaries of an ischemic condition of the pyramidal tracts. Somewhat more frequently than in arteriosclerosis of the spinal cord —it can be said in passing—spinal intermittent limping has been observed upon a basis of syphilitic endarteritis.

Along with the just described paretic paroxysms of spinal origin we can observe those of plainly cerebral origin, although much less frequently, in which, however, the coöperation of muscular effort as an exciting cause is less plain, or indeed, cannot be recognized. I have in mind particularly the temporary paresis of an arm or leg or even of both extremities on one side of the body which usually greatly frighten the patient, but after a few hours, or even minutes, entirely disappear. Still more interesting is the temporary motor aphasia, of which I might mention three observations: in one, the disturbance lasted only a few minutes, in the others, from 15 to 20 minutes. One of these last cases seemed to illustrate the influence of functional use, in that the woman affected afterward explained that on the day on which it occurred she had felt very much fatigued from long talking to visitors.

All these transitory phenomena, however striking their method of occurrence may be, always stand on the border between the functional and the organic. They can be considered as the original expression of *Grasset's* has it, "avertissements sans frais" (free warnings). The disturbances, however, to which we will now turn, depend upon material alterations which, although not very extensive, are nevertheless of far-reaching importance.

I cannot go any farther into arteriosclerotic diseases of the spinal cord (as a forerunner of which we have become acquainted with spinal intermittent claudication) but will only state that a degeneration in the region of the lateral columns due to arterial ischemia in consequence of atheromatous obliteration of vessels can produce the symptom-complex of spastic spinal paralysis. These cases which have been isolated by *Demange*, *Oppenheim* and others as senile spastic paraplegia, affect usually only the lower extremities, very rarely the arms also. A more extended affection of the vascular apparatus of the spinal cord can also produce sensory disturbances and sphincter paralysis.

These findings are much less frequent than the extensive symptoms which small, unapparent losses of substance due to arteriosclerosis in the brain can produce, thanks to the multiplicity which, as a rule, characterizes their occurrence. These are the so-called "Lacunæ" ("lacunes de désintégration cerebrale," "état lacunaire du cerveau") first thoroughly studied by *Pierre Marie* in Paris. From miliary, to the size of a pea, of irregular outline, these losses of substance pervade in greater or less number circumscribed areas of the cerebrum, the interbrain, and the midbrain. In the middle of each one of these small spaces located close together, we notice upon microscopical examination an arteriosclerotic but still pervious vessel; they cannot hence be considered as small areas of softening due to the closure of vessels. Rather have the newest histological investigations shown that inflammatory processes of the adventitia and of the peri-arterial lymph spaces are responsible for the destruction of the surrounding brain tissue.

The status lacunaris of the brain is properly to be distinguished from the so-called "status cribrosus" of *Durand-Fardel*, which results from dilatations of the peri-vascular lymph sheaths, while it is pathogenically more nearly related to a lesion of the cerebral cortex described by *Marie*, *Alzheimer* and others, which gives to this last a worm-eaten appearance ("état vermoulu") since it is due to the destruction of cortical substance about diseased arteries. On the other hand, the "cerebral porosis" of certain pathological anatomists (also called "cystic degeneration" or "état de fromage de Gruyère") is nothing else but a post-mortem artefact produced by the bacillus ærogenes capsulatus, by the aid of which these changes can be experimentally produced also.

A special variety of the hemiplegia of old age depends, too, upon arteriosclerotic formation of lacunæ. This lacunar hemiplegia (to anticipate our later remarks, Lecture XVII) can usually be distinguished without difficulty from the hemorrhagic, thrombotic and embolic hemiplegias by a number of clinical criteria. These criteria are particularly in the character and course of the attack, which almost never presents the picture of a true apoplectic seizure, namely, if it does not occur at night (which it often does) it usually runs its course with retained consciousness and at most, some vertigo and a temporary confusion are its accompaniments. In the rare cases, however, in which the hemiplegic paralysis begins violently (this is the case when the lacunæ involve the internal capsule) the loss of consciousness is of very short duration—lasts less than an hour. The resulting hemiplegia is, further, usually of little intensity and markedly transitory character. Although it also leads to exaggeration of reflexes, it never causes contractures, but more or less

completely disappears after hours, days or weeks. (A longer duration of the hemiplegic symptoms again points to the seat in the capsule of the tissue defects.) The lower extremities are usually longest affected, since a certain regularity in the distribution of the paralytic symptoms is not to be mistaken. Among these, most important is the absence of aphasic symptoms, while there may be some disarthria; further, we miss regularly hemianopsia, conjugate

Fig. 69.

Status Lacunaris Cerebri. Typical attitude and "démarche à petit pas."

deviation, and appreciable disturbances of sensibility, while quite frequently the facial nerve is unaffected. Ankle clonus and the *Babinsky* and *Oppenheim* phenomena can often be observed; on the other hand the abdominal reflexes are scarcely ever disturbed. Now, however, after the hemiplegic symptoms have disappeared with remarkable rapidity and completeness, a change which gives to such patients a quite characteristic attitude progressively sets in. While, in spite of persistent anomalies of reflexes, the paretic symptoms at

most manifest themselves in the fine movements, for example, in buttoning the clothes, the gait takes on an exceedingly typical character (Fig. 69). In the yards and gardens of asylums for old people and almshouses, such patients can be seen going about, their bodies bent forward, the knees in half-flexion, not spastic, indeed, but with inflexible joints, and slowly dragging one foot after the other, the soles scraping the ground. A certain similarity between this "démarche à petit pas" (gait of little steps) as *Brissaud* has called it, and the gait of patients with paralysis agitans (see Lecture V) is not to be denied, though the differential diagnosis presents no difficulty to those familiar with both conditions.

We leave now the lacunar hemiplegias to turn to a disease picture which in a large percentage of cases also depends upon arteriosclerotic tissue defects, sometimes, indeed, from disseminated arteriosclerotic foci of another character, for example, miliary hemorrhages or fibro-hyaline obliteration of vessels. This is pseudo-bulbar paralysis, which depends upon the fact that small but multiple lesions in both hemispheres interrupt the course of the nerve fibers between the cortex and the bulbar nuclei of the muscles of mastication, deglutition and speech. Unilateral location of the foci alone is so rare that we need not consider here such cases described as curiosities only. This is explainable from the fact that the lower facial, the hypo-glossal and the motor portion of the trigeminus are connected with both hemispheres, and that as a rule one of these is sufficient to maintain the functions of the muscles supplied by these nerves. Bilateral multiple losses of substance are seldom located in the cortex, more frequently in the centrum ovale, the central ganglia (particularly in the putamen of the lenticular nucleus), in the internal capsule and in the crura cerebri.

In typical cases of this pseudo-bulbar paralysis—we prefer this expression to the more cumbrous "cerebral glosso-pharyngo-labial paralysis"—the patients have formerly suffered from the previously described lacunar hemiplegia. Now comes a repetition of the same attack, this time, however, on the opposite side. This shows us that the arteriosclerosis has led to the gradual production of small tissue defects in the second hemisphere. And now we notice that this second half-sided paresis is accompanied by definite disturbances of articulation and deglutition, which, however (in contradistinction to the first time), persist further, while the weakness of the extremities gradually passes off this time also. Besides this typical beginning, as rarer methods of occurrence of arteriosclerotic pseudo-bulbar paralysis are observed, its sudden and definite origin after a single attack; slow development in steps broken by remissions; finally, a transition form with abrupt commencement, then total disappearance and progressive reappearance of the symptoms anew.

Let us look more closely into this last. The speech becomes drawling, monotonous, there is sometimes aphonia. While the formation of vowels suffers less, the consonants are pronounced badly and with trouble, so that in severe cases the speech becomes unintelligible. In speaking the breath often fails, and to complete the sentence the patient must begin over again several times; the enunciation takes on a chopped-off or semi-explosive character. According to whether paresis of the lips or that of the palate dominates, disturbances in the

formation of labials or nasal character predominate. The mobility of the tongue can be so greatly affected that, following the laws of gravity, it lies immovable upon the floor of the mouth. Usually, however, it can be protruded, at least to a certain extent, but there are disturbances in lateral movements or in forming a trough, or frequent repetition of its protrusion leads to rapid paralysis. Also the movements of the palate are either suspended or only slow and incomplete; in the larynx, however, paresis of the vocal cords but rarely reaches a high degree. The muscles of mastication are usually again greatly affected, a close approximation of the teeth is impossible, the mouth often remains permanently half open. Eventual paralysis of the pterygoids manifests itself through the impossibility of pushing the lower jaw forward or laterally. Sometimes, also, the muscles opening the mouth are weakened and complete opening, or opening it against resistance is impossible. The taking of nutrition sometimes develops exceedingly marked disturbances. Here along with weakness of the masticatory muscles, that of the muscles of the tongue, lips and cheeks plays a rôle; food cannot be pushed between the teeth, falls out of the mouth, and must often be forced into the pharynx by the aid of a finger. If paralysis of the palate and of the pharynx are added, portions of food get into the nose or the larynx. Still, in mild cases, the act of eating, namely, when the patient eats slowly and selects solid and semi-liquid food, is relatively well accomplished.

What constitutes the chief difference between the disturbances just related and those which we find in true bulbar paralysis, is the fact that the paralyzed muscles in pseudo-bulbar paralysis show no degenerative atrophy and no reaction of degeneration. Also, *mutatis mutandis*, the same difference which spastic spinal paralysis presents in contradistinction to spinal progressive muscular atrophy. True conditions of contracture can, indeed, little by little develop in the lips, the tongue and the palate, in patients with pseudo-bulbar paralysis. Increase of the masseter reflex is scarcely ever absent and is understandable. Paradoxical, on the other hand, is the very frequent disappearance of the palate and pharyngeal reflexes even when the voluntary movements of these parts are still intact.

The expression of countenance has something very characteristic. The lower part of the face acquires little by little a mask-like stiffness through the combination of paralysis and contracture. Saliva flows from the half open mouth. The physiognomy takes on a lacrimose expression. With this contrasts, indeed, the better mobility of the forehead, and the movable, often expressive eyes.

Much rarer than pseudo-bulbar paralysis, is another disease condition produced by bilateral disseminated tissue defects, the senile incontinence, type *Homburger*. One case observed by me corresponds anatomically exactly with the statements of this author, that is, there was a symmetrical status lacunaris, in the optic thalamus and in the corpus striatum. Clinically, such cases are characterized by the automatic character of the discharge of urine. At more or less regular intervals approximately the same quantity of urine, with constant retention of a residual portion, is discharged, and so suddenly that the patient wets himself. The ability to urinate spontaneously is preserved at the start but is lost later.

After a longer or shorter time all the somatic consequences of multiple arteriosclerotic focus formation in the brain are usually combined with a psychical decadence, becoming ever plainer. We enter with this, the subject of the arteriosclerotic dementias. I intentionally use the plural, since quite different symptom-complexes occur. In patients who have suffered from lacunar hemiplegias, in pseudo-bulbar paralysis and in the incontinence just related, we are struck particularly by the triad: weakness of memory, morbid emotivity, and loss of interest. The whole mental habitus becomes something silly. The patient, for example, pours out hot tears for a dog that was run over, about which he read in the newspaper, while the fortune or misfortune of his family does not impress him. Also the hypochondriacal complaints which are brought forward in such numbers by these old people, give the impression of mental weakness. They do not, as a rule, relate to serious troubles, for example, to the loss of memory, power of attention, etc., but to trivialities, as to the hardness of their stools, etc. Upon this quite characteristic basis but in a rather manifold combination, a whole line of further psychical anomalies develop themselves, for example, ideas of unseen influence, suicidal impulses, even hallucinations, so as observed, very heterogenous individual pictures occur. Many of these patients must be placed under psychiatric care; I will not enter upon the further disease pictures well known to asylum physicians. Only one special arteriosclerotic psychosis would I mention here, since to know and to recognize its early stages is of importance for the non-psychiatrist also. I mean "arteriosclerotic pseudo-paralysis." (Again a "pseudo"—you see, arteriosclerosis is a great simulator among diseases.) As a fact the similarity of this syndrome to metasyphilitic dementia paralytica is quite marked (see Lecture XIII, page 205). One of the newest descriptions of arteriosclerotic pseudoparesis, whose recognition we owe to *Klippel*, *Alzheimer* and *Binswanger*, is by *Charles Ladame*. According to this author, the clinical symptoms of these patients (they are usually men between 50 and 60 years old) are in the somatic sphere, premature senility, bodily deterioration, sluggishness in the pupillary reaction for light, convergence and accommodation, exaggeration of the tendon reflexes, hardening of the arteries, increased blood pressure, sometimes also, arteriosclerosis of the kidneys. In the psychical sphere isolated defects of intelligence, good preservation of orientation in general and of consciousness of self and of illness, reduction of the power of fixing the attention, partial defects of memory; sometimes very good judgment forming capacity in certain directions, demeanor externally well regulated (they present a "deceptive front"). Delusional ideas (mainly of a hypochondriacal nature), illusions and hallucinations, occur. The mood shows a mixture of anxiety, indifference, euphoria, great egotism, ideas of suicide. There is marked impulsive automatism. The differential diagnosis from progressive paralysis must usually be based upon the condition of the pupils, the defective mental condition with retained personality, appreciation of disease, the age of the person and his general habitus. The prognosis is in the great majority of cases bad. The anatomical substratum of this psychosis is not a simple one. It can be made up of the most varied results of the cerebral vascular disease, including *Marie's* lacunæ, punctate or larger hemorrhages of the brain, cortical or subcortical

foci of softening, or of the "subcortical chronic, progressive encephalitis" described by *Binswanger* a diffuse degeneration of the medullary layer as a consequence of insufficient blood supply.

Treatment

In conclusion a few therapeutic indications. Among the different clinical pictures which we have passed in review, of course only those which are usually the expression either of imperfect nutrition or of temporary vascular spasm, present a favorable outlook as to the possibility of being influenced by treatment. In the other forms, the treatment can only seek the prophylactic end of preventing further progress of the lesion if possible, but this end is important enough. Let me emphasize that in spite of their organic basis many of the troubles of the arteriosclerotic pseudo-neurasthenics are to be favorably influenced by psycho-therapeutic reassurance, since along with the material basis the depressing feeling of threatened "breakdown," and anxiety about previously unexperienced disagreeable sensations exercise a morbid influence. Under all conditions, further, the greatest importance must be laid upon the dietetic régime; absolute milk diet is to be considered only for a short time and in the rare cases of particularly obstinate sleeplessness or headache, though it has here sometimes a striking effect. In general a bland, predominantly ovo-lacto-vegetarian diet suffices. We must endeavor to so arrange it that the patient will take it for a long time without repugnance, and hence must proceed not too rigidly. Meat, for example, may be permitted at the mid-day meal, but only one dish, and the white meats are to be preferred. Fresh water fish may be permitted, sea fish forbidden. Bouillon should be replaced by cereal and vegetable soup; plenty of vegetables, easily digested farinaceous foods and potatoes, fruit raw or cooked, are permitted. Alcoholic beverages, strong coffee, and particularly the use of tobacco should be restricted as much as possible, and all the stronger condiments avoided. The patient should take plenty of milk, eventually buttermilk, yoghurt, kefir. In general, as table beverages, alkaline mineral waters, as those of Vichy, Passugg, Neuenahr, Fachingen, etc., are to be recommended, particularly during iodide cures.

Now, as far as these cures go you know that of late the indications for iodide of potassium in arteriosclerosis have been much narrowed by internists. We neurologists, however, consider this agent now, as formerly, so valuable a remedy that it should not remain untried in any form of nervous manifestations in arteriosclerosis. Upon what the action of iodine depends is not yet definitely concluded; it appears (from the investigations of *Müller*, *Inada* and others) determined that by reducing the viscosity of the blood it facilitates the flushing of the nervous centers; also a certain vasodilating action is attributable to it. According to *Pouchet* it has also a decidedly stimulating effect upon the lympho- and leuko-cytes and aids phagocytic processes, by which a modern explanation is furnished for its ancient reputation as a "summum resorbens." Very questionable, on the other hand, is the specific curative effect upon parts of the vessel walls already affected with arteriosclerosis affirmed for it, while again,

a prophylactic action in this direction is plausible from the fact that administration of iodides often lowers the blood pressure.

Since iodide of potassium or iodide of sodium must be prescribed always for a long time in arteriosclerotic nervous diseases, it is recommended that for the prevention of iodism daily doses of from 0.5 to 1.0 should not be exceeded, and that after each 20 days of use there should be a 10 day pause. The addition of sodium bicarbonate usually causes the drug to be better tolerated by patients with sensitive stomachs. As a specially well tolerated combination I would mention that of potassium iodide and strychnine: besides the alkaline iodides, in the better class of practice a number of organic preparations of iodine, such as iodipin, sajodin, iodon, etc., may be used; also these preparations are usually well tolerated and iodism is almost excluded, but in activity they do not compare with the alkaline iodides.

While we are speaking of drugs we may consider the question of hypnotics, to use which we are often compelled by arteriosclerotic insomnia. They appear to me unobjectionable provided that we avoid such remedies as act unfavorably upon the vessel walls (chloral hydrate, amylenhydrate, paraldehyde, for example), and on the other hand follow the end of the reëstablishing the ability to sleep spontaneously. The remedies of choice are the difficultly soluble hypnotics of the type of trional and veronal; we must resort to them when we cannot accomplish our ends with dietetic and hydriatic measures, or with nightly doses of bromides from 1.0 to 2.0 bromide of potassium (we refrain from giving larger doses of bromides since the arteriosclerotic seems to have a predisposition for bromism). 1.0 (grs. xv) of trional or 0.5 of veronal (gr. viii) dissolved in some hot liquid is given one hour before bedtime; this medication can be given every evening if it is desired that the patient should get the full hypnotic effect of the normal dose, and also when necessary, should sleep on into the next day. Under these conditions, after a few days a certain sleepiness often appears and this gives us the signal for the reduction of the dose—of veronal, for example, to 0.3 (gr. 5). Later it is attempted from time to time to intermit a night, then to give the hypnotic every second, then every third night, and finally to withdraw it altogether. For senile "sleep inversion," *Homburger* has proposed a method of "reinversion" of the type of sleep which I can thoroughly recommend; forcibly keeping the patient awake by day is carefully avoided; on the other hand, he is kept in bed, and we limit ourselves at first to procuring some sleep at night with hypnotics. If now the duration of the sleep by day and its depth diminishes, the evening dose of hypnotic can be increased and the patient kept up longer and longer by day.

Of hydrotherapeutic measures, hot salt and mustard foot baths which should be used daily when there is a tendency to congestions about the head, vertigo, headache, etc., may be mentioned in the first place. Further tepid baths of not too long duration and tepid rain douches with rubbing down afterward, work favorably. Carbonic acid baths should be used only with great caution, as they are poorly borne by many patients. Bath cures proper, unless they are indicated on account of some other result of arteriosclerosis are not called for. The patient is best sent to some quiet resort of medium altitude (500 to 800 meters—1,500 to 2,500 feet) which affords plenty of opportunity for being out-

doors, is protected from the wind, and has but moderate range of temperature. Sometimes general massage of the body, after the Swedish method, acts quite strikingly both upon the circulatory conditions and upon the general health.

The symptomatic treatment of the spastic phenomena, paresis of the muscles innervated from the bulb, incontinence, etc., is the same as that already described in the preceding lectures for these symptoms from other causes.

LECTURE XVI

The Acute Infectious Diseases of the Central Nervous System

A. "Essential Infantile Paralysis" (The Heine-Medin Disease). (Anterior Polyomyelitis)

Under essential infantile paralysis we include disease conditions which attack the childish organism in stormy fashion, to localize themselves shortly in definite parts of the central nervous system (in the vast majority of cases in the anterior horns of the spinal cord) and there to lead to characteristic inflammatory foci with subsequent degenerative processes. The clinical corollary of these lesions consists, as a rule, in flaccid paralyses which show no tendency to progression, but, on the contrary, usually undergo decided regression, soon after the onset of the disease. The disease often occurs in epidemic form; it has been produced experimentally in animals. While the first accurate description of the disease originated with the Stuttgart physician *Jakob v. Heine* (1840), the Stockholm pediatrist *O. Medin*, on the basis of careful studies on the Swedish epidemics of 1887 and 1895, first showed with all definiteness the infectious nature of the disease.

The classical, until a short time ago, almost exclusively considered, form of the *Heine-Medin* disease is infantile spinal paralysis (poliomyelitis anterior acuta infantum). This includes over 90 per cent. of all the sporadic and epidemic cases of this nosological group. We are hence certainly justified in our clinical discussion, in keeping in mind at first this typical manner of occurrence. Only afterward will the varieties of the disease due to variation in the location of the anatomical substratum, be considered.

Course and Symptomatology

While in general the separation from each other of different stages of disease in affections of the nervous system is rather artificial, and the boundaries between these stages can scarcely be drawn sharply, in acute anterior poliomyelitis we can without doing violence to the facts hold fast to the usual differentiation into four periods, the infectious stage, the paralytic stage, the stage of regression, and, finally, the stage of trophic disturbances and contractures.

1. INFECTIOUS STAGE

The age at which poliomyelitis acuta anterior occurs by preference is the second year of life. It finds, however, relatively many victims also in the third

and fourth years. The morbidity then rapidly decreases so that cases beginning between the 8th year and puberty are comparative rarities. The first year of life also furnishes but a very small contingent.

The first signs of the disease have so little that is characteristic that in the first stage the diagnosis can only be made definitely when the case occurs while an epidemic of the *Heine-Medin* disease prevails. The children fall ill with rapidly rising fever, which, however, only exceptionally exceeds 40° C. (104° F.), become somnolent (but scarcely ever comatose), often show vomiting and diarrhœa, not rarely coryza or angina. Sometimes the children complain of headache and pain in the back or limbs, frequently there is so much hyperesthesia of the integument that they scream when taken hold of. A sensitiveness to pressure and to tapping over the spinous processes, with stiffness of the spine is frequent (perhaps to be considered as a local symptom). Further, frequent disease manifestations of the infectious stage are convulsions, twitching, grinding the teeth, rolling the eyes, profuse sweating.

Ed. Müller attributes great importance to the absence of leucocytosis in the blood picture, in the diagnosis of the febrile stage of acute poliomyelitis, indeed, he has found usually decided leucopenia, a reduction of the number of leucocytes to from 3,000 to 5,000. For him, leucopenia, hyperidrosis and hyperesthesia represent the three cardinal symptoms of the early stage of essential infantile paralysis, a view whose justification is, however, not yet furnished. On examination of the spinal fluid, however, nothing definitely characteristic has been found; sometimes there is moderate lymphocytosis, sometimes, on the contrary, leucocytosis with almost complete absence of lymphocytes. The duration of the infectious or febrile stage is, as a rule, from 24 to 48 hours.

2. THE STAGE OF PARALYSIS

In typical cases, the stage of paralysis in acute poliomyelitis, abruptly terminates its febrile early period. The temperature falls, paralytic symptoms immediately appear, and within a few hours reach their maximum in intensity as well as in extent. This last, however, varies within wide limits. Most frequently the paralysis assumes a paraplegic type, that is, affects both lower extremities; there follow according to frequency, paralysis of a single leg (monoplegia cruralis), that of one arm (monoplegia brachialis), that of both arms (diplegia superior); further, tetraplegia, triplegia, and finally, hemiplegia, which latter is denominated "alternating" when it affects the right arm and left leg, or vice versa. The trunk and neck muscles are, as a rule, unaffected; still, upon careful examination of the abdominal muscles a unilateral or bilateral paralysis of them is not rarely observed, in the first case the umbilicus is drawn toward the healthy side.

Cases in which the cranial nerves are also paralyzed (eye muscles, facial, hypoglossal) naturally extend beyond the bounds of anterior poliomyelitis into those of polioencephalitis; a more extensive affection of the motor nuclei of the brain axis can, indeed, lead to rapid exitus from respiratory or cardiac paralysis.

This paralysis is now a thoroughly flaccid one, and can through the high degree of this hypotonia, change the extremities into unresisting, dangling

appendages, into "jumping jack limbs" ("membres de polichinelle"). The reflexes in the affected muscles are of course lost. Typical, further, is the escape of the bladder and the rectum; where any incontinence or retention of urine and fæces is noted, it is, as a rule, an inconsiderable and temporary phenomenon. Objective testing of sensibility discloses no anomaly; on the other hand, older children sometimes complain of severe pains in the affected limbs. These pains, which we consider as due to irritation by the inflammatory focus of the pain fibers which cross in the gray commissure (see Lecture X, page 145), have as an evidence of the intensity of this inflammatory process, a prognostically unfavorable indication. The paralytic symptoms usually remain for some days in unaltered intensity and extent; then, however, the spontaneous occurrence of marked improvement announces the beginning of the third typical phase of the disease.

3. THE STAGE OF REGRESSION

It is noticed that in single muscles of the paralyzed extremities the power of voluntary movement appears again, so that the extent of the paralysis, as compared to the picture presented in the preceding stage, slowly diminishes, or, as it is usually expressed, the paralysis "concentrates itself." Even before this regression or concentration has reached its maximum, however, the study of the electrical reactions permits us to predict which muscle groups will most rapidly regain their function, and in which others the paralysis will continue longest, or will declare itself permanent. The seriously or incurably affected muscles namely, show early (from the 10th to the 14th day after the beginning of the disease) reaction of degeneration, and in general, one can hold fast to the rule given by *Duchenne*, that muscles which 3 weeks after the onset of the acute spinal paralysis no longer react to faradism must be considered as lost. Exceptions to this rule I have indeed occasionally had the opportunity of observing, and otherwise also, there are sometimes findings which do not correspond to the general principles of electro-diagnosis, in that, for example, muscles permanently paralyzed show only a qualitative reduction of galvanic and faradic irritability which continually progresses to entire loss of reaction. The regression which probably occurs from disappearance of the inflammatory œdema and of the infiltrative processes, can proceed to complete recovery; still, such cases, which are somewhat more frequently observed in epidemics of the *Heine-Medin* disease than among sporadic cases of essential infantile paralysis, form only a small minority. In general, namely, acute anterior poliomyelitis after greater or less "concentration" leaves behind as corollary of the total destruction of definite centers of the anterior horns, the final elimination of certain muscles. There is, indeed, a certain regularity in the choice of localization of the residuary muscular paralysis. In the upper extremities, most frequently permanently affected are the deltoid, the infraspinatus, the serratus, the trapezius, the teres minor. In the lower extremities, the quadriceps, particularly, however, the extensors of the foot and toes. We will consider the deformities to which these paralyses lead, on describing the next stage. Corresponding to its anatomical substratum (the destruction of the tropho-motor

centers of the anterior horn) the disease leads to rapid progressive degenerative atrophy of the affected muscles, whose contractile substance finally is entirely replaced by fat and connective tissue. Upon superficial observation of the little patient during the stage of regression a decided subcutaneous development of fat in the paralyzed region, predominating to a certain extent over the muscular wasting, may lead to deception. Upon an average the "concentra-

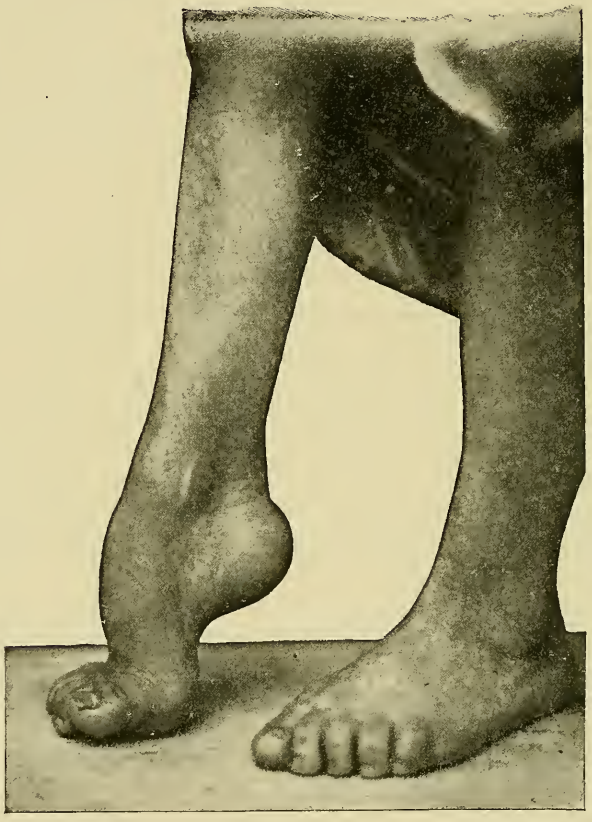

Fig. 70.

Pes Equinovarus as a Result of Anterior Poliomyelitis.

tion" of the paralysis is completed after 8 or 10 months, though a much longer extent of the stage of regression (up to a year and a half, or more) is not at all rare.

4. STAGE OF TROPHIC DISTURBANCES AND CONTRACTURES

The stage of regression passes over now into the stationary and final period of poliomyelitis, in which the bodily deformities resulting from the combination of paralysis, contracture and atrophy dominate the clinical picture. The degenerative muscular atrophy reaches its highest degree which no subcutaneous development of fat can mask any longer, so that the limbs affected appear

markedly thinned, in part, of skeleton-like emaciation. At the same time, severe disturbances of nutrition affect the bones of these extremities; these, in the further growth of the child, remain far behind those of the healthy limbs in development, and as the X-ray study of such cases shows, there may be regressive and degenerative processes of their cortical layers, which in time may become so thin that very slight causes lead to the so-called spontaneous fractures. Also in the ligaments there are atrophic processes which may lead to an excessive relaxation of the joint (flail joint), so, for example, in the shoulder-joint, the head of the humerus is seen to fall out of its socket, the natural rounding of the shoulder is interrupted by a deep furrow, and the palpating finger can easily be pressed in between the scapula and the joint surface of the humerus. The separation of the joint surfaces from one another leads in time to deformities which, however, never reach such a degree as in syringomyelia and tabes dorsalis. Of great practical importance are, further, the shortening contractures which occur in the unparalyzed (or only partially paralyzed) antagonists of the totally paralyzed muscles, as expression of the mechanical and trophic predominance which the unaffected or relatively unaffected muscles attain in time. Through this there arise characteristic anomalies of position, so, for example, pes equinovarus which, by far the most frequent deformity, occurs on account of contracture of the tibialis and the triceps suræ, the peronei, and extensor of the toes and great toe being eliminated. In degeneration of the calf muscles, on the other hand, there is pes calcaneus. Preponderance of the muscles of the sole over the more or less weakened anterior and posterior muscles of the leg, leads to "paralytic pes cavus," atrophy of the peroneal muscles, the anterior tibial being preserved, to pes valgus. In the hand, infantile paralysis but rarely leads to characteristic anomalies of position, if at all to "claw hand" described in Lecture VI, page 108. Rather frequent is scoliosis, which is to be considered sometimes as a primary, sometimes as a secondary result of poliomyelitis. The first is usual; the lateral curvature of the spinal column arises from the necessity of compensating the shortening of one leg, by altering the position of the center of gravity; the latter can result from the comparatively rare atrophic paralysis of the muscles of the back. With the scoliosis there are often deformities of the pelvis, which in female patients may in later years lead to serious difficulties in parturition.

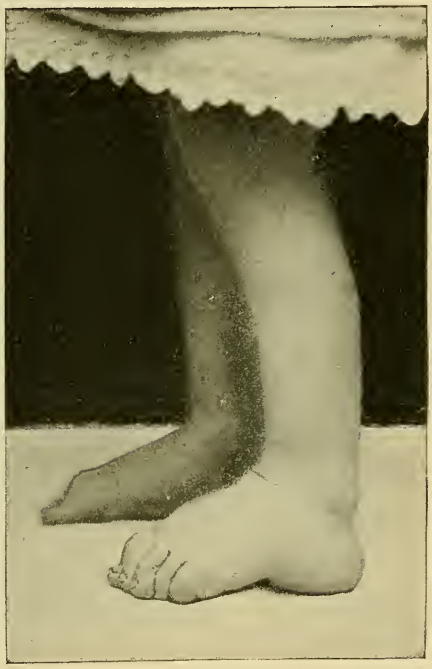

Fig. 71.

Paralytic Pes Cavus as a Result of Anterior Poliomyelitis.

In the skin of the paralyzed extremities also, there are atrophic and vasomotor anomalies. It is cool, usually livid in color or marbled, thin and smooth; often its hair is abnormal. The repeatedly made observation that the exanthem of measles and scarlet fever in children who have had poliomyelitis avoids the paralyzed limbs, is interesting. The integument of such individuals shows a

Fig. 72.

Infantile Spinal Paralysis. Late Stage; Extreme Atrophy of the Lower Portion of the Trunk, the Pelvic Region and the Legs; Marked Pes Varus, Right Side.

great tendency to form bed sores, which furnishes a warning as to very careful application of orthopedic apparatus, if it is needed. There is also a marked tendency to chilblains.

ANOMALIES IN THE COURSE OF ACUTE ANTERIOR POLIOMYELITIS AND OTHER VARIETIES OF THE HEINE-MEDIN DISEASE

Along with the just described "classical" picture of acute atrophic spinal paralysis in childhood, there are observed not infrequently anomalies in its course which are characterized by the absence of some of the disease stages. We have already mentioned the mild cases in which the stage of regression goes on to

complete recovery, so that the period of trophic disturbances and contractures does not occur; these are the so-called "temporary infantile spinal paralyses." It can further happen that a "concentration" of the paralysis does not occur, in that the paralysis at once establishes itself within its final limits (in these cases usually relatively moderate). Most frequently, however, the infectious stage entirely escapes clinical recognition. Here belongs the so-called "morning

Fig. 73.

Infantile Spinal Paralysis. So-called "Hand Walker" as a Result of Severe Dorsal and Lumbar Poliomyelitis.

paralysis" in which the child is put to bed healthy and lively, and upon awakening is found paralyzed.

Less frequent than the just mentioned anomalies in course, are those cases in which the same infectious agents which lead to acute anterior poliomyelitis on account of another anatomical localization of the inflammatory process, produce different clinical pictures. Of course, their identity with the typical infantile spinal paralysis, as long as we are unable to recognize bacteriologically their cause,* cannot be proved in the sporadic cases, it is shown much more, from the observation of such cases in epidemics of the *Heine-Medin* disease.

* Serological diagnosis after *Römer, Netter, Levaditi*, and others, does not come into practical consideration. According to these authors, the blood of individuals affected with poliomyelitis, or disease etiologically related to it, contains specific anti-bodies which are able to neutralize an emulsion prepared from nerve centers containing the virus of this disease, in experiments upon monkeys.

We must thank, above everybody else, the Swedish observer, *Wickman*, for our knowledge of these rare localizations of the poliomyelitis virus; still, before him, *S. Auerbach*, *P. Marie*, *Strümpell* and others have affirmed the etiological identity of certain meningeal or cerebral diseases with epidemic spinal paralysis.

In the meningitic form the original clinical picture of meningitis can either be complicated later with poliomyelitic paralyses or there can be the simple and persistent picture of a meningitis which in autopsies by *Wickman* proved to be serous meningitis. This author has successfully opposed the view championed by several observers, for example, *Pierre Marie*, according to which there is an etiological connection between essential infantile paralysis and epidemic cerebrospinal meningitis which we will study shortly (see page 248).

In the cerebral form, instead of poliomyelitis, there is "polioencephalitis," which produces, in an acute manner, the picture of "infantile cerebral paralysis" which will be described at length in Lecture XXII. It need only be emphasized here that the cerebral form of the *Heine-Medin* disease usually begins acutely with vomiting, convulsions, stupor and fever, from which, after a few days, the children recover, but with a spastic hemiplegia which, as a rule, is more marked in the arm than in the leg. The tendon reflexes are exaggerated, *Babinski's* reflex is present, and in time, contractures appear, etc. Not rarely there occur later in such children the phenomena of hemiathetosis and hemichorea already described in Lecture V, as well as epileptic attacks. The paralysis itself is never complete; rather is it a more or less marked paresis. Also it is never accompanied by reaction of degeneration and degenerative atrophy, but only with non-degenerative "cerebral" atrophy. The bones of the affected extremities are somewhat retarded in development and the skin shows similar but much less intense trophovasomotor disturbances than those in acute poliomyelitis anterior.

Another variety takes the form of ascending or *Landry's* paralysis, in which, in rapid succession, the lower extremities, the trunk and the arms are attacked, and finally, the respiratory centers are paralyzed. Almost all of those cases of infantile paralysis which end fatally ("quite few") run their course under this picture. In this connection must be mentioned further, the already shortly sketched, bulbo-pontine form in which the facial, the hypoglossal and the eye muscle nerves are also affected; this form can also occasionally manifest itself as a true acute bulbar paralysis. Further, there is a cerebellar form of the *Heine-Medin* disease to which particularly *Oppenheim* has directed attention, and which appears as an acute cerebellar ataxia, and finally, a polyneuritic form, in which sensitiveness to pressure, and swelling of the nerve trunks, severe spontaneous pains, paresthesias, sensory disturbances, etc., can be recognized.

Etiology and Pathological Anatomy

The bacteriology of acute infantile paralysis has so far produced no useful results, since the finding of cocci and diplococci, reported by former authors were not obtained under such precautions that the causal connection between these microbes and the spinal cord affection must be accepted. Also the newest investigations of *Flexner* and *Lewis* and of *Levaditi* only permit one thing to

be definitely affirmed; namely, that the cause of the *Heine-Medin* disease must be an excessively minute organism which can pass through a *Berkefeld* filter.*

On the other hand, the communicability of the disease has been proved by the investigations of *Landsteiner*, *Flexner*, *Levaditi*, *Römer*, and others. If an emulsion prepared from the spinal cord of a child dead of acute poliomyelitis, is injected into the peritoneum, a vein, or better, into the brain of a monkey, the animal becomes paralyzed and his spinal cord, prepared in an emulsion, develops the same pathogenic peculiarities toward others of his kind. Other animals, however, are almost without exception, immune; only in the rabbit can the disease be here and there produced. The experimental poliomyelitis or spinal paralysis of monkeys presents its first symptoms from 7 to 10 days after the injection. The stage of infection usually shows itself as a general tremor, to which flaccid paralysis of the legs which may extend to the arms, trunk and the neck, is soon added. The question as to the natural port of entry of the *Heine-Medin* infection in man is not yet entirely explained. Probably the exciting agent enters through the mucous membrane of the pharynx, the nose or the intestinal canal and makes its way along the nerve trunks to the central organs.

Of the anatomical changes which it there causes, we will keep in mind only anterior poliomyelitis as the typical lesion; this shows itself as a hematogenous myelitis which localizes itself exclusively, or almost exclusively, in the territory of the anterior spinal artery. This artery sends a special branch into the anterior horn alternately according to level, into the right or into the left. This bears the name of Arteria sulco-commissuralis and forms within the anterior horn an exceedingly rich and luxuriantly branched network, which surrounds the group of root cells in basket fashion. The greatest part of the posterior horn, the white matter, as well as the most peripheral portion of the anterior horn near the boundaries of the white columns, are, on the other hand (by means of radiating branches) nourished from another vascular system, the "vasa corona"; as the arterial crown of vessels which uniting the region of the anterior and that of the posterior spinal artery surrounding the periphery of the spinal cord are called (see Fig. 74). Investigations by *Lamy*, *Rothmann*, and others, have shown that a suspension of lycopodium injected into the abdominal aorta in animals almost exclusively passes into the sulco-commissural artery and its branches, which in a certain sense acts as "an embolus catcher." These experiments make it comprehensible that it can also be a "bacteria catcher" and the particular vulnerability of the anterior horn to the cause of poliomyelitis may be connected with these peculiarities.

In the first stages of acute anterior poliomyelitis, there are found, as *Wickman* and I, among others, have shown, a great infiltration of small, round cells about the vessels of the anterior horn, and smaller or larger extravasations of blood. The gray matter appears pale, its medullated fibers swell up and

* *Flexner* and *Noguchi*, in 1913, described a very small globular organism which they had succeeded in cultivating from the central nervous system in monkeys and in human beings afflicted with poliomyelitis, and in a very recent communication announce that by a special procedure they have preserved cultures of this organism for eight months and have found it still capable of producing poliomyelitis in monkeys. (Journal of Experimental Medicine, October, 1913, and January, 1915.)—*Translator.*

become varicose, or, on the contrary, they disappear. Sometimes also necrotic foci filled with granular cells, round cells, and all sorts of detritus, are found. A greater or less number of the motor cells of the anterior horn show decided alterations: now they are formless, decidedly swollen and no processes are recognizable any longer; again, they do not take any stain, appear nearly homogeneous "shadowy"; still again, there is definite destruction of the *Nissl* granula or "tigroid scales" (the so-called "tigrolysis"); finally, sometimes, there is marked atrophy, so that the cells are reduced to small, easily overlooked clumps ("pyknosis").

Naturally, cases of infantile spinal paralysis in which the disease has run its course, come much more frequently to autopsy than fresh ones. Here there

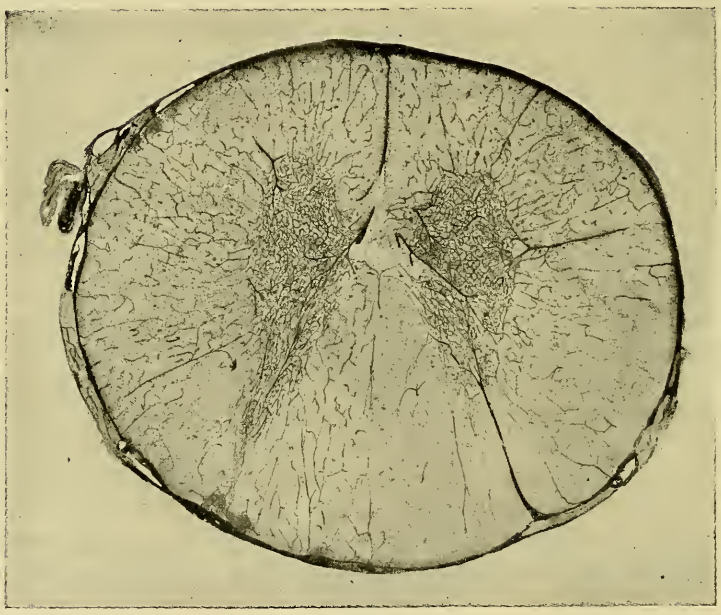

Fig. 74.

The Blood Supply of the Spinal Cord. Injected preparation. (Carmine gelatine.)

is replacement of the parts formerly affected by the inflammation by a more or less thick, gliotic scar tissue, more or less rich in vessels. The more extended the affection of an anterior horn has been, the more shrunken it appears, on which account sometimes, one-half of the spinal cord appears smaller than the other. Only a more or less considerable part of the cells of the anterior horn remain of characteristic appearance; the others have either entirely disappeared or have been changed to inconsiderable clumps (see Fig. 75).

Prognosis

The prognosis as to life is in general, and if the sporadic cases are chiefly considered, favorable. Only rarely does exitus occur in the infectious stage under hyperpyrexia and heart weakness, also the bulbar or ascending forms

of poliomyelitis as well as the meningitic varieties of the disease, all of which endanger life through their localization, belong to the exceptions. In the epidemic poliomyelitis, however, the prognosis is much more serious. So, according to *Harbitz, Scheel* and *Wickman,* in the epidemics in Sweden and Norway in 1905, among 2,078 cases there were 290 deaths, which is a mortality of about 14 per cent. As to the prognosis with regard to recovery, complete restoration is a great rarity; much more frequently there is partial recovery, that is, extensive improvement. The more rapid the regression of the paralysis,

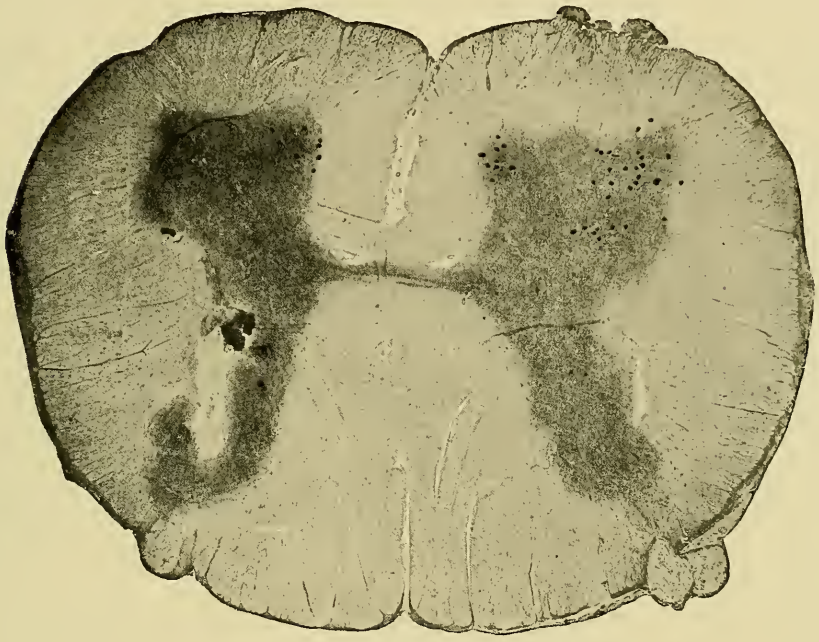

Fig. 75.

Acute Anterior Poliomyelitis in Childhood.

Late stage, carmine preparation from the spinal cord of a seventy-year-old man. (The cells of the anterior horn, still recognizable microscopically, are brought out by retouching.)

the better the outlook for a favorable result. Of the greatest importance, further, as already emphasized (page 237), is the result of the electrodiagnostic testing. Permanent severe paralyses are absent in only about a third of the cases.

In various instances the occurrence of another spinal cord disease in later life in patients who have had infantile spinal paralysis has been observed (combined system diseases, diffuse myelitis; particularly, however, spinal progressive muscular atrophy and chronic anterior poliomyelitis).*

* Chronic or subacute anterior poliomyelitis, an exceedingly rare disease, which we have already mentioned under the differential diagnosis of spinal progressive muscular atrophy, can occur also without previous acute poliomyelitis, and usually in middle life. Its etiology is obscure. Without febrile symptoms, pain, sensory or sphincter disturbances, but usually with fibrillary contractions, there appears an increasing weakness in the lower extremities, which

Differential Diagnosis

Most important from a prognostic point of view is the distinction between poliomyelitis and polyneuritis, since the last (which is not so rare in children as was formerly assumed) presents a good outlook as to complete recovery. In poliomyelitis, the paralysis reaches its maximum in a few hours, shows from then on a decided tendency to regression and is scarcely ever progressive. On the other hand, the paralysis in acute polyneuritis only reaches its maximum, as a rule, after days or weeks. Continuous extension or advance by stages in acute atrophic spinal paralysis is of the greatest rarity, on the other hand. One can count less upon the results of testing sensibility, since it should be remembered that the polyneuritis of childhood is not rarely purely motor (see page 41). Decisive as to polyneuritis is palpable thickening of nerve trunks or the occurrence of "glossy skin." The involvement of the cranial nerves speaks on the whole more for polyneuritis, optic neuritis almost certainly so. Affection of the muscles of the neck and trunk, in case there is no history of diphtheria, speaks for poliomyelitis and against polyneuritis. In the first the decided affection of the roots of the extremities is more frequent than in the last, which usually more markedly affects their ends. While after about one year poliomyelitic paralysis has almost always reached its stationary stage, the regression of polyneuritic paralysis can extend over a much longer time. In poliomyelitic paralysis, atrophy, and reaction of degeneration are usually developed proportionately. In polyneuritis reaction of degeneration can be found even in muscles which can still be moved voluntarily, hence electroprognosis is little certain here. The reappearance of faradic irritability which has been lost, turns the balance in favor of the neural seat of the paralysis. Œdema speaks for polyneuritis, and, finally, in this last, the fever may keep up longer and reappear from time to time.

Acute myelitis occurs exceedingly seldom in children; in differential diagnosis it hence plays a rôle almost only as opposed to the acute poliomyelitis of adults ("acute atrophic spinal paralysis" of adults). This last affection is exceedingly inferior in frequency to infantile poliomyelitis, but nevertheless is not such a very great rarity and has occasionally been confirmed by autopsy (*Strümpell, Schultze, Williamson* and others). That occasionally during epidemics of the *Heine-Medin* disease cases affecting adults have been observed justifies us in considering the poliomyelitis of adults in connection with infantile spinal paralysis. Many times, however, the acute atrophic spinal paralysis of adults has been observed in connection with influenza, gonorrhœa, puerperium, typhoid, etc., so that perhaps the most varied agents come into question in connection with it. Clinically it differs from the infantile form

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in the course of several days or weeks increases to an atrophic paralysis. The arms, usually soon after, share the same fate; the condition remains stationary several months, then improves gradually; indeed, there may be complete recovery. Important in prognosis is the degree of reaction of degeneration (always to be found). Death under bulbar symptoms only rarely occurs. Anatomically there are inflammatory degenerative lesions in the anterior horns of the spinal cord. In contradistinction to spinal muscular atrophy, in chronic poliomyelitis the paralysis precedes the muscular wasting. Further, it always begins in the lower extremities and runs a more rapid course. Its therapy is mainly an electric one; this corresponds to the electrotherapy of infantile spinal paralysis.

through longer duration (1 to 2 weeks) of the acute stage, through, as a rule, greater extent of the sudden flaccid paralyses and through slight tendency to regression. On the other hand, corresponding to its occurrence in completely developed individuals (usually in the third decade) the trophic disturbances and the tendency to contractures are less marked. From acute myelitis this acute poliomyelitis is distinguished by the absence of sensory and sphincter disturbances—ataxia, spastic symptoms, *Babinski* reflex, etc. (see Lecture IX).

Treatment

In the infectious stage of poliomyelitis acuta anterior, there is little to be done therapeutically, indeed, meddlesome interference is inadvisable. It appears to me much more correct to secure for the affected children as complete bodily rest as possible, than to disturb them by frequent warm baths and hot applications, as is the custom in America, for example. These procedures are intended to relieve the congestion of the spinal cord; this can be accomplished, however, by the application of several leeches along the vertebral column, by mustard paste to the calves, as well as by stimulating the function of the intestines. Among purgatives, calomel is preferred with the idea that it exerts an antiseptic action. Upon the same ground quinine salol, aspirin, etc., are recommended. I must, however, confess a great scepticism as to all these remedies. Only after the patient has entered the stage of paralysis can we interfere with somewhat more confidence in order to aid the reparative processes. Here strychnin is the medication of choice, best in the form of daily subcutaneous injections which, of course, must be carefully adapted to the age of the children. For example, in the second year of life, in which the disease occurs most frequently, 0.0005 (gr. $\frac{1}{120}$), given in one dose during the day. Further, favoring diaphoresis with hot infusions (for instance, lime flower tea), hot packs, hot air apparatus ("Phœnix," etc.) are to be recommended. On the other hand, too early beginning electrical and massage treatment is quite properly opposed by both neurologists and pediatrists. It is best to hold to *Oppenheim's* rule not to begin until after two or three weeks have elapsed, while at the start the patient is kept continuously in bed, which restriction is only to be relaxed later in the stage of regression. When we have once begun with electricity and massage, this treatment must be kept up for a long period, until definite entry upon a stationary condition is no longer dubious, on an average, for about a year. The electro-therapeutic methods here applicable have already been described in Lecture III and Lecture VII, namely, galvanization and faradization of the nerves and muscles in the paralyzed regions, and the galvanic current passed through the spinal cord. One must begin with very weak currents and gradually accustom the little patient to stronger ones. Massage, also, must be carried out in a mild, careful manner, and it must be attempted to stretch the muscles which are exposed to contracture as well as to preserve the mobility of the different joints. Vibration massage of the articulations tends to counteract trophic disturbances of the capsule and ligaments. The carrying out of active gymnastic exercises is also important: those movements which the children may

little by little become able to practice again, must (systematically with avoidance of all overexertion) be carried out, and against regulated increasing resistance. Massage and exercise therapy combats sluggishness of circulation, and so brings the muscles, integument and skeleton under better conditions of nutrition. The very popular salt-bath cures, and rubbing with liniments which stimulate the skin (linimentum ammoniæ, linimentum saponis, spiritus juniperi, etc.), accomplish the same thing. Of modern procedures, "diathermy," after *Nagelschmidt*, *Bergonie*, etc., appears to act favorably. In no case should it be neglected to reduce conditions favoring the production of deformities as much as possible, by the application of suitable splints, bed-rests, etc. If the patient has entered the stage of trophic disturbances and contractures, he should come under the care of the orthopedist. This is not the place to discuss at length the often very successful operations (arthrodesis, transplantations, plastic operations on tendons, etc.) or the various orthopedic apparatus, as they have been contrived and introduced by *Vulpius*, *Hoffa*, *Nicoladoni*, *Hübscher*, *Schulthess*, *Hessing*, and others.

B. Epidemic Cerebro-Spinal Meningitis

In contradistinction to epidemic infantile paralysis, epidemic meningitis is a bacteriologically well characterized, acute infectious disease. It is to-day established beyond a doubt that the meningococcus intracellularis (discovered by *Weichselbaum* in 1887) is its specific cause. This is a micro-organism, in its biscuit shape and its position within the leucocytes resembling the gonococcus which, inoculated into dogs, monkeys, or goats, produces a meningitis. Nevertheless, mixed infections with other pathogenic germs, particularly with the pneumonococcus and the diplococcus crassus, occur. Epidemics of the last sort have been observed particularly in barracks. The method of infection is connected with living in close association and occurs (since the meningococcus may remain viable in the naso-pharyngeal mucus even of healthy individuals, for months) regularly by "drop infection" in coughing, hawking, sneezing. *Westenhoffer* has recognized that its passage into the meninges takes place from the pharyngeal tonsil and the lymphatic tissue. Children with hyperplasia of these structures should be considered as predisposed. Ordinary anginas favor the infection, hence the preponderant occurrence of epidemics in winter, and in the cold, damp spring months. That traumatic influences which affect the skull (in children, sometimes boxes on the ear from the teacher or from the parent, have been held responsible for the outbreak of meningitis), are to be considered as exciting causes, is not certainly proved, but quite plausible. Besides the lymph channels the blood vessels are also open to invasion of the meningococcus, as culture experiments after puncturing the vein have shown. The first meningitis epidemic was observed in our country (Geneva) in 1805.

The pathological anatomy of the affection, apart from inconstant myocarditic changes and rather regular inflammatory lesions of the tonsils and the middle ear, consists predominantly of meningitic processes. The brain membranes, particularly their basal region, usually are much more decidedly affected

than the membranes of the cord. There is dependent upon the intensity of the case, and upon the stage in which it comes to autopsy, formation of a serous, sero-fibrinous or fibrino-purulent exudate between the pia mater and the arachnoid. The inflammation, however, also extends along the vessels or the nerve roots to the central organ itself. In the brain, abscesses, porencephalic defects, inflammatory hydrocephalus, etc., can occur.

Symptomatology and Course

Epidemic cerebrospinal meningitis usually seizes its victims brutally without prodromal symptoms, with sudden rise of temperature and repeated chills. In young children convulsions are not very rarely observed as an initial symptom. Only in the minority of cases a more or less marked feeling of illness, eventually with stupor and vomiting, precedes the disease proper. This, as a rule, in from 24 to 48 hours after the onset of the fever, has reached its full height, and then presents the following symptoms in more or less intense degree:

Headache of great violence and persistence, which appears to have its maximum intensity in the occipital region; very frequently, also, the vertebral column is the seat of spontaneous pains, particularly in the cervical and lumbar regions. Both the occipital region and the spinous processes of the vertebral column show great sensitiveness to percussion and pressure. Besides this, there is general hyperesthesia, all handling of the patient elicits expressions of pain; there is also great sensitiveness to optic and auditory stimuli. *Leichenstern's* phenomenon seems to indicate hyperesthesia of the skeleton; the patient when tapped on the bones of one of his extremities draws himself up violently, often with a cry. The "hydrocephalic cry" of *Trousseau*, a shrill scream which some patients even in deep stupor emit from time to time, is perhaps also to be considered as an expression of pain.

The "stiff neck" which has given the disease its popular name * imposes itself as pathognomonic even on the laity. Even on slight development of this dreaded symptom it is noticeable that the patient does not, as is generally the rule in other severe illnesses, let the chin sink more or less upon the chest, but that he deflects the head, and the occiput bores into the pillow. Active as well as passive movement of the neck is hindered in an increasing degree; our attempts to bend his head forward are answered by the patient with expressions of severe pain. Later, the rest of the vertebral column becomes stiff as a board. This high degree of hypertonia extends farther to the muscles of the extremities: in the lower extremities it is accompanied by a drawing them up on the body (tendency to contracture in flexion) through which can be produced the anomaly of position which has been called by the French on account of its analogy to the shape of a gun-flint, "attitude en chien de fusil." Here belongs also *Kernig's* sign, which depends upon the impossibility of bringing down the extremities of the patient placed in a sitting posture to the plane of the bed from their position of flexion at the hip and knee. Further

* German "Genickstarre."

phenomena characteristic of meningitis are masklike rigidity of the countenance, sometimes a moderate degree of trismus, with audible grinding of the teeth, particularly frequent, however, the "boatlike" drawing in of the abdominal walls.

The tendon and skin reflexes are usually exaggerated, their loss is an ominous symptom. Vasomotor irritability is also increased, from which the particular form of dermographia (see Lectures XXIII and XXVII) can appear, which *Trousseau* denominated "tache cérébrale." Stroking the integument leaves behind flame-red streaks which only pale after considerable time. A further cutaneous symptom of epidemic spinal meningitis is herpes, which in the majority of cases appears on the lips and face between the second and sixth day of the disease, while scarlatiniform erythemata, multiple hemorrhages into the skin, or a roseola resembling that of typhoid fever are quite atypical findings.

Further symptoms of the disease are vomiting, stupor, less frequently delirium, further, retention of urine and stools or ischuria paradoxa (dribbling of the urine when the filling of the bladder has reached a certain degree), often rapid emaciation, occasionally albuminuria and glycosuria. Characteristic of the fever is a continuous course at the start with temperatures between 39° and 40° C., which later a stage of alternating remissions and exacerbations follows; its rise to a hyperpyretic degree is prognostically bad and often immediately precedes death. In cases which enter upon recovery, the fever disappears by lysis. As to the pulse-rate, it should be emphasized that it is relatively low compared to the temperature, as a rule (for example, 100 for 40°, 80 for 39°), and that in convalescence a slow pulse is the rule (50 and lower). A rapid and considerable increase in frequency during the disease points to threatened exitus.

As to whether the slow pulse should be considered a vagus symptom, views differ. On the other hand, other cranial nerves in many cases are affected in unmistakable manner, particularly the optic and the auditory. While the meningitic optic affection (which shows itself ophthalmoscopically as papillitis), in cases which survive, seldom leaves behind amaurosis, auditory neuritis usually causes impaired hearing, sometimes, indeed, deafness. The pupils are usually narrow and react sluggishly for light and accommodation; on the other hand, they often dilate markedly when the integument is irritated. This last symptom, described by *Göppert*, can be obtained by stroking the skin with a finger-nail. Paralyses of the facial or of the external eye muscles are rarer.

In a limited number of the cases, pus collections in the eye, in the middle ear, in the brain substance occur, partly by direct conduction, in part by metastasis. With the last, there are produced spasms of constant localization (for example, in one side of the face, in one arm, etc.), which leave behind a paralysis of the muscles affected. Also a meningococcic endocarditis occurs. In the blood there is regularly a hyperleucocytosis; according to *Göppert* and others, high leucocytosis (over 24,000) is prognostically serious. It can reach 60,000. As long as the leucocytosis is not diminishing new exacerbations may be expected, even when the disease picture otherwise has consid-

erably improved. Finally, at the height of the disease a considerable polynucleosis shows itself in the spinal fluid (see Lecture XII, page 187). In the stage of convalescence (according to *Achard* and others) the polynuclear leucocytes are replaced by numerous lymphocytes. Bacteriologically the meningococcus can be recognized in the cerebro-spinal fluid, as *Heubner* first showed.

According to the course of epidemic cerebro-spinal meningitis, different forms of it have been distinguished. In the "foudroyant" form (meningitis cerebro-spinalis siderans) the patients die within a few hours after the beginning of the first symptoms; indeed, they can after relatively slight prodromes (slight fever, headache, nausea, etc.) with a loud scream fall dead to the ground, in which case we speak of apoplectiform meningitis. In a few cases of the "foudroyant" form there has been no rise of temperature. The rudimentary or abortive cases, in which the chief symptoms of the disease are only present in an undeveloped manner, form the other extreme. There is slight fever, some headache and nausea, feeling of tension in the neck and in the limbs, sensitiveness to touch, noises, etc. Such cases recover in from 8 to 14 days; when they appear sporadically, they are scarcely ever diagnosed, but their connection with cerebrospinal meningitis is only recognized during epidemics. In the acute forms, however, death in coma occurs in from 1 to 3 weeks after the outbreak of the disease. Finally, there is a prolonged, remittent, or even intermittent form, which with alternating improvement and relapse, can extend over weeks or even months. Termination in recovery is possible; usually, however, death in a so-called "status hydrocephalicus" finally occurs, after the most extreme emaciation and exhaustion, tonic contractures and flexion of the extremities, deep coma, frequent vomiting, etc. In convalescence after epidemic cerebrospinal meningitis, the danger of relapse is always great.

Prognosis

After what has been said above, the prognosis of cerebrospinal meningitis is naturally a very grave one. The mortality varies from epidemic to epidemic within comparatively wide limits (30 to 70 per cent.); as an average it is about 40 to 50 per cent. The more violent the commencement, the less the hope of survival. Even among the so-called "recovered" a large number remain severely injured for life (blindness, partial or complete deafness, mental reduction). Others escape with less impairment (tinnitus aurium, squint, tendency to headaches or neuralgias).

Treatment

From the point of view of prophylaxis, isolation of the patients, according to the general rules in infectious diseases, is to be ordered. The isolation of "carriers," that is, healthy persons who have meningococci in their nasopharyngeal secretions and who contribute to spreading the epidemic, is unfortunately only exceptionally attainable (for example, in epidemics in barracks).

In general one must content himself with having the relatives of the patient use disinfecting washes, irrigations, etc., for careful cleansing of the naso-pharyngeal space. If a serum prepared by *Wassermann* and *Kolle* for this purpose is specially efficacious, we must wait and see; favorable results have been obtained from it by *Kutscher*.

Also with regard to the result of specific treatment with meningococcus-sera (such have been prepared by *Jochmann, Flexner, Kolle, Wassermann,* and others, positive statements are not yet possible, since in consequence of the very variable malignity of different epidemics (see above, page 251) the comparison between the mortality in cases in which the sero-therapy was carried out and those in which it was not gives too great room for subjective impressions. Nevertheless, it is imperatively necessary to clear up the subject by further researches. According to *Peritz* the following rules should regulate such investigations:

If on the first lumbar puncture in a suspicious case, the cerebrospinal fluid is turbid, serum (it should not be older than three months) should be at once injected, and only when the fluid is clear should we wait for bacteriological examinations. When the puncture is made, as much fluid as possible is withdrawn, and then 30 cc of the serum is injected into the subarachnoid space, even when less fluid than this has been withdrawn. When a larger quantity of fluid has escaped, as much as 45 cc of the serum can be given. In very severe cases this last is particularly desirable. An appreciable resistance to the injection gives a signal for its cessation (*Levy* recommends limiting the first injection to 10 cc in small children and to 20 cc in adults). In malignant cases, if there is no improvement the injection should be repeated in twelve hours. Except in the very mildest cases the injections should be repeated daily for four days. If then diplococci are still found in the spinal fluid, the injections should be continued; when, however, after disappearance of diplococci from the fluid and in spite of four injections, the subjective symptoms, including the fever and stupor continue, we should wait four days, and then if there is no improvement repeat the four injections. Each exacerbation should naturally be treated on the same day. Each time the process flares up again, we should at once begin the four-day treatment and proceed otherwise as on the first occasion. This plan of treatment is to be continued until the patient is free from symptoms, the diplococci have disappeared from the spinal fluid, or the disease has entered upon the chronic stage.

As you see, this procedure demands numerous lumbar punctures and that these last by themselves, that is, even without serum therapy, present a rational method of treatment—corresponding to the rule "where there is pus, evacuate"—is evident without further discussion. They are undertaken then quite generally in epidemic meningitis, and usually as a routine procedure, that is, daily or every second day, 10 to 20 cc of fluid are removed. A few authors have, indeed, no hesitation in removing sometimes far greater quantities of fluid *Zupnik*, for example, 70 to 90 cc. The headache and stupor often markedly decrease after this procedure.

TECHNIQUE OF LUMBAR PUNCTURE

The puncture of the subarachnoid space, first done by the American neurologist *Leonard Corning* and soon after introduced into clinical practice by *Quincke*, is made in the lower portion of the lumbar region of the vertebral column. This localization suggests itself from the necessity of avoiding injury of the spinal cord with the puncture needle; the cord, however, does not extend below the second lumbar vertebra. Further distally, indeed, the "cauda equina" lies in the subarachnoidal space, but the nerves composing it are always pushed aside by the needle so that appreciable wounding of them does not occur. Slight pricking of them produces only a short, lightninglike pain. We punc-

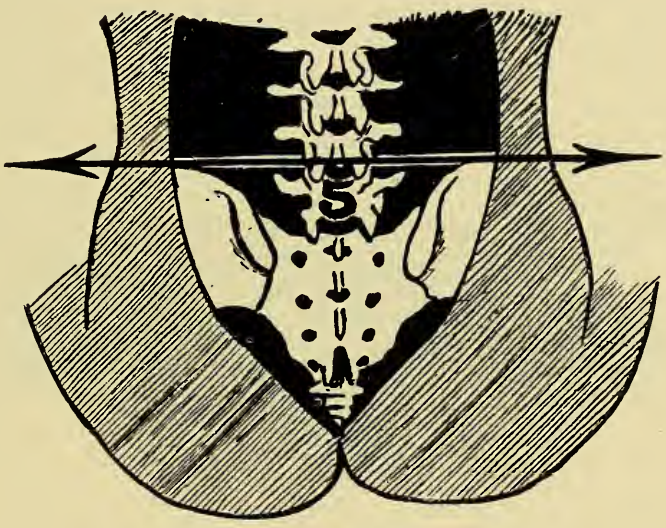

Fig. 76.

Anatomical Points for Guidance in Making a Lumbar Puncture.

ture usually between the 4th and 5th vertebræ, though the puncture can be made between the 2d and 3d or between the 5th lumbar vertebra and the sacrum. Topographic orientation is very easy, since a line drawn through the highest points of the ilia passes over the point of the spinous process of the 4th lumbar vertebra (see Fig. 76). We use a hollow needle, at least 8 cm long and of 1 mm external caliber and 0.6 mm lumen, shortly but sharply beveled at the point; a mandrin should be inserted during the puncture. This can be made in the median line in the space between the 4th and the 5th lumbar vertebræ; it is in general better, however, to make the puncture about 1 cm to the right of the middle line, since by so doing, on the one hand the tough interspinous ligament is avoided, and on the other the needle enters the somewhat more roomy lateral part of the intervertebral foramen. Of course, such a "paramedian" puncture demands that the needle be directed somewhat toward the median line, as shown in Fig. 77. For the rest, in children the needle should be introduced at the ideal cross-section of the

trunk; in adults it must usually be introduced somewhat more frontally. The operation is further made easier by strong flexion of the vertebral column, since by this the space between the arches of the vertebræ is enlarged. Patients who can sit up must hence bring their heads forward as low as possible; where this is not possible, as is almost always the case in meningitis, the patient is laid on his side and his knees bent up as far as possible. The skin over the point of entrance is disinfected by painting it with fresh tincture of iodine, and eventually is anesthetized with the chloride of ethyl spray. The hands are carefully disinfected (by soap, scrubbing and alcohol), and the previously boiled needle is introduced in the direction indicated. The ligamentum flavum, which is stretched between the arches of the vertebræ, gives an elastic resistance which, however, suddenly yields, and the point of the needle enters the subarachnoid space. If, however, the needle is wrongly directed

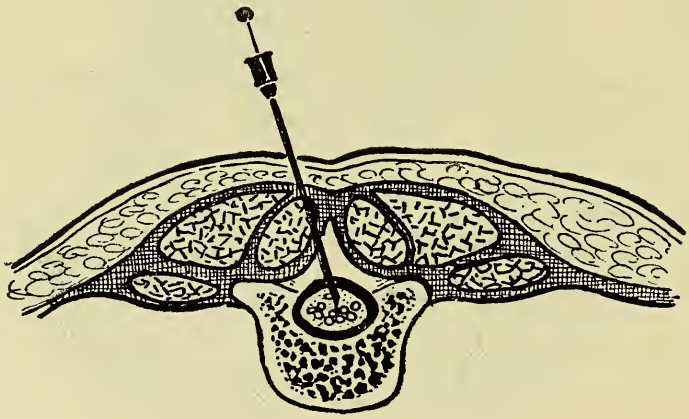

Fig. 77.

Introduction of the Needle in Lumbar Puncture.

and strikes the bone instead of the yellow ligament, it is somewhat withdrawn and directed either more frontally or more caudally. If the instrument has been properly introduced, the mandrin is withdrawn and the fluid escapes in drops or, when the pressure is increased, in a stream. If the needle gets stopped up by coagulum, which is not rare, the obstruction is to be removed by reintroduction of the mandrin. If, instead of fluid, blood appears, the needle has probably entered a vessel of the venous plexus which covers the posterior aspect of the body of the vertebra; in this case the needle is withdrawn several mm until no more blood escapes. That anomalies of the vertebral column or adhesions of the arachnoidal sac render lumbar puncture impossible is a rare happening. The depth to which the needle must penetrate varies from 2 cm in small children and 6 or 7 cm in adults. After lumbar puncture the patient should remain in a horizontal position for at least 24 hours.

Bier, *Vorschütz* and *Eckert* have recommended the combination of lumbar puncture with "stasis." Two hours after the puncture an elastic band is placed

about the neck, tight enough to produce a slight cyanosis of the face, and kept on about 20 hours.

In general use are hot baths—which, according to *Heubner's* directions, should be given daily—in which beginning with 35° C., the temperature should be raised slowly to 40° and more. On the other hand, cold baths and affusions, as are customary in typhoid, are recommended. Application of the ice-bag or of the cold cap to the head and along the spine usually reduce the headache. Further, derivative procedures, as leeches or cupping over the mastoid and along the vertebral column, inunction of gray ointment over the neck, blisters, frequent purgation (calomel), mustard paste to the calves, etc., can be used. Of drugs, antipyretics and sedatives come into consideration (salicylate of sodium, antipyrin, pyramidon, chloral, morphin, etc.). More important, however, is the carrying out of the difficult and responsible nursing; namely, by the choice of concentrated food, its frequent introduction (when it is necessary with the œsophageal tube), the rapid loss of strength is to be combated as much as possible. In convalescents the most careful oversight and tonic treatment is in place (fresh air, sea and mineral baths, arsenic, strychnine, etc.).

Note.—On account of the discovery that when hexamethylamin (urotropin) is given by the mouth it appears in the spinal fluid, this drug is quite generally administered in infectious diseases of the central nervous system, particularly in poliomyelitis and cerebrospinal meningitis. Its value is, however, uncertain.—*Translator.*

LECTURE XVII

Encephalorrhagia and Encephalomalacia

(Cerebral Hemorrhage and Cerebral Softening)

GENTLEMEN: We turn now to the exceedingly important disease pictures which arise from more or less extended, circumscribed destructions of the brain substance upon a basis of vascular lesions. Now it is rupture of an artery, again occlusion of an artery (from endarteritis, thrombosis or embolism). In the first case a pouring out of blood destroys the cerebral parenchyma, in the last this undergoes ischemic softening. Now, before we consider separately encephalorrhagia (cerebral hemorrhage) and encephalomalacia, we will take up the most typical of the symptom complexes through which they manifest themselves clinically—cerebral hemiplegia. Since now, however, of these cerebral hemiplegias by far the greatest number have their focus in the internal capsule, our description of the semiology will be based upon this form.

Capsular Hemiplegia

This is, as a rule, the result of a cerebral hemorrhage. That the internal capsule is the favorite seat of this last is due to the anatomical relations of the vascular distribution. The middle cerebral artery, the most important branch of the internal carotid, gives off on the base of the brain, branches mounting vertically to the optic thalamus, the corpus striatum, and the internal capsule, the lenticulo-optic artery, and the lenticulo-striate arteries. One of these last which passes along the surface of the lenticular nucleus to the internal capsule, perforates this and finally ends in the corpus striatum, has been given by *Charcot* the name "artery of cerebral hemorrhage" (see Fig. 78). These perforating basal vessels (in contradistinction to the arteries of the cortex which anastomose with one another) are so-called "end-arteries"; since, besides this, they come off at nearly right angles from the largest branch of the carotid, there is in them a pressure nearly equal to that in the carotid, and every increase of this pressure is transferred directly to them, but not to other cerebral vessels of similar caliber. *Mendel* has been able to bring these relations to demonstration manometrically in a model of the cerebral vascular system made out of rubber tubes. Variations of pressure are now, as we will later see, of decisive importance in the production of cerebral hemorrhages.

There lies, however, in the posterior limb of the internal capsule nearly the whole of the motor tract for the opposite side of the body together in a

relatively very small space (see Fig. 79). Complete hemiplegia, hence, is in by far the majority of cases the consequence of a lesion of this posterior limb of the internal capsule, that is, there is a cross paralysis of the lower facial (the upper escapes in consequence of its bilateral representation in the cortex) of the hypoglossus, of the arm and of the leg. If the disease focus, however, extends also to the posterior third of the posterior limb of the internal capsule,

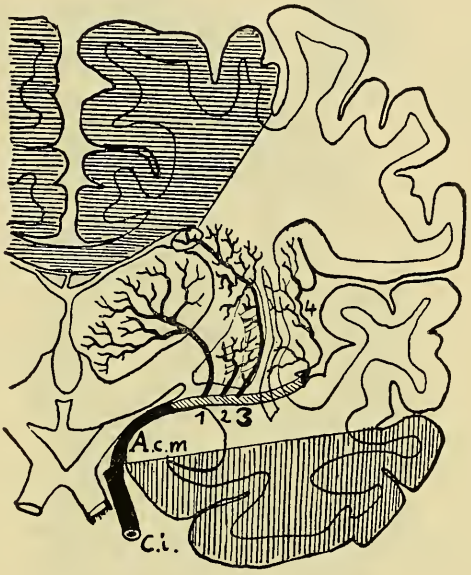

Fig. 78.

Arterial Supply of the Cerebrum and the Basal Ganglia.

= Distribution of the Anterior Cerebral Artery.

= Distribution of the Middle Cerebral Artery.

= Distribution of the Posterior Cerebral Artery.

C.i. = Internal Carotid.
A.c.m. = Middle Cerebral Artery.
1 = Lenticulo Optic Artery.
2 and 3 = Lenticulo Striate Arteries.
3 = So-called "Artery of Cerebral Hemorrhage."

where the sensory tracts lie together immediately after their exit from the optic thalamus, there results besides a crossed hemianesthesia of the whole body. Sometimes the destructive focus extends even to the posterior end of the internal capsule to the so-called "Carrefour sensitif" (sensitive crossway). where two important sensory tracts, the optic and the auditory, branch off, from the total contingent of sensory tracts to proceed to the visual and auditory centers of the brain cortex (see Fig. 79).

In such cases, now, there occur also hemianopsia and unilateral deafness,

both on the opposite side; since each visual area receives the visual impressions from the opposite halves of the visual field of both eyes, as is shown in Fig. 80, and each auditory cortical area the end neurons of the auditory tract from the opposite ear.

Apart from the eventual sensible and sensory accompanying symptoms, capsular hemiplegias present also the following peculiarities:

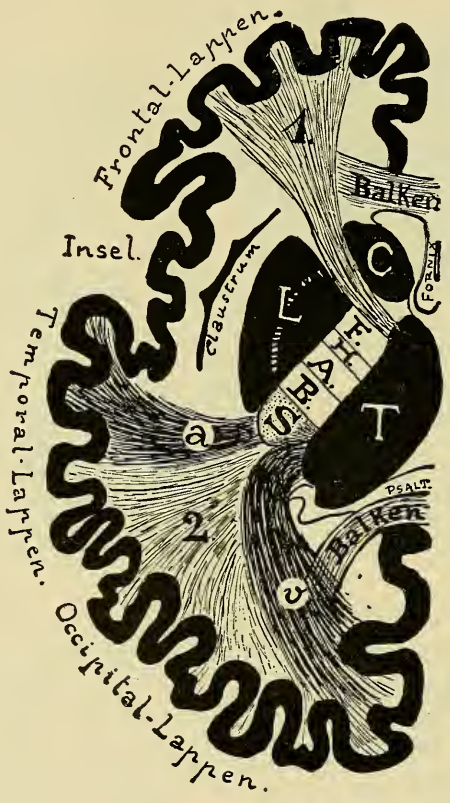

Fig. 79.

The Internal Capsule and Corona Radiata.

T = Optic Thalamus.
L = Lenticular Nucleus.
C = Caudate Nucleus.
F = Supranuclear tract for the Facial.
H = Supranuclear tract for the Hypoglossal.
A = Supranuclear tract for the Arm Muscles.
B = Supranuclear tract for the Leg Muscles.
S = Sensory tract (Tractus Thalamo-Corticalis).
a = Auditory tract to the Temporal Lobe.
v = Visual (*Gratiolet's*) tract to the Occipital Lobe.
1 = Fronto-Pontine tract and fibers to Thalamus.
2 = Occipito-Temporo-Pontine tract and fibers to Thalamus.
Balken = Corpus Callosum.
Insel = Island of Reil.
Frontal-Lappen = Frontal Lobe.
Temporal-Lappen = Temporal Lobe.
Occipital-Lappen = Occipital Lobe.

Besides the upper facial muscles and on account of analogous relations of innervation (connection of the corresponding nuclei of the brain axis or spinal cord with both halves of the cerebrum) the masticatory, deglutitory, eye and

trunk muscles remain unaffected. (For the rest, as to the upper facial, it is, however, to be stated that often there is slight diminution in the contractility of the frontalis and the orbicularis palpebrarum on the side opposite to the lesion; the eyebrows hang perhaps somewhat deeper, or the eye can be kept closed for a shorter time than upon the homolateral half of the face.) Also,

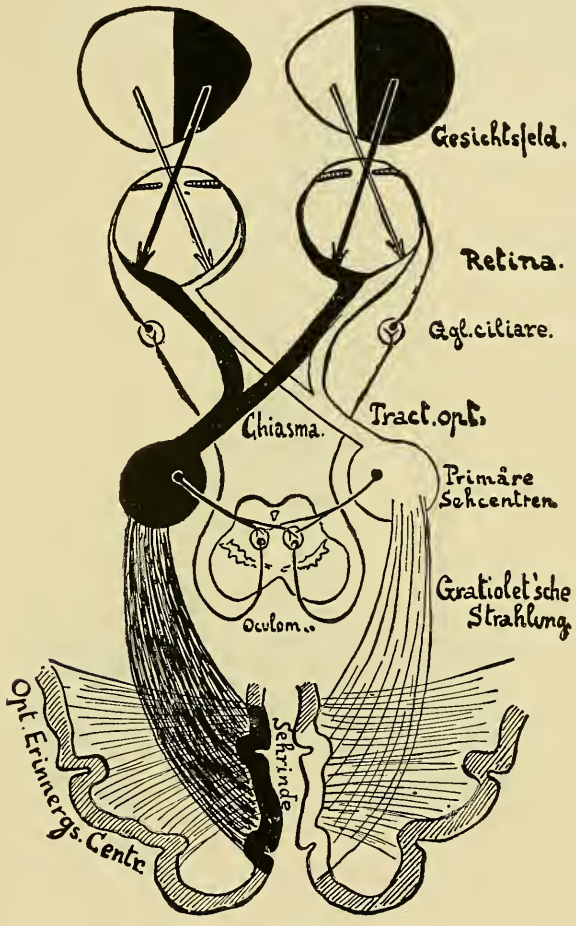

Fig. 80.

Visual Tract and Pupillary Reflex Paths.

Opt. Erinnergs. Centr. = Center for Optic Memories.
Sehrinde = Cortical Visual Center.
Gratiolet'sche Strahlung = Optic Radiations.
Primäre Sehcentren = Primary Visual Centers.
Gesichtsfeld = Visual Field.

in complete hemiplegia different groups of muscles in the extremities are regularly unequally affected. While power of movement, little by little, returns in a great number of muscles, others do not usually recover again (peroneal muscles, flexors of the knee, extensors of the elbow, extensors of the hand and fingers, external rotators of the arm, supinators of the forearm). The arm, as a rule, is more affected than the leg. The positions, however, which the

extremities assume in consequence of the preponderating action of those muscles which regain their motility are often later fixed by contractures. On this account the foot is held in equinovarus position and the knee extended so that in walking, the leg dragging, must be swung forward in a lateral arc ("circumduction," "helicopodia"). The arm is fixed in adduction, the elbow, hand and fingers in flexion.

These contractures occur from the fact that the anterior horn cells, liberated from the inhibiting influence of the pyramidal tract, are stimulated in a

Fig. 81.

Cerebral Hemiplegia from Capsular Hemorrhage on the Right Side. Circumduction of the Left Leg in Walking and Typical Contracture Position of the Left Arm.

tonic manner by the impulses entering through the posterior roots, and a summation of these stimuli is produced. Since, however, there are certain definite muscles which regain their motility and therewith (thanks to the supranuclear hypertonic nature of cerebral hemiplegia) are predisposed to contracture, this striking regularity must depend upon relations of innervation not yet sufficiently explained anatomically. Probably motor paths from the subcortical centers (for example, the roof of the mid-brain, the tegmentum and *Deiters'* nucleus) pass in a preponderating manner directly into contact with the anterior horn cells of these groups of muscles, so that in the reparatory effort of the organism it is not difficult for them to obtain again, in a roundabout way, a share of the cortical innervation. Cases of cerebral hemi-

plegia in which other muscle groups retain preponderance and develop contractures (so that, for example, the leg is fixed in flexion) form very rare exceptions. The hypertonia of capsular hemiplegia is accompanied by increase of the tendon reflexes. To this hyperreflexia and the pathological reflex phenomena (*Babinski's*, *Oppenheim's* and the *Mendel-Bechterew* reflexes), clonus, certain associated movements, and so forth, which so often accompany them, all that we said in Lecture VII in connection with spastic paraplegia applies. In a great number of capsular hemiplegias besides, one or another of the following phenomena can be demonstrated: The patient is laid upon a firm surface, his arms crossed and his head partially supported; if he is now requested to raise himself up, he bends his leg on the paralyzed side at the hip-joint so that the heel is raised from the supporting plane. The same associated movement (the "hip flexion phenomenon") appears when the patient, seated, attempts to lie down again. As "platysma phenomenon" ("signe de peaucier") is denominated the energetic contraction of the platysma of the healthy side, which becomes visible when the cerebral hemiplegic opens his mouth. Finally, an interesting modification of the radius reflex, which consists in the occurrence of a flexion of the fingers instead of the normal flexion of the forearm at the elbow, when the radius on the paralyzed side is tapped at its distal extremity, may be mentioned. The skin reflexes (namely, the abdominal and cremaster reflexes), on the contrary, are almost always reduced or absent on the paralyzed side; this symptom can be used while the patient is lying unconscious after an apoplectic stroke, to determine on which side of the body the hemiplegic paralysis will be found after he comes out of the coma. This phenomenon is explained in quite plausible manner as follows: The centripetal fibers concerned in the abdominal and cremaster reflexes enter the cord through the posterior roots of the lower dorsal and upper lumbar regions and their centrifugal fibers come through the anterior roots from the same part of the spinal cord. But the stimulating process is not transmitted here through the direct spinal reflex arc, but through the intermediary of interposed neurones which first proceed in a frontal direction into the cerebrum and then again run caudally. We cannot propose any further hypotheses concerning this, in any case, complicated mechanism. The conjunctival reflex is also often absent on the paralyzed side.

Capsular hemiplegia is a nondegenerative paralysis. Nevertheless, there are certain exceptional cases in which in the paralyzed or even in the recovered muscles, the so-called "cerebral atrophy" which cannot be referred to inactivity and is accompanied by reduction of the electric irritability, although without reaction of degeneration, develops. With this, arthropathies like those in tabes have been observed. Still more isolated are the observations of atrophy of bone upon the paralyzed side. More frequent, on the contrary, than all these trophic disturbances, are anomalies of the vascular innervation: the paralyzed parts are cyanotic and feel cold; sometimes there develops quite decided localized œdema.

The clinical picture just sketched you need not expect immediately after the occurrence of a cerebral hemorrhage or of a sudden plugging of a vessel, as I have already indicated in speaking of the condition of the skin reflexes.

Much more do these happenings accompany a serious, though also usually transitory injury of the whole brain, which, since *Hippocrates*, has been termed "apoplexy" (*αποπλήχτειν*—to strike down). We will proceed to the clinical description of this condition.

The Apoplectic Attack

The most striking symptom of the apoplectic attack is the sudden loss of consciousness, which is, indeed, sometimes preceded by certain warnings, as feeling of dizziness, paresthesias in the limbs, headache, nausea, darkening of the field of vision; the patient is restless, begins to totter, and then suddenly falls into a lifeless heap. He lies there unconscious, usually with congested and swollen face and stertorous breathing. There is incontinence of urine and stools, only exceptionally retention. The limbs are all flaccid, the tendon reflexes are lost. Nevertheless, we are enabled now and then to recognize, even at this time, which side of the brain has been affected. For example, if when raised up and then suddenly let loose, the extremity on one side falls more heavily to the bed than the other and remains stretched out straight upon it, this is the one in which hemiplegia will later be manifested. On this side, also, sometimes the hand brought into supination falls back into pronation much quicker than on the other side. Further, the limbs on the side opposite to the lesion, when the patient is exposed, chill more rapidly than the others. Also, observation of the face may permit localization of the disease focus, since the sail-like flapping of the cheeks in expiration in the comatose patient (so-called "blowing smoke") is more marked upon one side (the paralyzed one). Occasionally it is remarked also that the head and eyes of the apoplectic patient are directed toward one side, and when the face is brought to the front again it always returns to this forced position. This is *Vulpian's* "conjugate deviation" ("déviation conjuguée de la tête et des yeux"). This, also, can well be applied in the localization of the disease focus, since the prostrate and relaxed apoplectic turns his head and eyes to the side of the body unaffected by the hemiplegia.* We have already spoken of the absence of the abdominal and cremaster reflexes on the hemiplegic side and its importance as an early symptom.

The temperature of the body usually falls below the normal in the apoplectic attack, to rise later to a febrile degree. When the course is favorable it returns to normal after some variation; a continued mounting of the fever is very unfavorable prognostically. It depends so very frequently upon broncho-pneumonic processes, that many neurologists would make trophic disturbances in consequence of the cerebral attack responsible for the rapid occurrence of these last. "Acute decubitus," developing occasionally immediately after the ictus (in the region of the sacrum), an ominous sign, encourages a similar hypothesis.

The apoplectic coma usually lasts some hours, but unconsciousness for

* In contradistinction to this, patients who show unilateral convulsions from irritative cerebral lesions, in the case of conjugate deviation, look toward the convulsed extremities.

several days is nothing unusual. The longer this condition, which must be considered as a sort of concussion of the brain, lasts the worse in general is the prognosis. It is particularly unfavorable in cases in which even during the coma the tendon reflexes are exaggerated, the muscles spastic; previous experience justifies the diagnosis of the excessively dangerous breaking through of the blood into the lateral ventricle, under these circumstances.

Sometimes a short, so-called "stage of reaction" precedes coming out of the coma; during this the apoplectic becomes restless, sweats profusely, is somewhat delirious, complains of headache, etc. When these general symptoms have passed away and the patient is again conscious, there follows, by degrees, the change of flaccid into spastic hemiplegia introduced by the return of the tendon reflexes and their exaggeration upon the paralyzed side. The *Babinski* phenomenon appears on this last, not at all infrequently, already in the coma; in traumatic unilateral cerebral hemorrhages I have been able many times to elicit it, even in the hours just after the accident. With the development of spasticity there is often a limitation of the extent of the paralysis. Indeed, not so very rarely, the disturbance of motility may disappear in relatively short time. This is the case when the hemorrhage does not directly sever the cortico-spinal tracts, but has occurred in their neighborhood, for example, in the lenticular nucleus. The compression to which the neighborhood of the hemorrhagic focus is exposed for some time, the collateral œdema, etc., are to be held responsible in such cases for the indirect symptoms of interference with the motor tracts. Only the pareses which persist 6 to 8 months after the stroke are to be considered as direct focal symptoms, and prognostically to be estimated accordingly.

This restitution may depend upon the disappearance of the so-called "Diaschisis." By this expression—since the fundamental work of *v. Monakow*—we denominate as sort of passive shock: the absence of stimuli which are connected with the destroyed nerve tracts paralyze, also, anatomically intact regions, which in their activity have adapted themselves to these stimuli. The action of this diaschisis is in its nature fleeting, temporary, not residual.

Pathogenesis and Etiology of Cerebral Hemorrhagic Foci and Areas of Softening

Since now we have studied from an exclusively symptomatological point of view the typical clinical pictures of capsular hemiplegia and of the apoplectic attack which accompanies it, as a rule, it is now our task to subject to a systematic description the different pathological conditions which lead to encephalorrhagia and encephalomalacia.

1. *Arteriosclerotic changes in the brain vessels* take the first place in the anatomical substratum of cerebral hemorrhage. We can here refer to the general remarks which we made in Lecture XV (page 224), in speaking of cerebrospinal arteriosclerosis. We would emphasize again that between arteriosclerotic changes of the peripheral arteries and those of the cerebral vessels there is no necessary parallelism, and that a high degree of alteration of the one set in no way excludes the good condition of the other, and vice versa.

Particular interest, however, from the point of view of the pathogenesis of cerebral hemorrhages is presented by the characteristic lesions whose exceedingly frequent occurrence in the small and medium-sized cerebral arteries (particularly, however, in the lenticulo-striate artery) *Charcot* and *Bouchard* pointed out in 1868; the "miliary aneurisms" which usually are a "sacciform" subvariety of vascular dilatations, have at most a caliber of 1 mm and are often found in very great numbers.

Having been produced by the influence of permanently increased blood pressure (here contracted kidney and hypertrophy of the heart, frequent accompaniments of arteriosclerosis play a very great rôle), they are exposed to the danger of bursting upon any sudden further increase of the blood pressure. In this sense act all influences tending to produce congestion, which either increase the arterial blood supply to the brain or hinder its venous return. On this account the following physiological conditions furnish the most frequent exciting causes of cerebral hemorrhages: Digestion, defecation, coitus, psychical excitement, overexertion of any sort. As to traumatic apoplexies, which occur, for example, after a fall on the head, it is to be remarked that they usually affect arteriosclerotic individuals, that also rupture of a brain vessel from the action of external force, as a rule, presupposes a morbid brittleness of the cerebral arteries. The apoplexies depending upon arteriosclerosis usually occur beyond the fortieth year. Many apoplectics, long before the occurrence of cerebral hemorrhage, are distinguished by the so-called "Habitus apoplecticus." They are thick-set, somewhat corpulent individuals, with short necks, red faces, and visibly swollen and tortuous temporal arteries.

The pathologico-anatomical picture of encephalorrhagia is, according to the stage in which the patient comes to autopsy, a varied one. In fresh hemorrhages a more or less extended (usually walnut-size) black-red coagulum is found. About this the brain substance is broken up and infiltrated with blood. In somewhat later stages the saturation of the neighborhood with œdematous fluid, colored citron-yellow from admixture of blood-coloring matter, is characteristic. Later, the following changes occur: Contraction of the blood clot, liquefaction and absorption of the destroyed brain substance, reactive proliferation of the glia at the periphery of the focus. If the last has become small, there results finally, as a residuum of the hemorrhage, an ochre-colored "apoplectic scar." If it is a large extravasation, an "apoplectic cyst" filled with serous fluid is left behind. The most important secondary lesion of capsular hemorrhagic foci is descending degeneration of the pyramidal tracts.

Much more rarely than to rupture of a vessel, arteriosclerosis leads to thrombotic closure of one of the brain arteries. In such cases the obliteration is introduced by a specially intense fibrocellular proliferation of the intima.

2. *Syphilitic Endarteritis Obliterans.*—In this vascular disease, studied by *Heubner*, *Friedländer*, and others, there is inflammatory infiltration of the intima with round cells, as well as considerable increase of the endothelium. By the intrusion of newly formed vessels from the vasa vasorum, there is produced within the diseased brain artery an organized granulation tissue, which (occasionally with the addition of thrombotic blood coagulation) finally com-

pletely occludes the vessel lumen and interrupts the blood stream. Now, if no collateral circulation occurs (and in the "end arteries" of the brain this last is excluded), the brain area, cut off from its nourishment, undergoes ischemic necrosis and softening. Fresh areas of softening are white or, in consequence of infiltration by blood from the vessels of the healthy neighboring tissue, red in color. Older foci take on a yellow or brown color, on account of alterations in the blood pigment. Syphilitic endarteritis affects on the average younger individuals than arteriosclerosis. It occurs (upon a hereditary basis) even in children; most of the cases, however, are in the third and fourth decades of life. Sometimes it leads to rupture of the diseased vessel instead of to obliteration. All hemiplegias in young individuals are directly suggestive of syphilis.

Once in a way other infectious diseases also lead to affections of the intima of the brain arteries, in which there is sometimes formation of a thrombus in the diseased vessels; the very rare encephalomalacias in typhoid fever, diphtheria, etc., are thus explained.

3. *Cerebral Embolism.*—This variety of vascular occlusion depends upon the lodgment of a coagulum originating somewhere else in the organism in an artery of the brain. The place of origin of the emboli brought to the brain is usually the left heart; the diseases responsible for their production are vegetating or ulcerous endocarditis of the aortic and mitral valves, further heart aneurisms and certain cases of heart weakness in which thrombi are formed in the recess or in the auricles of the left heart, and later portions broken off and thrown into the circulation. Also arteriosclerotic disease and aneurisms of the aorta and of the carotids occasionally furnish the material for brain emboli.* These last, in by far the majority of cases, are caught in the *Sylvian* artery, occlude it, and effect destructive softening of the basal ganglia and the internal capsule—while in a remarkable manner the cortical regions supplied by this artery often escape destruction through collateral circulation. The left *Sylvian* artery is more frequently affected than the right.† It is usually young individuals in whom brain emboli occur. As exciting factors those producing elevation of blood pressure, like in encephalorrhagia, play an unmistakable rôle: defecation, coughing, vomiting, overexertion, psychical excitement, coitus, etc.

Differential Diagnosis Between Hemorrhage and Softening

It is sometimes an exceedingly difficult task to form the decision as to whether an apoplectic attack or a cerebral hemiplegia depends upon a hemorrhage or is the consequence of an embolic, thrombotic, or endarteritic vascular occlusion. As guiding principles in this differential diagnosis, the following points may be of use:

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* As to septic emboli from the pulmonary vein (in gangrene of the lung, etc.), see Brain Abscess, Lecture XIX.

† As to embolism and thrombosis of the basilar artery ("acute apoplectic bulbar paralysis"), see Lecture VII.

1. Embolism and syphilitic endarteritis usually affect younger, arteriosclerotic thromboses as well as cerebral hemorrhages, older individuals.

2. Heart murmurs speak for embolism; it is to be remarked, however, that quite frequently with the appearance of the apoplectic attack, heart murmurs previously present, disappear, and do not reappear for some time. Hypertrophy of the left ventricle turns the balance in favor of hemorrhage.

3. The urinary findings characteristic for chronic interstitial nephritis (increased quantity, small amount of albumin, hyaline, and granular casts, a very few leucocytes) speak rather for hemorrhagic apoplexy. Contracted kidney occurs in about 30 per cent. of the cases of cerebral hemorrhage, while blood casts (an expression of a hemorrhagic kidney infarct) point to embolism.

4. In a small number of cases, examination of the eye grounds can furnish diagnostic aid, namely, when hemorrhages into the retina, albuminuric retinitis, or embolism of the central artery of the retina, can be found. The last speaks for an analogous process in the brain, the two first anomalies for cerebral hemorrhage.

5. The forerunners of the apoplectic attack (described on page 262) are of much longer duration in thrombotic or endarteritic vascular occlusions than in hemorrhage. In embolism they are but inconsiderable, or are absent entirely.

6. In the attack itself, congestion of the face makes probable a cerebral hemorrhage; its pallor, a closure of a vessel; nevertheless, these rules admit of exceptions which are in no way rare. The fall of the temperature at the start is nearly always absent in vascular occlusion. Clonic spasms speak decidedly in favor of embolism, particularly when they are unilateral. Conjugate deviation with relaxed limbs is characteristic of hemorrhage. Coma sometimes is absent in autochthonous vascular occlusion, and when present is, as a rule, of shorter duration, but deeper than in embolism; very profound and long-continued coma, however, indicates bursting of an artery. The seizure clears up most slowly in this last lesion.

7. Accompaniment of a right-sided hemiplegia appearing after the ictus by aphasia (see Lecture XVIII) speaks in general for cerebral softening rather than cerebral hemorrhage. A complete, or almost complete, disappearance of the hemiplegia or an only partial development of this (for example, brachial monoplegia) speaks in the same sense.

Apoplexies which leave behind a hemiplegia lasting only one or several days should always arouse a suspicion of general paresis. In young individuals there come also into question, as already said (see Lecture VIII, page 140), the apoplectiform attacks of multiple sclerosis.

Prognosis

The clinical distinction between the several anatomical substrata of apoplexies and hemiplegias is not only of scientific interest, but also of great importance prognostically. Now, the decision that an endarteritic obstruction has arisen upon a syphilitic basis, arouses the hope that by commencing an energetic anti-syphilitic treatment a recurrence will be prevented, while arterio-

sclerotic thrombosis gives in this respect a very dubious outlook; repeated attacks are here the rule; in many cases there are, finally, bilateral hemiplegias, pseudo-bulbar paralytic phenomena, deep dementia, marasmus, etc. After passing through a cerebral hemorrhage the patient, while not safe from recurrences, is much less threatened by them. Also, in contradistinction to arteriosclerotics with brain softening, his intelligence remains intact after the seizures. After embolism there are no further attacks, as a rule. Decisive for the prognosis as to life is here the underlying disease. Further, the paralytic symptoms resulting from cerebral hemorrhages, according to experience, show a much greater tendency to disappear little by little than those from foci of softening; this is connected with the fact that there the indirect and distant symptoms play an important rôle, here a very minor one. The defect symptoms which persist about a month after the occurrence of embolism or thrombosis can usually be considered as final, while in hemorrhage a permanent condition can only be assumed after 6 or 8 months. If there is decided regression of the paralyses immediately after recovering from a sanguineous apoplexy, this is prognostically very satisfactory. The appearance of contracture, on the other hand, excludes finally the hope of complete restoration.

A very important prognostic question is as to whether the patient will survive the apoplectic attack or not, in case he does not die at once ("apoplexie foudroyante"). That a spastic condition of the muscles from the start clouds the prognosis, we have already said; also, we have emphasized the unfavorable indication of acute decubitus. Further, very bad symptoms are, long duration of the coma (over 24 hours); its increasing depth (ingravescent apoplexy); the occurrence of a pneumonia; finally, "*Cheyne-Stokes* respiration" (a periodical increase, then decrease, of the depth of inspiration which occasionally can lead to complete intermissions in the breathing).

Treatment

In fresh apoplexy, after we have taken care that the patient is put to bed, avoiding all jarring (with careful supporting of the head), and all tight clothing has been loosened, in our further proceedings the differential diagnosis between hemorrhage and vascular occlusion, into which we have already entered quite thoroughly, are of importance; besides, the condition of the heart and the vessels must always be taken into consideration. The time when every one affected by an apoplectic stroke was bled schematically has gone by. We consider blood-letting as indicated only when it must be assumed that there is cerebral hemorrhage, the face being red, the pulse tense. In many cases the application of leeches behind the mastoid processes suffices; when the carotids are very full of blood and there is great congestion of the face, however, withdrawing from 125 to 250 cc of blood (venesection or puncture of the cubital vein) is entirely in place. On the other hand, if the clinical picture, in agreement with the previous history, speaks for embolism or thrombosis, if the face is pale, the pulse is weak or intermittent, we should refrain from bleeding, and must make use of stimulants. As such, there come into consideration subcutaneous injections of ether, acetic ether, camphorated oil,

or caffein, as well as oxygen inhalations. Putting an ice-bag upon the head of the apoplectic is a measure sanctioned by tradition; the possibility that in cerebral hemorrhage it acts through the skull in promoting hemostasis is, indeed, very problematic, though it is not entirely excluded; in embolism and thrombosis, however, it can in any case do no harm, so that we need not object to this ordinary measure, which gives a certain satisfaction to the friends. Adrenalin injections are to be avoided in cerebral hemorrhage; the disadvantage of the considerable increase of blood pressure which they cause outweighs the possible advantages of a vasoconstriction (which, if we draw conclusions from animal experiments, is least to be expected in the brain arteries). That the injection of ergot preparations is able to stop the cerebral hemorrhage is very questionable; nevertheless, there is no objection to trying them.

From the start we should exert our efforts toward the avoidance of the dangers of acute decubitus and pneumonia. From the point of view of both of these it is to be recommended when the coma lasts longer than 2 or 3 hours, to turn the patient on his side (naturally very carefully and with proper assistance), and when it is necessary, after a few more hours, to change him to the other side. Eventually pneumatic or water cushions. The skin exposed to bed-sores is bathed from time to time with alcohol. Further, it is well, with a wad of cotton on the end of an applicator, to mop out the mucus which tends to collect in the pharynx, from time to time, or to promote its being swallowed by encouraging the swallowing reflex.

The retention of this reflex almost without exception, as a rule, permits giving the comatose apoplectic some nourishment without having to use the œsophageal tube; naturally only small portions must be given at one time (beef-tea, egg-nog, "hygiama," meat-juice, olive oil, etc., a tablespoonful at a time). Also analeptics and expectorants; for example, strong coffee with aromatic spirits of ammonia, can be given as needed from time to time, by the mouth. When there is retention of urine, we should not forget to catheterize at proper intervals; in the more frequent incontinence, the utmost cleanliness should be observed. Enemata are needed in most cases; for "depletion by the intestine," instead of the usual injections (soapsuds, glycerin, etc.), enemata of sulphate of sodium or sulphate of magnesium solutions can be tried. (After emptying the bowel, a nutrient enema, consisting of gruel, yolk of egg and peptone may be given.) If it is desired to act depletingly not only through the intestine, but also by the skin, hot packs to the lower extremities are simplest; more energetic effect may be obtained, however, by the application of mustard plasters to the calves.

In apoplexies of traumatic origin, with persistent or ingravescent coma, surgical interference (puncture, or even incision into the hemorrhagic focus) has been practiced occasionally with fortunate outcome.

In the stage of reaction we should not fear to combat the often great jactitation by injections of morphine. If the patient regains consciousness, it is our first task to explain to him the situation, and to instill into him hope and equanimity by pointing out the improvement that is to be expected. To obtain for him good nights he should be given the alkaline bromides, chloral, codein, veronal, trional, etc. For the rest, however, with the excep-

tion of cases of syphilitic etiology (in which we usually give iodipin injections of 10 to 20 cc of a 25 per cent. solution in the first days after the attack, and begin soon after with injections of biniodide of mercury—see Lecture XIII, page 210), it is better to avoid further medication. Only after about 14 days the arteriosclerotic apoplectic is allowed to begin a course of iodide of potassium, as we have described it in Lecture XV. Naturally, the therapeutic and special dietetic indications given for arteriosclerosis cerebro-spinalis apply to the later treatment of these cases in the fullest degree. During the earlier days after recovery from an apoplectic attack, the patient is kept on a liquid and semi-liquid diet (farinaceous soups, purées, compôtes, pap, etc.), and it is sought to procure regular stools without straining, by mild purgatives (cascara, purgen, pil. rhei, comp., pulv. glycyrrhiz., etc.).

A few days after the patient has regained consciousness very careful passive movements of the paralyzed limbs, with gentle massage of their muscles, is begun. Above everything, it is necessary to oppose, as far as possible, the formation of contractures and faulty positions of the extremities; on this account we should see to it that the foot is kept continuously in a position at right angles to the leg, and that the arm and the fingers lie as far as possible stretched out (see Lecture III, page 54). Only after 2 or 3 weeks should electric treatment be begun, also (and apart from faradization of the anesthetic skin regions with the wire brush), exclusively, galvanism. Stimulating the muscles by the faradic current is contraindicated. By the comparison of otherwise analogous cases of cerebral paralyses which have been treated by faradism, by galvanism, and without any electro-therapy, one may at any time convince himself that the faradization of the muscles can favor the development of hemiplegic contractures. The constant current appears, on the other hand, when it is used particularly to stimulate the extensor muscles in the upper, the peronei and flexors of the knee in the lower extremity, not only to encourage voluntary motor innervation (to open up new paths—"bahnen"), but directly to oppose the occurrence of contractions.

When motion has returned again in the leg, the patient can be gotten out of bed, at first for a very short period. For a number of weeks, however, he should spend the greater part of the day in bed, as too early and too frequent attempts at walking usually favor the development of contractures. Only very gradually should long remaining up and going about be permitted. The arm should, at the start, be kept in a sling in order to prevent swelling and cyanosis of the dependent hand as well as dragging upon the joint, which last can favor the development of arthropathies. Now, salt baths of gradually increasing concentration may be given, provided that they are neither too cold nor too hot—34° to 35° C. (93° to 95° F.). During the bath, cold compresses are applied to the patient's head to avoid congestion. In the bath, systematic exercises, never pushed to fatigue, are undertaken; soon, also, outside the bath. In these less stress is to be laid upon the development of strength than upon the practicing familiar acts (eating, buttoning and unbuttoning the clothing, writing, etc.).

But rarely orthopedic after-treatment of hemiplegic paralyses and contractures comes into question. At most, it is a matter of supportive apparatus

with arrangements for extension; there is scarcely ever any indication for bloody interventions, as tenotomy, tendon transplantation, etc.

(Supplementary.) Atypical and Extracapsular Hemiplegias

Gentlemen: It is necessary now to call your attention to some varieties of cerebral hemiplegia, which are separated more or less widely, pathologico-anatomically and semiologically from the typical capsular hemiplegia with whose consideration we have commenced this lecture. If in this task I do not strictly confine myself to cerebral hemorrhages and cerebral softening, nevertheless this bringing in of other pathologico-anatomical conditions is amply justified from the point of view of differential diagnosis.

1. *Cortical Hemiplegia.*—This depends chiefly upon embolic or thrombotic processes, though it can also occur from tumors, diseases of the meninges and the bones of the skull. It is, on account of the great extent of the cortical motor zone, usually incomplete, that is, sparing some portions of the limbs. When it begins in an apoplectiform manner it usually causes conjugate deviation (see page 262). The combination with cortical aphasia (see Lecture XVIII) is very frequent. The so-called "intra-cortical" hemiplegia presents a variety of cortical hemiplegia. *Spielmeyer* has shown, namely, that chronic diseases and resulting atrophies of the cerebral cortex (for instance, in epileptics) entirely isolate the (for the rest intact) pyramidal tracts from the other cells and cell associations of the cortex, and can hence paralyze them. Intra-cortical hemiplegias arise naturally, never in an apoplectiform manner, but are always chronic and progressive.

2. *Peduncular Hemiplegia.*—Hemorrhage into one crus cerebri (into the crusta or ventral portion) produces (as do also tumors or aneurisms of the same region) the so-called *Weber's* symptom-complex, or alternating oculo-motor hemiplegia. In this, there is found on the side of the lesion, a paralysis of the oculo-motor; on the opposite side, however, a paralysis of the face and the extremities. This occurs from the fact that the root fibers of the third nerve pass close to the pyramidal bundles which are proceeding distally in the ventral portion of the crus, but do not cross until lower down, and are destroyed in common with these latter.

3. *Pontine Hemiplegia.*—If the lesion is situated a little farther distally, namely in the lower third of the pons, there results from this another alternating hemiplegia, the so-called "*Millard-Gubler* symptom-complex" (alternating facial hemiplegia). On the side of the lesion the facial, on the opposite side the limbs, are paralyzed. The explanation is furnished by the following anatomical relations: the pyramidal fibers intended for the facial undergo decussation in the middle third of the pons, while the rest of the pyramidal fibers only cross the middle line distally from the pons. A lesion in the lower third of the pons varolii hence affects, along with fibers for the contralateral extremities, those intended for the facial muscles of the same side.

4. *Hemiplegia Cruciata*, an excessive rarity, can be produced by hemorrhages into the medulla oblongata, when these are located laterally at the pyramidal crossing through which the tracts for the arm are affected before,

those for the leg after, passing over to the opposite side. The paralysis then affects the contralateral arm and the homolateral leg.

5. "*Lacunar Hemiplegia*" has already been mentioned in Lecture XV, when discussing arteriosclerotic brain diseases. This form of hemiplegia is characterized, as was there emphasized, first by great capacity for rapid restitution and slight tendency to the formation of contractures, on the other hand, however, by a manifest tendency to new attacks in the originally unaffected hemisphere. In the last instance paretic spastic disturbances occur then, also in the bilaterally innervated muscles (see above, page 258), which manifest themselves in pseudo-bulbar phenomena. Sensibility and skin reflexes are scarcely disturbed in this form; also, when it begins suddenly, the ictus is not severe, consciousness is usually preserved or but slightly clouded, eventually loss of consciousness, but of only very short duration.

6. *The So-called "Hemiplegia Sine Materia."*—Cerebral hemiplegias have now and then occurred without any lesion in the central nervous system being discoverable at the autopsy. Many times it has been in nephritics who died in uremia. The uremic poison seems here for some reason entirely obscure to us, to have acted in producing one-sided paralysis. The older authors thought of one-sided vascular spasm and spoke of "serous apoplexy." Other cases of so-called hemiplegia sine materia exposed in the older literature, however, are probably to be considered as unrecognized lacunar hemiplegias.

7. *The So-called "Homolateral Hemiplegia."*—There are found in the literature some observations in which the disease focus has been found, not on the opposite side, but upon the same side as the hemiplegic paralysis. Usually there have of course been mistakes in clinical or pathologico-anatomical observation: so, namely after embolism, as already said, spasmodic movements may occur in the paralyzed extremities immediately after the ictus; these may be wrongly interpreted as signs of voluntary muscular power, while the contralateral extremities, in consequence of the coma, lie motionless and are considered to be paralyzed. Further, on autopsy a gross lesion of one hemisphere, which, however, does not involve the motor tracts, may so fix the attention of the examiner that he overlooks inconspicuous foci located in the pons or medulla of the other side. However, after eliminating these cases erroneously designated as homolateral hemiplegia, there still remain a few unassailable observations: on several occasions an absence of the pyramidal decussation, a relatively rare fiber anomaly, has been found in such cases.

LECTURE XVIII

Aphasia, Apraxia and Agnosia

Gentlemen: In our lecture to-day we will occupy ourselves with the exceedingly instructive and interesting disturbances, which can appear in different brain diseases in consequence of the more or less elective destruction of a definite "memory." Where it is a "memory" necessary for the speech arrangement, we see an aphasia result from its elimination; if there are disturbances of associated memories, affecting movements adapted to an end but not in the service of speech, the clinical picture of apraxia occurs; if, finally, the recognition of objects of any sort is interfered with or rendered impossible, we speak of agnosia. In each of these phenomena, however, we have to construct a large number of symptomatological sub-groups, which, for the determination of the seat and the nature of the underlying pathological processes are in part of decisive importance.

A. Aphasia

Already, in 1825, the French clinician *Bouillaud* emphasized the difference which existed between a paralysis of the speech mechanisms and another condition in which these muscles had suffered loss, not in their power to act in itself, but only in their applicability to the service of forming words. Eleven years later *Marc Dax*, based upon the frequent coincidence of such a loss of speech with right hemiplegia, placed the seat of "word memory" ("mémoire verbale") in the left hemisphere. The anatomical proof of the general correctness of this view was furnished first by *P. Broca* (1861-1865), in that he showed that the motor speech center was to be sought in the foot of the third lower frontal convolution on the left side. Further, he recognized exceptional cases with right-sided location of this center. Soon after, observations on a different sort of aphasia, in which the lesions occurred in the left temporal lobe and which were characterized not by the impossibility of forming words, but by substitutions, leaving out and mutilation of words ("paraphasia") and jargon, were made known. The English neurologist *Charlton Bastian*, laid stress on the fact that these patients also presented disturbances in the understanding of words, in that they perceived what was spoken to them only as noise, but could not understand it as speech. Finally, in 1874, *Wernicke* proved that this last phenomenon, which he called "sensory aphasia," occurred through the destruction of a "center for the memory of sounds" in the superior left temporal convolution, and that the accompanying paraphasic phenomena are explainable from the loss of the regulating influence which this "*Wernicke's* zone" (as we call it to-day), exercises upon *Broca's* convolution.

Newer investigations make it exceedingly probable that *Broca's* center is not limited to the foot of the third frontal convolution, but extends to the neighboring parts of the Island of *Reil*, the second frontal convolution and the precentral gyrus. Further, *v. Monakow* has shown that *Broca's* center cannot be considered simply as a depot for "kinesthetic" memory pictures* for the synergies necessary for speech. The Zurich neurologist demands with justice, a greater consideration of the dynamic factor along with the anatomical one, in the valuation of facts in the aphasia question. In this he stands upon the ground of the so-called "Diaschisis theory." This last assumes that when any part of the cortex of the brain is eliminated, not only this itself is inhibited in its function, but also a sort of shock-like inhibition is exerted upon other cortical areas connected with it. This shock is, however, not to be considered as active; rather does the absence of stimuli from the destroyed center simply affect a passive paralysis of those cortical regions which have regulated themselves in their activity by these stimuli, but which, through the new conditions, have suddenly been put out of setting. This "Diaschisis" action is in its essence of temporary nature. In the extended *Broca's* region lies now a place of production of motor aphasia, in that, this part of the cortex acts as director over other cortical regions, which probably distributed through large portions of both hemispheres affect the proper carrying out of the acts of speech. Hence, lesions of *Broca's* zone alone (chiefly traumatic) produce motor aphasia only temporarily and in the restitution of the most important speech functions in such cases, the passing away of diaschisis is to be perceived—the cortical apparatus coming into question learns in time to work even without the regulation of *Broca's* center. Before *v. Monakow*, the disappearance of aphasic phenomena was explained in general by the vicarious action of symmetrical parts of the other hemisphere (to a certain extent, "reserve centers"); his views appear to me, however, to indicate an important progress. This last can in no way be said of the attempt of *P. Marie* to deny to *Broca's* zone any significance in the production of motor aphasia.

In the rare cases of *Broca's* aphasia in which the autopsy disclosed no disease of the foot of the third frontal convolution, there may have been very fine alterations, for example, senile cortical atrophies, only to be discovered microscopically, if not purely functional aphasias; there is, for example, a hysterical aphasia studied by *Marinesco* among others. As to the still rarer observations in which clinically there was no aphasia but *Broca's* cortical area on autopsy is found destroyed, we by no means need to overturn the whole *Broca-Wernicke* localization theory on this account. The following reflection suffices: The left-handed person has his speech sphere as *Broca* emphasized, on the right side. Nevertheless, he has seen left-handed people also become motor-aphasic when the left lowest frontal convolution was destroyed. Even so, is the left-sided cortical localization of the psychical speech mechanism in right-handed people a rule, which does not exclude certain exceptions, and on this account it is no wonder that right-handed people sometimes have not be-

* That is, memory pictures, which render possible the repetition of formerly carried out analogous movements.

come aphasic after destruction of the left lowest frontal convolution;—they had had, probably, their speech regulating centers exceptionally developed on the right, since in general an originally bilateral plan for this cortical sphere can be assumed with the greatest probability. In destruction of *Broca's* convolution there may have been aphasia, which later, however, disappeared, so that the person making the autopsy if he has no accurate information as to the past history, registers a lesion in the third frontal convolution in a patient not aphasic.

THE PATHOLOGICAL PHYSIOLOGY OF THE APHASIC PHENOMENA

I beg you now to carefully observe Fig. 82. You will note in it the following:

The center for motor aphasia is connected not only with the cortical centers for the lip, tongue, and larynx muscles; that for sensory aphasia not only

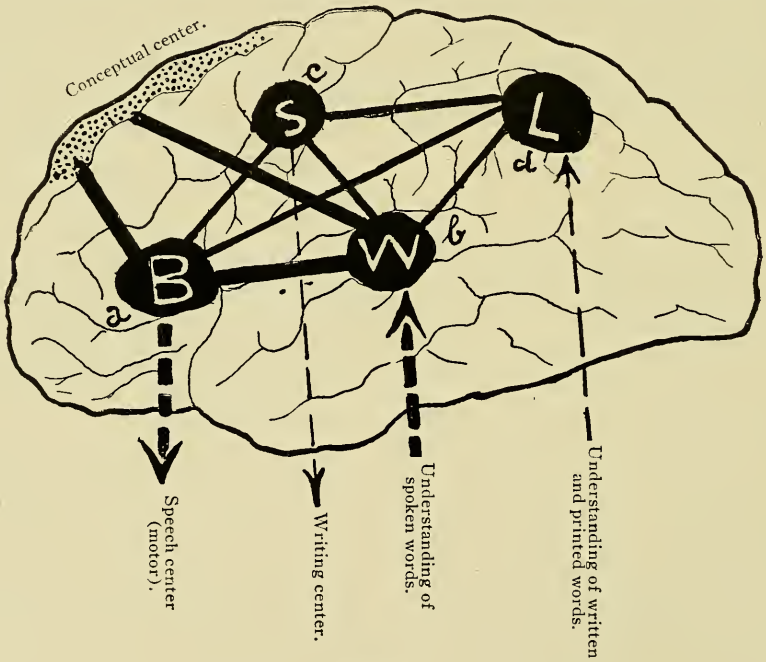


Fig. 82.

The Cortical Speech Centers and Their Connections.

B = *Broca's* Center. W = *Wernicke's* Center. L = Reading Center. S = Writing Center. a = Third (lower) Frontal Convolution. b = First (upper) Temporal Convolution. c = Anterior Central Convolution. d = Angular Gyrus.

with the auditory cortical zone, but these two cortical regions communicate by association fibers, 1, with one another, 2, with the higher psychical centers of the frontal region (the so-called "conceptual center"), 3, with the cortical apparatus which render writing and reading possible. The left angular gyrus

presents a reading center proper, the memory field for the recognition of letters, whose destruction produces alexia, word blindness. The writing center, on the other hand, contrary to the former view, coincides with the "hand and finger" center of the anterior central gyrus.

The network of association fibers indicated schematically in our figure comes into consideration for those psychical functions which we can designate shortly as "internal speech." Under this designation we understand everything which must go on beneath the threshold of consciousness in our brains, before we translate a thought into words and project these words outward by way of mouth or writing, or before we can take other cognizance of spoken or written expressions. This internal speech develops ontogenetically, in that the

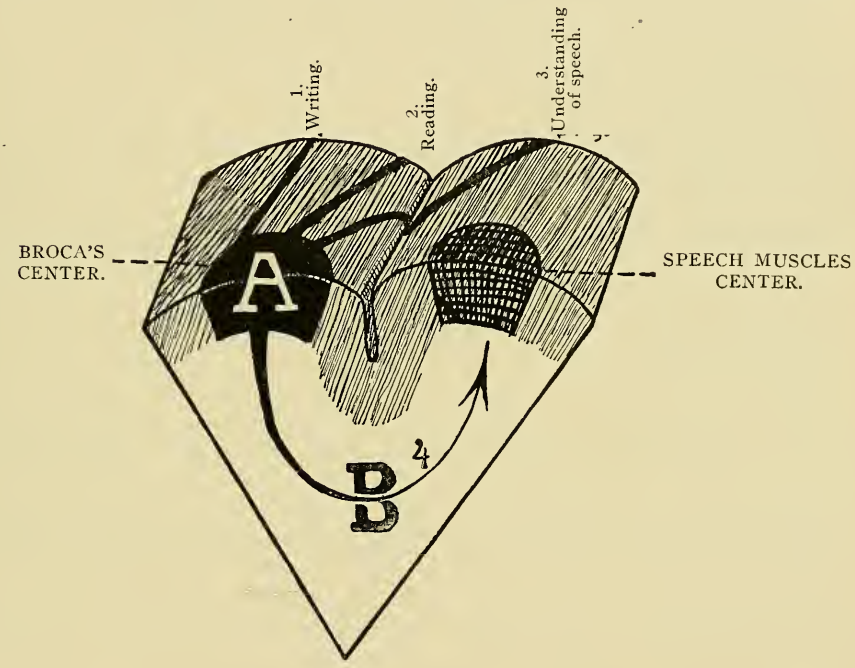


Fig. 83.

Seat of a Cortical Motor Aphasia (A, Blocks 1, 2, 3, and 4) and of a Sub-Cortical Motor Aphasia (B, Blocks only 4).

child first repeats words heard, then connects a conception with them, then upon the emergence of this conception reproduces the words himself, later, on learning to write and read, connects definite symbols with the individual sounds, etc.

Of decisive importance for the understanding of the aphasia question, is now the fact, that all the association fibers in the service of internal speech run through the brain cortex, while those neurones serving external speech make their way through the medullary substance. In Fig. 83 you see this schematically indicated, and learn from it that a cortical lesion of *Broca's* center (A) must produce more symptoms than a subcortical one (B). By

the first, the association net-work for internal speech is broken, by the last, however, in no way touched, so that subcortical motor aphasia indicates a pure aphasia an exclusive suppression of external speech, a simple "word dumbness." *Mutatis mutandis* in the same manner cortical sensory aphasia differentiates itself from the subcortical pure "word deafness."

When now, we turn to the study of the individual aphasic symptom-complexes we will proceed according to the following plan:

1. *The four chief forms of Aphasia.*
   (a) Cortical motor aphasia, *Broca's* aphasia.
   (b) Subcortical motor aphasia, pure word dumbness.
   (c) Cortical sensory aphasia, *Wernicke's* aphasia.
   (d) Subcortical sensory aphasia, pure word deafness.
2. *Other forms of Aphasia.*
   (a) Total aphasia.
   (b) Conduction aphasia.
   (c) Transcortical aphasias.
   (d) Aphasias of "single senses."
3. *Other symptom-complexes resembling the aphasic phenomena.*
   (a) Alexia.
   (b) Agraphia.
   (c) Amusia.
   (d) Amimia.

CORTICAL MOTOR APHASIA, BROCA'S APHASIA

Through the destruction of the motor cortical zone of *Broca*, it becomes impossible for the patient to translate conceptions into words. Indeed, such a patient can emit sounds and his speech muscles are not paralyzed like those of one with anarthria as a result of bulbar paralysis; still their regulated co-operation necessary for speech has become impossible. The French language can express the difference between this expressive speech disturbance of the motor aphasic and the articulatory speech disturbance of the anarthric in much shorter and more complete fashion than the German: "l'aphasique ne sait plus parler, l'anarthrique ne peut plus parler" (the aphasic does not know how to speak any longer, the anarthric cannot speak any longer). There are graduated differences, hence, a quite considerable variety of clinical pictures occurs. Either the patient has entirely lost the power of speech, has retained a few stereotyped expressions, or, in slight cases, there has remained to him a rudimentary ability of emitting speech, so that, for example, he uses all verbs in the infinitive ("negro fashion") or even leaves them out ("dispatch style").

Along with the loss or the disturbance of speech, writing is also disturbed to a corresponding degree. Ability to copy can, however, be retained, as a tracing of the letters which is accomplished without the assistance of memory pictures. Reading, on the other hand, is always more or less impaired and often

special methods are needed to make evident this disturbance. So, for example, printed words can be understood, written ones, on the other hand, cannot; or the understanding of words may be suspended if (with the help of the well-known child's block alphabet) the syllables are separated and arranged at horizontal or vertical intervals. The understanding of speech appears in a general manner undisturbed; when, however, one speaks very quickly to the patient, or uses a complicated sentence in the conversation, one can make out an increased difficulty of understanding. Though, however, the disturbances of speaking and writing stand out most prominently, nevertheless the destruction of a nodal point in the cortical association net has also not left reading and understanding entirely unmolested. To have indicated this last point was namely, the service of my teacher, *Dejerine.*

SUBCORTICAL MOTOR-APHASIA, PURE WORD DUMBNESS

Here, as is visible in Fig. 82, the path between *Broca's* center and that for the speech muscles has been destroyed. Internal speech, reading, writing, understanding of speech, are intact. Though the patient cannot bring out the words in contradistinction to patients with *Broca's* aphasia, he has preserved the corresponding motor memory pictures; he can even with his fingers give the number of syllables in the expression which he endeavors in vain to emit. (*Dejerine-Lichtheim* phenomenon.)

Pure subcortical motor aphasia has now certain relationships with those conditions with which we have become acquainted as anarthria, or dysarthria (Lectures VII and XV). Common to both symptom-complexes is the retention of internal speech; while, however, in anarthria the formation of sounds is disturbed, we find in pure motor aphasia that the formation of the word from the individual sounds is impaired. Of these sounds, often a large number can be emitted explosively as the so-called "word débris." For the inexperienced, confusion may easily arise between these two conditions.

CORTICAL SENSORY APHASIA, WERNICKE'S APHASIA

As already said, the sound pictures rendering possible the understanding of speech, are preserved in the posterior sensory or *Wernicke's* speech center of the left superior temporal convolution. Destruction of this center causes, hence, no loss of the power of speaking, but of the understanding of speech, no expressive, but a perceptive disturbance. In this, the patient, indeed, hears the words spoken to him, but cannot comprehend their sense, since they no longer rouse into consciousness the conceptions corresponding to them. His native language sounds to him as a foreign language, of which he knows nothing, or has learned but little, does to the healthy person. That sensory aphasia is often accompanied by paraphasia, that such patients continually make mistakes in speaking and instead of the words which are proper, emit other, often similarly sounding words, is connected with the following: We unconsciously first pronounce internally the word which we wish to speak by means of *Wernicke's* sound picture center. When, now, this no longer functionates, there

is easily the emission of wrong words, which the patient again cannot notice, since in consequence of his sensory aphasia, he does not understand his own words.

Where the understanding of speech is not entirely lost but considerably affected, it is particularly the familiar expressions (such as: "How are you?" "What is your name?" etc.) whose sense the patient is still able to comprehend. Sometimes the patient also guesses the sense of the sentence from one characteristic word which he understands. If now, we vary the sense of the sentence while retaining this characteristic word, a corresponding modification of the answers of the patient does not occur. For example, "Have you children?" "Yes." "How many children?" "Yes." "Where are your children?" "Yes."

In contradistinction to the motor aphasic who speaks little or not at all, the patient with *Wernicke's* aphasia usually talks a great deal, but badly. We denominate his paraphasia verbal when he confuses words (for instance, says "dog" instead of "bed"); as literal when he confuses letters (for example, "winnow" instead of "window"), while we speak of "Jargon aphasia" when he brings out a succession of senseless syllables. In this last case there is sometimes a clinging to one syllable which is repeated over and over—"perseveration."

The understanding of writing is also suspended in cortical sensory aphasia or at least impaired. In complete "word blindness" the patient can no longer read, and the letters are for him only senseless black figures upon a white ground, which, however, he may trace without understanding. Writing voluntarily or after dictation is, on the other hand, lost or impaired (this last in the form of literal or verbal paragraphia). Other symbols besides letters such patients may still understand, for example, very often numbers, and further, they can play cards and dominoes. A patient of *Dejerine's* who could not read the letters R. F., said instantly "République Française," as soon as the coat of arms was drawn about the two letters.

SUBCORTICAL SENSORY APHASIA—PURE WORD DEAFNESS

In this very rare form, of which I have so far been able to observe no case, and a knowledge of which we owe, among others, to *Dejerine*, *Wernicke*, *Liepmann* and *Sachs*, internal speech is intact, reading and writing may be possible without disturbance and only the understanding of speech is suspended. Besides this, paraphasia is absent in pure word deafness since the sensory cortical center is still able to exert its stimulating and controlling action upon the motor center. This form occurs only through the cutting off of the otherwise intact *Wernicke's* zone from the stimuli coming from the general auditory center.

I might here introduce an anatomical point not unimportant for localization. It has indeed been proved by autopsy that a subcortical lesion of the left temporal lobe can be the substratum of pure word deafness. Still, incomplete destruction of the sensory speech center can probably also produce pure word deafness, just as occasionally incomplete disturbances of the motor

speech center can cause pure word dumbness. I assume that in such cases the association fibers between the cortical field of the cochlear nerves and *Wernicke's* center, or respectively between *Broca's* convolution and the speech muscle center, are interrupted intracortically, without on this account the function of the speech centers under consideration having been destroyed.

TOTAL APHASIA

We speak of total aphasia when a pathological process has destroyed the anterior as well as the posterior speech center and on this account ability to speak as well understanding of speech, and also ability to read and to write are destroyed.

CONDUCTION APHASIA

This designation *Wernicke* has introduced for certain speech disturbances which are characterized by paraphasia, paragraphia, and impairment of speech repetition. He assumes that here there is interruption of the direct communication between the sensory and the motor speech centers, in which particularly a lesion in the Island of *Reil* would come into consideration. This clinico-anatomical hypothesis, however, has been proven untenable by *v. Monakow.*

TRANSCORTICAL APHASIAS

According to *Lichtheim* these arise by the blocking of the paths between the conceptional center on the one hand and *Broca's* or *Wernicke's* center on the other. Hence, a motor and a sensory variety are differentiated. In the first only voluntary speech and writing are suspended or impaired; speech repetition, reading aloud, writing from dictation are, on the other hand, retained. In the latter there is more or less pronounced word deafness and alexia along with paraphasia and paragraphia, but speech repetition, reading aloud and writing from dictation, may be possible, although without understanding. With *Dejerine,* I consider these forms only as intermediate stages of cortical aphasias which are improving. As an "attenuated form of transcortical motor aphasia" *Liepmann* has designated the so-called amnestic aphasia or verbal amnesia, in which finding the word has become very difficult, the suggested word, however, is at once recognized as correct and is repeated smoothly and perfectly. Further, chiefly substantives and verbs for the concrete are not found, while the forms of speech and further inflection and declension are preserved.

APHASIAS OF SINGLE SENSES

Best known is the optic aphasia of *Freud;* for objects simply held before the patient he cannot find a name, which, however, promptly occurs to him when he is given an opportunity to observe the object through some other sense than that of sight, for example, to feel a spoon, to hear a bell, to taste a piece of sugar. At the basis of this disturbance lies an interruption of the

connection between the centers for the recognition of objects in the occipital lobe and *Wernicke's* sound memory center in the temporal lobe. The existence of an optic aphasia in the strict sense of the word is, however, according to *Gustav Wolff*, not definitely proved—neither are the other analogous single sense aphasias (auditory, tactile aphasia). As a matter of fact, many of these observations have entirely lacked certain proof that the patients actually had recognized the seen, touched, heard things, and only could not find their names, also that they were free from mind blindness, mind deafness and touch agnosia. (See below.)

ALEXIA

This disturbance leads in slight cases to blindness for letters, in severe ones to word blindness, and can, apart from accompanying cortical aphasias, appear more or less independently. If a focus is seated in the reading center of the left angular gyrus, the alexia is accompanied by agraphia, since the association fibers to the center for writing movements in the precentral convolution are interrupted. We speak then of "cortical alexia." If, on the other hand, the focus is subcortical, beneath the angular gyrus, so that only to the optic word pictures, which come from the visual cortex in the cuneus, the entrance is closed, there occurs "pure alexia." Also, conditions which *mutatis mutandis* correspond to those which are present in cortical and subcortical aphasias.

AGRAPHIA

Disturbance of the writing center, shown in Fig. 82, causes no isolated agraphia, but a paralysis of the right hand. With the left, however, such a patient may be able to write or to learn quickly to do so, provided that his *Broca's*, *Wernicke's* and reading centers have remained intact. Isolated agraphia we will indeed become acquainted with later as a component of apraxia.

AMUSIA

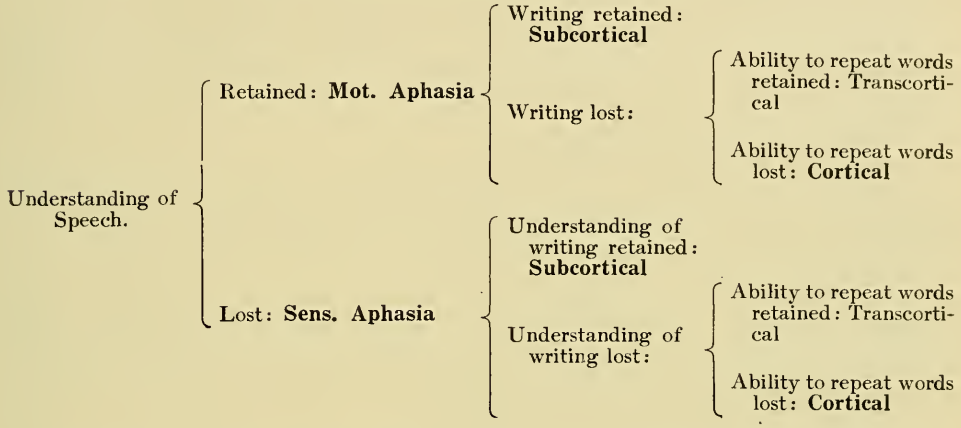
As motor amusia the inability to sing, as sensory, the inability to appreciate a melody, is designated. Also musical alexia or blindness for notes is usually herein included. The amusic disturbances are rare accompaniments of the corresponding forms of aphasia. The musical centers appear to be situated very close to the corresponding speech centers, but do not entirely coincide with them.

AMIMIA

The condition that in the patient also expression and gesture speech is lost (motor amimia) and even understanding for this has disappeared is found in total aphasia. If in this manner all possibility for comprehending one's fellows is excluded, we speak of asemia or asymbolia.

EXAMINATION FOR APHASIA AND DIAGNOSIS OF THE INDIVIDUAL FORMS

When we have before us an aphasic patient we must bend our endeavors toward making as rapid a diagnosis as possible of the form of aphasia present. After that, we will take up its symptomatological study. I would recommend that you proceed according to the following scheme which I have prepared after the manner of the botanical determinative books.*



In general it is, especially for the beginner, of advantage in the thorough examination of an aphasic to stick to one of the schemata proposed by different investigators and to carefully register the result of the examination. I give you here for this purpose the *Stewart* examination schema somewhat modified by myself, which you will find useful in all cases:

1. Can the patient spontaneously give utterance to understandable words? How extensive is his vocabulary? Can he utter all words, or only some?

The patient is allowed to talk spontaneously and it is observed whether he speaks fluently, whether he misplaces words or syllables, whether he speaks in connected sentences, or talks unintelligible stuff.

2. Does he understand the words which he hears? He is asked to touch alternately his nose, his ear, his eye, his chin, etc., in order to test his understanding of substantives. He is then requested to whistle, to smile, to close his eyes, etc., by which his understanding for verbs is determined. It should be observed if he carries out the first order correctly, then, however, continually repeats the same act, even when requested to do something else. If there is no trouble in carrying out these simple orders, he is given more complicated sentences, for example, "I beg you, when I have come around to the other side of the bed, to touch my arm twice with your left hand."

3. Does he understand written questions or orders which are placed be-

* In this I ignore the problematical "conduction aphasia," and on account of its small practical importance I have put in parentheses what is said about the differential diagnosis of the so-called "transcortical" aphasia.

fore him? In testing this, simple sentences are first used (for example, "How old are you?" "Show me your tongue." "Give me your left hand") and only after this, more complicated constructions. Further, not only handwriting, but also print, is used and it is noticed whether separation of the individual syllables or their unusual arrangement disturbs this understanding of them (see page 277).

4. Can he write spontaneously?

If his right hand is paralyzed, he may try with his left hand. It is observed whether he scribbles senseless signs.

5. Can he transcribe printed letters into written ones, and vice versa?

For this test one of the so-called "letter games" with which short words like "Monday," or short sentences as "Where is the tree," are put together, or allowed to be put together, is used.

6. Can he write from dictation?

7. Can he find objects whose names he hears?

A heap of objects, like a key, a piece of money, a lead pencil, a match, etc., are placed before him and he is solicited to pick up one at a time.

8. Can he repeat words which he hears?

He is first tried with simple words and sentences, for example, "Cat," "Dog," "Sister," "Good morning," etc. If he has lost the power to repeat words spoken before him, it is ascertained whether the patient can sing, what is sung for him, which is sometimes the case.

9. Can he name objects which he sees?

Different objects are pointed to (see under 7) and he is asked "What is that?"

10. Can he read aloud?

11. Can the patient, in case it is impossible for him to pronounce a word, give the number of its syllables on his fingers?

12. Does he understand gestures and mimic movements?

Without speaking to him, he is solicited to imitate touching the nose spreading the fingers, sticking out the tongue, etc.

In order not to fatigue the patient these different tests are to be undertaken not all at one sitting, but with extensive periods of rest between. In polyglot patients it is well to test the different languages separately when possible. The same thing applies to dialect and to written speech.

PROGNOSIS OF APHASIA

The estimation of the etiological factors is very important in the first place (whether hemorrhage, embolism, tumor, trauma, etc.). Of decisive importance is the intellectual condition. The onset of dementia makes the hope of disappearance of the aphasic disturbances illusory. Particular emphasis, however, is to be attached to the prognostic differences as to restoration of function which appear to exist according to the form of the aphasia or according to the localization of its anatomical substratum. The best outlook is presented by pure word dumbness, while the severe cortical aphasias with complete loss of spoken and written speech present little prospect of improvement.

In sensory aphasias the prognosis is the worse the more intensely marked paraphasia and jargon aphasia are. Alexia and agraphia in improvement of *Wernicke's* aphasia, show themselves much more obstinate than word deafness.

Treatment

The exercise treatment of these conditions furnishes the greatest test of patience which comes to the physician. *Gutzmann* properly warns imperatively against beginning the exercises too early. We should wait at least half a year after the cessation of the stormy symptoms and the entire disappearance of all other disturbances. If exercise is commenced too early in apoplectics, there is the danger that during the exercises, in consequence of the exertion, a new hemorrhage may occur. *Gutzmann* usually first prescribes the exercises when, after one or two years, there is no further spontaneous improvement of speech. If they are applied earlier, it is very difficult to decide how much is owing to the exercise treatment. *Gutzmann* further lays the greatest stress upon slow progress, as only by this can the patient be kept in good spirits, which is absolutely necessary for the result of the exercise treatment. Sensory aphasics with speech impulsion must first practice holding their tongues, endeavoring to regain again to some extent the normal inhibition by the will. Even in the severe, prognostically unfavorable, *Broca's* aphasias, treatment should not be given up, since it is of great importance for the patient to give expression to his will, at least by a few words, even when they are used ungrammatically. By this, the lability of mood which not rarely becomes exaggerated to outbreaks of rage, is combated. There occurs, from the fact that the aphasic is not in a condition to give expression to his thoughts, wishes and conceptions, an internal tension, for which even a rudimentary ability to speak, gives an outlet. Also to this same end, such patients must systematically practice gesture speech. A picture book with simple representations of all the conceptions and wishes lying within the circle of thought of the patient, should always accompany him, so that he can make known his wishes to those surrounding him by indicative gestures, though when apraxia is present, the descriptive gestures cannot be learned.

B. Apraxia

As apraxia* we denominate, since the monumental work of the Berlin psychiatrist *Liepmann* which first appeared in 1900, a disturbance in which the extremities, especially the hands, are capable of executing correct single movements, but not those movements directed to a definite end. A patient affected with this disturbance has lost the memory of the synergies and combinations of single movements necessary for the proper use, for example, of a pair of scissors, for beckoning, for shaking hands, etc. He sticks a toothbrush like a cigar into his mouth, and so on. There are hence great analogies with motor aphasia, or to better express it, aphasia from a physiological point

* Other names, as Asymbolia, Parektropia, Parekinesia, are applied only by a few authors, and have not come into general use.

of view can be considered a special form of apraxia, as apraxia of the speech apparatus.

We distinguish three varieties of apractic disturbances:

1. IDEATORY APRAXIA

The idea-plan for the composite act to be carried out is disturbed, so that the resulting unsuitable action gives the impression of extreme mental distraction. *Liepmann* cities the following typical examples: The patient sticks a match along with his cigar into his mouth instead of striking it; in sealing a letter, puts the seal in the flame and then presses it on the stick of sealing wax; attempts to cut off the end of his cigar by wedging it between the match case and its cover. The single acts which make up these perverted maneuvers are in general correctly performed. This ideatory apraxia usually appears only in complicated movements and in proportion to their complexity. The limbs are, as a rule, equally affected; it is not that they respond incorrectly to the will, but they receive perverted impulses from it.

2. IDEO-MOTOR APRAXIA

In this form, both the plan of movement and also the limb center with its content of kinesthetic memory pictures are intact, but their connections are severed. Simple acts for whose accomplishment memories preserved in the limb centers suffice, are correctly executed; only those requiring a great number of successive movements are incorrectly accomplished, since the proper directions and commands are no longer carried from the ideation centers to the limb center which, to a certain extent, has become autonomous with regard to the whole brain. *Liepmann* calls these forms of apraxia the "ideo-kinetic," *Heilbronner*, the "transcortical." They can affect single limbs or one half of the body. According to *Liepmann* the following kinds of faulty reactions occur:

a. Movements which do not resemble any purposive movements, flourishing the hand, spreading the fingers, so-called amorphous movements.

b. Executing the wrong movements.* (Beckoning instead of threatening, taking hold of the ear instead of the nose, etc.)

c. The movement occurs in another set of muscles; standing stiff instead of giving the hand, which may simulate leaving out of movements.

d. Often motor helplessness and actual abandonment of movement appears.

In many cases of the faulty reactions there is decided perseveration (compare the analogous condition in aphasia, page 278).

3. MOTOR APRAXIA

In this, the motor innervation is disturbed by an injury to the limb center itself which is not sufficient to paralyze this, but enough to extinguish its

* Can also be called "Parapraxia."

kinesthetic memory pictures. On this account the movements of the extremity are carried out as if the patient was trying them for the first time, so the coarse movements are very awkward, the finer acts, however (writing, threading a needle, sewing), cannot be executed. *Liepmann* calls this variety of apraxia the "limb kinetic," *Heilbrunner*, the "cortical," *Kleist* speaks of "innervation apraxia."

The Diagnosis of Apraxia

As a scheme for examination I recommend to you that of *Dromard* and *Pascal*. The patient is solicited first by simple spoken orders, eventually by making before him the necessary gestures, to carry out the following categories of movements:

1. *Auto-kinetic movements:* Walking, standing up, sitting down, dressing and undressing himself.

2. *Simple movements:* Close the eyes, open the mouth, show the tongue, cross the arms, raise the right hand, stretch out the left hand, spread the fingers, raise the right leg, etc.

3. *Movements requiring reflection:* Show the right eye, touch the left ear, twist the beard, scratch the head, etc.

4. *Expressive movements:* Throw a kiss, "thumb the nose," make the military salute, cross himself, etc.

5. *Descriptive movements:* Show how one plays the piano, turn an organ, grind coffee, how to catch a fly, etc.

6. *Complicated movements:* Present a glass of water, tie a knot, light a candle, seal an envelope.

In order to diagnose apraxia it is necessary to exclude word deafness, that is, to determine that the patient understands the command; also agnosia, that is, to make sure that he correctly recognizes the objects, for instance, does not take the toothbrush for a cigar. Against confusion with atactic, choreic and athetoid movements one must naturally be on his guard.

In the diagnosis of the lesions upon which the apractic symptom-complexes depend, the following rules are applicable:

a. Ideatory apraxia occurs in diffuse processes (senile dementia, progressive paralysis, diffuse arteriosclerosis of the brain).

b. Motor (*"limb kinetic"*) *apraxia* depends, as already said, upon disease processes in the cortical centers of the extremity in question, which without being sufficient to paralyze them, nevertheless, destroy their kinesthetic memory pictures.

c. Ideo-motor (*ideo-kinetic*) *apraxia* occurs under two different conditions:

1. First in general when the sensori-motor cortex is retained in its integrity, indeed, but is cut off from its connection with other parts of the cortex, namely from the areas for word and object conception in the temporal and occipital lobes, hence, foci in the parietal lobe cause an apraxia of the opposite hand.

2. An ideo-motor apraxia of the left hand is also observed in destruction of the fibers of the corpus callosum in which its sensori-motor cortical field

in the right hemisphere remains intact, but it is deprived of its relations with the sensori-motor cortical field of the left hemisphere. The left hemisphere controls also (at least in right-handed people) the activity of the right.

SPECIAL FORMS OF APRAXIA

Along with the gross apractic symptom-complexes there occur also special apractic disturbances, of which the most important is apractic agraphia, which, in contradistinction to the aphasic form, can occur as an isolated phenomenon. I would mention further the observations preserved in the literature of apraxia of lid closing, apraxia of the head muscles, apractic disturbances in the finger speech of deaf mutes, and "instrumental amusia" in musicians; also amimia, already mentioned, may be thought of again in this connection.

C. Agnosia

If, in spite of undisturbed perception by the senses, the recognition of objects of the external world is impossible, we speak of agnosia. If we would extend this conception to the conventional phonetic or graphic speech symbols, we would have to denominate the phenomena of word deafness and alexia, already known to us as special instances of auditory and optic agnosia. This, however, contradicts the usual use of the word which, under "agnosia," understands only "object agnosia." We distinguish auditory, optic and tactile agnosia; there are also taste and smell agnosias, but they are so difficult to differentiate from ageusic and anosmic disturbances, and in any case so rare, that they are practically of no importance.

1. AUDITORY AGNOSIA: MIND DEAFNESS

In this (in consequence of disturbed associations between the areas for auditory sense perceptions and the "concept center") the identification of auditory impressions is suspended. With closed eyes such a patient is unable to recognize a watch by its ticking, a dog by his bark, a bunch of keys by their rattling, a stream of water by its splashing, etc. Mind deafness is an accompaniment of sensory aphasia in large foci in the left temporal lobe.

2. OPTIC AGNOSIA: MIND BLINDNESS

From the cortical end stations of the optic tract (in the so-called cuneus on the median surface of the occipital lobe), association fibers run to the lateral convex surface of the occipital lobe. If the cuneus is the perception center for the optic impressions, these parts of the cortex represent the optic conception centers. In them the memory pictures of the meaning of an object seen are laid up. Bilateral destruction of these cortical optic-memory fields (or of the association fibers passing to them from the cuneus) produces "mind blindness," in which the patients see the objects as flat or physical pictures,

but cannot recognize them. Where unilateral disease of the occipital lobe has produced optic agnosia it has been in the form of tumors which also must have injured the opposite occipital lobe through pressure. If we show a mind-blind person an object (spoon, pencil, bottle, key, hat, box, scissors, photograph, glove, etc.), he can tell its form; often also, its color; he can further pick out similar objects without difficulty; he has, however, lost the understanding of the use of the object.

3. TACTILE AGNOSIA

This form of agnosia occurs in disease foci of the posterior central convolution and the parts of the parietal lobe just behind it, when the patient is required to recognize objects by touching them with the opposite hand. The testing must particularly be carried out with each hand. Naturally, not only must the eyes of the person being investigated be blindfolded, but also characteristic noises (for example, the rattling of a bunch of keys) must be avoided. In diagnosing tactile agnosia it is further necessary to determine that the individual modalities of superficial and deep sensibility (movement, touch, pressure, cold, warm sensations, etc.) are not disturbed to such an extent as by this alone to render recognition illusory.

LECTURE XIX

Tumor Formations, Inflammations, and Disturbances of Circulation in the Brain and its Membranes

A. Brain Tumor

GENTLEMEN: When we speak of brain tumor in the clinical sense, we understand thereby not only neoplasms arising from the brain substance itself, but all intracranial tumor formations which act upon the brain; that is, also those which take their origin from the meninges and from the bones of the skull, and to these tumors we reckon along with the neoplasms proper, in a pathologico-anatomical sense; also tuberculous and syphilitic granulomata, as well as many parasites (as cysticercus and echinococcus), and, finally, other space-reducing processes within the skull; for example, aneurisms of the *Sylvian* artery or of the carotid, certain hematomata of the dura mater, also the so-called "serous meningitis," cannot be separated clinically from brain tumor.

In serous meningitis there is a great serous effusion into the subarachnoid space leading to symptoms of pressure upon the contents of the skull; that is, the analogue of serous pleurisy or peritonitis. It is often accompanied with increase of the ventricular fluid (acquired hydrocephalus), and develops partly in consequence of infectious diseases (for example, measles or pneumonia), partly upon a syphilitic basis, partly in connection with inflammatory diseases of the ear or nose, partly after injuries to the skull; often, however, its etiology cannot be made out. Along with the diffuse forms of serous meningitis there occur circumscribed encapsulated collections of fluid in the subarachnoid space, usually at some place upon the convexity of the brain; such observations have, for example, been reported by *Fedor Krause*, *Iselin*, and myself. Only exceptionally does serous meningitis run a course similar to that of purulent meningitis (see below), although much milder; usually, however, it gives the symptomatology of brain tumor, and hence furnishes the chief contingent of the so-called "pseudo-tumor cerebri." (Under the last classification would fall also a number of cases which furnish clinically the typical picture of a brain tumor, but on autopsy show neither a tumor nor meningeal effusion, and which have been conceived to be the result of a molecular "brain swelling."—*Reichardt* and others.)

Of tumors in the interior of the skull, glioma, gliosarcoma, and sarcoma of the brain occur most frequently; sarcomata often take their origin also from the meninges or from the connective tissue envelope of the nerve roots (particularly the facial and the auditory, so-called "tumors of the cerebello-

pontine angle"). Fibromata have also been observed here. From the inner surface of the dura arises endothelioma, which sometimes spreads out as a flat mass ("endotheliome en nappe"), sometimes, however, forms a spherical tumor; where this last, through the inclusion of calcareous material, presents a sandy cross section and very firm consistence, we speak of psammoma. Upon the clivus at the base of the skull through proliferation of cells of the chorda dorsalis, which are there included in the bone substance, chordoma arises, while osteoma and chondroma from other places in the skull bones can intrude into the cranial cavity. By epidermal germ dispersion in the interior of the skull, and indeed in the ventricles, arise cholesteatoma, dermoid cyst and teratoma. Very rare is lipoma, which can develop in the region of the corpus callosum; still rarer, epithelioma of the ventricular ependyma, of which I have observed one case. Relatively more frequent is adenoma of the hypophysis. The other epithelial tumors of the interior of the skull have scarcely ever arisen autochthonously—rather have been introduced by metastasis; this applies especially to the carcinomata. Among the infectious granulation tumors, solitary tubercle of the brain takes the first place, particularly in childhood. Not very rare is also the gumma, which most frequently arises from the membranes of the cerebral convexity. As a rarity, actinomycosis has been described. Finally, as to the parasitic cysts, thanks to the strict meat inspection, they rarely occur in our neighborhood any more; the cystic stage of the tænia solium (cysticercus) is, on the contrary, occasionally observed in the brain in the north of Europe, in the south the echinococcus ("hydatid cyst") somewhat more frequently. As far as they are not parasitic or infectious new growths, the etiology of brain tumors is naturally just as obscure to us as that of neoplasms in general; nevertheless, it is certain that among exciting causes skull trauma has strikingly often been found in the history. Men are more frequently affected than women.

Symptomatology

The morbid manifestations produced by brain tumors may be divided into general symptoms and focal symptoms. The first are to be interpreted predominantly as "brain pressure" phenomena; that is, produced by the narrowing of the space within the cranial cavity and the resulting compression of the brain, and hence, to a certain extent, independent of the seat of the tumor. The last, on the contrary, are directly connected with the special location of the disease process, and hence put us in a position to diagnose the seat of trouble, which is naturally of prime importance in the eventuality of surgical interference.

Among the different brain pressure phenomena, headache occupies the first place; this is usually characterized by terrible severity as soon as the space within the skull has been narrowed to a considerable degree. It can be continuous, but is usually described as remittent—exacerbating. A migraine-like headache, occurring paroxysmally and accompanied with vomiting, also occurs, especially in the early stages. Its character is usually grinding and boring, and it is usually felt all over the head. In those cases, how-

ever, in which it is localized in a definite region, this by no means necessarily corresponds to the seat of the tumor; so, indeed, in tumors of the posterior fossa of the skull there is often pain limited to the back of the neck and head; sometimes, however, on the contrary, frontal headache. Sensitiveness on percussion or to pressure, localized at some point on the skull, has great value, as it not rarely corresponds exactly with the disease focus below. One should, however, never be satisfied with the result of one examination, but should only use diagnostically such symptoms as have proved to be constant on long observation. A in no way frequent, but very important, symptom on skull percussion is further the finding over a definite region of the skull a "cracked pot" sound which seems to depend upon a lack of firmness in the skull sutures, and is most frequent in young patients.

Upon Roentgenological examination, a definite bulging and thinning of the bone in the part of the skull, corresponding to a tumor, is sometimes to be made out; on the other hand, there is sometimes thickening of the skull walls, especially in the region of such tumors as arise from the dura. Only a very small percentage of brain tumors manifest themselves in the X-ray picture, by destruction of the skull wall; most frequently hypophyseal tumors which erode the sella turcica. A particularly interesting brain pressure symptom is dilatation of the veins of the diploe (*Schüller*). Scarcely recognizable in the normal X-ray picture, in brain tumors, they are so distended that they appear as wide stripes on the plate. This dilatation can be plainest in the neighborhood of tumors.

"Cerebral vomiting" is a very frequent brain pressure symptom. It appears either very suddenly without any warning, explosively, or a short period of nausea, or an attack of vertigo, precedes it; in any case, however, it is independent of taking food. For the rest, vertiginous phenomena which are to be considered as general brain pressure symptoms are much less frequent in intracranial tumors than those locally caused; that is, by direct action upon the cerebellum or the vestibular apparatus (compare Lecture XX). Only in the last instance does rotary vertigo proper, eventually with constant direction of the turning, usually occur, while in brain tumors located elsewhere, diffuse and less characteristic disturbances of the space consciousness are more frequent.

Of the greatest value for the diagnosis of the space-reducing brain tumors is "papilloedema," whose pathogenesis has been made plain to us by the ophthalmologists *Schmidt* and *Manz*. The optic nerve possesses a connective tissue sheath whose cavity communicates with the subarachnoid space of the brain. In increasing brain pressure the cerebrospinal fluid consequently presses against the "lamina cribrosa" of the optic nerve head which is pushed forward and rendered œdematous. In this, also, there is strangulation of the central vein of the retina, which favors stasis and serous infiltration. Fig. 84 shows you the ophthalmoscopic picture of such a papilloedema in a case of cerebellar sarcoma. The papilla loses its sharp outline and appears enlarged; it protrudes knoblike toward the vitreous, and is occupied by a fine radial streaking which extends into the neighboring retina; its color is gray-white, the "physiological excavation" is not sharp, the vessels are in places "veiled"

by the œdema, and altered in their caliber, the veins dilated, the arteries narrowed; the fine dots of the lamina cribrosa are invisible. If the brain pressure disappears, even a high degree of papilloedema may entirely pass away; its long continuance, however, leads to an atrophy of the optic nerve which is incurable.

The influence of increased intracranial pressure upon the psychical condition of the patient expresses itself in the more advanced stages of brain tumor by stupor; the psyche usually remains intact only in certain basal tumors or in those of the cerebellar region. Slight confusion manifests itself

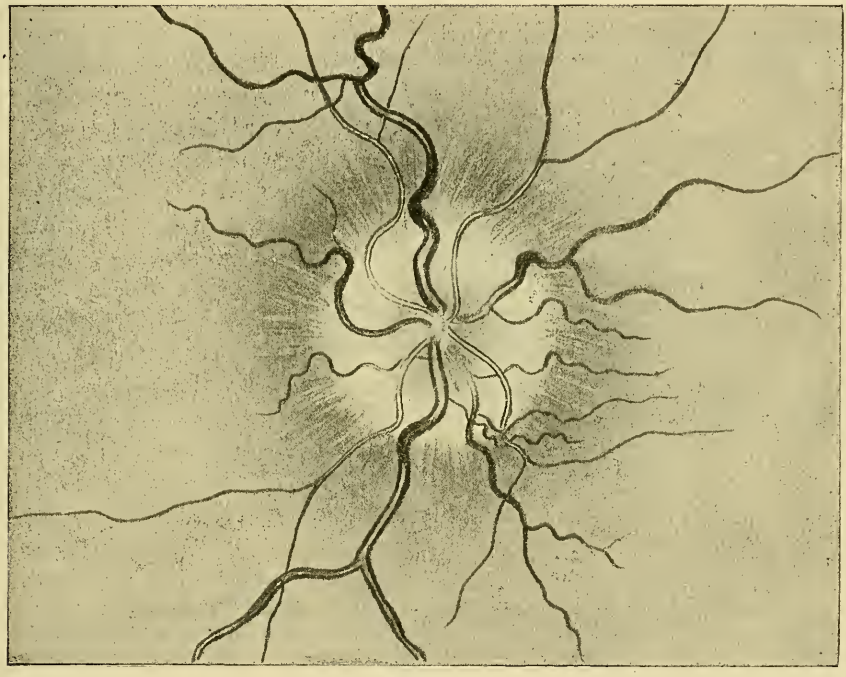


Fig. 84.

Papilloedema. Sarcoma of the Left Cerebellar Hemisphere.

in that the patients, when questioned, must visibly "pull themselves together" in order to answer, make a somnolent impression on speaking, and left to themselves, are entirely apathetic with regard to the happenings in the external world. Higher degrees of confusion are accompanied by somnolence, so that the patient, even when taking nourishment, can scarcely be kept awake; in the late stages this torpor passes finally into complete coma, with involuntary discharge of fæces and urine. Along with these more or less permanent disturbances of consciousness, paroxysmal attacks of loss of consciousness also occur. These sometimes take on an epileptiform character through accompanying spasms, or if the loss of consciousness is not complete, a hysteriform.

Anomalies of pulse and respiration in brain tumors usually only appear in advanced stages. As "pressure pulse," is denominated a pulse slowed to about 45 to 55 beats a minute, but full and tense; only exceptionally has still

more marked bradycardia (even 20 beats to the minute) been observed. In the terminal stage, in the place of bradycardia, a tachycardia with arhythmia ("paralytic pulse") occurs. That in tumors which are located in the neighborhood of the bulbar vagus centers, the pulse anomalies appear earlier is easily understandable. The same remark applies to respiratory disturbances which in general also appear as late symptoms only; slowing and deepening of the breathing, finally the so-called "*Cheyne-Stokes*" respiration. This is, as we know, a type of breathing characterized by alternating periods of

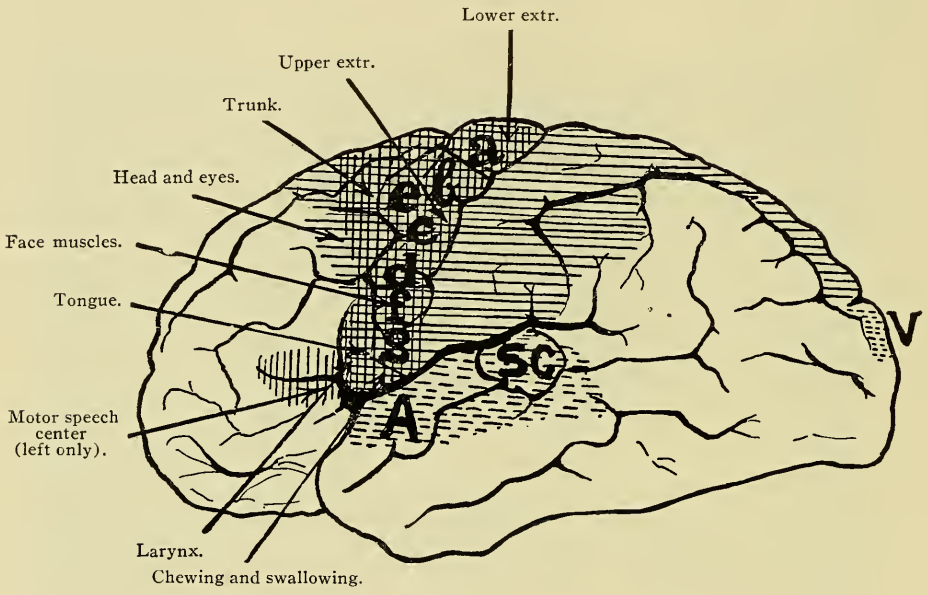


Fig. 85.

Motor Sensible and Sensory Cortical Fields.

A = Auditory.
V = Visual Cortical Field.
S C = Sensory Speech Center (only Left).
a = Center for the Foot.
b = Elbow.
c = Hand.
d = Finger.
e = Shoulder.
f = Upper Facial.
g = Lower Facial.

deepness and shallowness of the breathing with regular interpolation of an apnœic stage, which occurs also in other disease conditions (namely in uremia).

A direct measurement of the brain pressure is permitted us by lumbar puncture (see above, page 253), in which a water manometer attached to a cannula mounts higher than normally. If namely in healthy people the manometric pressure measured while lying down is usually from 40 to 150 mm water, in cerebral tumor it usually runs from 250 mm to 900 mm. We can generally, however, dispense with exact measurements and satisfy ourselves with determining that in intracranial hypertension the cerebrospinal fluid, instead of being discharged in drops, spurts from the cannula in a stream as soon as we withdraw the mandrin. Absolutely to be avoided is lumbar puncture in all cases in which there is a possibility of a tumor being located in the posterior fossa of the skull; since in this eventuality sudden death from

the effect of rapid decompression upon the vital centers of the medulla is risked. Also in tumors located in the frontal lobe, as soon as the raise of pressure within the skull has reached a great height, lumbar puncture is not without danger. Its diagnostic application is hence to be limited as much as possible in brain tumors. This last remark applies with even more force to brain puncture after *Neisser* and *Pollack*. As *de Quervain* very properly emphasizes, this measure is too dangerous and in its results too uncertain to be properly reckoned among the diagnostic aids of the physician; on the other hand, it can occasionally be a valuable aid to the surgeon when everything is prepared for operation.

Now as to the focal symptoms of brain tumor, the accurate determination of the seat of the tumor, as deduced from the knowledge of them, is often

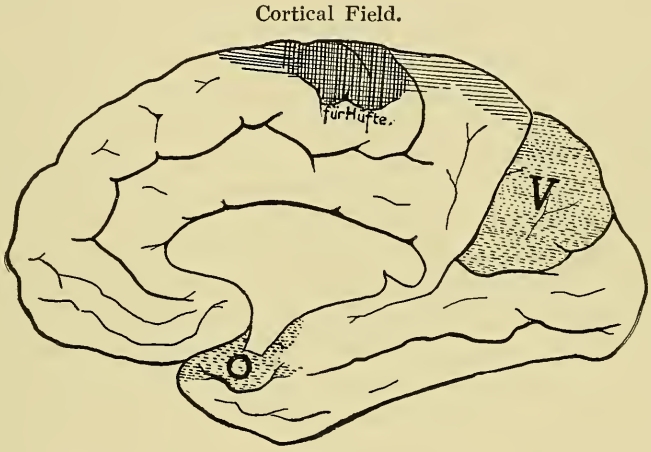


Fig. 86.

Motor Sensible and Sensory Cortical Fields.

V = Visual. O = Olfactory Cortical Field. Für Hüfte = For Hip.

a difficult and responsible task. It is impossible for me, within the limits of these lectures, to enter into the particularities with regard to methods of localization which you will find assembled in my "Compendium of Topical Brain and Spinal Cord Diagnosis." Only the most important points will be here considered.

Tumors of the motor region of the cortex lead, in the majority of cases, to the so-called "cerebral monospasms" as well as to "cortical (*Jacksonian* epileptic attacks" (both representing cortical irritative symptoms), on the other hand, also, to "cortical paralyses" (to be considered as defect symptoms in consequence of destruction of cortical motor centers). Figs. 85 and 86 will show you the distribution of these centers. As cerebral monospasm we designate tonic-clonic spasms which affect a muscular center up to this time normal or already paretic (face, arm, hand, etc.), and leave behind them permanent paresis, or a permanent increase of an already present paresis. *Jacksonian* or cortical epileptic attacks begin usually as monospasm, and,

as a rule, tonically. The contractions, however, do not remain limited to their starting-point, as does an ordinary cortical monospasm (to the muscles about the mouth, for instance), but they extend to the arm, and finally, also, to the leg of the same side of the body (naturally the seat of the tumor is on the side of the brain opposite to the spasm). When the attack begins in the arm, after the whole arm is affected, the face, and then the leg, become involved. As the crural type is designated the succession, lower limb, upper limb, face. The tumor sets the pyramidal cells in its neighborhood into a condition of irritability which spreads itself like waves over the surface of the water, gradually to the neighboring cortical regions. The muscles, indeed, are attacked in an order which corresponds to the arrangement next to one another of their cortical centers. In severe cases, *Jacksonian* convulsions pass over also to the extremities on the other side and can be accompanied by loss of consciousness. A very intense irritation can also propagate itself through the commissural tracts of the corpus callosum, etc., to the cortical motor field of the other hemisphere, and can finally benumb the centers for the higher psychical functions (in the first place the frontal cortex). Cortical monoplegia usually develops as a gradually increasing spastic paresis; according to its extent, we speak of monoplegia cruralis, brachialis, facialis, facio-brachialis, facio-lingualis. Facial paralysis is limited to the lower division of the muscles, since the upper is innervated from both hemispheres so that the loss of one-sided innervation is comparatively unimportant clinically.

While on account of their surgical interest the tumors of the cortical motor region a short time ago stood so much in the foreground of all brain tumors that *v. Bergmann* defined brain surgery as the "surgery of the central convolutions," to-day the tumors of the posterior fossa of the skull are also relatively frequently operated upon. The description of their symptomatology we will reserve for the next lecture devoted to cerebellar affections. Also the most important facts with regard to the not very rare tumors of the hypophysis of late equally attacked by the surgeons, we will put off until a later lecture (XXIV).

The remaining localizations of intracranial growths indicate usually inoperability; the knowledge of their special clinical symptom-complexes is hence somewhat less important, and we can content ourselves here with a comprehensive synopsis.

Tumors of the frontal lobe often produce a marked defective intelligence, somewhat like that of progressive paralysis; again, the so-called "Witzelsucht" ("moria")—a tendency to silly jests with loss of ethical feelings and pleasure in offensive demeanor. In affection of the lowest frontal convolution on the left side, that is, of *Broca's* center, motor aphasia occurs, as already mentioned in Lecture XVIII. Occasionally paralysis or contracture of the trunk and neck muscles can be observed. Tumors on the lower surface of the frontal lobe lead, as a rule, to protrusion of the eyeball, to anosmia and to an atrophy of the optic nerve which often progresses so rapidly that a papilloedema does not develop. Where, however, the last occurs, it is found not rarely only upon one side (that of the tumor).

Tumors of the region of the optic thalamus set up lateral hemianopsia

and (by their effect upon the internal capsule) a progressive spastic paresis of the extremities (both on the opposite side). Further, the suspension of certain "psycho reflexes" have been described as a thalamus symptom. On unintentional laughing and crying the lower facial muscles of the opposite side remain masklike, stiff and immovable, while voluntarily they can be put in action in every manner. Finally are to be mentioned: A crossed, persistent hemianesthesia (which is usually much less marked for touch, pain and temperature than for deep sensibility) and exceedingly severe continual pain, which is refractory to analgesics and exacerbates from time to time ("central pain"), which is projected into the contralateral half of the body.

Tumors of the temporal lobe are sometimes accompanied by sensory aphasia (see above, page 277), those of the occipital lobe by hemianopsia (see above, page 259). Also tumors of parietal situation can produce half-sided blindness by destruction of the optic radiations of *Gratiolet*. With the last localization the symptom-complex of alexia, already mentioned, is not very rarely observed, very frequently, however, the so-called astereognosis, that is, the inability to recognize objects by touch when the eyes are closed, although with this the elementary sense perceptions are tolerably preserved, and in the "pure" cases, which have been called "touch paralysis," are found entirely intact.

Characteristic of tumors of the corpus callosum are: Apraxia (see above, page 283), bilateral hemiparesis without increase of reflexes, and without *Babinski's* symptom. According to the more or less median situation of the lesion, the intensity of the paresis on the two sides is even or uneven; also, unilateral hemiparesis can be combined with contralateral motor irritative symptoms (for example, convulsions or hemichorea). In corpus callosum lesions, sensibility, as a rule, remains intact, also the functions of the cranial nerves; only in involvement of the most anterior portion of the corpus callosum the facial is found paretic. Tumor formation in the internal capsule produces a chronic progressive hemiplegia whose clinical end result closely approximates that of the acute form. In tumors of the region of the corpora quadrigemina, pupillary paralysis, paralyses of the external eye muscles, ataxia, impaired hearing, visual disturbances, nystagmus directed vertically and outward, are found. That the origin of a tumor pressing upon the lamina quadrigemina is in the pineal gland (epiphysis cerebri) is indicated by the occurrence of conditions of sexual excitement, abnormal development of hair and adiposity, in young individuals, also, occurrence of a hyperplasia of the genitals and an abnormal increase of the length of the body (the so-called "dyspinealism," see Lecture XXIV).

The tumors of the base of the brain manifest themselves in general by early and marked involvement in the clinical picture of the cranial nerve roots. Of irritative symptoms, hyperesthesia and neuralgiform "root pains" in the region of the trigeminus are to be specially mentioned. The irritative symptom of the motor trigeminus is trismus, the cramp of the masseters leading to grinding the teeth, that of the facial the twitching of the face muscles. Also sensory irritative phenomena (*e.g.*, tinnitus aurium) are not rare. The paralytic symptoms can affect every brain nerve. They are, as a

rule, multiple, but, on the other hand, usually develop only on one side. Most frequently affected are the facial, abducens, and oculo-motorius whose alteration, as a rule, first manifests itself by ptosis. Destruction of the pyramids is quite rare; slighter affection comes only little by little into plain view—through the *Babinski* reflex, spasticity, etc. Only lesions in the neighborhood of the crus cerebri have a great tendency to destroy the cortico-spinal tracts, so that in this location of the tumor the *Weber* symptom-complex hemiplegia alternans oculomotoria (see page 270) comes under observation. Papilloedema usually occurs early. If the tumor is located in the caudal division of the base of the brain, bulbar paralytic symptoms arise (see page 120).

Prognosis and Treatment

The prognosis of brain tumor is in general a very gloomy one. Spontaneous improvement and recovery has been observed in aneurisms (obliteration), in cysts and serous meningitis (resorption), in tubercle, cysticercus, echinococcus (calcification), in gumma (regression); the process of cure may be hastened in syphilitic new formations by anti-syphilitic treatment, also mercury and iodide cures have been of benefit in serous meningitis. In true neoplasms there are cases in which in benign tumors recovery up to a certain point has occurred through these growths undergoing calcification or ossification, very great rarities, so that death in coma or from rupture of a vessel is the nearly regular ending of the frightful disease picture (although sometimes it is only slowly progressive, and eventually interrupted by stationary periods). In most cases, hence, operative interference, in spite of the very great dangers which it presents in many instances, offers to us the only therapeutic way standing open.

In principle, curable by operation are, properly, only tumors on the cerebral convexity and upon the cerebellum (provided that they are cystic, or, if solid, are well limited and can be extirpated in toto *); perhaps, also, hypophysis tumor. Very many cases can, however, derive great benefit from a palliative operation; this is the so-called "decompressive craniectomy."

With regard to the indications for surgical interference, I would impress upon you the following: In too many cases decision as to an operation is put off for a long time to try combined mercury and iodide treatment (which usually has also in glioma and sarcoma a decided but unfortunately only temporary effect—probably on account of resorption of the secondary ventricular dropsy) until it finally proves itself necessary. Such treatment is usually ordered even in the absence of a history of syphilis, while on the other hand it should not be forgotten that former syphilitics also can have non-luetic brain affections. It is always to be remembered that direct danger to life, unbearable intensity of the headache, threatened loss of vision or dementia, present pressing indications for decompressive craniectomy. *Babinski* has properly emphasized that this indication should be accepted much more

* Gliomata show such indefinite limitation from the healthy surrounding tissue and such a gradual passage over into it, that their operative prognosis, in spite of their pathologico-anatomical benignness, is unfavorable.

frequently than it generally is. Even in malignant neoplasms it can act palliatively; in benign tumors inaccessible to extirpation, which, however, are capable of coming to a standstill in growth, it may prevent the occurrence of irreparable destruction; in serous meningitis it may be directly curative. Where the brain pressure is immediately threatening it may be necessary to advise first operative decompression, and then a mercury cure. In tumors not exactly localizable, temporal craniectomy recommends itself, since the muscles of mastication oppose in suitable manner the occurrence of brain prolapse. If the disease is located in the posterior fossa, the occipital region is, as a rule, the place of choice. The neck muscles also affect a good closure. Less favorable conditions in this respect is afforded by parietal craniectomy which is undertaken for tumors of the motor region.

B. Brain Abscess

The infection lying at the base of brain abscess usually enters the organ by continuity, more rarely by metastasis. The first applies to traumatic and otitic brain abscesses which are observed after septic wounds of the skull and suppuration in the ear; in these the germs pass by the lymph channels, along the nerve sheaths, or by means of septic venous thromboses into the brain substance. Metastatic infections reaching the brain by way of the blood vessels, namely, after lung abscesses, empyemata of the pleura and putrid bronchitis, lead to cerebral pus collections. Brain abscesses can be multiple or solitary; in the last case they sometimes reach the size of an apple. Their pus contains staphylococci, streptococci, colon bacteria, different anaërobic germs, etc. Otitic abscesses locate by preference in the temporal lobe or in the cerebellum; traumatic or metastatic, naturally show a very variable localization.

The clinical symptoms of brain abscess, like those of brain tumor, may be divided into general symptoms and focal symptoms. In regard to the last, there is entire agreement between the two diseases, and also the general symptoms show very great analogy. Only papilloedema is much rarer in abscesses than in brain tumors; in them fever, which sometimes has a septic character (with rapid rise of temperature and chills), sometimes, however, expresses itself only in moderate evening rises of temperature, plays a decidedly greater rôle. That does not mean indeed, that cases with entirely afebrile course are great rarities; on the contrary, they represent a very considerable minority which may well depend upon a relatively frequent formation of thick "abscess membrane" which prevents the resorption of the septic substances. The encapsulation can go so far that the abscess becomes "latent," and either causes no longer any difficulties or manifests itself only through the focal symptoms which have become stationary. Such a becoming latent, however, cannot be considered equivalent to a cure, as even after latency for years acute lighting up again of the process still threatens. On this account the treatment of brain abscess (in so far as it can be diagnosed and an operatively accessible point can be localized) in general can be suitably and conclusively summed up in the old surgical axiom, "Where there

is pus, evacuate." The diagnosis, however, in those cases in which a skull trauma, suppuration in the ear or in the mastoid, a lung abscess, etc., cannot be recognized, and where fever and chills are absent, is mostly exceedingly difficult with regard to its distinction from brain tumor; indeed, sometimes an impossibility even for the experienced neurologist, or it must be made by exploratory trephining and puncture of the brain. Occasionally the differential diagnosis from purulent meningitis, upon the description of which we will immediately enter, is difficult.

C. Purulent Cerebral Meningitis

What we have said as to the etiological factors in brain abscess applies also to purulent meningitis. It can also arise from traumatic, otitic or metastatic causes. The last method of origin, however, is observed here much more frequently than in brain abscess; along with the affections of the lung and pleura mentioned, in purulent meningitis, ulcerous endocarditis, puerperal infection, further typhoid, pneumonia, influenza, etc., play a not inconsiderable rôle. A relatively frequent manner of meningeal infection by continuity is further the progressive infection of the so-called emissary veins of the skullcap and the lymph vessels in erysipelas. Other important etiological factors are, finally, the septic diseases of the orbit, the nose, the frontal sinuses, and the ethmoid cavities, as well as the rupture of a brain abscess into the subarachnoid space.

The often exceedingly voluminous pus collection in cerebral meningitis usually affects the convexity of the brain in such preponderant fashion that it is often directly denominated "meningitis of the convexity."

The clinical picture of this affection shows so great a similarity to that of epidemic meningitis that, referring to Lecture XVI, we can dispense with a thorough description of the individual symptoms and content ourselves with a summary sketch of its symptomatology. The initial symptoms of purulent meningitis are very often veiled by those of the underlying disease, for instance, of erysipelas or of purulent otitis. Where, however, they have not developed from such a primary infection, they are usually characterized by quite masked and sudden beginning.

Fever is present in by far the majority of cases and of rather continued character. As a rule, the temperature varies between 38° and 40° C. (100.4° and 104° F.); apart from the initial chill sometimes occurring, many patients in the further course of the disease have marked shivering from time to time. Headaches of terrible severity and diffuse distribution are present from the start, and show themselves even in half coma by the patient frequently putting his hand to his head. The pulse is rapid, respiration hurried. Cerebral vomiting is only frequent in the initial stages. Stiffness of the neck is sometimes developed quite early, also other conditions of muscular tension; boat-shaped retraction of the abdomen, trismus, grimacing, contractures in flexion of the extremities, *Kernig's* symptom. On the other hand, general convulsions coming on in attacks, are rare, more frequent, localized twitching of one extremity, one-half of the face, etc. The pupils are at the start narrowed

and frequently unequal; in otitic meningitis the myosis is more marked on the side of the affected ear; in the terminal stage they are wide. Of other ocular symptoms, there occur occasionally convergent and divergent strabismus, as well as tonic spasm of the orbicularis palpebrarum. Further disease signs are, hyperesthesia of the skin, the so-called "taches cérébrales" of *Trousseau* (see Lecture XVI, page 250), retention of urine and fæces, particularly regularly, however, deliria, which manifest themselves sometimes by the murmuring of incoherent words as well as by disorientation for time and place, sometimes, however, by screaming, motor jactitation, even furibund conditions. Here and there papilloedema, usually of moderate grade, occurs.

The disease results fatally in the great majority of cases. In the terminal stage the clinical picture changes, in that, in the place of contractures, paralyses (monoplegia, hemiplegia, paraplegia, even tetraplegia), in the place of delirium, a torpor deepening to coma; instead of myosis, mydriasis; instead of retention, incontinence of stools and urine, instead of quickening of the pulse and respiration, their slowing occur, and finally death from respiratory and cardiac paralysis follows. Occasionally at the last, *Cheyne-Stokes* breathing is observed.

Cases going on to "recovery" almost always leave behind serious defects, as deafness, mental weakness, idiocy, etc.

The treatment agrees in general with that of epidemic meningitis. Naturally, upon the beginning of meningeal symptoms, the underlying disease must be searched for, and energetically combated; for example, the mastoid trephined, the frontal sinus opened, furuncle of the face incised, etc. Repeated lumbar punctures are always in place, although they only rarely produce lasting effect; nevertheless, they quite frequently affect considerable alleviation, especially as to headache and delirium. When the puncture fluid, at first cloudy, later distinctly purulent, becomes by degrees again free from leucocytes and clearer, this is naturally favorable prognostically. Definite surgical treatment (trephining, opening the dura, washing out, drainage) has of late been recommended by *Girard* and others; the results are, however, unfortunately very uncertain, as a rule. For the rest, see Lecture XVI, page 254.

D. Tuberculous Meningitis

A special clinical and anatomical consideration is due, to the by far most frequent variety of metastatic inflammation of the pia-arachnoid, tuberculous meningitis, which occurs, as a rule, in young individuals affected by lung, bone or joint tuberculosis. It is particularly often, also, a symptom in acute miliary tuberculosis. Tuberculous meningitis arising by continuity is much behind the metastatic form in importance and frequency; tuberculous caries of the skull-bones, ear tuberculosis, solitary tubercle of the brain, etc., usually furnish a starting-point for it. As exciting causes, injuries to the head (even bloodless ones), insolation, taking cold, overexertion, as well as psychical excitement, come into consideration. So, I have observed in a girl with slight affection of the lung apices, a tuberculous meningitis terminating fatally and con-

firmed at the autopsy, which occurred in immediate connection with a violent quarrel with her unfaithful lover, as acute delirium, and which was at first quite pardonably interpreted as hysteria by the family physician. In contradistinction to purulent meningitis, tuberculous meningitis in the great majority of cases spreads itself over the region of the base of the brain. It is hence designated as basilar meningitis. The extension of the inflammatory process into the fissure of *Sylvius* is almost the rule, while the convexity of the brain is usually but inconsiderably affected. In the region of the disease, an opalescent or cloudy "gelatinous" infiltration of the pia and arachnoid is found; the fluid is seropurulent only in places. At most varied points, however, particularly in the neighborhood of the vessels, a more or less abundant deposit of miliary, whitish, tubercular nodules is noted, which anatomically present the well-known structure of tuberculous granulations (giant cells, etc.). Also *Koch's* bacillus can be recognized in most cases.

The disease picture of tuberculous meningitis has so great similarity to that of purulent meningitis that in order to avoid repetition, I will limit myself more or less to emphasizing the differences in its course and symptomatology.

Only exceptionally does tuberculous meningitis begin suddenly. In the great majority of cases there is a definite prodromal stage of from one to several weeks. The patient is peevish and irritable, sometimes also apathetic and soporous. In many cases the alternating redness and pallor of his face is striking to those surrounding him. He loses all appetite and emaciates rapidly; usually there is obstinate constipation, somewhat less frequently, nausea or vomiting. Soon there are irregular rises of temperature with slight shivering, further, headache, stiffness of the neck, confusion at night—and so it proceeds finally to the completely developed meningitic symptom-complex, in which headache, cerebral vomiting, stiffness of the neck, and tension in other muscles, vasomotor disturbances, hyperesthesia of skin and muscles, *Kernig's* and *Leichenstern's* signs (see Lecture XVI, page 249), reach a very high degree. In children opisthotonus is so excessively developed that the occiput almost rests upon the back; there is frequently also very great contracture in flexion of the lower extremities. The delirium manifests itself usually in restless grasping movements of the hands, muttering, etc. ("muttering delirium"). In contradistinction to purulent meningitis, the relatively slight elevation of temperature (usually between 38° and 39° C.), is striking; frequent, also, is the absence of quickening of the pulse, if not its slowing. Lumbar puncture shows in the initial stages almost always a lymphocytosis of the spinal fluid, further on in its course, however, the leucocytes may preponderate just as much as in purulent meningitis. Naturally, a positive finding of tubercle bacilli in the spinal fluid is decisive. As a criterion of the basal localization is, further, the much greater frequency of cranial nerve symptoms important: paralyses of the abducens, oculomotorius, facialis, deafness, optic neuritis. Symptoms from the side of the pyramidal tracts (*Babinski* and *Oppenheim's* reflexes, ankle clonus) not rarely occur. The ophthalmoscopic finding of tubercles in the choroid is, on the other hand, a very great rarity and scarcely ever available for confirming the etiological diagnosis (for which in children the *v. Pirquet*

cutaneous reaction may be called into aid, in case the primary tuberculous focus is latent).

The prognosis is a very bad one. Of the bacteriologically confirmed observations of tuberculous meningitis, only very exceptionally have cases survived the disease, and here even the recovery was usually with defect; also some of these patients perished later with recurrence. The treatment agrees in general with that of other meningitides.

E. Internal Hemorrhagic Pachymeningitis

In the etiology of this disease, chronic alcoholism occupies the first place in frequency and practical importance; besides, it occurs in different disease conditions accompanied by the "hemorrhagic diathesis" (scurvy, leucemia, hemophilia, pernicious anemia, etc.), as well as after the most varied exhausting infectious diseases. Exciting causes for this disease, which affects by preference those of advanced age and the male sex, are, above everything else, contusions and wounds of the head. For the rest, many cases remain etiologically entirely unexplained. According to *Obersteiner*, hemorrhagic pachymeningitis occurs in about 20 per cent. of paretics as a complication of the end stage.

Anatomically, internal hemorrhagic pachymeningitis presents itself as follows: There arise upon the inside of the dura mater, flat, membrane-like deposits, extraordinarily rich in vessels, of inflammatory granulation tissue, which later changes into connective tissue. The walls of these vessels show more or less marked evidence of degeneration and a great tendency to rupture, which have as a result the formation of blood coagula upon the membrane covering the dura. New granulation tissue develops in these coagula, and so there arises from hemorrhages which later show themselves only as brown pigment, strata layer built upon layer. This hemorrhagic pachymeningitis develops (usually bi-laterally, although, as a rule, asymmetric) particularly over the convexity of the hemispheres, namely over the parietal lobes; it is more rarely found at the base of the skull.

The disease in slight cases can run its course without symptoms and is then counted among the so-called "accidental autopsy findings." This applies, for example, to most of the cases found in paretics. The more severe cases, on the contrary, produce grave symptoms, which indeed have in themselves little typical, and are subjected to such great variations in their character and grouping that confusion with leptomeningitis, brain tumor, hemorrhages, etc., is very frequent. In the early stages severe headache is the chief symptom. Since the patient besides this usually complains of vertigo, shows different speech disturbances and defects of memory and in his movements can give evidence to a certain slowness, weakness and clumsiness, we are greatly reminded of arteriosclerosis of the brain, cerebral syphilis or dementia paralytica. Also conditions of psychical excitement and epileptiform attacks are described. Later the condition is complicated by suddenly occurring (probably corresponding to further hemorrhage) focal symptoms which sometimes disappear again, sometimes persist: Monoplegias, hemiplegias, also tetraplegias, con-

jugate deviation of the eyes and head, sometimes also spasms in the individual extremities or in the face. Stiff neck is rare, *Kernig's* symptom somewhat more frequent. The pupils are usually narrow, sometimes unequal, and react slowly; the tendon reflexes are usually exaggerated, ankle clonus and *Babinski*, on the other hand, but rarely present. There can be also a weakening and diminution of the reflexes. The intelligence diminishes rapidly. In cases running a fatal course, the symptom complex of brain pressure is gradually established (slowing of the pulse, papilloedema, etc.) and the patient dies in coma.

A standstill or partial regression of the disease picture can occur in cases which have not progressed too far. Along with treatment of the underlying disease, in all cases an energetic iodide cure should be tried. To be recommended further, are free purgation, bland diet, abstinence from alcohol and from smoking. Apoplectiform incidents are to be treated after the principles laid down in Lecture XVII.

F. Thrombosis of the Brain Sinuses

The thrombotic occlusion of the different brain sinuses (particularly of the transverse, superior longitudinal and cavernous sinuses) can arise autochthonously or secondarily, that is by the extension of infection or through metastasis. In the first instance there is the so-called "marantic" thrombosis which is observed in decrepit individuals, in chlorosis, pernicious anemia, following exhausting infectious diseases, particularly, however, in the "atrophy of nurslings." Secondary or infectious sinus thrombosis arises in consequence of erysipelas, suppurations in the petrous bone, in general sepsis, pyemia, etc. The thrombotic occlusion of a large sinus manifests itself clinically by severe headache beginning suddenly, by confusion and by spasms in the extremities; in infectious thromboses these symptoms are accompanied by chills and fever. Characteristic, however, are the local signs of stasis; in thrombosis of the cavernous sinus the lids are swollen, the eyeballs protrude; in thrombosis of the transverse sinus the neighborhood of the mastoids is œdematous and marked by dilated veins; in thrombosis of the longitudinal sinus, the temporal veins are swollen, and there is also venous stasis in the nasal mucous membrane and epistaxis. Papilloedema is frequent only in cavernous thrombosis, rarest in transverse thrombosis. Extension of the thrombosis into the jugular vein is accessible to direct palpation.

The prognosis of these conditions is very serious, as even mild sinus thromboses, if they reach a higher degree, produce œdema of and hemorrhages into the brain. Only clotting of limited extent is susceptible of recovery, a result which we seek to assist by the application of leeches to the neck, iodide cures, etc. Infectious thromboses, on the other hand, demand operative removal, which, however, only in the minority of cases is able to prevent death from diffuse meningitis.

G. Non-Suppurative Encephalitis

The non-purulent inflammations of the brain, if we ignore the cerebral localization of the *Heine-Medin* disease, and the inflammatory forms of in-

fantile cerebral paralysis, are great rarities; their knowledge we owe to *Strümpell*, *Wernicke*, *Oppenheim*, and *Cassirer*. Anatomically, the diseased parts present the histological criteria of inflammation (infiltration with small cells, dilatation of the vessels, œdematous swelling, degeneration of the parenchymatous elements, etc.) and manifest themselves macroscopically by redness and swelling. Usually there are also numerous punctiform or miliary extravasations of blood upon cross section (the so-called "Flea-bite Encephalitis").

Clinically, different forms may be separated, of which the two following are the most important:

1. POLIOENCEPHALITIS SUPERIOR HÆMORRHAGICA (WERNICKE)

The patients are taken ill suddenly, with headache, vertigo, vomiting, confusion, and somnolence, sometimes also with conditions of delirious excitement, and with this there develops a rapidly progressive, finally almost total ophthalmoplegia. The gait is extremely atactic. Tremor is almost the rule, choreic unrest is sometimes to be observed. The disease, which mainly affects confirmed alcoholics of middle age, but can arise upon the basis of meat poisoning, usually leads to death under increasing stupor and heart weakness in one or two weeks. Recoveries are rare.

Upon autopsy, in the neighborhood of the third ventricle and of the aqueduct of *Sylvius*, particularly in the region of the eye muscle nuclei, a severe hemorrhagic inflammation is found.*

2. THE ACUTE HEMORRHAGIC ENCEPHALITIS OF ADULTS (STRUMPELL)

This is in the majority of cases an accompaniment or the result of acute infectious diseases, namely, of influenza. It can, however, represent a primary acute infection. *Klieneberger* saw it arise after salvarsan injection. Rather suddenly there arise with high fever, headache, unconsciousness and hemiplegic disturbances, which end in a few days in death. Only a few cases pass on to recovery, leaving behind more or less marked hemi-paresis. Pathologico-anatomically a hemorrhagic inflammatory process forms the basis of the disease. This, as a rule, is limited to one hemisphere, but is rather diffusely spread throughout this and affects both the white and the gray matter.

Therapeutically we can, though with justified pessimism, in both forms, hold fast to the recommendations of *Oppenheim:* Icebag to the head, leeches to the temples or to the mastoid region, free purgation (calomel) inunctions of collargol or a mercury cure, iodide of potassium, antipyretics, wet packs to the body, hot foot baths, sinapisms to the extremities, etc.

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* "Polioencephalitis inferior hemorrhagica," which is located in the gray matter of the medulla, occurs in infectious diseases and gives the clinical picture of "acute bulbar paralysis." (See Lecture VII.)

H. The Circulatory Disturbances of the Brain

We will take into consideration here only those circulatory disturbances of the brain which occur in the form of attacks and rather rapidly disappear again; hyperemia of the brain, anemia of the brain, and the so-called concussion of the brain.

Determination of blood to the brain occurring in attacks forms the basis of the so-called "congestion of the brain" which occurs under the influence of heat, psychical and especially sexual excitement, etc., in predisposed individuals (vasomotor neurasthenics, arteriosclerotics, inveterate smokers, drinkers, people with "true plethora") and which can be experimentally produced by the inhalation of amyl nitrite. In it, the face becomes intensely red, there is a feeling of burning in the conjunctivæ, the carotids and the temporal arteries pulsate appreciably; the patient usually experiences tinnitus aurium, a sense of pressure in the head, vertigo, spots before the eyes, sometimes, also, nausea, becomes excited, sometimes, slightly dull and confused. After a few minutes, or only after a half hour to an hour, the hyperemia ceases and the symptoms disappear. Its danger lies in arteriosclerotics, naturally, in the condition that it favors the occurrence of cerebral hemorrhage.

Paroxysmal diminution of the blood in the brain finds its clinical expression in the temporary reduction of the brain functions to the minimum necessary for maintaining the vital functions; there is fainting, syncope. After indefinite prodromal symptoms and a general feeling of illness, there occurs, with smallness and rapidity of the pulse as well as great pallor of the face and outbreak of a cold sweat, often also, with repeated yawning and usually decided nausea (rarely actual vomiting), a condition of complete muscular relaxation and marked clouding of consciousness or even complete unconsciousness, which lasts for a half hour or more, then to make way for the normal condition. The prognosis of these conditions which occur almost exclusively in anemic, weak or cachectic persons, is in general good; a fatal ending is only to be feared from syncope after severe hemorrhage.

Commotio cerebri represents a special variety of circulatory disturbance in the brain, which occurs after violent concussions of the head in accidents of the most various sorts, and indeed also without direct traumatization of the skull (for example in persons who have been in the neighborhood of a violent explosion). This "concussion of the brain" occurs at once and disappears after a short time (some minutes to a few hours). Only very exceptionally in such cases in which the autopsy has shown an absence of any organic injury of the brain (for instance, compression or hemorrhage) does death occur from cardiac and respiratory paralysis; the patient in commotio cerebri suddenly becomes unconscious, breathes, as a rule, shallowly (more rarely deep and stertorously), has a small, generally slow pulse, is pale and presents coldness of the surface. He shows complete muscular relaxation and does not react to painful stimuli (pinching, stabbing, etc.). The pupils are now narrow, again dilated, but react to strong light. Already during unconsciousness, particularly, however, when the victim comes to again, there is frequently retching or vomiting, and afterward there persists for a long time, headache, staggering

gait, and slight clouding of consciousness. More severe symptoms, for example, convulsions, do not belong within the limits of cerebral concussion, but indicate organic injuries of the brain. As a characteristic result cerebral commotion almost always leaves behind the so-called "retrograde amnesia," that is, the memory not only of the accident itself, but even the period (of about a quarter of an hour) preceding it, is completely obliterated from the recollection of the patient. Sometimes, however, these memories return after a time, in which case, on retrospective diagnosis, we must think of concussion of the brain, when required to give an opinion on the case.

Treatment of hyperemia of the brain consists in loosening the clothing about the neck, raising the head, giving hot foot baths, hot applications to the calves, mustard plasters to the feet, cold applications to the forehead and neck; when necessary, also, in bleeding (best by puncture of a vein). In syncope and concussion of the brain, the patient is placed in a horizontal position, is allowed to inhale ammonia, the face is sprinkled with cold water, the temples rubbed with alcohol, the respiratory muscles are faradized, and, when necessary, the heart's action is stimulated by injections of ether and camphor.

LECTURE XX

Diseases of the Cerebellum

GENTLEMEN: The progress which brain surgery has made during recent years has rebounded to the advantage of the cerebellum in special degree. This, however, places not only upon the neurologist, but also upon the general physician, the duty of being alive to the earliest possible recognition of cerebellar affections, since the result of an operative procedure depends chiefly upon the time at which we entrust the patient to the hands of the surgeon. On the other hand, it should not be overlooked, that every operation in the posterior fossa of the skull is accompanied by dangers to which no patient should be lightly exposed. On this account the differential diagnosis of cerebellar diseases is of great practical importance.

From a clinical standpoint we are justified in including certain extra-cerebellar affections of the posterior fossa of the skull under "cerebellar affections," since they manifest themselves preponderatingly through cerebellar symptoms, I mean: 1. Tumors of the cerebello-pontine angle. 2. Serous meningitis in the posterior fossa of the skull. Both disease conditions, as well as abscesses and tumors of intra-cerebellar location, come within the province of surgery, while softenings, hemorrhages, atrophies and ageneses of the cerebellum, as well as cerebellar encephalitides and cerebellar syndromes in malaria, are to be cared for by the physician.

We will take up these disease processes in the order of their practical importance.

A. Tumors

The cerebellum is, during childhood and youth, the location of predilection for tumor formation in the nervous system. Along with true neoplasms—gliomata, sarcomata, fibromata—cystic tumors occur, and as an infectious granuloma, solitary tubercle is to be considered. Besides, there must be mentioned as a rarity, a parasitic tumor, echinococcus of the cerebellum.

Symptomatologically, the brain pressure syndrome which we have already described under tumors of the cerebrum, makes itself first apparent; still, with this, some important criteria for the localization of the pathological process, whether it is in the cerebellum or in its immediate neighborhood, usually must be considered. So, headache in cerebellar affections is characterized by extraordinary violence and persistence probably on account of the richness of the tentorium in sensory fibers. Further, this pain presents its maximum severity in the occipital region and the back of the neck, from whence it radiates sometimes into the upper part of the back. Many times, also, the patients complain

of frontal headache; still, this is diffuse, while the occipito-nucheal pain usually predominates on the affected side. It can be accompanied by more or less marked stiffness of the neck. Very frequently the back of the head is sensitive to percussion and pressure; still, I place particular value upon finding the characteristic pain which a pressure upward upon the mastoid process of the affected side produces. Papilloedema appears particularly quickly and markedly in cerebellar tumors, and even when they are unilateral it is generally bilateral. Finally, a "spinal fluid phenomenon" has been described in cerebellar tumors; this consists in the excessively rapid fall of pressure after a lumbar puncture and the abrupt cessation of the flow of the cerebrospinal fluid. Evidently the tumor presses the medulla suddenly into the foramen magnum and in this way interrupts the communication between the intracranial and the spinal fluid. In spite of all the assertedly infallible precautions which a few specialists on lumbar puncture have recommended, we had better avoid this dangerous diagnostic method just as soon as we have become convinced that there is a suspicion of tumor of the cerebellum, since some cases of sudden death have occurred under its application.

Among the local symptoms of cerebellar tumors, cerebellar ataxia stands in the foreground, as it presents the cerebellar symptom par excellence. By this I mean only, that without ataxia no cerebellar affection can be diagnosed with certainty; in no way, however, that typical cerebellar ataxia is foreign to extra-cerebellar lesions. The interruption of one system of cerebellifugal or cerebellipetal tracts suffices to deflect the regulating activity of the cerebellum, whether this lesion has its location in the midbrain, in the pons, in the bulb, or even in the spinal cord. It is all the same to a reflex apparatus, whether the interruption has destroyed the reflex center, the afferent or the efferent fiber complex. We have already, under the description of *Friedreich's* disease in Lecture VIII, mentioned the clinical peculiarities of cerebellar ataxia and its physio-pathological basis (page 123).

While incoördination in consequence of a total lesion of the posterior roots of the spinal cord affects movements in their entirety, cerebellar ataxia—particularly on the trunk and in the lower extremities—shows a decided predilection for the composite movements, that is, for those movements which require the coöperation of extended groups of muscles. In such patients it is observed that the simple muscular movements, for example, flexing or extending the foot, the knee, the hip, adduction or abduction of the thigh, can be executed correctly; that, however, their dynamic and static combination is disturbed. So are produced a zigzag gait, reeling, more or less marked swaying, as a consequence of an interruption of the synergies which are necessary for the immobilization and steadying of the body and its members in walking and at rest. Here, also, must be considered that phenomenon to which attention has been called by *Babinski* under the name "cerebellar asynergy." When the patient attempts to raise himself while lying on his back, he elevates his lower extremities instead of his trunk; when going forward he allows his trunk to lag behind to a certain extent, and so is in danger of falling backward.

As a rule, cerebellar ataxia affects the upper extremities to a much less degree; sometimes, indeed, it appears to avoid them entirely. It must be con-

sidered, however, that in man the arms are subjected to the coördinating influence of the cerebellum to a very limited extent on account of their little importance in the maintenance of the equilibrium. Nevertheless, I have nearly always been able to detect certain atactic disturbances in the movements of grasping with the hand. As to this, we owe to *Babinski* an ingenious method of bringing out latent disturbances of coördination in the arms. The patient is ordered to carry out in rapid succession muscular actions opposed to one another, for example, pronation and supination, and it is often noticed then that the individual with cerebellar disease can no longer effect so subtile cooperation of the antagonistic muscles. This phenomenon is called "adiadochokinesis" (*ά*=privative; *διαδοχή*=succession).

In clinical importance cerebellar hypotonia is inferior to cerebellar ataxia, since the first may be entirely obscured by simultaneous lesion of the pyramidal tracts; such a lesion, however, is one of the most frequent indirect effects of tumors of the cerebellum, while, on the other hand, marked spastic conditions are in no way uncombinable with typical cerebellar ataxia. Where, however, cerebellar hypotonia is present, it manifests itself like spinal hypotonia (see above, page 173) as well in a relaxed condition of the muscles as in the possibility of bringing the extremities into very abnormal positions on account of the relaxation of the antagonists, for example, overextension or overflexion.

A method of examination recommended by *Stuart* and *Holmes*, and of which I regularly make use, deserves to be mentioned. If a movement of flexion which the patient is attempting to carry out is opposed and the resistance is suddenly withdrawn, the flexion takes place in an extreme degree and the reaction, which normally always occurs, is either absent entirely or there is only a trace of it. This is a symptom which is capable of demonstrating to us the hypotonia of the extensor muscles.

I would lay particular emphasis upon the different conditions of the reflexes in spinal and in cerebellar hypotonia. The first is accompanied by areflexia or hyporeflexia, as we saw in Lecture XI, the last is entirely independent of the intensity of the tendon phenomena which can be found not rarely retained or even exaggerated. Where, however, in cerebellar tumors, a reduction or suspension of the reflexes is found, it appears to be dependent upon mechanical indirect action. Indeed, the increased brain pressure by extension to the closed sac which the spinal dura mater forms about the posterior spinal nerve roots may produce radicular lesions which can lead to symptoms of their anatomical destruction (*Hoche, Wollenberg* and others).

In general, ataxia and hypotonia are most marked in tumors of the vermis. In tumors of the cerebellar hemispheres, they, as a rule, are present in less intensity, and indeed, when the process is unilateral, in the form of hemiataxia and hemihypotonia of the same side. According to my personal experience, strict unilaterality of these symptoms is much rarer than their preponderance on one side. This last is the rule in all those cases in which the neoplasm indeed affects one hemisphere to a greater degree, but oversteps the middle line to a greater or less extent; in this class are probably the majority of cerebellar tumors—if we leave out of consideration those of the cerebello-pontine angle. As a matter of course, the recognition of atactic or hypotonic disturbances

on the relatively avoided side requires particularly careful methods of investigation.

Paradoxical are certain observations of tumors of the vermis without ataxia or hypotonia. It has been attempted to explain these rare cases in different ways. For example, *Nothnagel* believed that here the tumors were of such slow growth that a compensatory function could be established by an entering into action of other brain parts. In the case of solitary tubercle the absence of atactic disturbances has been attributed to the intactness of the axis cylinders which pass through the focus. Though these attempts at explanation may be satisfactory for a certain number of cases of tumors of the vermis which have run their course without ataxia and hypotonia, they are not suitable for other cases which, on the contrary, were characterized by an extremely rapid growth and plainly destructive nature of the process. If later pathologico-anatomical investigations do not solve the riddle, we must satisfy ourselves with the hypothesis that these exceptional cases affected individuals in whom the cerebellum, "ab ovo" possesses a particularly small functional importance.

Vertiginous phenomena are exceedingly characteristic for cerebellar tumors, even in the initial stages, and indeed they are manifested as true rotary vertigo, "systematic vertigo," produced by irritation of the neurones from the nuclei of the vestibular nerve, which pass through the cerebellum. The patients have the distinct sensation of rotation both of their own bodies and of the surrounding objects in a definite direction. These sensations set up nausea, often accompanied by pallor, sweating, vomiting, etc. They depend upon the fact that by the irritation of the tumor the stimulation of the vestibulo-cerebellar apparatus (which is normally produced only by the hydrostatic relations in the semicircular canals of the labyrinth) is in contradiction to the actual position of the body and so falsifies the ocular, articular and muscular impressions. Particularly interesting are the attacks of vertigo which, under the names of "cerebellar" or "vestibular" attacks, "cerebellar fits," "cerebello-pontile seizures" have been described by *Ziehen*, *Dana*, *Hunt*, and others. The sudden and extremely violent vertigo which characterizes them is often accompanied by loss of consciousness; as a rule, however, by definite atactic disturbances, severe vomiting, headache, tinnitus aurium and nystagmus.

This last symptom is particularly frequent in cerebellar tumors and can in general be considered as a symptom of irritation of "*Deiters'* nucleus" or of the "posterior longitudinal fasciculus." The one, indeed, lies close to the cerebellum, the other, however, passes directly under the vermis in the most dorsal portion of the tegmentum, to the eye muscle nuclei.

Via the nucleus of *Deiters* and the posterior longitudinal fasciculus, also, in the artificial provoking of a nystagmus by the so-called *Barany's* test, the impulse is transmitted to the abducens and oculo-motorius. In healthy people, by syringing the external auditory canal with cold water, a nystagmus toward the opposite side is produced; with hot water there is one toward the side of the injection. In tumors of the cerebellum, however this "caloric nystagmus" may be absent.

A spontaneous nystagmus of patients with cerebellar tumors can be but rarely noticed when looking to the front and must be provoked by having the

patient look far to the right or to the left. On this, a decided oscillation of the eyes is observed when the patient looks in the direction of his cerebellar lesion. I lay great stress upon this point. On the other hand, I have never been able to confirm another phenomenon mentioned by different authors, namely, the greater extent of the excursions in the eye corresponding to the diseased half of the cerebellum. As *Oppenheim* has emphasized, a nystagmus which is absent in a standing position can sometimes be elicited by having the patient lie upon his side.

Upon further growth of the cerebellar tumor a number of indirect symptoms usually manifest themselves in variable combination. Among the most frequent is abducens paralysis or, rather, a paralysis in looking toward the cerebellar focus. Of course the last can give rise to "conjugate deviation" toward the opposite side. Trochlear paralysis occurs rather regularly in neoplasms of the anterior end of the vermis, besides this, also, sometimes a paralysis of the superior, inferior, and internal recti muscles, on the other hand, almost never one of the internal eye muscles. Defect and irritative symptoms on the part of the cranial nerves from the 5th to the 12th develop more rapidly when the neoplasms are situated outside of the cerebellum than when they are intracerebellar; they are, however, quite frequent here also. Disturbances of speech, swallowing, respiration and articulation can produce in such cases the picture of bulbar paralysis already considered in Lecture VII. Possibly from dysarthria produced in this manner, the scanning speech which has been described by *Dreschfeld* and *Bruns* in cerebellar tumors, and which perhaps represents nothing more than a cerebellar ataxia of the speech muscles, must be distinguished. The investigations of *Rothmann* and *Katzenstein*, who have found in animals a region in the cerebellum whose extirpation produces an ataxia of the vocal cords, indicate this.

Frequently in cerebellar tumors sudden death from pressure upon the medulla has been observed; not rarely also in the course of the affection, a unilateral paralysis or paresis. According to whether the pressure has occurred anterior or posterior to the pyramidal decussation, the hemiplegia or hemiparesis affects the limbs of the same side or those of the opposite side.

Certain forced attitudes and compulsory movements are particularly marked when the neoplasm acts upon the anterior or the middle cerebellar peduncle. They are, rolling about the long axis or bending the neck and trunk toward a definite side, as, for example, in the observations of *Russell* and *Bruns*. Nevertheless it is impossible to give generally applicable rules with regard to the direction in which these involuntary attitudes and movements take place, whether toward the healthy or toward the diseased side, and its physio-pathological substratum is still quite obscure and the subject of controversy. No more definite is our knowledge of the pathogenesis of the choreic-athetoid movements which according to *Bonhöffer*, *Pineles*, and others, may occur on the same side upon destruction of one brachium conjunctivum. Also the conditions to which the vertical divergence of the eyeballs (of *Magendie*) owes its origin, are unknown to us. This symptom is supposed to be found along with conjugate deviation in affections of the middle peduncle of the cerebellum.

A definite category of tumors of the posterior fossa of the skull manifests itself by a rather stereotyped clinical picture, namely:

TUMORS OF THE CEREBELLO-PONTINE ANGLE

These are neoplasms (usually fibroma or fibrosarcoma) which take their origin from the connective-tissue sheaths of the auditory or the facial close to the exit of these nerves and present the following symptoms: Paresis in looking in one direction, nystagmus, absence of the corneal reflex, disturbances of sensibility in the distribution of the trigeminus, nervous hardness of hearing or deafness, adiadochinesis, all upon the side of the disease focus, and besides this, papilloedema, cerebellar ataxia and occipital headache. Of course the symptomatology of these tumors is not always such a complete one; on the other hand, however, further phenomena may be added to it, for example, convulsions, neuralgiform pains in the trigeminus region, anosmia, etc.

Differential Diagnosis

Confusion of cerebellar tumors with multiple sclerosis has frequently occurred. We know, indeed, that this last has in common with cerebellar neoplasms, the nystagmus, cerebellar ataxia, vertigo occurring sometimes in attacks, that further, scanning speech also occurs in cerebellar tumors, and that their pressure upon the motor tracts of the brain axis may produce increase of reflexes and spasticity. We might add that upon a few occasions an intention tremor has been observed in cerebellar neoplasms (whether as a symptom of irritation of the pyramidal fibers or not?). Very important for differential diagnosis is examination of the eye grounds; though there is exceptionally a swelling of the papilla in multiple sclerosis also (probably from complicating hydrocephalus). The headache in multiple sclerosis is scarcely ever as severe as in cerebellar tumors.

Particularly difficult is sometimes the differentiation of cerebellar tumor from cerebellar abscess and serous meningitis of the posterior fossa, in case the latter affections do not come on acutely with fever after an otitis or a head trauma. The absence of papilloedema in doubtful cases speaks always for abscess, while in serous meningitis this symptom occurs to almost the same extent as in cerebellar tumor. In favor of the diagnosis, serous meningitis, an outspoken remitting then exacerbating course may be in any event decisive. In the great majority of cases indeed, it is unfortunately impossible with tumor symptoms to exclude a "pseudo-tumor meningiticus." This, however, matters little, since therapeutically after failure of medical treatment directed against serous meningitis, an operation for this is indicated, and its finding after opening the skull may prove a pleasing surprise for us.

We might mention the also difficult differentiation from aneurisms of the basal arteries. Characteristic of the last is a vascular murmur within the skull synchronous with the pulse. A suspicion in this direction might be justified when there is advanced arteriosclerosis and increased blood pressure, also in alcoholism or syphilis, finally, after injuries to the skull (fractures of the base).

Treatment

We will proceed here according to the principles laid down already when speaking of tumors of the cerebrum, that is, at first we will try the resorption-favoring action of mercury and iodide against the scarcely ever to be excluded eventuality of serous meningitis. But not for too long a time. Where, after 3 or 4 weeks, definite reduction of the objective and subjective symptoms is not apparent, the continuance of internal treatment should not be persisted in, but trephining into the posterior fossa of the skull should be advised.

Even if the tumor then proves to be irremovable, nevertheless a long period of improvement may be the result of the decompression, in any case we can hope to relieve the patient of his unbearable headache and to prevent threatened blindness. Now there is in any event danger connected with simple decompressive craniectomy; so, a boy with glio-sarcoma of the vermis died suddenly in collapse, 24 hours after making an opening in the occipital bone, although he apparently recovered well from the operation, and the section showed, besides the tumor, marked hyperemia of the choroid plexuses with great hydrops ventriculorum—perhaps to be considered as a reaction to the operative removal of pressure. Many neurologists and surgeons have reported similar experiences and many on this account prefer, even in cerebellar tumors, to make the decompression in the temporal region of the skull; indeed, I have seen also in a case of sarcoma of the left cerebellar hemisphere subjected to temporal craniectomy, after a few days exitus from vagus paralysis in consequence of hydrops of the 4th ventricle. Naturally it may be objected to temporal craniectomy, that in it we either in advance renounce a curative operation in favor of a palliative one, or postpone the first by addition of the last and subject the patient to two operations instead of one. In so unfavorable a disease, in any case some courage is needed. Now as to the curative effects of the radical procedure, they are, as already expressly emphasized, dependent upon the time of the intervention, and many bad results would perhaps be avoided by early diagnosis. The pathologico-anatomical nature of the tumor is also important; cysts and fibroma give the best prognosis; after them, solitary tubercle (which can spontaneously cease growing), gliomata which pass over into normal tissue without sharp limitation, the worst; again, the location of the tumor plays a great rôle—so the operation for cerebello-pontine tumors in spite of their usually good limitation and often easy separation is, on account of the neighborhood of the medulla and the difficulty of access to the tumor, always a severe procedure, which is usually followed by death from heart failure. This happened in two cases observed by me, though the operation was undertaken with local anesthesia, and *F. Krause* in 50 operative cases, has lost 46 (that is, 92 per cent.). The operation in two stages is preferred by most surgeons upon intervention in the posterior fossa of the skull.

B. Abscesses of the Cerebellum

They occur like cerebral abscesses, either by metastasis (particularly after lung abscess and gangrene) or after injuries to the skull, which sometimes

have occurred years or decades before. Most frequently, however, cerebellar abscess is the result of purulent middle ear inflammation extending through the petrous bone and so presents the counterpart of otitic abscess of the temporal lobe. The development can be very acute, or the symptoms may arise gradually in the course of several weeks; finally there are very gradual forms which stretch over years. Through encapsulation such abscesses may indeed become latent; in these, however, sudden lighting up of the acute process which then usually leads to perforation of the abscess wall and to death from purulent meningitis, is frequent. The symptoms are headache, stiff neck, vertigo, vomiting, dysarthria, disturbances in swallowing, paralysis in looking toward the affected side (important for localization) ataxia. Rarer are nystagmus, hemiataxia upon the side of the abscess, papilloedema and loss of the patellar reflexes. Fever is important diagnostically, but no regular phenomenon; indeed, according to *Macewen*, subnormal temperatures occur. Against brain puncture which naturally is capable of confirming the diagnosis in the first place, *Fedor Krause* warns on account of the danger of an infection of the meninges. Operation is, of course, strictly indicated, but unfortunately offers only slight probability of cure.

C. Serous Meningitis of the Posterior Fossa of the Skull

The sacculations of the pia arachnoid in the neighborhood of the cerebellum—first, the "cisterna acustico-facialis" at the cerebello-pontine angle, second, the "cisterna cerebello-medullaris" between the medulla and the cerebellum—present a peculiar seat of predilection for retention cysts. These last are of inflammatory nature, depend upon the closing off of these preformed arachnoidal spaces along with corresponding inhibition of the resorptive processes and are caused by traumatic, otitic and syphilitic infection. Fig. 87 shows a case of such cystic serous meningitis formerly published by me, in an ideal sagittal cross-section in which trephining, with separation of the flat adhesions which stretched over from the region of the pyramis and the lobuli biventres cerebelli to the posterior limit of the medulla, had as a result disappearance of all the symptoms pointing to a space-narrowing process in the posterior fossa of the skull. When, six months later, attacks of vertigo and headache, dysdiadochokinesis, and slight cerebellar gait occurred again, an iodide cure caused rapid disappearance of these symptoms. There was plainly only a slight accumulation of exudate within partial adhesions in the operative field. Slight cases of serous meningitis can also, from the beginning, be brought to recovery by purely internal treatment (in which, even in nonsyphilitic cases, along with iodide of potassium, mercury certainly works favorably); however, such treatment if it gives no result should not be too long continued (especially when there is definite papilloedema). Also, a decided tendency to relapse may justify a decision to operate even in such cases as promptly react to resorbents, all the more, since the results of these operations are very good. This is shown not only by my case, but also by those of *Placzek-Krause*, *Oppenheim*, *Borchardt*, and others. Symptomatologically a differentiation from tumor of the cerebellum or of the cerebello-pontine angle is usually not possible. The

diagnosis can nevertheless be made from the anamnesis, the remittent-exacerbating course, also ex juvantibus. In my case, besides this, the relatively slight degree of cerebellar ataxia after one year's duration of the disease, the absence of spontaneous nystagmus, the exceedingly marked facial paresis; further, the comparatively equal distribution of the symptoms upon the right and upon the left, had so impressed upon me the idea of an extracerebellar and median location of the expansive process that I could make a probable diagnosis of serous meningitis and hydrops of the 4th ventricle. This last con-

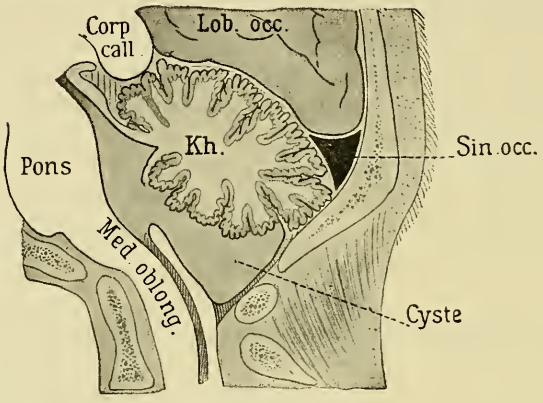


Fig. 87.

Serous Meningitis of the Posterior Fossa of the Skull. Ideal Sagittal Section.
Kh. = Cerebellum. Cyste = Cyst.

dition, as Fig. 87 shows, was in fact present and is a usual accompaniment of cysts in the region of the cisterna cerebello-medullaris.

D. Hemorrhages and Softenings

Hemorrhages into the cerebellum occur under the same conditions as those into the cerebrum (miliary aneurisms of the artery of the dentate nucleus play often an important rôle), but are, however, much rarer. The lamellar structure of the cerebellum brings it about that the blood breaks through outward or into the 4th ventricle. Softenings in the cerebellum are even rarer. This depends on the one hand upon the fact that the cerebellar arteries are given off from the basilar artery at an angle which does not favor the entrance into them of embolisms; on the other, there are numerous anastomoses between the individual arteries of the cerebellum. Slight hemorrhages and softenings in the cerebellum may manifest themselves clinically only by a short period of unconsciousness or an attack of vertigo and leave behind no residua, so that such lesions occasionally are noticed only as accidental autopsy findings. As prodromal symptoms of cerebellar hemorrhage, *Mingazzini* has mentioned occipital headache in two-thirds of his cases lasting for months, even for years. *Remak* has observed occasionally repeated severe vomiting. *Lussana* considers severe and repeated attacks of vertigo a sign of arteriosclerosis of the cerebellum. In very severe cerebellar hemorrhage, death may occur suddenly; *Mingazzini*

describes a case in which an old woman suddenly turned around several times, and then fell dead. In other cases of cerebellar hemorrhage, as well as in many instances of cerebellar softening, definite functional disturbances follow an ictus, for example, staggering gait, conjugate deviation of the eyes, nystagmus, forced turning of the head and neck. These phenomena can disappear by degrees.

E. Agenesis and Atrophies

Congenital cerebellar defects may remain latent even when they affect an entire half of the organ. If they extend, however, to both halves, they usually manifest themselves by cerebellar ataxia. A few cases, however, do not show this even. Sclerotic atrophies of the cerebellum can equally, although more rarely, remain latent clinically, especially when they occur at an advanced age and may be considered as degenerative processes without inflammatory basis. In general, however, they cause more severe atactic disturbances than the congenital ageneses, and even when they are only unilateral. Sometimes, besides this, other symptoms are also noticed, for example, speech disturbances and choreiform movements. Often such patients have epileptic attacks which indicates involvement also of the cerebral cortex—even when this escapes recognition by the histological methods at our disposal. In any event, combination of atrophy of the cerebellum with sclerotic atrophy in the cerebrum and spinal cord is not at all rare. Here the clinical picture is richer in symptoms, in that tremor, nystagmus, weakmindedness, paresis, etc., are also present. To enter into the different disease pictures which it has been attempted to isolate as cerebello-spinal, olivo-ponto-cerebellar, cerebro-cerebellar atrophies, etc., does not correspond to the practical end held in view in these lectures. It is a difficult and imperfectly investigated subject which is connected at many points with the hereditary forms of ataxia which we have discussed in Lecture VIII.

That even total defects of the cerebellum can remain latent, in case the rest of the central nervous system presents no defect, is explainable from the slight functional dignity of the cerebellum. This is taken strictly, neither motor nor sensory in function; neither is it possible, at least in man, to separate limited centers of specific activity. In animals there is, nevertheless, according to *Bolk*, *van Rynberk*, *Rothmann*, *Lourié*, and others, a certain cortical localization possible, which, however, with ascent in the animal kingdom, diminishes in definiteness. The results of the best experimenters, however much they differ from one another in point of detail, agree in showing that the cerebellum is only able to act in a modifying manner upon the cerebrospinal function. *Luciani* first attempted to determine precisely the influence of the cerebellum upon the rest of the nervous system and distinguished a sthenic, tonic and static action. The cerebellum, according to this author, reinforces the potential energy of the brain-spinal cord innervation, increases the neuromuscular tonus, and aids the continual blending of the motor impulses. The elimination of the cerebellum has on this account asthenic, atonic and astasic disturbances as a result. Then *Thomas* has defended the view that the triad of *Luciani* depends only indirectly on the suspension of cerebellar function.

He has shown that the cerebellum is a reflex center in the service of preserving the equilibrium, that it receives peripheral and central impulses and reacts to both, that it is not the seat of a special sense, but that of a special reaction, that this last, finally, serves equilibration in the different positions, both on reflex, automatic and voluntary movements. The animal without a cerebellum owes, according to *Thomas*, to his cerebellar ataxia, the weakness, incompleteness and disharmony of his muscular contractions also; must, indeed, to a certain extent, try his muscles out anew. Later, *Hermann Munk* has laid stress upon the fact that the ataxia of the animal without a cerebellum, in whom injury to neighboring structures has been avoided, is limited to the muscles of the vertebral column and the limbs. It affects the "composite movements" in the service of maintaining the equilibrium on standing and walking. This finding agrees best with the clinical studies on cerebellar ataxia. The preponderant importance of this last in the circle of all the cerebellar affections, even the ageneses and atrophies, stands in agreement with the result of my experiments in cutting the spino-cerebellar tracts in animals. As these tracts represent the afferent portion of the reflex for the cerebellar coördination of the trunk, so the cerebellar influence in maintaining the equilibrium of the head and neck is regulated through the vestibulo-cerebellar fibers. This, the experiments of *Ewald* and others have made clear.

F. Infectious Diseases of the Cerebellum

There is a cerebellar form of acute infantile paralysis into which we have already gone when mentioning the *Heine-Medin* disease. An encephalitis can also localize itself in the cerebellum; its differentiation from abscess of this region can be very difficult, even almost impossible, as is sometimes even that from cerebellar serous meningitis, although in such encephalitis a papilloedema occasionally occurs (*Oppenheim* and others). Italian neurologists (*Pansini, Forli*, etc.) have pointed out, finally, an acute cerebellar symptom-complex in consequence of malaria, in which after a sudden febrile beginning (with plasmodium in the blood) there appear cerebellar disturbance of gait, muscular atony, nystagmus, tremor, dysarthria, and swaying of the head. This syndrome reacts well to quinine, speedily disappears, but has a great tendency to recurrence.

LECTURE XXI

Malformations, Congenital and Early Acquired Defective Conditions

A. Hydrocephalus

In considering the disease condition known as hydrocephalus characterized by an excessive collection of fluid within the skull, we will in the first place confine ourselves to the congenital form. It is by far the most frequent, but is not always separable from the early acquired forms.

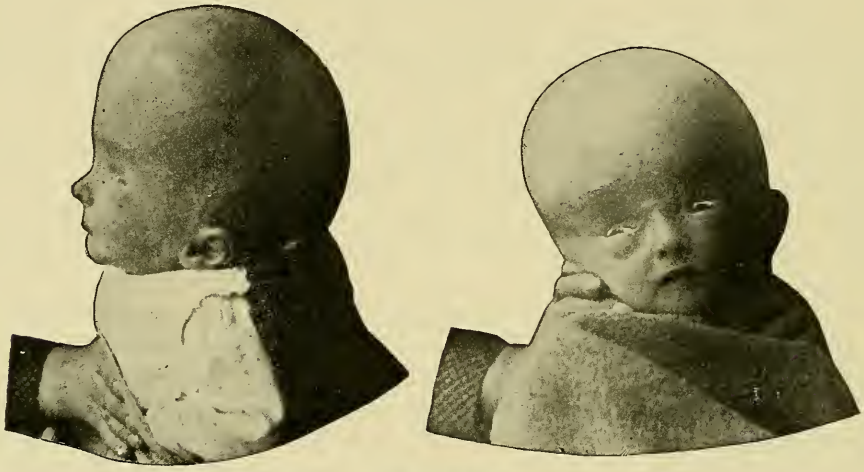
The pathological anatomist distinguishes a meningeal or external hydrocephalus from a ventricular or internal hydrocephalus. The first is rare, reaches on an average only moderate intensity, and is clinically not to be differentiated with certainty from internal hydrocephalus. As a rule, external hydrocephalus, also called "intra-meningeal hygroma," is a consequence of malformations of the brain, for example, microcephalus; we speak then of a "hydrocephalus ex vacuo." The possibilities for the production of a ventricular hydrocephalus are manifold. So, it can represent a standstill of brain development at the stage of membranous vesicle; on the other hand, however, it may be the result of a fœtal or early infantile ependymitis or meningitis (the choroid plexuses of the ventricle are morphologically the result of the invagination of the delicate membranes of the brain). Further, it can be a hydrops by stasis, through interruption of the venous discharge channel from the choroid plexus and the ventricle walls; finally, it is the result of the obstruction of those orifices through which the intra-cerebral cavities communicate with the subarachnoid space.

In the different directions hereditary syphilis appears to play the most important etiological rôle; *v. Barensprung* found, among 99 hereditary luetics, 4 congenital cases of hydrocephalus; *Elsner*, among 18 hydrocephalics, 3 children with manifest signs of syphilis; *Dean*, in 14 hydrocephalics, obtained 4 times a positive *Wassermann* reaction. In the second place, parental alcoholism must be mentioned. Rarely can trauma or acute infection which have acted upon the pregnant mother be made responsible. The connection with mental shocks during pregnancy is scientifically not proven. A few authors have called attention to hereditary family cases of the disease.

The ventricle walls can be so thinned in internal hydrocephalus that the brain has become a fluctuating vesicle. The amount of fluid varies between 50 to 100 cc and 10 liters—the average, according to *Oppenheim*, is about 1 liter.

No wonder that the skull can reach enormous dimensions. The circum-

ference of the head of the normal new-born infant, taken about the glabella and the external occipital protuberance, measures 34 cm, and in the course of the first year reaches 45 cm. In hydrocephalus circumferences of 60 and 70 cm are not rare. A 16-months-old patient of *Frank* had, indeed, a measure of 154 cm. Since only the skull proper enlarges while the face remains small, the head assumes a characteristic pear-shape; the thin bones of the skullcap can become parchment-like, the whole cranium is translucent by transmitted light. The sutures and the fontanelles gape open, the skin of the forehead and the scalp becomes thin and atrophic, and all the more prominent appear the dilated veins. Since in young children the neck muscles are not able to support the head of adult size, it wobbles helplessly about. Sometimes,



Figs. 88 and 89.

Congenital Hydrocephalus.

on palpation, fluctuation is perceived, while on auscultation a vascular murmur is heard.

The skull deformity exerts peculiar morphological reactions upon the eye and ear. The eyeball is forced down and forward, the upper lid can only incompletely cover it. A large portion of the sclerotic is always visible and the upper part of the cornea rising over the border of the lower lid gives a picture of the "rising sun." The (often much deformed) ears are placed strikingly far back and low on the head (see Figs. 88 and 89).

These great morphological anomalies correspond to marked functional disturbances. As to the intellectual capabilities, it is indeed a fact that after recovered slight hydrocephalus, striking mental capacity has been observed (which is known to have been the case in *Cuvier* and *Helmholtz*), but permanent more or less gross defects are the rule and feeble-mindedness, imbecility or idiocy is the usual lot of hydrocephalics who do not die in early childhood. Of 41 hydrocephalics, according to *Wyss*, only 5 could go to school. Disturbances of the cranial nerves are often observed, for example, dilatation or contraction of the pupils or lack of all reaction, nystagmus, strabismus,

grimacing (irritative facial symptoms?). In a remarkable manner hearing is almost always retained and only exceedingly rarely is there change in the optic nerve (papilloedema, atrophy). Spastic conditions of the muscles of the trunk and extremities are frequent; they correspond to the clinical picture which we will describe in the next lecture on infantile cerebral paralyses and *Little's* disease. Epileptiform attacks, paroxysmal vomiting and the occurrence of severe headaches should indicate sudden increase of brain pressure. Young children give evidence of the last by whimpering and putting the hand to the head. Unusual symptoms are disturbances of coördination, tremor, paralyses, and (apart from the not infrequent hyperesthesia of the integument) disturbances of sensibility. Frequently, on the contrary, there is incontinence of fæces and urine, as well as a decided tendency to bed-sores; the last occur upon the skull also (parietal bosses, occiput).

A word on the hydrocephalic fluid which is obtained on lumbar or brain puncture. It is, as a rule, colorless to a slight amber tinge, without clouding, only rarely it contains a few flocculi. The content of albumen (globulin) is practically none, or very little (not over 1 per cent.). The specific gravity lies between 1,001 and 1,009. The fluid contains further glucose and milk sugar. The cytological findings are limited to the occasional presence of a few leucocytes.

How relatively frequent hydrocephalus is at birth is shown best by the statements of the obstetrician *Runge*, according to which a disturbance in delivery in consequence of hydrocephalus comes under observation about once in 300 deliveries. In this connection it should be remembered that a great many congenital hydrocephali are at birth of still too moderate dimensions to cause any disturbance. In any event, the post-natal growth of congenital hydrocephali plays a great rôle; this can amount to 1 cm a week and corresponds to an extra-uterine duration of the original disease process. The mortality of hydrocephalics is a very considerable one. At birth there may be bursting of the head if the child is not sacrificed by the obstetrician by perforation and cranioclasis to prevent rupture of the uterus. Hydrocephalics born alive often die during the first days, weeks or months; the remainder of the more severe cases are taken away before the third year of life; only slight cases can recover; usually, however, it is a "recovery with defect." Not to be underestimated is the danger of relapse, renewed progressive advance which threatens during all of childhood. As immediate causes of death should be mentioned: increasing brain pressure and coma, bed-sores and infection, laryngismus stridulus; status epilepticus, attempts at operative treatment, particularly, however, intercurrent affections, as gastro-enteritis and bronchopneumonia. In one case there was bursting of the head. Rare but interesting are the so-called "spontaneous recoveries" in which the hydrocephalic fluid, by wearing away of its enclosing case, makes a path out either through the nose, through the orbit, or through the coronal or sagittal suture.

From the point of view of differential diagnosis, there come into consideration before everything else the rachitic enlargement of the skull, in which, however, the cranium assumes the characteristic "box form," the open fontanelles are not bulging, and the hydrocephalic position of the eyes is absent;

also the so-called "steeple-skull" (oxycephalus) should not lead to confusion with hydrocephalus, although it is affirmed that this deformity represents the reaction of the rachitic bones of the skull to a moderate hydrocephalic pressure (*Meltzer*). It is to be emphasized that oxycephalus is almost never accompanied by idiocy, but, on the contrary, very frequently by optic atrophy. We would mention further the peculiar skull-form of hereditarily syphilitic children, which *Fournier*, on account of the abnormally high and bulging forehead, has called the "Olympic" forehead ("front olympien"), and would allude further to the idiots with sclerotic hyperplasia of the brain and great skull dimensions denominated by *Virchow* "cephalones," and finally, the so-called "cleido-cranial dysostosis," a heredo-family anomaly in formation described by *Marie* and *Sainton*, in which persistence of the fontanelles and excessive width of the skull is accompanied by congenital defects of the clavicles.

The non-congenital hydrocephalus of childhood is most frequently of meningitic origin, which can manifest itself clinically by acute onset with fever, stiff neck and convulsions. After the passing off of this infectious stage, during which the spinal fluid is usually characterized by richness in albumen, and has also been found to contain bacteria (for example, meningococci), there occurs a progressive increase of fluid within the skull which, by degrees, may produce the same symptom-complex as the congenital form. Only in children beyond the second year of life the firm closure of the sutures and fontanelles which has already occurred, causes a plainly different disease picture. Only very exceptionally are the sutures forced open; as a rule, indeed, the skull responds to increase of pressure in its interior by abnormally rapid growth; nevertheless, such monstrous dimensions as those in hydrocephalus of the first period of life are not nearly reached, neither does the typical eye position occur. All the more intensely, however, does the increase of the intracranial hypertension make itself felt; severe headaches, tinnitus aurium, vertigo, vomiting, confusion, optic atrophy, spastic rigidity of the extremities, namely, the lower ones, epileptiform attacks, etc., make the differential diagnosis from brain tumor uncommonly difficult. Here and there, indeed, the hydrocephalus distinguishes itself from tumor formation by considerable variation in the intensity of the disease manifestations.

Into the treatment of hydrocephalus we can enter with but very faint hope. In every case we begin with iodide and mercury, which even in non-syphilitic cases appear to exercise a favorable influence. From the start large doses are called for (0.15 to 0.2 KI per diem for children in the first year, 0.2 to 0.25 in the second year, ungt. hydrarg., 1.0 a day by inunction). Sublimate baths (1.0 $HgCl_2$ per bath) have but slight activity, internal mercurial treatment is not to be recommended on account of the tendency of such children to gastro-enteritis.

Derivative measures whose activity cannot be entirely denied are penciling the scalp with tinct. iodi, and the exposure of the occipital region to the sun for 15 to 20 minutes a day as recommended by *Somma*.

More rational, however, is repeated lumbar puncture (25 to 50 cc spinal fluid drawn off at intervals of 3 weeks), while the incision of the dural sac

to produce permanent drainage, as recommended by *Quincke*, does not appear to promise much result. Puncture of the ventricles is reserved for those cases in which increasing symptoms of brain pressure or papilloedema appear to call for relief, but in which on account of imperfect communication between the ventricles and the spinal dural sac this cannot be accomplished by lumbar puncture. Ventricular puncture is, indeed, a dangerous procedure, which frequently leads to fatal collapse or to status epilepticus; in any case the following precautions should be observed: the most rigid asepsis, puncture 3 to 4 cm laterally from the large fontanelle, that is, below the longitudinal sinus; slow withdrawal of at most 100 cc fluid; accurate watching of pulse and respiration; in threatening collapse a partial replacement of the fluid by physiological salt solution. After the operation an elastic, yielding bandage should be placed about the skull, but the dangerous compression by adhesive plaster, after *Trousseau*, should be avoided (brain pressure, bed-sores). In conclusion, the methods of "permanent drainage of the ventricle," recommended by surgeons, may be mentioned; these are: draining into the subdural space (*Kocher*), under the scalp (*Mikulicz*), into the longitudinal sinus (*Payr*), and finally, the so-called "Balkenstich" (puncture through the corpus callosum) (*Anton* and *Bramann*).

Since in marked hydrocephalus the existing defects in the psychical sphere, even though the disease process may cease, can only to the slightest extent be removed by any method of treatment, an ethically thinking physician will enter into the task of making possible the further vegetative existence of a paralyzed and idiotic being only with repugnance.

B. Cranial and Spinal Ectopies

Upon the basis of preformed defects of the skull partial escape of its contents can occur intra-uterine and be present at birth. We speak of a cranial meningocele when only the brain membranes are displaced outward (in this the dura mater is usually absent), of hydrencephalocele when the hydropic ventricle also protrudes. By encephalocele is meant, on the contrary, an ectopy of solid brain substance which does not include any part of the ventricle. These malformations can be located on the most varied parts of the skull. They are most frequently found in the occipital region, sometimes above and sometimes below the squama occipitalis (see Fig. 90); again they appear between the nose and the orbit or between the nose and the frontal bone. Ectopies into the nasal cavity, the mouth and in the temporal region are rarer. Cystic protrusions of the brain and brain membranes are greatly increased in dimensions after birth. Pedunculated are the meningoceles, which occur particularly in the occipital region. This form is the mildest, since it

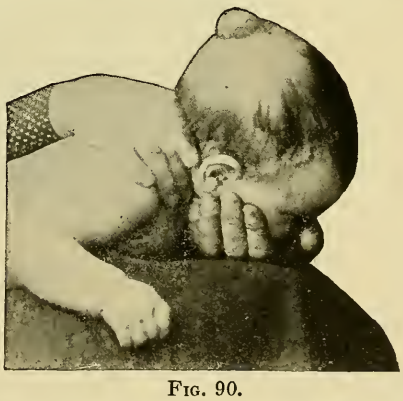


Fig. 90.
Superior Occipital Hydrocephalocele.

is compatible with normal brain activity. Often enough, indeed, it is combined with hydro- or microcephaly which, along with other developmental disturbances, are almost regular accompaniments of the ectopies containing brain substance. The worst prognosis is given by the hydrencephaloceles; their post-natal growth provokes brain pressure symptoms which lead to death within a few months.

Good results can be expected only from the operative removal of simple meningoceles; these should be early transferred to the surgeon, as otherwise

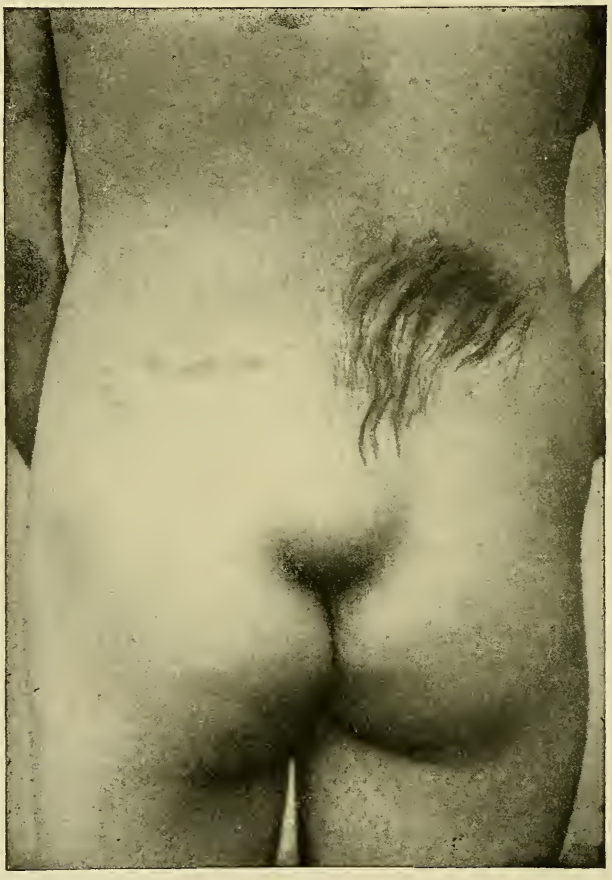


Fig. 91.

Occult Spina Bifida with Local Hypertrichosis.

there is a risk of rupture of the cyst with unavoidable meningeal infection. Spontaneous cures are excessive rarities. As clinical criteria of meningoceles it is noticed that they fluctuate always in contradistinction to the encephaloceles, and they are distinguished from hydrencephaloceles by being usually pedunculated and often translucent; also they decrease in size on pressure, and can even be replaced. In the last method of examination there is, indeed, need for great caution on account of the danger of provoking brain pressure.

We include the spinal ectopies under the common designation of "spina

bifida." If the skin over the hernia of the spinal cord or of the meninges is present, even though of abnormal appearance (*e.g.*, showing hypertrichosis or drawn in like a stellate cicatrix), we speak of occult spina bifida (see Fig. 91); if the hernia is exposed, however, of open spina bifida, or "Rachischisis" (Fig. 92). For the rest, we also distinguish here the simple meningoceles from the forms which contain nervous tissue. The spinal analogue of hydrencephalocele is called myelocystocele. The most pronounced anomaly, however, is presented by myelomeningocele, in which the spinal cord, fissured and drawn apart, is exposed externally and closes dorsally the summit of the meningocele sac as the so-called "Zona medullo-vasculosa." Spinal ectopies can reach the size of a child's head. They are usually situated in the lumbo-sacral region which corresponds with the embryological fact that closure of the spinal canal takes place from above downward.

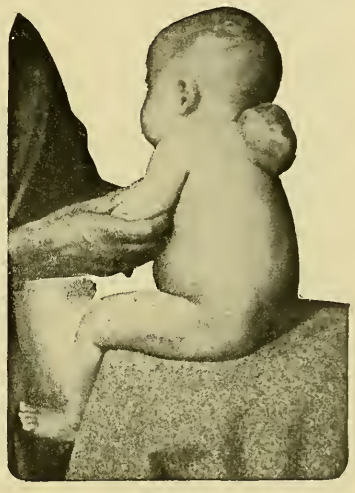


Fig. 92.

Open Spina Bifida (Cervical Meningocele).

Clinically, in so far as there is not occult spina bifida, the tumor naturally first strikes the eye. It is characterized usually by definite fluctuation and partial replaceability. In the last test great caution is necessary on account of the danger of brain pressure; on this the fontanelles are often noticed to protrude. On the other hand, when the child cries or strains, the tumor sac becomes more tightly stretched.

Myelomeningocele, which is almost always accompanied by serious paralytic symptoms, is easily to be recognized by the deep red granulating "zona medullo-vasculosa"; the differentiation of the other types, even with the assistance of the X-ray, is scarcely possible before the operation. Of the symptoms of motor involvement, in consequence of spina bifida, symmetrical paralyses in the legs are next to be mentioned. Clubfeet are practically always present. Many times not only are the feet and legs paralyzed, but there is complete paraplegia—with the usually deep situation of the tumor, of flaccid character. The electrical reactions may be lost, the patellar reflexes are usually markedly reduced, the Achilles reflexes are generally absent. In cervical spina bifida I have also seen spastic paraplegia—a very rare occurrence. Even when the process is located high up, the arms usually remain free; on the other hand, bladder and rectal paralyses belong to the typical symptom-complex of spina bifida. In their most severe form, these symptoms are found in the spinal hernias situated low. Disturbances of sensibility are usually but slight, and only exceptionally is total anesthesia found in the paralyzed limbs. Finally, we may mention certain trophic disturbances (ulcers on the genitals, on the heels, over the tumor itself), etc., here and there also perforating ulcers on the feet.

Occult spina bifida takes a special position, in that its symptoms develop only in late childhood, even after the tenth year. There occurs, then, in

previously healthy individuals, little by little, either bilateral pes equinovarus, weakness of the sphincters appears, which can increase to total incontinence, or, and this is the usual thing, both these morbid conditions develop spontaneously, accompanied by more or less severe pain in the legs. This is probably a result of the traction which the spinal cord fixed at the point of the hernia must suffer from displacement by growth. In many such cases lumbar hypertrichosis (see Fig. 91), a scarlike depression or a slight bulging of the skin in the lower part of the back would point to the diagnosis of occult spina bifida, which naturally must be confirmed by X-ray examination.

The "myelodysplasia" of *Fuchs* can be considered as a rudimentary form of occult spina bifida under which is understood the following symptom-complex: 1. Weakness of the sphincters which leads to continued nocturnal enuresis. 2. Syndactylism, or membrane formation between the individual toes. 3. Disturbances of pain sense in the toes. 4. Anomalies of the skin and tendon reflexes on the abdomen and in the legs. 5. Occasional deformities of the bones of the foot, eventually combined with weakening of the peroneal muscles. 6. Roentgenological evidence of imperfect closure of the sacral canal, defects of the vertebral arches, etc. 7. Exceptionally, tropho-vasomotor disturbances on the toes.

Spina bifida is often complicated by other malformations and defect conditions, for example, by hydrocephalus, ectopy of the bladder, defects of the abdominal walls, etc., which naturally unfavorably influence its prognosis. For the rest, one can say, the greater the involvement of the spinal cord, the worse the prognosis, even after operative procedures. Without operation the outlook is very bad; *Wernitz*, in 90 patients with spina bifida, saw only 20 live to be over 5 years old; the majority died during the first month of life. That any one reaches middle age is excessively rare. Spontaneous disappearance of the tumor by contraction is so extraordinary that this eventuality need not be reckoned with; the same thing applies to spontaneous recovery by rupture of the cyst. The last accident is usually, indeed, fatal. If the cyst ruptures before birth, the children may be born with a fistula which, left to itself, is usually sooner or later the port of entry for a meningeal infection. Every case which is not accompanied by other serious malformations should be brought under surgical treatment.

C. Congenital Nuclear and Muscle Defects

In the cranial nerves, and the muscles supplied by them, congenital ageneses are not so very rarely observed. Most frequent is congenital ptosis, which may be uni- or bilateral. Rarer are the following congenital paralyses: total external ophthalmoplegia, paralyses of looking in one direction, abducens paralysis, paralysis of the superior rectus; facial paralysis, paralysis of the muscles of mastication and of the tongue; single or combined, unilateral or bilateral, symmetrical or asymmetrical. There is also a so-called "congenital bulbar paralysis." For these absences of function, as anatomical investigations have shown, there may be, as a basis, either defects of the muscles concerned or those of the nuclei of the cranial nerves supplying them. Now, there is

in these last, however, an "infantile nuclear atrophy" which is to be referred not to agenesis, but to an early atrophy of these structures; these two forms, indeed, are in principle not to be separated from one another, since for the early infantile atrophy a congenital defect should be a prerequisite also. In the paralyzed muscles the electric irritability is reduced or lost, still in congenital defects reaction of degeneration is not found.

The prognosis of all these conditions is, that they will remain stationary. The relatively slight functional disturbance which results from defects of the eye muscles is noteworthy: double vision scarcely ever occurs, and in congenital abducens paralysis, contrary to what is observed in the acquired form, there is usually no contracture of the antagonists; the eye, on account of this, remains in the right position as long as the glance is not turned toward the paralyzed side. Therapeutically, only plastic operations, as, for example, transplantation of the frontal muscle into the levator palpebræ superioris, come into consideration.

The congenital muscular defects on the trunk and extremities are, as far as anatomical investigations up to this time permit a decision, always of peripheral nature, that is, not dependent upon ageneses of the nuclei in the spinal cord. These conditions, which *Erb*, *Damsch*, and I have recognized, attract clinical interest less by their symptomatology than through their relations to progressive muscular dystrophy. It is striking that the muscle defects are observed in the first place in such muscles as usually atrophy frequently and early in progressive muscular dystrophy. By far the most frequent is defect of the pectoralis, which *Schlesinger*, among 54,000 patients of one Vienna clinic, found 5 times; I have seen 6 cases in the course of 10 years; over 200 are described in the literature. The fact that, like in the scapulo-humeral type of dystrophy, the clavicular portion of the pectoralis major usually is preserved, is remarkable. Although rarer than isolated muscle defects, defects of whole groups of muscles also occur; also, these affect predominantly such muscle complexes as are known as typical localizations of the dystrophies; in a boy observed by me, for example, the left pectoralis major was absent (exceptionally, in toto), also the triceps brachii, a part of the trapezius and the rhomboids. I do not go so far as *Erb*, and particularly *Damsch*, who raise the question as to whether congenital muscle defects may not be the result of an intra-uterine variety of dystrophy. It appears to me, however, that between the total inhibition of formation, on the one hand, and the congenital predisposition to later dystrophic destruction, there is only a difference in intensity. Repeatedly it has been observed for the rest, that people with congenital muscle defects in the course of their later lives develop progressive muscular atrophy. Remarkable is the excessively frequent accompaniment of congenital muscle defects with other malformations of all sorts, which in defects of the pectoralis are usually located about the shoulder girdle, thorax, or arm.

It is also striking here how small the disturbances of function are; so, one patient with left-sided pectoralis defect, fenced with the left hand, another shone as a rider and swimmer. These are, of course, examples of the formation of vicarious synergies by the retained muscles or portions of muscles.

Here, also, treatment has to take hold. In the boy above mentioned, by faradization of the retained portions of the shoulder girdle and by suitable exercises I brought him to the point of being able to execute the previously impossible, for him, "dip" on the parallel bars.

LECTURE XXII

Infantile Spastic Hemiplegia and Diplegia; Little's Disease; Idiocy

GENTLEMEN: We will to-day review a number of disease conditions which etiologically and pathogenetically quite heterogeneous, are distinguished by common clinical features. These are always the result of injurious factors which have affected the nerve centers either prior to birth, during this, or in the course of earliest childhood, and have led to spastic-paretic symptoms of hemi- or diplegic type; the syndromes which have so arisen are not progressive, but represent either a stationary residual condition or even manifest tendencies to spontaneous improvement. Within these rather wide nosological limits we include as "*Little's*" disease the quite frequent cases which are characterized, 1, by diplegic type, and, 2, by the so-called *Little's* etiology. As early as 1846 the English obstetrician *Little* had pointed out the predisposition which premature births, multiple births, and difficult deliveries seem to exert toward the development in the children in question of bilateral "spasticity of the limbs."

In order to introduce some system into this rather complicated matter, I must first make you acquainted with the different pathologico-anatomical bases of these symptom-complexes. I will divide them according to *B. Sachs*, into three categories: prenatal, natal, and post-natal lesions. It is to be remarked, however, that in individual cases, usually not the autopsy, but a sufficiently clear and extended anamnesis renders possible its inclusion in one of these groups.

1. Prenatal Lesions

a. Porencephaly.—As "true" or "primary porencephaly" we denominate a craterlike depression of the brain surface which sinks in toward the ventricular cavity to communicate with this latter. This defect arises in the intra-uterine period through the abnormal depth or infolding of the primary fissures at an early embryonic period, or from fœtal encephalitis and meningitis. These last sometimes lead also to "secondary" or "pseudo-porencephalies," cystlike defects of substance, in consequence of the shrinking of cicatrices on the convexity of the brain. Porencephalies may have a unilateral or a bilateral symmetrical location.

b. Lobar Sclerosis.—In this condition, described by *Virchow* as "congenital encephalitis," there is a gliotic contraction and induration of the totality

or of the greatest part of one or, indeed, of both hemispheres. It seems to present a parallel process to the encephalomalacia of the developed brain. In its early stage, the glia tissue remains more readily preserved after processes leading to ischemia and hence, after destruction of the nervous parenchyma, can proliferate reactively, and after that contract. The diffuse fœtal scleroses, hence are very closely related to the prenatal porencephalies, but owe their origin to a less degree of intra-uterine disturbance of the blood supply. Lobar sclerosis can also affect only one or both halves of the brain.

c. Tuberous Sclerosis.—This anomaly was formerly considered as the hypertrophic contrast of atrophic lobar sclerosis. According to the newest investigations of *H. Vogt*, however, it is a malformation resembling that in tumors. There are found in the cortical regions, particularly in the sensorimotor zone, superficial knotty prominences which may reach the size of a nut and consist of exceedingly proliferated glia. *Vogt* has pointed out also the frequent finding of congenital heart and kidney tumors (rhabdomyoma, hypernephroma, liposarcoma), as well as congenital adenoma sebaceum of the skin, in patients with tuberous sclerosis. These last render possible the diagnosis of this lesion *intra vitam.*

d. Cysts and foci of softening in the brain arise from the causes mentioned when speaking of lobar sclerosis, rarely before birth; where this is the case, obstruction of arteries, usually upon a syphilitic basis, are to be held responsible as causes for the circumscribed destruction of brain substance. These are usually unilateral lesions.

e. Spinal foci of disease which lead to descending degeneration of the pyramidal tracts; these are usually, as was recognized in two cases anatomically examined by *Dejerine*, also of syphilitic origin.

2. Natal Lesions

These furnish, when bilateral, the anatomical substratum of *Little's* disease.

a. Development of the cortico-spinal tracts interrupted by premature birth. This defect appears in many cases which later strikingly improve spontaneously, capable of being compensated to a certain degree through supplementary development of these tracts.

b. Hemorrhages into the brain substance, or upon its surface, or their residues in the shape of cysts and areas of softening, on the one hand, of adhesions, with the meninges on the other. The hemorrhages occur (often bilaterally) by tearing of the blood vessels in a difficult labor or through too sudden passage of the still soft skull through the superior strait of the pelvis in premature delivery. Factors favoring them are asphyxia, twisting of the umbilical cord, brittleness of the vessels on account of fœtal or maternal diseases.

c. Cerebral meningitis and encephalitis post partum, which may occur from the infection of hematomata, in case the abrasions, bruises, and circulatory disturbances which the skull and its contents suffer in difficult labor, do not suffice to open the way for the invasion of microörganisms. The results of such meningeal infections can be, among other things, the abnormal folding

of the cerebral cortex known as "microgyria," external, or even internal, hydrocephalus, as well as pseudo-porencephaly.

d. Hemorrhages into the spinal cord or its meningeal envelopes. These have been observed after breech delivery and extraction, after turning, etc.

3. Post Natal Lesions

1. *Pseudo-porencephalies* arising from embolic, encephalitic, and meningo-encephalitic processes, also through trauma (falls on the head, striking it on sharp edges, etc.).

2. *Lobar sclerosis;* 3, *tuberous sclerosis*, rarer than that of prenatal origin, but still agreeing with it pathogenetically.

4. *Cysts and Foci of Softening.*—These changes are relatively frequently the bases of spastic diplegias and hemiplegias arising in the course of earliest childhood. They depend upon vascular lesions (rupture or thrombosis of pathologically altered vessels, embolism).

5. *Acute Polioencephalitis of Childhood* (*Strümpell*).—The relatively rare localization of the infectious process also lying at the base of acute poliomyelitis, in the cerebral cortex already considered by us (see Lecture XVI, page 242), occurs unilaterally and leaves behind infantile spastic hemiplegia.

Etiology

Apart from the last-mentioned infectious disease, as well as from the already mentioned "*Little's* etiology" (premature delivery, multiple births, pathological delivery, with the injuries resulting from them) specified in speaking of the natal lesions, congenital syphilis plays the chief rôle* among the causes of infantile spastic hemi- and diplegia. Most of the just sketched pathologico-anatomical anomalies can even at the autopsy be brought into connection with existing heredo-syphilis, which, on account of the well-known teratogenic action of the luetic virus, on the one hand, and its injurious effect upon the vessels on the other (endarteritis syphilitica), is not astonishing. That luetic patients do not bring into the world hemiplegic or diplegic children much more frequently than is actually the case, *Sachs* explains by the frequency of abortion in such families. For the rest, enough cases are found in which there have been a number of abortions, while the child finally carried to term presents one of the above-mentioned prenatal lesions. In the prenatal, as in the postnatal forms, *Box* has recognized that they frequently give a positive result with the "four reactions" (see above, page 187). Also tuberculosis of the parents is not rarely found, as is saturnism and alcoholism of the ascendants. In congenital cases exhausting diseases of the mother during pregnancy can often be held responsible. There is often a history of physical or psychical trauma which has been suffered by the pregnant woman. Value is to be placed upon these anamnestic statements mainly when it is recognizable that a gross trauma has affected the uterus. *Sachs* mentions also uremia of

* Indeed, it is to be held responsible for many cases of *Little's* disease (predisposition to premature delivery, brittleness of the vessels in luetic fœtuses).

the mother. First-born children are predisposed to injuries at birth and to *Little's* disease. On the other hand, as *Ganghofner* and *Freund* have shown, intra-uterine injurious factors frequently affect the last children in a large family (exhaustion of the maternal organism through overexercise of the generative function).

We will now proceed to the description of the individual forms of infantile hemi- and diplegia.

A. HEMIPLEGIA SPASTICA INFANTILIS

(Unilateral Form of Infantile Cerebral Palsy)

The great majority of cases arise postnatally during the first year of life, and usually acutely febrile with hebetude, headache, often also vomiting and convulsions. After these symptoms pass away a spastic half-sided paresis, usually with inclusion of the face, becomes evident. Generally the leg is less affected than the arm; the patients learn to walk again with the leg slightly bent at the knee, adducted, and rotated inward with the foot fixed in equinovarus position; the latter drags and is "circumducted"; the arm, however, and particularly the hand, remains incapacitated. In general, the same tendency to contracture is evident in it, as in the cerebral hemiplegia of adults, the arm is pressed against the side, flexed at the elbow-joint, and there is pronation and flexion of the hand and fingers; only a fixation of the hand in a position of hyperextension is sometimes found. Hypertonia, exaggeration of the tendon reflexes, *Babinski's* phenomenon, associated movements, etc., are naturally present. In contradistinction to what is the case in adults, marked trophic disturbances usually manifest themselves in the paralyzed extremities; they are delayed in development, the X-ray picture shows a more or less marked osteoporosis; along with the skeleton, the muscles also appear atrophic, which is not only to be considered as a result of inactivity, but as a "cerebral muscular atrophy." Reaction of degeneration is never present. Fingers and toes are often the seat of athetoid, more rarely of choreic movements (see Lecture V, pages 80 and 85); unilateral tremor also appears. Sensibility is, apart from atactic disturbances of the paralyzed limbs, usually intact. Aphasia is, even with paralysis of the extremities on the right side, very rare, which is connected with the early age of the patient. The patient, early deprived of his cerebral cortex on the left side, becomes not only left-handed, but also learns to talk with his right hemisphere. Accompaniment of hemiplegia spastica infantilis by disturbances of intelligence and epilepsy is an exceedingly common occurrence.

B. DIPLEGIA SPASTICA INFANTILIS

1. Bilateral Forms of Infantile Cerebral Palsy

Among these forms, according to *Freud's* example, 3 types may be distinguished:

a. The Bilateral Hemiplegic Type.—This presents a doubling of the infantile hemiplegic symptom-complex (in which, however, differences in intensity

between right and left usually exist), and is by far the most severe variety of infantile cerebral diplegia. Also bilaterally innervated cranial nerves can be paralyzed, on which account pseudo-bulbar paralytic phenomena (see above, page 229) occur. The arms are more severely affected than the legs and the psyche intensely injured; athetosis is frequent (see Fig. 93).

b. Paraplegic Type.—The majority of these cases are congenital, whether they owe their origin to prenatal lesions or are to be referred to injuries at

Fig. 93.

Diplegia Spastica Infantilis ("Bilateral Hemiplegic Type"; Idiocy, Athetosis).

birth. The last represent a particularly large contingent of the cerebral form of *Little's* disease.

Many cases are noticed even in nurslings, since when dressing them, bathing them, etc., the legs are held abnormally stiff and immovable. Usually, however, striking disturbances first become apparent when the child should begin to take his first steps. It is then noticed that the thighs are held in forced

adduction pressed together, or indeed crossed like a pair of scissors, the knees are moderately flexed, the feet in equinovarus position with the toes turned in; upon attempts at walking, the knees and the toes rub together. The legs are decidedly hypertonic, and oppose great resistance to passive movements. If the child is placed upon a chair they do not hang down, but remain more or less stretched out straight.

As the child grows older, these disease manifestations not rarely improve, so that, indeed, occasionally in the 6th or 8th year of life locomotion may be nearly normal. Still the children usually long retain walking on the toes and a slowed and laborious rhythm in progression. The legs are delayed in development not at all, or only inconsiderably, in contradistinction to the hemiplegic form. Indeed, muscular hypertrophy, as a result of the hypertonia, has been observed (*Ibrahim* and others). Exaggeration of reflexes, *Babinski's*, often also *Oppenheim's*, and the *Mendel-Bechterew* symptoms, are to be found in the legs; on the other hand, the arms usually remain free from every anomaly, or they are only slightly affected (somewhat slow movement, exaggeration of reflexes). Sensibility is, as a rule, intact; on the other hand, strabismus and weakmindedness are present.

You will note that in the syndrome just sketched the paretic symptoms are much less manifest than the hypertonic ones. *Freud* sustains with good arguments the view that the paralytic symptoms are the more marked the deeper subcortical the hypertonic phenomena, the more decided the more superficially cortical the lesion lies. No wonder that the "paraplegic rigidity" is so preponderantly frequent in *Little's* birth-palsy, which usually presents the correlate of meningeal hemorrhages. Where, however, in cerebral spastic diplegia in childhood, paresis preponderates over hypertonia (also a "paraplegic paralysis" exists), there are usually deep-seated prenatal lesions.

c. "*General Rigidity.*"—This also usually falls under the "cerebral cases of *Little's* disease." All four extremities are affected; the arms, however, much less than the legs; also there are never in them fixed contractures, as in infantile spastic hemiplegias. Hypertonia considerably preponderating over the paretic phenomena stands in the foreground of the clinical picture. Almost always disturbances of speech and intelligence as well as defect symptoms upon the part of different cranial nerves, for example, strabismus, are present.

2. Spastic Spinal Infantile Paraplegia

Here belongs chiefly the "spinal type of *Little's* disease," in which along with the characteristic paraplegic rigidity of the legs, no other anomaly can be found, namely, disturbances of intelligence, strabismus, speech disturbances, choreic or athetoid phenomena, epilepsy, etc., are without exception absent. In the spastic paraplegias arising prenatally the paresis of the legs stands far in the foreground as compared to their rigidity.

Freud assigns to infantile palsy also cases of persistent uni- or bilateral chorea as well as "double athetosis" in early infancy (see Lecture V, page 86). The clinical elements of hypertonia and paresis are here replaced by

spontaneous movements. The recommendation to record these forms as "infantile cerebral palsy without paralysis", I cannot accept.

Prognosis

In the prognostic estimation of the spastic paralyses of childhood, it is well to draw a distinction between the motor symptoms and their eventually accompanying symptoms (among which the psychical anomalies and epilepsy

Fig. 94.

Diplegia Spastica Infantilis ("Paraplegic Rigidity," Feeble-Mindedness, Speech Disturbance).

are full of significance). These last always give a very poor outlook as to improvement or recovery, epilepsy having, indeed, a tendency to grow worse with advancing age, and its appearance being threatened up to adult life, even in such cases which at first do not present this complication. The hemiplegic and diplegic symptoms, on the contrary, are never progressive, frequently, indeed, regressive. Intact intelligence improves the prognosis very considerably, since treatment, as we will see, cannot dispense with the coöperation of conscious impulses of the will and its consequent exercise on the part of the patient. Choreic and athetoid phenomena usually persist, but

the patients often know how to reckon with them in a remarkable manner, and, in spite of them, use their limbs for all sorts of acts. *Little's* and the postnatal forms, particularly, however, "paraplegic rigidity" and the spinal form, are to be estimated as entirely more favorable than the results of intrauterine disease; where in the last there are serious defects, death usually occurs during the first months of life. I have also repeatedly seen children with congenital cerebral palsy develop later (at the time of puberty) severe organic nervous diseases or die (for example, with fibro-sarcoma of the lumbar cord, or purulent meningitis of the convexity of obscure pathogenesis). However, the prognosis of spastic hemiplegia and diplegia of infancy, as to life, is favorable.

Treatment

A causal treatment comes under consideration in a large number of cases of uni- and bilateral spastic paralysis in children, in which heredo-syphilis has played a part. Anti-syphilitic treatment should be begun as soon as possible and should not be carried out, as is so often the case, with inadequate methods. The sublimate baths (0.5 to 1.0 hydrarg. bichloride per bath) preferred by many pediatrists, for example, are at most of use as an adjuvant. Of a 1 per cent. solution of iodide of mercury (hydrarg. biniodid, 0.01; sodii iodid, 0.01; sodii chlorid, 0.08; aq. dest., 10.00), 0.2 to 0.5 cc, is injected every second, third or fourth day in very young infants; in older infants 0.5 to 0.1 cc is injected every second day; a course of treatment lasts 6 weeks. Internally, hydrarg. protiodid or calomel can be given in milk; the doses are for the first three months: 0.001 (gr. $\frac{1}{60}$), t. i. d.; for the rest of the first year, 0.003 to 0.006 t. i. d. (gr. $\frac{1}{20}$ to gr. $\frac{1}{10}$). For the second year 0.0075, for the third 0.01, t. i. d. As a substitute for the inunction treatment *Bruno Bloch* recommends the "plaster treatment" as very simple: one extremity or a correspondingly large portion of the trunk is covered with mercurial plaster. The plaster is left on for a week.

In cases beginning with fever during the early years of life antiphlogistic treatment is in place as long as the acute stage lasts; ice bag to the head, free purgation by several doses of calomel, eventually the application of leeches behind the ears. For the convulsions, enemata of the following composition may be given with advantage: Chloral. hydrat., 0.4 (gr. vii); pulv. camphoræ, 0.1 (gr. $1\frac{1}{2}$); vitell. ovi I; aq. dest., q. s. ad 200.0 (℥ vi).

As to the causal therapy of the initial stage of *Little's* disease, the operative removal of meningeal hematomata after protracted labors has been undertaken by *Cushing* and other surgeons: the children did not develop *Little's* disease (perhaps they would not have done so anyhow).

The treatment of the later stages in all spastic hemiplegias and diplegias of childhood is above everything else by curative gymnastics and exercise therapy. All the measures to be applied by neurologists, orthopedists and surgeons have the one end of shaping the conditions for the regression of the spastic resistance and the compensation of the paresis as favorably as possible. As already said, what may be expected from our therapeutic endeavors, apart from the seriousness of the lesion depends, above everything else, upon the

mental level of the patient. At the start we have to place our chief emphasis upon the application of physical agents (electricity, massage, hot baths); as far as electricity is concerned, the rubbing the hypertonic muscles with the anode of the galvanic current often acts quite favorably. For this application a rather large, flat electrode is selected and all sudden variations of current strength which may act as irritants are to be avoided by careful use of the rheostat; intensity 3 to 5 milleamperes. Those muscles, on the other hand, which are not greatly hypertonic but paretic, can be treated by the labile application of the cathode; only one should limit himself to such strength of current as suffices to produce a definite contraction on cathodal closing. On the other hand, I avoid in general the faradic current, since with it not only the muscles which it is desired to influence, but also those tending to contracture are affected by diffusion of the current. As to bathing, according to *Heubner*, hot baths are much to be recommended: 3 to 4 times a year during a period of 4 to 6 weeks, a bath lasting for 10 to 15 minutes is given every day; beginning with a temperature of 37° C. (98.6° F.) and gradually raising it to 40° C. (104° F.). Also the use of natural hot baths, especially those of higher temperature, for example, Baden-Baden, Aix-les-Bains, Teplitz, Hot Springs of Virginia, Arkansas and California are indicated. Massage can be carried on in the bath; it consists in careful (not jerky) stretching of the spastically shortened muscles whose hypertonia, as *Hoffa* has shown, can be diminished by tapotement of the ends of the tendons; their antagonists, on the other hand, are stroked and kneaded.

It quite frequently occurs that the systematic exercise of active movements, which must be begun as soon as the intelligence of the child permits it, must be preceded by orthopedic measures; above everything tenotomy, in its modern modifications (oblique, stair-like incisions), with application afterward of suitable bandages, splints, etc., also shortening of tendons, tendon transplantations, etc.

On the other hand osteotomy of the thigh below the trochanter, which has been recommended in *Little's* disease with the view of moving the center of gravity of the body farther backward, appears to have little justification. *Förster's* operation (rhizotomia posterior) has met with considerably more encouragement. In it, a number of posterior spinal roots corresponding to the spastic muscles are cut in order (according to the mechanism explained in Lecture I, page 7) to obtain relaxation of the hypertonia. Like all other operations, however, this last also promises success only with exceedingly careful after-treatment, long and continuous gymnastics and exercise therapy, in connection with which appliances like the resistence apparatus of Zander & Herz, etc., can find frequent application.

(Supplementary.) The Idiocies.

We have seen how often infantile cerebral paralyses are accompanied by idiocy. This last can also occur without any, or with such slight paralytic manifestations that it entirely dominates the picture. The accurate presentation of its semiological peculiarities belongs to the domain of psychiatry;

I would, however, sketch broadly the psychic relations common to all forms of idiocy and give you also some insight into their most important clinical varieties.

Under idiocy in the broad sense are included the conditions of psychical arrest of development characterized by want or defectiveness of the intellectual functions; the term "idiocy" in the narrower sense is reserved for those severe forms in which the individual is unable to direct his own life within the bounds of society. In the most extreme cases of this sort all ability to receive impressions and to form conceptions is absent; there is "mind blindness" and "mind deafness." The speech does not even reach rudimentary development, in short, the mental level is considerably below that of the higher mammals. On the other hand, however, the defective condition may be slight, so that a smaller or larger number of concepts develop, a certain amount of education is possible by a rational way of bringing up, ability to speak is developed to a greater or less extent, etc. There are the most manifold gradations until the slighter grades which are denominated "imbecility" are reached (in this latter the mental condition, in contradistinction to idiocy, permits the exercise of some calling, the individual can still be designated as "intra-social"), and to the very slightest "debility" or feeble-mindedness, whose separation from physiological stupidity is quite indefinite.

Fig. 95.

Pithecoid Idiot.

Frequent accompaniments of idiocy are: Epilepsy (in about one-third of the cases); genital infantilism; physical stigmata of degeneration (Gothic palate, asymmetry of the skull=oblique skill, plagiocephaly, "pithecoid"=ape-like formation of the face (see Fig. 95), anomalies of the teeth, hare-lip, cleft palate, prognathism, hyperdactylism, syndactylism, malformations of the ears, etc.); ambidextrism (in about one-sixth of the cases); backwardness in development of the body, automatisms of movement (showing the teeth, boring the fist into the mouth), grimacing, rocking the body, etc.); reduction of pain, temperature and muscle sense. According to their conduct with regard to the external world idiots are divided into apathetic (anergetic or torpid) and erethristic (energetic, agile or versatile).

All the pathologico-anatomical lesions mentioned on speaking of infantile cerebral palsy when they affect the mechanisms of the mental functions, furnish

the substratum for more or less profound idiocies as do also malformations of the brain, like true porencephali, or focal diseases in their earlier period of development, cysts, foci of softening, pseudo-porencephaly, further, microgyria depending upon meningitis, hypertrophic tuberosis and atrophic lobar sclerosis. Other malformations and arrests of development of the brain found in idiots are, among other things, defect of the corpus callosum and *Sachs'* "Agenesis corticalis," in which the cortex may appear macroscopically normal, the cortical ganglion cells, however, prove to be rudimentary. "Hydrocephalic idiocy" we have already considered in the preceding lecture. There remains to us now the task of going over a few forms of idiocy particularly marked by their physical accompanying symptoms.

Amaurotic Idiocy

This is a marked family disease which was recognized in its clinical peculiarities in 1887 by the New York neurologist, *B. Sachs*, although six years earlier the English ophthalmologist, *Warren Tay*, first saw and described the alteration of the eye grounds pathognomonic for this affection. On this account it is usually spoken of as the "*Tay-Sach's*" disease.

In typical cases at an age of from 3 to 6 months an infant up to this time normal, is affected by a torpor increasing until at length he lies almost continuously in a condition of complete apathy, relaxation and immobility, in which however, breathing, the heart beat, and the taking of nourishment is still undisturbed. All the muscles are hypotonic, if the child is set upright the head rolls, without support, in all directions. Now, however, spastic phenomena become more and more mixed with this picture of hypotonic akinesia and finally occupy the foreground. At first they are intermittent, tonic extension spasms, finally a continual spastic condition in the place of the former relaxation. Now the nutrition suffers through involvement of the swallowing and sucking mechanisms and death occurs after skeleton-like emaciation. It can be stated, as a rule, that this occurs before the completion of the second year. Only once did such a child live to be 8 years old.

Parallel with the progressive psychic and motor disturbances proceeds the loss of vision with a peculiar alteration of the eye grounds; this is a clouding of the retina distributed symmetrically in both fundi surrounding the yellow spot, of whitish color but with a cherry-red point in the middle (see Fig. 96). Finally, optic atrophy is added to it.

It is noteworthy that the typical cases practically always occurred in Jewish families originating in Poland, which raises the suspicion of descent from a far-removed common ancestor. According to *Apert's* collection, only two of the 166 cases which he recognized as undoubted instances of the disease formed an exception to this rule. It is paradoxical, however, that the overwhelming majority of cases of amaurotic family idiocy have not come under observation in Poland, but among Polish Jew emigrants, and for the most part in America, though also in England, Germany, Austria, France, and even in Australia. Perhaps the "transplantation" into other conditions of life is an exciting cause. The atypical cases in which the characteristic macular altera-

tions are absent (here belong also the so-called "late form" described by *Spielmeyer* and *Vogt*), do not manifest this ethnological predilection, or only to a very small degree.

The pathological anatomy of the *Tay-Sach's* disease is very accurately known. All the gray matter of brain and spinal cord shows marked cytological alterations (swelling of the ganglion cells, disappearance of the *Nissl* granules and of the fibers passing through the cell body, vacuolation, etc.). Also the retinal cells of the macula and its neighborhood are affected by the degenerative

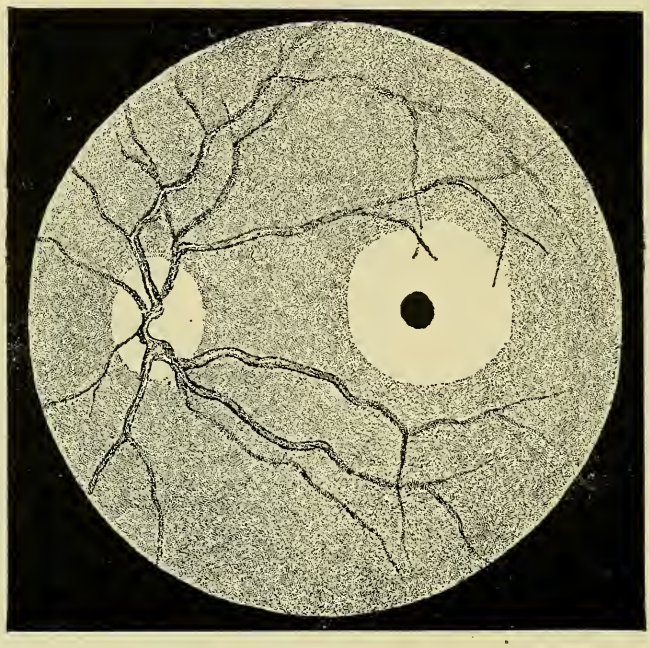


Fig. 96.

Eye-ground in Amaurotic Idiocy. (After *Sachs*.)

process, through which they become opaque. Hence the white circle about the fovea centralis. Only in the last (which has no ganglion cells) the choroid is visible afterward as before, and by contrast imposes itself as *Tay's* "cherry-red spot." As chemical correlative of the disappearance of the tigroid substance, that of nucleo-proteid in the whole nervous system has been shown, so it is probably a constitutional disease of metabolism of the ganglion cells.

Microcephalous Idiocy

This form, on account of the abnormal smallness of the skull, takes on a particularly characteristic picture. We must distinguish true microcephaly (simple pure microcephaly, micro-encephaly) from pseudo-microcephaly. At the base of the former there lies a genuine hypoplasia of the brain; in the second, the inhibition of growth and development of the brain is caused by gross intra-uterine brain diseases. In any case, the anomaly of the skull forms

the secondary correlative of an abnormal condition of the brain. *Virchow's* hypothesis, according to which premature synostosis of the sutures of the skull is responsible for the arrested development of the brain, has long been abandoned. With it also, fortunately, the "*Lane-Lannelongue*" operation in which by removal of segments of bone from the roof of the skull or even the separation of the calvarium (craniamphitomie) the compressed brain could be furnished relief. Fig. 97 represents a plaster model of one of the most celebrated

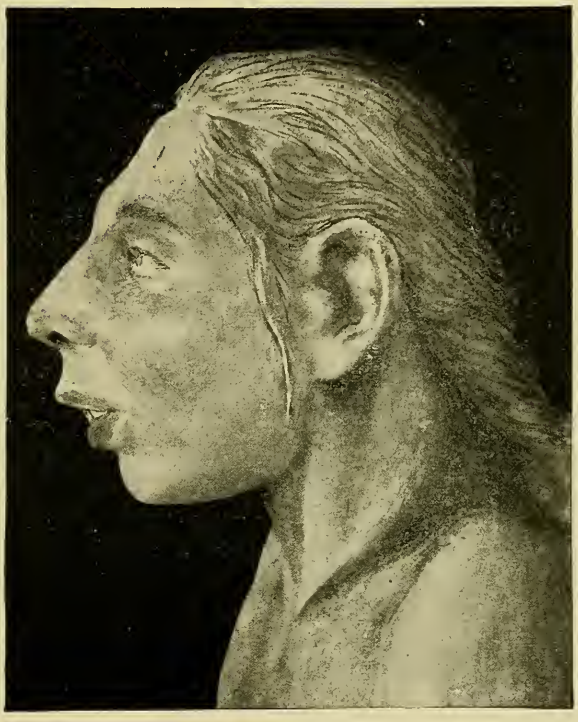


Fig. 97.

Microcephalous Idiot. (Modeled after Life.)

cases of family true microcephaly, which is preserved in the Pathological Institute at Basle. The peculiar form of face which is shown in our picture has led to the designation "Bird head" or "Aztec Type." The lowest brain weights observed were 15.9 grm. in a 7 weeks' old boy; 283 gm. in a 43 years' old woman. Microcephaly must not be confused with the "nanocephaly" which occurs in small individuals.

Mongoloid Idiocy

As the most important clinical peculiarities of this form of idiocy discovered by *Down* in 1866, the following group of symptoms may be mentioned. First, the peculiar physiognomy which has given the disease its name, the "Tartar" or "Kalmuck" type (see Fig. 98); flat face, wide-bridged nose, prominent cheek-bones, almond eyes (often with epicanthus), with reddened

lids and margins, and without lashes, a grayish-brown complexion, with red cheeks which produce the impression of being painted like a clown, round skull, flattened at the back. Then the hypertrophy of the tongue with enlargement of the circumvallate papillæ and remarkable wrinkling of the surface ("scrotal tongue"). Abnormal flaccidity and softness of the muscles, ability to bring the joints into abnormal positions, like a "snake man." Often there are congenital anomalies of the internal organs (atresias, umbilical hernia, congenital defects of the heart, arrests of development of the teeth, indications of dwarfism with normal ossification shown by the X-ray, but with atrophy of the end and shortening of the middle phalanx of the little finger, more rarely, a tendency to partial giantism).

Fig. 98.

Mongoloid Idiot.

The growth of hair is not affected, the eyebrows, indeed, are abnormally heavy. The defect in intelligence in "Mongoloids" is, as a rule, combined with quite considerable ability to fix the attention and reactibility, with happy mood and a great tendency to imitation. This exceedingly characteristic form of idiocy is not very rare in our neighborhood; *Konrad Frey* found in the Aargau Idiot Asylum at Biberstein, among 60 inmates, 3 Mongoloids; in England and Scandinavia also 5 per cent. are found, in Germany, only 2 per cent., on the other hand, according to *Kowalewsky*, in the Government of Petersburg, the percentage among the inmates of asylums is 10, in the Government of Kazan, indeed, 25. Whether this last is connected with the decided Mongol admixture in the population cannot be certainly stated.

According to *H. Vogt*, Mongolism is connected with retardation in the later stages of development of the brain; according to *Buschan*, *Weygandt*, *Frey* and others, it could be attributed primarily to disturbances of internal secretion (thymus?). The brain is often small, sometimes it presents defects in development (for example, partial defect of the corpus callosum). Often the convolutional type renders recognizable an abnormally simple development with very coarse and wide convolutions; the cortical cells are imperfectly differentiated, the cortex abnormally rich in vessels.

Cretinistic Idiocy

Those forms of idiocy which occur in connection with alteration of the thyroid gland or of its "internal secretion" and are accompanied by peculiar changes in the skeleton, demand particular attention. We include them under the common designation of "Cretinism." It may be remarked here that in the earlier, and unfortunately also in the newer literature, much confusion has been produced by bringing together under this designation different diseases of other sorts (running their course partly with and partly without idiocy) on account of superficial resemblances in their external appearance. Above everything the just described "Mongolism," further, fœtal chondrodystrophy or achondroplasia, a form of disproportioned dwarfism (micromelia) dependent upon a congenital defect of the zones of direction of the cartilages of the skeleton, which does not imply any disturbance of the intelligence or any alteration of the thyroid.

We distinguish 1, sporadic, and 2, endemic cretinism. The first, again, occurs in two forms: a, infantile myxœdema, and b, thyreoaplasia congenita.

We will become acquainted in the lecture after the next with the disease picture of myxœdema which occurs from the destruction or the serious interference with the function of the thyroid gland and in which the mental functions are more or less seriously affected. If this myxœdema occurs in childhood there results besides this, a retardation in the growth of the skeleton, a proportioned dwarfism on account of injury of the cartilage, the bone marrow and the periosteum, through which infantile myxœdema takes on the picture of a sporadic cretinism.

Similar, only much more marked, are the anomalies in congenital defect of the thyroid gland (thyreoaplasia). Here also the hypothyreotic pathogenesis of idiocy is very plain.

More complicated relations exist in endemic cretinism which is at home in such regions in which goiter and deaf-mutism occur in great frequency. The Alpine and sub-Alpine valleys are afflicted with this severe scourge. In Switzerland, in Tyrol, Styria, Savoy, Piedmont, in the Veltlin. For Switzerland *Heinrich* and *Eugen Bircher* have established the striking fact that there is a connection between cretinism and the geological formation of the soil, that above everything the marine formations of the Paleozoic, the Triassic and the Tertiary, are affected, while all the fresh-water formations, also the eruptive and the crystalline rocks and the sediments of the Jura and the lower chalk remain free from endemic goiter, cretinism and deaf-mutism (*E. Bircher* classes these 3 conditions together as cretinistic degeneration). That the cause is to be sought in the drinking-water is certain; for instance, it has been possible to check the endemic cretinistic degeneration in the Canton Aargau by supplying the villages of the endemic district with water from healthy localities. Further, in the rare endemics in lowlands (for example, on the island of Schutt, or along the course of the Mur in Hungary) it has been found that the affected regions get their water from rivers which rise in cretin regions. The nature of the disease agent and its method of action, which for the rest presupposes a personal predisposition, on the other hand are not yet

explained, in spite of recent hypotheses. In no case can endemic cretinism in its whole clinical picture be referred to a pure thyreoaplasia or hypothyreosis. The injurious agent must primarily have other points of attack than the thyroid gland, since the symptom-complex is a much more complex one than in both the varieties of sporadic cretinism, and the reaction to thyroid administration, as we will soon see, is scarcely ever appreciable.

The weakmindedness can reach an excessive degree: there are cretins who can never be taught to take nourishment for themselves, but must even be fed with the tube. Along with these, however, there are often slighter forms of idiocy, imbecility, and debility (feeblemindedness). The apathetic torpid form

Fig. 99.

Alpine Cretin.

of idiocy predominates. There are, however, very disturbed, obstinate and troublesome cretins who, as the accomplished describer of Alpine cretinism, *Peter Rosegger*, expresses it, "are capable of the 7 deadly sins." Where they can learn to talk it is imperfect, stammering. Smell, taste, sensibility, and especially hearing, are more or less seriously affected; vision, on the contrary, is usually good. The head is usually abnormally large, more rarely, microcephalous, frequently asymmetrical. The nose is wide, deeply drawn in at the root, the eyes are widely separated from one another, small, and slit-like; especially is the whole face widened. A low forehead, a dry, wrinkled skin of an unclean color, sparse or absent growth of beard, a wide mouth with irregular teeth, a short neck on which a goiter is often prominent, complete the

grotesque physiognomy (see Fig. 99). The body structure is characterized in general by disproportioned dwarfism, the lack of development in length affects chiefly the lower extremities, but, however, never reaches the excessive degree observed in achondroplasia; the trunk is not only too long in proportion to the limbs, but it is usually very massive and plump, with the exception of the sometimes deformed pelvis. Very many cretins are the bearers of great inguinal hernias. In the X-ray picture an irregular delay in the formation of bone nuclei and persistence for a long time of the epiphyseal cartilages (until the middle of the third decade) is noted.

Along with these typical forms there are in cretin countries a great many cases in whom there are only a small number of the anomalies mentioned: they are called "cretinoids," or "half cretins."

The Treatment of Idiocy

A causal treatment is only possible in idiots where there is either congenital syphilis or where thyreogenic causes prevail. In the last there is a great difference between endemic cretinism and the sporadic forms, however. Infantile myxœdema reacts even in the psychical condition, often with surprising improvement, even recovery, to the administration of thyroid gland preparations, for example, the thyroid gland tabloids of Burroughs, Wellcome & Co., or the Thyraden tablets of *Knoll* (each containing 0.3 grm of the thyroid gland substance of the sheep or of the hog, per tablet). The children are given one-half to one tablet (that is, 0.15 to 0.3 gland substance) per day. In thyreoaplasia improvement is the rule under organo-therapeutic treatment, but it is usually less marked than in infantile myxœdema and there is practically never recovery. Endemic cretinism is still less suitable for thyroid treatment. In slight cases, indeed, *Magnus-Levy* and *Wagner v. Jauregg* have obtained improvement, but this almost always fails to appear. The transplantation of viable thyroid into the spleen of a cretin as *Payr* has undertaken, has only a certain influence in encouraging growth, none upon the psyche. The pedagogic treatment of psychical defect conditions of every pathogenesis is only hopeless in those of the highest degree and accomplishes indeed, in the majority of cases, as among others, *Bourneville*, has shown upon the enormous material of the Bicêtre Asylum at Paris, satisfactory and encouraging results. The instruction of idiots has become a scientifically well-founded specialty. In order to develop the muscular sense in these children we usually begin by instructing them in the use of their extremities (by the aid of their faculty of imitation) for all actions of daily life (also eating, etc.). At 4 or 5 years the education proper begins, which first has to render of use those capabilities which have remained least rudimentary, also the instincts present (for example, the desire for tid-bits), are utilized for the accomplishment of results, for fixing the attention, etc. All this can only be accomplished in a way promising results in an institution. On this account idiots should in all cases be removed from home in the 5th year. With tireless and intelligent management a large part of them learn in time to read and write and many of them can in any case learn to employ themselves as helpers in healthy manual occu-

pations (farming, gardening, etc.). In Basle the endemic cretins have to such an extent a monopoly of the peddling of sand that the terms "Sand-männli" and "Sandwybli" (little sand men, little sand women) in the popular speech are equivalent to "cretin," and they are able to carry out this modest but useful occupation with entire satisfaction. Further mental development of idiots is terminated only at about 20 years.

LECTURE XXIII

Dysglandular Symptom-Complexes

GENTLEMEN: We will occupy ourselves in this lecture with several disease pictures for whose clinical symptoms acting particularly upon the nervous system, as in certain forms of the idiocies just considered, anomalies of "internal secretion" of different glands, play the most important pathological rôle, and which on this account I hence bring together as "dysglandular."

As is known, specific substances which are secreted by the definite glandular structures into the blood represent an important physiological factor of our organism in which they have to stimulate in a chemical way, certain functions. From ὁρμάω = I stimulate, is derived the name "hormone" proposed by *Starling* for such substances. A particular affinity of a great number of hormones for the nervous system or certain parts of it, is just as important from a pathogenetic point of view, as the fact that the disturbance of function of a gland with internal secretion (they are also called "endocrine glands") can involve also one or more of the other glands, so that it is sometimes difficult to decide certainly as to which organ was primarily affected. I will confine myself chiefly to the discussion of such disease conditions in which the nature of the endocrine disturbances are relatively plain and generally recognized, namely: 1, *Basedow's* disease; 2, myxœdema,—both of thyreogenic origin; 3, *Addison's* disease—a consequence of disease of the adrenals, and, 4, different syndromes connected with disturbed function of the hypophysis. The future will show how many of the dyskinetic conditions with which we have already occupied ourselves can definitely be arrayed among the dysglandular affections. For tetany, as we have seen, this is very probable, but also in relation to paroxysmal paralysis, paralysis agitans, etc., the view that they are dependent upon disturbance in the action of hormones is coming more and more to the front.

I. Basedow's Disease

This affection was comprehended in its clinical individuality, and described simultaneously (about 1840) by the English clinician *Graves*, and the Merserburg physician *Basedow*. There are, however, exact descriptions of a few cases from an earlier time, among which those of the Italian *Flajani* (1802) deserve special mention. "Morbo di *Flajani*" is also a name for the disease used in Italy, while the English designation is "*Graves'* disease." Besides "maladie de *Basedow*," the disease is also called in French after two of its most striking symptoms, "goître exophtalmique," while the German term, "Glotzaugenkrankheit" has become almost obsolete.

Symptomatology

The fully developed cases of *Basedow's* disease are characterized by the unmistakable and characteristic combination of four cardinal symptoms about which again are grouped a number of less striking phenomena. Recognizing that opposed to these classical cases there are a great number of "Formes frustes" with rudimentary symptomatology, we will begin with the consideration of these cardinal symptoms (goiter, exophthalmus, tachycardia, tremor).

1. *The Goiter.*—This is in general not an excessive struma formation, but only a moderate hypertrophy to about double the size of the healthy thyroid. Usually at once perceptible upon near inspection of the neck, it is often only plain upon palpation. Either both lobes are equally enlarged, or the increase in volume is especially of one lobe, remarkably more frequent on the right. The goiter is usually soft and as auscultation for murmurs shows, is very vascular. Not rarely an arterial thrill may be detected upon palpation, and besides this, a definite pulsation of the gland. This richness in vessels well explains the great variations in the size of the struma, which can sometimes be noted when it is measured regularly. For the rest, it swells also upon exertion, excitement, etc., sometimes quite plainly. Within the soft tissue a few more resistant portions are to be felt in some places. In general the *Basedow* goiter, mainly in consequence of its general softness, produces much fewer subjective symptoms than an ordinary struma of the same size. Pressure symptoms are usually absent entirely; asphyxic difficulties proper, from compression of the trachea or the recurrent nerves only very rarely occur, while a feeling of fullness and tension in the region of the thyroid gland is somewhat more frequently complained of.

Fig. 100.

"Tragic Look" in *Basedow's* Disease.

2. *The Exophthalmus.*—The protruding eyes ("Glotzauge") of *Basedow* patients when typically developed, give an uncommonly characteristic expression of countenance; this has been called the "tragic look," and as a fact, the physiognomy in advanced cases reminds one of certain masks of ancient tragedies (see Fig. 100). In general, however, I should characterize the expression which the exophthalmus gives to the features of *Basedow* patients rather as that of anger, to which, also, the swollen neck contributes its part (see Fig. 101). In less intense development, the exophthalmus, produces rather the impression of a glistening eye ("Glanzauge") than of a protruding eye, while

on the other hand, particularly marked protrusion of the eyeball can cause inability to close the lid and has even been responsible for luxation of the eye from its socket. The exophthalmus is usually symmetrically developed, though in a considerable proportion of cases there is a distinct difference between right and left. That the intensity of the phenomenon shows great variation in the course of the disease is quite usual, so that even a marked exophthalmus can disappear again. Three phenomena are to be emphasized as characteristic accompanying symptoms of the protruding eyes in *Basedow's* disease: 1. "*Stellwag's* sign": winking is abnormally infrequent in such patients and the palpebral fissure is unusually wide, so that below and above the cornea a more or less wide strip of the sclerotic is visible. 2. *Graefe's* sign: in looking down a disturbance in the physiological synergy between the movements of the lid and those of the eyeball is apparent, so that the upper lid lags behind instead of sinking synchronously with the sagittal axis of the eye. 3. *Möbius'* sign: the movement of convergence of the eyes occurs imperfectly or is quickly impaired. This last-mentioned phenomenon is less constant than the symptoms of *Stellwag* and *Graefe*.

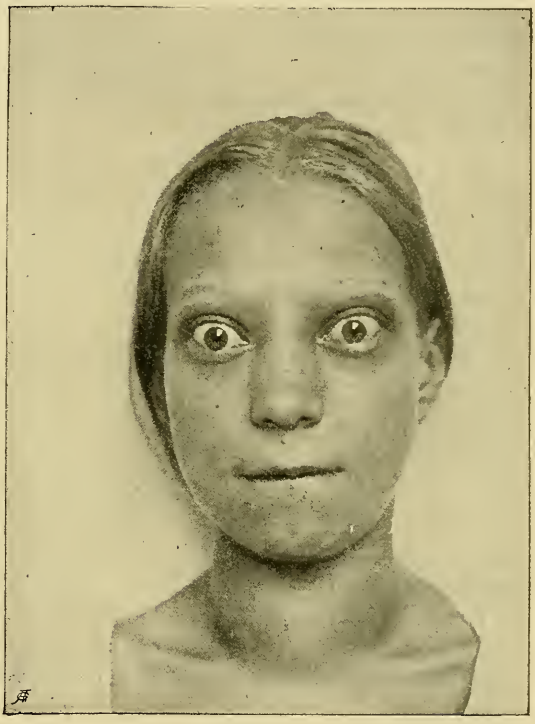


Fig. 101.

Basedow's Disease.

3. *The Tachycardia.*—The pulse is permanently quickened, so that even when lying quietly on the back figures of 100, 120, and even 160 beats to the minute are observed. Changes of position, exertion of any sort, but particularly psychical excitement, increase the pulse frequency excessively, so that even 200 beats a minute may occur (usually with a feeling of severe palpitation of the heart). The rhythm of the pulse, however, remains nearly always regular. Cardio-vascular symptoms accompanying this permanent tachycardia are quite frequent and manifold. I would mention above everything the increased vasomotor irritability of the integument which, among other things, manifests itself in so-called dermographism: stroking the skin with a blunt object produces, after a few seconds, wide red stripes in which the drawing or writing produced in this manner only pales and disappears after several hours. Often the dermographism is combined with a serous infiltration of the skin which leads to the production of wheals; we speak then of factitious urticaria. Congestion of blood about the head, temporary redness which can alternate

with marked pallor, intense feeling of heat in extended regions of the surface of the body, are further phenomena belonging here. The blood pressure I have sometimes found abnormally high; for example, from 140 to 185 (measured with *Gärtner's* tonometer on the patient's sitting quiet), and abnormally labile; still, these conditions vary from day to day within wide limits. Over the heart systolic murmurs are sometimes to be heard, namely with increased tachycardia. Their maximum intensity is usually at the base; these are (as the inconstancy and not rarely quick disappearance of these phenomena show), as a rule, functional murmurs due to relaxation of the atrioventricular orifices and a resulting insufficiency of the mitral and tricuspid valves; the character of these murmurs is soft and blowing. Also, by percussion and orthodiagraphy a certain, usually inconsiderable, degree of dilatation of the heart can be demonstrated in some cases.

4. *The Tremor.*—This is an exceedingly rapid and fine tremor (8 to 10 oscillations a second), which is to be perceived in the outstretched fingers of the patient, but which we can often plainly appreciate when we place a hand upon the head or upon the shoulder of the standing or sitting patient. Upon movement, intensity of this tremor usually increases, while it may cease upon complete rest.

When now the four cardinal symptoms are present, along with which, besides, a number of the ocular and cardio-vasomotor accompanying phenomena already mentioned are always to be found, we speak, as already said, of the classical form of the disease. Frequently, however, we meet with "formes frustes" in which either the exophthalmus or the enlargement of the thyroid, or even both, are absent, and only the heart and vascular anomalies and the tremor can be found. If, now, for a diagnosis of *Basedow's* disease, the presence of these two "obligatory" cardinal symptoms would suffice, they must present, of course, those clinical peculiarities which I have pointed out to you. In spite of this, however, the inclusion of rudimentary cases of this kind under exophthalmic goiter is not justified if they do not present besides some of the rather rich array of the so-called "secondary *Basedow* symptoms" with which we will now become acquainted.

The following disease manifestations of subjective and objective nature play, along with the cardinal symptoms in the clinical picture of *Basedow's* disease, a rôle varying from case to case and presenting manifold combinations, but yet quite important:

a. Muscular Weakness in the lower extremities, often in the form of a peculiar, paroxysmal paraparesis, so that the patient's legs, while walking or standing, suddenly give way at the knees. Rarely, however, does this weakness increase to a degree causing the patient to be bedridden, and is even then temporary, since this symptom in particular is subject to great variations during the course of the disease. Infrequently do these transitory pareses assume another localization, for instance, hemiplegic or monoplegic, or affect the neck muscles or the regions innervated from the brain. In the last case there may be not only temporary pareses, but, as *Stellwag* has shown, actual paralyses of the external eye muscles, while the muscles of accommodation and of the iris are always spared (except as an extreme rarity). Here belongs also the im-

possibility of drawing deep inspirations sometimes observed ("*Louise Bryson's* sign").

b. Disturbances of Menstruation.—Dysmenorrhœa is very frequent; amenorrhœa not exactly rare.

c. Diarrhœas.—*Charcot* and *Möbius* have pointed out the great tendency of *Basedow* patients to profuse diarrhœas occurring in attacks sometimes lasting for days and weeks, which begin without any apparent reason, defy opium, bismuth, tannin, etc., and then suddenly and for no apparent reason, cease. The evacuations (up to 10 and more per day) are very thin and often entirely painless.

d. Hyperidrosis.—Excessive sweat secretion may be general or it may be limited to a definite portion of the body (face, hands, feet); it may be continuous, or there may be profuse outbreaks of sweat occurring in attacks usually coincident with congestions and palpitation of the heart. Also in such patients who do not complain of excessive sweat, even when the temperature is low, the skin is found in the condition for which I would borrow the French expression "peau moite" (moist skin). *Vigouroux's* phenomenon is to be attributed to this abnormal moisture of the integument; this is reduction of the electrical resistance. If this last is in healthy persons about 4,000 ohms, in *Basedow* patients it often sinks to 1,000 ohms and below. To obtain a certain number of milleamperes in galvanization, we need in these cases to switch in only one-quarter as many elements as for a normal person, corresponding to *Ohm's* law:

$$\text{Current Strength} = \frac{\text{Electromotive Force}}{\text{Resistance}}$$

e. Psychic Changes.—Most *Basedow* patients show a marked hasty and restless manner, a "feverish motor impulsion," which, however, is accompanied by great defect in staying power when the task is once begun. They find it difficult to remain long seated or standing in the same place; their mood is extremely labile; their expressions of joyful, of sad emotions are boundless, and in conversation they are wordy, precipitate and inclined to deviation from the subject under discussion. Their surrounders often complain of the exaggerated sensitiveness, the irascibility, the capriciousness of such patients. Like the other disease symptoms these psychic symptoms are subject to the greatest variations in their intensity, can disappear in the course of the affection and reappear. More severe psychic disturbances, psychoses proper, develop in some, fortunately rare cases. *Homburger* has furnished proof that a specific "*Basedow* insanity," as was asserted by former authors, does not exist. Rather does *Basedow's* disease occur in connection with almost all forms of insanity, a coincidence which probably is to be explained through the sometimes severe neuro-psychopathic heredity of *Basedow* patients.

f. Neuralgic Phenomena.—Eye neuralgias, intercostal neuralgias, pain in the trigeminus distribution, particularly in the upper branches, are the most frequent manifestations of these troubles, not rare in *Basedow* patients. The pains are, as a rule, of only moderate intensity; paresthesias may occur also.

g. Signs of over-irritability in the neuro-muscular apparatus. Light tapping of the muscles usually calls forth a definite contraction, and by firmer

striking, one can bring out sometimes the phenomenon of "idio-muscular contraction" with which we became acquainted when speaking of tetany. On the other hand the appearance of either mechanical or electric overirritability on percussion or on galvanic stimulation of the nerve trunks is very rare, while only as a very unusual occurrence has the appearance of tetanoid spasms been described. As to the "choreiform movements" to which, among others, *Raymond* and *Dieulafoy* have alluded, I believe that they may have been only a specially striking degree of intensity, of the already mentioned psycho-motor restlessness of *Basedow* patients. In a great majority of cases you will find the tendon reflexes, very often also the skin reflexes, abnormally lively.

h. Trophic Disturbances.—Falling out of the hair, combined with dryness, brittleness and lack of lustre has been considered by *Curschmann* as a very important *Basedow* symptom and actually occurs in my experience in about 60 per cent. of the cases, however, usually only to a moderate extent, so that the patient's attention must be called to it. Less frequent are pigment anomalies, usually in the form of a hyper-pigmentation, about the face, neck, abdomen, etc. (similar but less intense than those which we will soon meet when describing *Addison's* disease), here and there, however, on the contrary, in the form of a pathological loss of pigment, vitiligo. As rarities may be mentioned, also, a firm but fleeting œdema on the body and the lower extremities, further, abnormal softness of the bone and atrophic changes in the breasts, while the observations of sclerodermic phenomena (see below, Lecture XXV) in *Basedow* patients (*Leube, Stähelin* and others) practically do not indicate anything more than a combination of two distinct disease conditions.

i. Anomalies of Metabolism.—The investigations of *Fr. Müller, Magnus Levy, Scholz,* and others have proved that in *Basedow's* disease the excretion of nitrogen, carbonic acid, phosphoric acid can experience a pathological increase, which probably depends upon increased destruction of albumin in consequence of an elevation of the total metabolism. No wonder that, as a rule, even when taking plenty of nourishment there is often a tendency to extreme and rapid emaciation (in one case of *Mannheim's* the weight fell 45 kilos. in 10 months). *Huchard* has referred to "crises of emaciation" occurring paroxysmally. On the other hand, there are *Basedow* patients who are corpulent and remain so. Polyuria, eventually with polydipsia, is not entirely rare, also alimentary glycosuria, while true diabetes mellitus has only been described in a few cases (mostly as a complication in the advanced stages). Rises of temperature during shorter or longer periods are also an unusual symptom, while transitory albuminuria of slight degree (without casts) is somewhat more frequently found.

k. Blood Changes.—As characteristic for *Basedow's* disease, *Kocher* considers the following hematological finding: actual increase of the number of lymphocytes, causing relative diminution of the polynuclear leucocytes, with red corpuscles normal in number and in hemoglobin content.

Course and Prognosis

The disease, which affects by preference the female sex in the second and third decade, but occurs even in children, begins usually, but by no means

always, by the generally gradual, only exceptionally sudden, appearance of a "cardinal symptom," and, indeed, in almost two-thirds of the cases with the heart troubles, more rarely with the struma or the tremor, most rarely with ocular symptoms. The further course is only in the mildest cases subchronic (of several months' duration), in cases of moderate severity chronic, remittent and intermittent, extending over years. Malignant cases with great intensity of the symptoms characterize themselves sometimes by acute or hyperacute course (in cases of *Trousseau* and *Fr. Müller*, after about 2 months, in one case of *Mackenzie*, in 3 days, after the abrupt beginning of the first symptom, death occurred). Usually, however, severe disease pictures develop through the chronic, progressive increase of originally slight disturbances. The prognosis depends in the first place upon the intensity of the clinical manifestations and upon the possibility of appropriate care and manner of life; if this last condition can be obtained, even apparently hopeless cases, though not cured, may again be brought into a tolerable permanent condition. Along with the duration of the disease, for the prognostic estimation of the individual case, the condition of the heart comes into consideration above everything else, since heart weakness, with its consequences (which we naturally do not include under the symptoms proper of *Basedow's* disease), also anasarca, ascites, œdema of the lungs, congestion of the liver, congestion of the kidneys, asystole, form along with general marasmus the chief proximate causes of death in the lethally ending cases. A specially wretched complication of the most severe *Basedow* cases may be mentioned here: the keratitis due to the excessive exophthalmus, the inability of the eyelids to close and the drying of the cornea, which may lead to the perforation of the eyeball and panophthalmitis with all its frightful consequences.

Pathogenesis and Etiology

How is this peculiar disease picture, so rich in symptoms, produced? This question (in consequence of the inconstancy and ambiguity of the sparse disclosures which pathological anatomy has furnished us) it has been attempted to answer in many different ways.

Many of these theories have long ago fallen into oblivion after their all too apparent conflict with the data of actual observation has been shown; for example, it was suggested that the symptom-complex was due to the compression of the vessels and nerves of the neck by the goiter.

There have remained as a subject of discussion only those views supported upon facts which either have asserted a primary nervous basis for *Basedow's* disease, or which have spoken for the thyreogenic autotoxic nature of its symptoms. To-day both views have justification; both are sanctioned by the result of clinical and experimental observation. It is not understandable why it is attempted to bring these two views into antithesis with one another.

That since *Charcot*, *Basedow's* disease has been included among nervous diseases has a good clinical basis. First, the hereditary and family relations to the psychoses and psychoneuroses which are recognizable in most cases; second, its frequent coexistence with almost all forms of psychoneuroses and insanity which either precede the onset of *Basedow's* disease begin

simultaneously with this last, or develop in its later stages; third, its occurrence as a complication in organic diseases of the nervous system (tabes, multiple sclerosis, paresis); fourth, finally, its beginning in countless cases in connection with a fright or some other psychical shock. But why cannot this neurosis be a secretory neurosis of the thyroid? Do we not know numerous cases of the quantitative influencing of the most varied secretory processes by functional as well as organic affections of the nervous system (nervous gastroxynsis, sialorrhœa, colica mucosa, etc.)? On the other hand, it cannot be denied that there are cases enough in which the functional anomaly of the thyroid gland appears to play the rôle of a "primum movens," the remaining nervous and vascular components of the syndrome that of secondary symptoms. These cases have been denominated as "Basedowified goiter" and separated from true goiter. We can easily class both categories together and assume that the anomalies of the thyroid gland function coming into question arise not exclusively upon the basis of the neurosis, but sometimes also upon that of the local affection. Indeed, for both varieties, according to the results of clinical experience and experimental investigation, the two following points may be regarded as proven:

1. In the center of the disease picture stands the altered thyroid gland function.

2. The point of attack of the toxic action proceeding from this lies in definite portions of the nervous system.

Recognition of the first forms the basis of the "thyreogenous theory" of *Basedow's* disease. This is supported by a number of experiences which I will shortly indicate. Cure of *Basedow's* disease by thyroidectomy; appearance of *Basedow's* symptoms in inflammation of the thyroid or carcinoma, after excessive use of thyroid tablets for therapeutic purposes; identity of the anomalies of metabolism on thyroid feeding with those of *Basedow's* disease (*Magnus Levy*); acute appearance of *Basedow* symptoms on the overwhelming of the organism with expressed thyroid gland products, as sometimes occurs in goiter operations, the so-called "acute post-operative thyroidism"; more or less extensive analogy of the experimental hyperthyreosis (transplantation, feeding, injection experiments by *Ballet*, *Enriquez*, *Lanz*, and others) with the picture of human *Basedow's* disease.*

As to the point of attack of the thyreogenous noxious agent, it is to be placed either in the sympathetic or in the medulla oblongata. The sympathetic theories (*Benedikt*, *Friedreich*, *Eulenburg*, *Abadie*) have much that is seductive, as the tachycardia, the vaso-dilatation, the ocular symptoms, the hyperidrosis, etc., can well be brought into connection with disease of the sympathetic, as you will soon recognize in the lecture on sympathetic affections. If we do not find either the assumption of simple irritation or that

* The value of this last demonstration has experienced a considerable limitation through the discovery of *E. Bircher*, that after transplantation of viable thymus into the abdominal cavity of dogs, *Basedow's* symptoms also appear. However, the relations between *Basedow's* disease and the most varied glands with internal secretion is an exceedingly interesting, though still insufficiently investigated subject. Whether the extirpation of remains of the thymus proposed by *Garré* will establish itself in the treatment of *Basedow's* disease remains to be seen.

of paralysis alone satisfactory, this is not in contradiction with the experiences of neuro-pathology, in which the most varied examples of combined irritative and defect symptoms in the same nerve territory are found. Rather must the absence of pupillary symptoms arouse consideration. Anatomical changes in the gangliated cord have been described in isolated cases, and the physiological proof has been furnished that the tachycardia produced by thyroid juice depends upon irritation of the accelerator nerves.

Of late, however, those who hold that the point of attack of the noxious agent in *Basedow's* disease is in the bulb have become more and more numerous. More important than the heterogeneous and not very convincing autopsy findings (into which I will not enter) are the experiments of *Filehne*, *Durdufi*, and *Bienfait*, who in animals set up a part of the *Basedow* syndrome by lesions of the restiform bodies, and especially those of *Tedeschi*, in which the experiment only succeeded when the animals were in possession of their thyroids.

Personally, I am of the conviction that still other parts of the nervous system come into consideration, since how can we otherwise explain the psychic and paraplegic disturbances?

Allow me to anticipate somewhat, and to remark that myxœdema, with which we will occupy ourselves after *Basedow's* disease, and which depends upon secretory insufficiency of the thyroid gland (whether on account of its operative removal or its pathological elimination), may be considered the exact clinical antithesis of the latter. In myxœdema metabolism is reduced, in *Basedow* increased; in the former the temperature is reduced, in the latter increased; the skin in the first is dry and thickened, in the second hyperidrotic and thin; on this account, in the first instance there is elevation, in the second reduction of the electrical resistance; in myxœdema there is corpulence, in *Basedow* almost always emaciation; in the one there is small and slow pulse, in the other bounding and rapid pulse; here mental slowness, there excitation; in the former the movements are stiff and slow, in the latter hasty and tremulous, etc. *Möbius* first expressed the idea that in *Basedow's* disease there is overproduction of a secretion which, produced in normal quantity, is intended to neutralize the toxic product of metabolism, which one can designate briefly as "myxœdema poison." Also in *Basedow's* disease there is a pathogenic action of this excess of hormone, its symptoms are of a hyperthyreotic nature.

The substance at fault is probably in the first instance iodothyrin, discovered by *Baumann*, for which also speaks the fact that iodin medication exerts an unfavorable influence upon *Basedow's* disease; indeed, as *Kocher* and others have shown, forced iodin cures lead to a symptomatologically similar disease picture, the so-called "iodin-*Basedow*." Indeed, iodothyrin is not the sole active constituent of the thyroid gland, but still other substances (organic bases, thyreoproteid, etc.) come into consideration. Just to this multiplicity of active substances, which in part seem to show antagonistic action, I have ascribed the fact that certain symptoms of *Basedow's* disease and of myxœdema are the same—irregularity of menstruation, pigmentation of the skin, digestive disturbances, falling out of hair. Hyperthyroidism may well be accompanied by dysthyroidism (faulty composition of the secretion).

Now as to the "formes frustes" without enlargement of the thyroid gland,

it must be pointed out that a secreting parenchyma may be found in chronic functional hyperactivity without hyperplasia. (I would remind you, for example, of gastrorrhœa acida.) As *Oppenheim* suspects, however, the obstruction of the intralobular lymph channels—a regular lesion of the *Basedow* thyroid can cause an inundation of the venous blood with the products of secretion of the thyroid. It is, however, also conceivable, that exceptionally the sympathetic or bulbar lesion-complex characteristic for *Basedow's* disease may be produced by other factors than thyroidism.

Treatment

The treatment of *Basedow's* disease in most cases makes great demands upon the persistence of the physician and of the patient. Though it is not rarely permitted to us, even after a short time, to note decided improvement, we should not be too ready to fold our hands and let the patient get out of sight. Only by therapeutic effort, extending over months if not years, will it be permitted to us, first to render durable the improvement obtained, then to increase it, and finally, often enough, to procure permanent recovery.

Of capital importance in this connection is careful regulation of all the conditions of life of the patient. Except in very mild forms, one will do well to begin the treatment with several weeks' rest in bed, which also, in the further course of the disease, may be occasionally introduced with advantage. The excitability and the tachycardia are usually quite plainly influenced by this simple measure. As an ideal régime is to be regarded, the ovo-lacto-vegetarian diet which permits the preparation of quite tasty and varied bills of fare; besides this, we forbid the stimulating spices.

The avoidance of irritants which the spices and the extracts of meat prove themselves, acts not only sedatively upon the nervous system *in toto*, but specially plainly upon the cardiac and vasomotor innervation; also tea, coffee, alcoholics and tobacco, are best forbidden upon the same grounds. Some concessions, especially with regard to the use of meat, can be made according to the individuality of the case. Where possible, however, it should always be brought about that the extractives are only taken in small quantities, in that, for example, the portions of fish, poultry, veal or beef allowed daily should be taken after the juice is expressed.

The tendency to diarrhœa is not to be too much considered in the choice of food, since the character of the latter appears to be of slight influence upon the intestinal troubles of *Basedow* patients. Where milk is unpleasantly laxative, I rub up 10 grms of pure powdered gum arabic in some cold milk, then add it to a ¼ liter of milk, boil it, and finally add 1 bitter and 1 sweet almond, each blanched and ground.

Next to rest in bed, it is well to prescribe freely sojourn in an elevated region. Besides moderate elevation, 2,500 to 4,000 ft., in most cases residence in the high valleys, from 4,000 to 6,000 ft., can be recommended with advantage. The Engadine furnishes all the degrees of elevation coming into consideration. But also the Black Forest and other wooded hill regions give satisfactory results, while residence by the sea is usually disadvantageous. A cer-

tain measure of bodily exercise in the open air, increased little by little, should be united in climatic cures with several hours daily reclining in the open air. Sports proper, as well as dancing, should be strictly forbidden; automobile riding also appears to me to act disadvantageously; excitement is to be avoided as much as possible, especially that in the sexual sphere.

Under hydrotherapy there come into consideration carbonic acid baths, cool frictions, tepid fan or rain douches of short duration, slowly cooled from 20 to 18° C. I thoroughly disapprove of "cold water cures" proper.

Electric treatment in different forms has been applied. In my experience, however, good results are obtained only from the stabile galvanization of the sympathetic in the neck (anode over the top of the sternum, cathode at the angle of the jaw, the current is slowly raised from 3 to 5 milleamperes, 5 minutes' application to each side), provided that daily treatment can be carried out for several weeks successively.

Our list of drugs furnishes some agents of valuable efficiency in the application of which you will do well to vary from time to time. In the first place, I would mention sodium phosphate, which, given in adequate doses (6, 8, 10 grm per day, dissolved in water, soup or milk), appears to me to act almost as a specific on most of the *Basedow* symptoms. Whether it exercises this action through its property of being an antidote to iodin, as *Kocher* thinks, or in some other pharmacodynamic manner, I will not attempt to decide, and only affirm the imperative fact; sodium phosphate must be given in relatively large quantities, since it is only to a small extent absorbed. More absorbable and assimilable are the organic phosphoric combinations, for example, calcium glycero-phosphate (0.25 grm 4 times daily). I only give it preference over sodium phosphate when this last sets up diarrhœa.

We give bromin and valerian preparations symptomatically for jactitation, for which the last is in general to be preferred, since besides this it is the most efficient internal medication which we have at our disposal for the cardiac and vasomotor disturbances of innervation. The result is, however, only to be expected with some certainty when the valerian preparation is given continuously and in large doses which I have denominated "valerian saturation." By preference I order valerian tea. It is most efficient when it is taken according to the following directions: In the morning the patient adds a heaping tablespoonful of valerian root to a large cup of cold water and lets it stand all day, occasionally stirring it; before bedtime, then, it is filtered through linen, expressed, and the concentrated infusion so obtained is taken. The next cup of tea is immediately prepared in the same manner, is put on the night table, and taken immediately after awakening. Boiled valerian tea, or that made with hot water, acts much more weakly, since many of the ethereal components and volatile acids are thereby driven off. The wretched taste of valerian tea has created a need for a number of new valerian specialties (Valyl, Bornyval, Gynoval, etc.) which, given in proper doses, also act quite well, but are rather expensive and only applicable to elegant practice.

I would warn against the use of digitalis and strophanthus except in such stages of the disease in which heart insufficiency and asystole dominate the picture. Apart from the above-mentioned indications in which they fill a

decided indication, these cardiac remedies accomplish nothing in *Basedow's* disease, if they do not, indeed, injure. Belladonna preparations and atropin, which are adapted to the symptomatic treatment of the hyperidrosis, have in certain cases also a favorable influence upon the disease picture as a whole, as *Gowers* and *Grasset* have shown. The use of ergot preparations and quinine appear to me to rest more upon theoretical grounds—attempting to act upon the vessels—than upon favorable and empirical results. Arsenic medication, on the contrary, properly enjoys the greatest popularity in the treatment of *Basedow's* disease; beginning with small doses as customary, we gradually increase to larger ones; for instance, acid. arseniosi, 0.002 to 0.005 (gr. 1/30 to gr. 1/12) twice a day, best in pill form, as Asiatic pill, or sol. Fowleri, 3 to 7 drops t.i.d., or sodium cacodylate, daily 0.05 to 0.15 (gr. ¾ to gr. 2) subcutaneously. Also courses of Levico-, Roncegno-, Val Sinestra or Durkheimer arsenic waters are in place.

Most interesting are the attempts at opotherapy based upon the thyrogenous theory of *Basedow's* disease. So it has been attempted to introduce into the patient the antagonistic "myxœdema poison." Already, in 1895, *Ballet* and *Enriquez* had injected the serum of dogs in whom the thyroid had been removed, with encouraging results; only since 1901, however, has a serum preparation more simple in application the "antithyreodin" of *Möbius-Merck*, which is obtained from the serum of sheep deprived of their thyroids and ordered in doses of from 1.5 to 6.0 a day, has come into commerce. In practice the use of the flesh of thyroidectomized animals (Sorgo) and the serum of myxœdema patients (*Burghardt-Blumenthal*) have fallen entirely out of use, still more so the anthropophagous methods of *Lanz* and *Möbius* (the administration of thyroid substance from cretins). On the other hand, the milk of thyroidectomized goats (introduced into commerce in a pulverized condition as "Rodagen") has come into use after the advice of *Lanz*. Finally, in France the glycerinized blood of thyroidless animals has been given as "Hémato-éthyroidine."

Most recently it has been endeavored to prepare a serum containing the specific cytolysines directed against the thyroid gland; the interesting results of various experimenters (*Mankowsky*, *MacCallum*, *Lépine*, and others) have not, however, proved of practical value.

The pathogenetically so rational opotherapeutic measures have not yet become established as a part of the therapy of *Basedow's* disease, and it is questionable if this will ever be the case. According to their nature, these substances exert only a temporary action, and are able to influence the severe cases, which, as has been recognized, are insusceptible to recovery—only during the time that they are being administered, and here, even, on account of the high price of these anti-thyroid remedies (a course of antithyreoidin may cost five hundred francs), they can rarely be chosen. Their indication is found above everything in acute intermediate cases where there is danger in delay, for example, from attacks of suffocation. A protracted use of the antithyroid remedies will only take precedence over the other methods already mentioned in those fortunately rare cases of very severe *Basedow's* disease.

A formidable rival to them, besides the of late recommended X-ray treat-

ment of goiter, has been developed in surgical treatment. With a description of this last we will close this lecture.

Practically abandoned is resection of the sympathetic in the neck which *Jaboulay* attempted to establish. This operation can, indeed, favorably influence goiter, exophthalmus and tachycardia, but only temporarily, and has also acted unfavorably. The same author has recommended "exothyropexia," in which the thyroid is drawn out through an incision, covered with a dressing, and left to the spontaneous atrophy which occurs in favorable cases; this operation also has been abandoned (on account of the dangerous results which have repeatedly followed it). *Th. Kocher* favors partial strumectomy in combination with ligature of the arteries, and with him the majority of surgeons agree. One must not conceive, however, that the results which this operation often produces in an astonishingly short time must be permanent, and only too often one sees rapid return of the troubles in their former, or even in greater intensity, which has often led to a new partial resection of the thyroid. The mortality of thyroidectomy in *Basedow* patients is about 5 per cent., mainly on account of the vascularity of the goiter and the labile condition of heart and vasomotor nerves. It will be well to reserve operative interference for severe cases which do not react to other methods, but here, however, not to put off this last resort until the disease has reached a stage at which life is directly threatened. Although there are surgeons who on principle advise against any strumectomy in *Basedow's* disease (for example, the well-known student of goiter, *Heinrich Bircher*), on the other hand, even in some cases, which from their nature could be very well cured bloodlessly, in practice, on economic and social grounds we cannot oppose operation, since these patients on the one hand are doomed to invalidism through the severity of their trouble, on the other are not in a position to afford a long rest cure, change of climate, etc.

LECTURE XXIV

Dysglandular Symptom-Complexes

II. Myxœdema

The pathological condition to whose description we must now proceed was first studied clinically by the Englishman *Gull*, in the year 1873, but received its present name 5 years later from *Ord*, since which time the name proposed by *Charcot*, "cachexie pachydermique," has become almost entirely obsolete. The most important step in the recognition of this disease was furnished by the discovery by the Geneva surgeons, *J.* and *A. Reverdin*, that after total extirpations of the thyroid gland, a condition entirely analogous to spontaneous myxœdema appeared ("cachexia strumi-priva," after *Kocher*). On this account the conception that spontaneous myxœdema also is a hypo- or athyreosis, a more or less complete insufficiency of the internal secretion of the thyroid gland imposes itself. *Ewald* has found pathologico-anatomical support for this view: He showed that the thyroid in almost all cases of myxœdema which had come to autopsy was atrophic, cirrhotic, showed a destruction of the parenchyma of the gland, with fibrous proliferation of the interstitial tissue, and also that in the macroscopically enlarged thyroid glands of myxœdema patients, the increase in volume is caused by connective tissue increase at the cost of the secreting epithelium. It is noteworthy that sometimes enlargement of the hypophysis has been found in myxœdematous conditions (perhaps to be considered as an attempt of the organism to effect compensation).

The chief symptoms of myxœdema are a peculiar alteration of the integument—by infiltration of the meshes of its connective tissue with a mucoid semi-fluid mass—and intellectual disintegration. Along with this there is also, in congenital myxœdema (thyreoaplasia) and infantile myxœdema, retardation of the growth of the skeleton; since, however, we have already become acquainted with both these forms as sporadic cretinism, we will consider to-day only the myxœdema of adults, and first in its spontaneously appearing form.

SPONTANEOUS MYXŒDEMA OF ADULTS

As exciting causes of this form, like in its pathogenetic antithesis, *Basedow's* disease, psychic traumata (fright, excitement, etc.) appear to play a certain rôle. Also infectious diseases and loss of blood (particularly in consequence of repeated difficult labors) have been accused. Certain relations to

the function of the female genital organs are made probable in that women are 4 times as frequently affected as men; virgins, however, very rarely. Also, a local predisposition without doubt exists. In England and France, for example, spontaneous myxœdema occurs more frequently than in Germany, Austria and Switzerland (although the two last countries show numerous endemics of goiter and cretinism). In England hereditary-family occurrence also does not appear to be very infrequent.

A rapid development of the symptoms is entirely unusual, rather do these generally establish themselves gradually and little noticed in the course of years, until in typical cases the following exceedingly characteristic disease picture is present: The skin takes on a pale yellowish coloration and an œdematous appearance, though pressure of the finger does not leave behind the pitting so characteristic of renal and cardiac anasarca; this is a hard and elastic infiltration, a pachydermia; the face upon whose waxlike pallor two rose-red spots appear over the cheeks, is swollen like a full moon; the thickened, deformed eyelids make the palpebral fissure appear very narrow; the cheeks appear puffed out, the lips form thick swellings, and are slightly cyanotic in color; the forehead lies in coarse, immovable folds, the physiognomy is expressionless, often dull. The fingers are like blocks, sausage-shaped, the wide, plump hands remind one of fencing gloves; the feet are deformed like paws. The infiltration of the skin can render the use of the extremities quite difficult. The mucous membranes undergo similar alterations to those in the skin; on this account the volume of the tongue increases considerably and the myxœdematous alteration of the laryngeal mucous membrane makes the voice hoarse, deep and monotonous. The sweat secretion ceases entirely, or at least becomes markedly limited; the integument often becomes cracked and falls off in scales. Its electrical resistance is considerably increased, its sensibility dulled. The hair of the head and body, the eyebrows and lashes, fall out or become very sparse; often, also, the finger nails and toe nails become brittle and are lost. The movements grow continually slower and weaker, although no paralysis occurs. Locomotion is carried out at a snail's pace. Nearly always (there are exceptions) mental impoverishment goes hand in hand with this physical decay. The patients lose every interest, become weak in memory and judgment. If one talks with them, they must think a long time for an answer, and make the impression that they are continually going to sleep during the conversation; in fact, they show abnormal sleepiness, torpor. They complain a great deal of feeling cold; the temperature of the skin and the central temperature are lowered, the last sometimes below 36° C. (96.8° F.). Headaches and tinnitus aurium occur in a portion of the cases, sometimes there is also a hemorrhagic diathesis (metrorrhagias, bleeding from the gums, etc.); further, occasional accompaniments of the disease are: chronic synovitis of the knee-joint, albuminuria with hyaline casts, and certain blood alterations, which *Vaquez* has studied; numerical decrease of the red blood corpuscles, with increase of their size and appearance of nucleated erythrocytes (infantile peculiarities of the morphological blood picture).

The sexual sphere is almost always affected; besides menorrhagia, amenorrhœa is also observed, libido disappears, frigidity appears; in men there is

also impotence. The heart action is usually weak, the pulse small, occasionally irregular; the blood pressure is often lowered. Particularly interesting, since they stand in direct contradiction to those of *Basedow's* disease, are the anomalies of metabolism in myxœdema patients, studied by *Magnus-Levy;* gas interchange is reduced to about half the normal, bodily weight decreases considerably; the appetite is sometimes reduced, but even where this is not the case a decided repugnance to meat is usually apparent. Palpation of the thyroid gland is made more difficult by the infiltration of the integument; usually no glandular body is to be felt; sometimes, however, an abnormally hard one which is then, as a rule, very small, rarely enlarged. The tendon reflexes are sometimes normal, sometimes weakened.

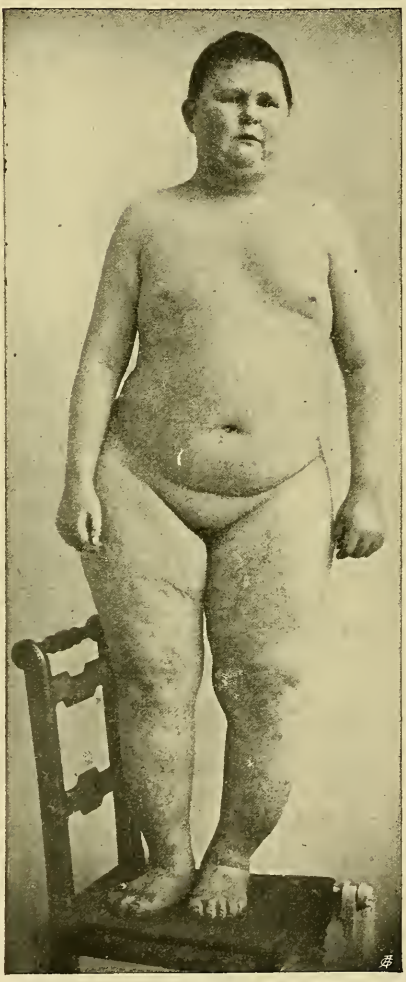


Fig. 102.
Myxœdema.

If the disease is uninfluenced therapeutically, it is slowly progressive, though often with remissions, which have been observed under the influence of summer temperature, sometimes also upon the occurrence of pregnancy. The patients usually die of intercurrent diseases (particularly phthisis), more rarely from the severe cachexia, which the terminal stage of myxœdema presents.

As in *Basedow's* disease, the "*Formes frustes*" of this disease also demand our special interest, since on the one hand they appear much less rarely than the severe form, and on the other they can be much more easily overlooked. They have been designated as "benign hypo-thyroidism." Sometimes they represent, like the "*formes frustes*" in *Basedow's* disease, the preliminary stage of the typical symptom-complex. The skin change can be entirely absent, or may be present only as a suggestion in the form of a slight, nonœdematous swelling of the face. There is a marked feeling of general weakness and relaxation in spite of very good appetite and digestive function. The urine often contains some albumin and a few hyaline casts, so that one might think of a chronic rudimentary uremia. In the psychical picture, in only a part of the cases a certain apathy and loss of interest makes itself apparent; more frequently, however, so great a sopor comes over the patient upon all occasions that he has the greatest trouble to keep awake, and usually, just on account of this trouble, seeks professional aid. In part of the cases there is falling out of the hair, which is often limited to the lateral parts of

the eyebrows. Sometimes there is a feeling of cold, weak pulse, sexual indifference. The thyroid gland is either not palpable, or very small and hard.

OPERATIVE MYXŒDEMA, CACHEXIA STRUMIPRIVA

This form of myxœdema, since the serious results of total thyroidectomy have been recognized and this procedure has been carefully avoided in goiter patients (this would be in any case in malignant goiters), is scarcely ever observed any more. Still, as *Kocher* has shown, it can arise exceptionally, also after partial thyroidectomy, in consequence of a subsequent atrophy of the portion of the gland parenchyma left behind. On the other hand, cases of total strumectomy have been seen which remained free from myxœdema or acquired only a "*forme fruste*"; they possess, evidently, which is not so rarely the case, accessory thyroid glands. Post-operative myxœdema usually develops 3 to 4 months after the extirpation, in a somewhat more rapid manner than the spontaneous variety. The first symptoms are feeling of weakness, shivering, then the skin becomes altered in typical manner, the movements are slow, the hair falls out, intelligence decreases; in short, the clinical picture corresponds completely with that of the spontaneous cases. Formerly the combination with tetany was frequent; the importance of avoiding not only the parenchyma of the thyroid glands, but also that of the parathyroids, in goiter operations, was not yet known. The younger the individual the more severe the cachexia strumipriva; in children it leads to the picture of sporadic cretinism with complete idiocy and dwarfism.

Treatment of Myxœdema

There is only one, but fortunately a quite efficient, remedy for myxœdema of every variety; the thyroid gland substance of animals, which can be replaced by certain substances isolated from it (iodothyrin, thyreoglobulin), but without therapeutic gain and with the disadvantage of increased cost. Without doubt, the fresh thyroid substance from sheep, cattle or hogs works most actively, in doses of 1.5 to 3.0 grm per day, which doses, however, are only reached by degrees (beginning with 0.5 grm). One lobe of the thyroid gland of a sheep corresponds to 1.0 to 1.5 grm. The thyroid must be eaten raw ("à la tartare," spread on bread); however, the patient soon resists this method of administration, and it is often accompanied by difficulties in obtaining the very easily decomposable gland always fresh. On this account the compressed tablets of dried thyroid substance present the most convenient and useful manner of administration. If they are not prepared from entirely fresh material under aseptic precautions, they can act injuriously on account of their content of ptomaines; hence, one should restrict himself to tablets of known efficacy (as, for example, the "Tabloids of Burroughs, Wellcome & Co.," or Knoll's "Thyradine"). Both these products contain 0.3 grm glandular substance per tablet. The daily dose is raised slowly and cautiously from 1 to 5 or even 10 tablets a day.*

* The dosage given here is for adults; that for children is given on page 343 when speaking of sporadic cretinism.

While using this remedy, continued control of the patient is necessary, since under no circumstances should the symptoms of intoxication known as "thyroidism" be allowed to occur; these manifest themselves in tachycardia, palpitation, jactitation, rapid loss of weight, reduction in strength, attacks of vertigo, vomiting, diarrhœas and exanthems. The patient is best protected from these unpleasant, often dangerous, incidents by discontinuous administration; 5 to 6 days organo-therapy, 4 days pause, etc. Decided improvements are obtained, often cures, which verge on the marvelous. The infiltration of the integument gives way to normal conditions, the temperature rises, the hair grows again, movability, activity of mind return, etc. If normal conditions have been reattained, a prophylactic permanent treatment with small doses of thyroid must be ordered. On the average it is correct to give one lobe of sheep's thyroid or 3 to 5 tabloids or thyraden tablets a week. The best criterion for determining the efficient dose is, as *Combe* has shown, the body temperature, which should remain permanently between 36½ and 37½° C. As a régime, this author recommends chiefly lacto-vegetarian diet. Meat is best permitted only with the juice squeezed out, without the bouillon.

III. Adrenal Insufficiency and Addison's Disease

Our knowledge of the functions of the adrenals (glandulæ suprarenales), whose rôle in the organism has long been enveloped in obscurity, has been much advanced during the last few years by many experimental investigations. Indeed, it has been known since 1855-56 that the adrenals are absolutely necessary for life. This was proven by the pathologist *Addison* in agreement with the physiologist *Brown-Séquard*. To-day, however, we know many new and interesting details as to the functions of these peculiar glands. Among other things that their secretion acts as an antitoxic both against exogenous poisons (as for example, certain alkaloids), and also against the toxines produced by muscular exertion, and that they produce lecithin and pigments in their cortical layer, in their medullary substance, on the other hand, adrenaline and angiotonic substance, which slows and strengthens the heart beat, causes the circular muscles of the blood vessels to contract, and decidedly raises the blood pressure. Through its special ability to produce adrenaline, however, the medullary substance of the adrenal shows that it belongs to the "chromaffin system" (called after its histo-chemical relations). To this last we also assign besides this a number of small structures, the paraganglia, which lie in the retroperitoneal tissue near the abdominal aorta, further, diffused cells within the sympathetic nerves and ganglia, and, finally, the so-called "carotid gland" in the neck. The medullary substance of the adrenal and the rest of the chromaffine system show close anatomical and physiological relations to the sympathetic nervous system and have, indeed, been denominated directly as the "accessory apparatus of the sympathetic." On the other hand, there appears to be reciprocal action between the adrenals (cortex and medullary substance) and the chromaffine system on the one hand, and the thymus, the lymphatic apparatus and the sexual glands on the other; for this speak among other things, the hypoplasia of the paraganglia and the suprarenal medullary

substance which is found in status thymico-lymphaticus, and the hypertrophy of the whole lymphatic apparatus which is found in adrenal tuberculosis; further, the occurrence of marked hyperplasia of the adrenal cortex ("struma suprarenalis") in pseudo-hermaphroditism, in inversion of the sexual characters, in precocious puberty, etc. For the neurologist, however, very special interest is presented by the exceedingly frequent coincidence of defective development of the adrenals with anencephaly, microcephaly, encephalocele, hemicephaly and other congenital brain defects, a correlation whose nature is still disputed but which is none the less striking; possibly the function of the suprarenal gland as the locality of production for the lecithin, necessary for the upbuilding of the brain, plays a rôle here.

All these things have mainly theoretic interest. Clinical importance is to be attached above everything to the symptom-complexes arising from insufficiency of the adrenal function, the most striking of which is that known to us since 1885 as "*Addison's* disease." About 90 per cent. of the cases of this disease show on autopsy destructive lesions of the adrenals (usually this is tuberculosis, more rarely malignant tumors, syphilis, interstitial inflammatory atrophies). In such cases, however, in which, after *Addison's* disease, the suprarenal glands have been found intact, their function has been plainly suspended by disease of the chromaffine elements of the paraganglia and of the sympathetic plexus of the abdominal cavity so closely related to them physiologically. *Von Neusser* and *Bittorf*, indeed, take the not yet generally accepted position that an affection of the splanchnic nerves, which, as *Biedl* has shown, contain the secretory nerves for the adrenals, can cause *Addison's* disease. On the other hand, the occasional cases of tuberculous destruction of the suprarenal glands without "*Addison's*" are brought nearer an explanation by an observation of *Wiesel*, who in a case of this character found vicarious hypertrophy of the paraganglia and other portions of the chromaffine system.

Addison's disease begins gradually; it may be in a manifestly tuberculous, or again in a previously healthy individual; usually abdominal pains or rapid loss of strength are the first symptoms; sometimes, however, the peculiar coloration of the skin, which *Addison* denominated "bronzed skin," and which also bears the name of "melanodermia," occurs at the start.

At first appear brown-black spots on such regions as are normally rich in pigment, or are usually exposed to the sun; for example, the groins, about the genitals, the axilla, about the nipples, on the face, the neck, the backs of the hands. These spots become continually darker, and are finally confluent, so that the patients assume a mulatto-like appearance; as, however, in mulattos, in *Addison's* disease, the matrix of the nails, as well as the palms and soles, usually remain free from pigment. On the face sometimes, upon a dark ground, still darker points are found, also the hair of the head and the beard can occasionally become darker. Very frequently the melanodermia extends also to the mucous membranes, that is, to the inner surface of the cheeks and the palate; here, however, the single dark spots do not usually run together.

The second cardinal symptom of *Addison's* disease is adynamia, an exhaustion occurring upon the slightest use of the muscles. The patients on this

account become continually less able to get about; they assume a "drooping attitude," finally become bedridden. With this, examination of the function of the muscles shows that there never is true paresis or paralysis.

There is always, too, a decided relaxation and loss of energy in the mental sphere. All intellectual exertion is avoided or fails, the patients become increasingly apathetic; the duration and depth of sleep are increased. Only rarely psychic irritative symptoms appear; for example, deliria, hallucinations, delusions. As an anatomical basis of these anomalies, a diffuse encephalitis in the frontal region has been accused by *Klippel* (encephalopathia Addisonia).

Sensory disturbances are present in only part of the cases. Usually these are pains which are located in the epigastrium, in the loins, in the hypochondria, in the joints, in the head, and sometimes show an irritative, radiating, neuralgiform character. More rarely, there is general hyperesthesia of the integument, never anesthesia or hypesthesia. The reflexes usually show no anomalies. Without exception there are gastro-intestinal disturbances, great loss of appetite (rarely and only episodically, excessive hunger), frequent vomiting, constipation, in the later stages diarrhœa. The pulse is small and weak. In women the menses usually stop. The blood pressure falls off decidedly; in the advanced stages the body temperature also (in this, subjective feeling of cold). There occur attacks of vertigo, tinnitus aurium, temporary amblyopia, syncope. The patients become continually more and more anemic and thin, finally cachectic. In the preponderant majority of cases, *Addison's* disease ends fatally, though its course is usually rather protracted and may extend over from 2 to 4, sometimes even over 10 or more years. Epileptiform convulsions with subsequent coma may precede death. Acute cases in which exitus takes place after a few weeks or months, are quite rare; still rarer, cases terminating in recovery, which is chiefly to be expected in adrenal syphilis. On the other hand, extended remissions occasionally occur in the course of the disease.

The most striking symptom of *Addison's* disease, the bronzed skin, greatly facilitates the diagnosis of this variety of adrenal insufficiency (one should indeed, never forget to exclude other diseases in which also a melanodermia may develop: pigmentary syphilides, melanodermia from pediculosis ("morbus vagabundus") malarial cachexia, cirrhosis of the liver with pigmentation, bronze diabetes, pellagra, *Basedow's* disease, chronic silver and arsenic intoxication. There is also an adrenal insufficiency ("hypoepinephria") without pigment anomaly, which most probably depends upon the fact, that in such cases the cortical layer is more or less spared by the destructive lesion (*Bittorf*). The symptomatology agrees otherwise so closely with that of *Addison's* disease that these rather rare observations may be considered as "formes frustes" of this.

As the opposite of adrenal insufficiency may be considered certain nervous symptom-complexes which, for example, occurring after hemorrhages into the adrenal (*Materna* and others) have been included under the name "Hyperepinephria" and considered as the result of an oversaturation of the organism with the hormones of the suprarenal glands. These observations are still too controverted to be described here.

It still remains for us to describe the treatment of adrenal insufficiency. Great hopes have been placed upon the use of extracts or organs, hopes which have proved to be much exaggerated, however. Both the chopped up fresh substance of the suprarenal glands of the sheep (2 to 5 grm a day) and dry adrenal extract (about 1 grm a day) have been administered; further, subcutaneous injections of adrenalin (1 cc of adrenalin solution (1 to 1000) in 250 cc physiological salt solution) have been tried, in this, on account of the vessel narrowing effect, the absorption is very slow. In many, but not in all cases, these remedies have effected definite, though temporary, remissions; a comparison with the stupendous results of thyroid medication in myxœdema is, however, not in the slightest degree possible. Adrenalin has, indeed, repeatedly harmed *Addison* patients. Hence, one must satisfy himself with increasing as far as possible the nutritive value of the diet with the avoidance of all overexertion, and with the administration of tonic remedies (iron, quinine, arsenic). Considering the antitoxic function of the adrenals, their insufficiency demands the strictest avoidance of alcohol and tobacco. Constipation should be combated with medicines only with the greatest caution, since otherwise the outbreak of uncontrollable diarrhœa is risked. Syphilitic patients are to be treated specifically; in adrenal tuberculosis *Strümpell* recommends a cautious trial of *Koch's* tuberculin.

IV. Acromegaly

In the year 1885 *Pierre Marie* called attention to a disease whose chief symptom he defined in the following manner: "A singular non-congenital hypertrophy of the extremities, superior, inferior, and cephalic," and to which he applied the appropriate name of "acromegaly." As a constant finding in the autopsies on such cases, disease of the hypophysis cerebri, the pituitary body, was found.

The disease begins, as a rule, between the age of 25 and 30 years and develops very slowly. The anomalies of growth accompanying it are often noticed indirectly by the patients from the fact that from year to year larger hats, gloves, thimbles, rings and shoes must be used. Also those surrounding the patient become aware of a gradual increase in volume of his nose, his chin, the distal portions of his limbs, and finally the picture becomes so characteristic that any experienced person can make the diagnosis at the first glance.

In the fully developed disease, the acromegalic presents coarse features with prominent brows and cheek-bones, great, prominent lower jaw, hanging lower lip, thick, knobby nose; the external occipital protuberance is excessively prominent and sometimes the ears take part in the hypertrophy. The tongue is enlarged, while a thickening of the mucous membrane of the palate, the tonsils and uvula as well as enlargement of the teeth, are rarities. Arm and forearm, thigh and leg are normal; so much the more striking are the disproportioned paws in which the extremities terminate. Usually this hypertrophy of the hands and feet extends chiefly in a transverse, only rarely in an axial direction ("type en large," "type en long"). The fingers and toes are wide, quadrangular, the nails on the other hand, which have not taken part

in the increase of volume, appear small. The hypertrophy affects both the skeleton and the soft parts (the skin, subcutaneous tissues, muscles). Less constantly than the limbs, is the thorax altered; in this case it increases particularly in antero-posterior diameter, so that a kyphosis in the upper dorsal region along with knob-like bulging of the sternum forward occurs; also the collar-bones become very prominent. The neck appears shortened, the head drawn down between the shoulders. The larynx is often abnormally enlarged, the voice is rough and deep, which is particularly striking in women. The skin is discolored, dry, often covered with warts, feels spongy on the hands and feet. Falling out of hair is rare. Of other anomalies there have been found occasionally, hypertrophies of the viscera, heart, liver, spleen, etc.; almost regularly, on the other hand, disturbances in the sexual organs; in women there is early amenorrhœa and sterility with atrophy of the breasts; in men impotence, sometimes also, atrophy of the genitals. In about one-third of the cases there is glycosuria with polyuria, polydipsia, polyphagia, sometimes with the criteria of diabetes mellitus. Abnormal fatigability and great feeling of weakness, are almost always present; sometimes there are neuralgic pains in different groups of muscles which has been designated the "hyper-algesic form" of acromegaly. Psychically the patient is apathetic and permanently out of sorts. The tendon reflexes are, as a rule, normal, sometimes reduced.

Acromegaly always has as its basis, disease of the hypophysis and indeed, as we know to-day (thanks to the most recent very thorough investigations, and contrary to the original opinion of *Marie*), in an alteration leading to overfunction of its glandular portion, a hyperpituitarism.* These are chiefly simple hyperplastic tumors, the so-called strumas of the hypophysis, further, adenomata with all transition forms to adenocarcinoma and carcinoma. That tumors of the connective tissue series have also occasionally been found (sarcoma) was for a long time brought forward as an argument against the consideration of acromegaly as hyperpituitarism. Still, *Hanau* and *Benda* have furnished the pathologico-anatomical evidence that these neoplasms which appear sarcomatous are nevertheless to be classed among the malignantly degenerated glandular tissue tumors and indeed, in the locations appearing like sarcoma, are infiltrated with glandular epithelium. Only rarely in acromegaly is a hypophysis tumor not present in such cases, in part, hypophyseal adenomata have been found in other locations (for example, in the cavities of the sphenoid bone), in part there has been an increase of the chromaffine cells of the pituitary body, which are to be considered as its functional element, without macroscopical enlargement of this latter.

In general, however, by means of the X-ray the hypophyseal tumor can be recognized *intra vitam;* the sella turcica shows abnormal dimensions and is usually enlarged in a sagittal direction. Large neoplasms can, besides this, manifest themselves clinically by pressure and neighborhood symptoms; severe headache, cerebral vomiting, vertigo, slowing of the pulse, only very rarely papilloedema; on the other hand (from chiasma lesions) bitemporal hemianopsia, or even bilateral amaurosis. Further, by paralyses of the eye muscle nerves

* From "glandula pituitaria," the old, to-day obsolete, designation of the hypophysis.

which pass by the hypophysis on their way to the orbit, as well as of the first branch of the trigeminus.

The more or less great prominence of these special tumor symptoms is naturally of prime importance in relation to the course and prognosis of the individual case. *Sternberg* has differentiated three varieties of course, perhaps somewhat too schematically; the usual form, with a duration of from 10 to 30, the malignant, with one of from 3 to 4 years, and finally, the benign form which may extend over even 5 decades. In general the disease proceeds but very slowly and often remains a long time, sometimes even definitely, stationary. It does not seem susceptible of recovery. The proximal causes of death are intercurrent diseases, brain pressure, diabetes, cachexia.

Therapeutically we are rather helpless. Extirpation of the hypophysis by the nasal or temporal routes (*v. Eiselberg*, *Cushing*, and others) is so excessively dangerous and offers so little hope of radical removal that it must be reserved for cases with marked symptoms of brain pressure, unbearable headache, threatened blindness. As to the X-ray treatment of acromegaly (acting upon the hypophysis from the naso-pharynx) no decisive opinion can yet be formulated. On the other hand, repeated courses of arsenic, raising it to large daily doses, appears to favorably influence the course in most cases (*Brissaud*). The neuralgiform pains and the diabetes demand symptomatic and dietetic treatment.

In differential diagnostic relations, as opposed to acromegaly, there come before everything else into consideration, the different varieties of giantism produced by a delay of the ossification in the region of the epiphyseal synchondroses, in consequence of which growth comes to a standstill not at all, or only very late, and the skeleton assumes excessive dimensions. There are distinguished partial giantism, which usually affects only one or two extremities (or indeed, only parts of them; for example, in one case of *Wieland's*, only the anterior half of one foot), and general giantism. The last stands pathogenetically plainly very near to acromegaly, in a great number of cases; in this, indeed, enlargement of the hypophysis cerebri has been confirmed both Roentgenologically and on autopsy. If hyperpituitarism occurs in children, it also leads, as *Brissaud* has shown, to giantism, if it arises, however, after ossification of the epiphyseal lines of the skeleton, it makes itself evident by acromegalic changes. In hypophyseal giantism, the genitals usually remain rudimentary, also the secondary sexual characteristics (beard, pubes, etc.), do not develop. With acromegaly, further, *Marie's* "pulmonary osteoarthropathy" (Ostéoarthropathie hypertrophiante pneumique") should not be confused. In this last there is an enormous drumstick-like thickening of the end phalanges with marked curvation of the nails and ankylosis of the finger and toe joints—changes which in patients with chronic bronchiectasies (more rarely with other diseases of the bronchi and lungs) occasionally develop.

OTHER DYSGLANDULAR SYNDROMES

If we now should go over all the dysglandular symptom-complexes not yet described, we would exceed the limits of a neurological text-book in a manner

which would be as little justified by the slight occurrence of nervous disease pictures in agenitalism and hypogenitalism (castrated and eunuchoid persons), in infantilism and its different varieties, and in the "pluriglandular insufficiency" resulting from the secretory disturbances of numerous glandular apparatus, as by the slight practical importance of these conditions. Hence we will content ourselves with a short consideration of two clinical pictures which indeed are very rare, but as the newest acquisitions to the subject which has occupied us for the last two lectures, attract actual interest.

Adiposo Genital Degeneration (Fröhlich's Disease, Hypophyseal Eunuchism)

This rare disease is due to hypophyseal tumors which cause no hyperfunction of the pituitary gland, but rather act destructively, also to traumatic lesions, as, for example, the entrance of a rifle bullet into the sella turcica in an observation of *Madelung*. It is hence a clinical expression of hypopituitarism, of hypophyseal insufficiency which is confirmed by the improvement of the symptoms upon the administration of hypophyseal substance. Sometimes disease processes which are located, not in the hypophysis itself, but in other parts of the base of the skull, by "neighborhood action" can lead to the cardinal symptoms of adiposal genital degeneration: excessive development of fat, along with arrest of development of the genitals and of the secondary sexual characteristics (respectively disturbance of function and atrophy of the sexual organs in case the disease occurs after puberty). Sometimes this hypophyseal eunuchism is combined with dwarfism. That the genital disturbance is common to hyper- and hypopituitarism, may appear paradoxical; still, *Basedow's* disease and myxœdema have traits in common, for example, disturbances of digestion, irregularity of the menses, falling out of the hair.

Dyspinealism

Only most recently has it been made known through *Marburg* in Vienna that the epiphysis, the pineal gland, is a blood gland, whose hormone during childhood has important influence upon the mental and physical development of the individual. On this account, in children, certain tumors in the region of the corpora quadrigemina, along with the correlates of disturbances directed into neighboring neurones (paralyses of the pupil and the external eye muscles, ataxia, hardness of hearing, visual disturbances) produce the following symptom-complex: Hyperplasia of the genitals, abnormal growth of the body in length, unusual growth of hair, sexual and mental precocity. *Marburg* assumes in these cases an insufficiency of the pineal secretion; the last acts, in that it normally delays the sexual development, antagonistic to the hypophyseal secretion.

LECTURE XXV

Diseases of the Sympathetic, Angio- and Tropho-Neuroses

Gentlemen: That portion of our nervous system of whose disturbances we will speak to-day, has other attributes than the brain, spinal cord, motor, sensible and sensory nerves. Separated from our consciousness and our will, to a certain extent independent of the cerebrospinal apparatus, it appears not by far to reach the importance of the latter. Indeed, it plays—in the language of *A. v. Kolliker*—while the brain like a mighty ruler is enthroned high up in the state chambers of the skull, only the rôle of a servant, who exercises in the lower rooms of the body his appointed functions. And still, this is not without importance, and the brain itself is in the last instance dependent on it. Indeed, for the whole life of the individual as well as for that of the race, a system which regulates the heart's action, the whole circulation, the secretion and the processes of nutrition as well as reproduction, has a fundamental importance.

This nervous system has, as the sympathetic or autonomic, been contrasted with the cerebrospinal, as the visceral with the somatic and as the vegetative with the animal nervous systems. Of late, however, it has become customary no longer to use promiscuously the words "sympathetic" and "autonomous," but to apply the term "sympathetic" to the system of the gangliated cords, and to reserve the word "autonomous" for the visceral nervous apparatus in the head and the pelvic organs (cranial and sacral-autonomous systems). A separation into these two subvarities is justified among other things by differences in their susceptibility to toxicological influences which have become known to us, particularly through the brilliant work of the English physiologist *Langley*, to discuss which at length, however, would lead us too far afield.

We cannot, however, neglect to sketch briefly the principles which control the anatomical structure and the physiological differentiation of the visceral nervous system. Its peculiar criterion as compared to the somatic nervous system is the fact that numerous nerve cells are interpolated in its course far into the periphery. This interpolation occurs in two ways in that, on the one hand sympathetic cells are grouped in compact ganglia which, connected with one another by sets of fibers, form the so-called "gangliated cord"; on the other hand, however, both in the neighborhood of the viscera and also in the substance of these themselves, still further collections of cells are to be found. As you know, the gangliated cords extend along in front of the vertebral column on either side from the neck to the coccyx and are in communication through

the "rami communicantes albi" (white communicating branches) with the spinal roots and the ganglion cells of the spinal cord; on the other hand, they give off the "rami communicantes grisei" (gray communicating branches) which enter the spinal nerves and intimately mixing with their fibers pass farther, finally to reach the blood vessels, and to undertake their innervation. The sympathetic ganglia not belonging to the gangliated cord (the latter is called by *Gaskell* "the lateral," the former, the "collateral" ganglion system) are connected with it through the visceral plexuses. These last are distinguished

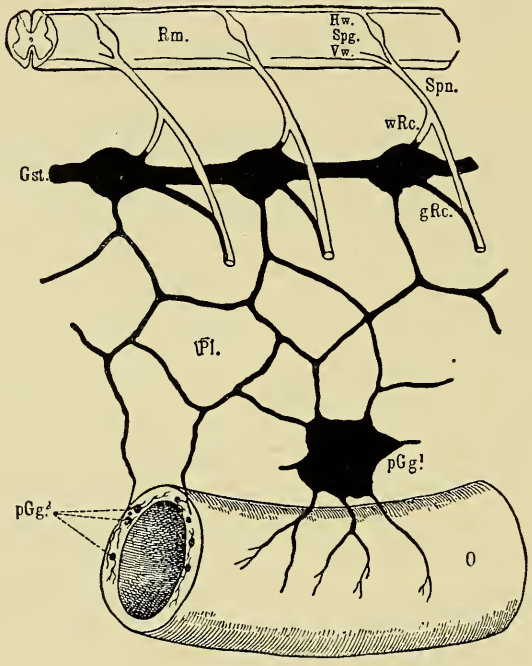


Fig. 103.

Plan of the Sympathetic Nervous System.

Rm = Spinal cord; Hw = Posterior root; Vw = Anterior root; Spg = Spinal ganglion; Spn = Spinal nerve; Gst = Gangliated cord (Lateral ganglion system); pGg^1 = Peripheral ganglion in the neighborhood; pGg^2 = Peripheral ganglion in the wall of the innervated organ (Collateral ganglion system); Pl = Sympathetic plexus; O = Innervated organ (hollow organ).

from the plexuses of the somatic nerves, on the one hand by their net-like structure, on the other by their gray color (without medullary sheath). Collateral ganglia are, for example, the cœliac, solar, mesenteric ganglia in the abdominal cavity, *Ludwig's*, *Bidder's*, *Remak's* ganglion in the heart wall. It is important that a different and to a certain extent antagonistic, functional significance is attributed to the lateral and to the collateral sympathetic ganglion system. The neurones arising from the cells of the lateral system of ganglia, that is, from the gangliated cord, subserve vaso-constriction, contraction of the circular muscles of the hollow organs and acceleration of the heart. On the other hand, those from the collateral collections of cells, effect

vaso-dilatation, contraction of the longitudinal muscles and heart inhibition. Specially in regard to this last it must be emphasized that the cardiac plexus receives an important addition from the vagus nerve (which, however, also anastomoses with the sympathetic plexuses of the lungs and stomach, as well as with the cœliac plexus) and we already learned in Lecture II that irritation of this nerve slows the movements of the heart. Is there in this an encroachment of the cerebrospinal system upon the function of the visceral? Are there in this way vegetative functions exercised directly by the brain without the intermediary of the sympathetic? Not at all. The vagus has in its rôle as a heart nerve, only to be considered in principle a "ramus communicans albus." The inhibitory apparatus proper of the heart are intracardiac ganglia—belonging to the collateral system—which give off their "post-ganglionic" inhibiting fibers to the heart muscle.

A. Diseases of the Sympathetic

The pathology of the sympathetic must, up to the present, be designated as that department of nervous diseases in which there is the least actual knowledge, and it has been attempted to supply its place by more or less well-founded hypotheses. So of late, some authors of the Vienna School (*Eppinger*, *Hess* and others) sketch a symptom-complex which may owe its origin to a hyperfunction of the vagus or of the autonomous system, under the name of "vagotony." Such patients manifest narrowness of the pupils (irritation of the ciliary ganglion), salivation and lacrimation (irritation of the chorda tympani and of the lacrimal nerve), hyperidrosis (irritation of the sweat nerves), bradycardia, irregular respiratory pulse, respiratory arhythmia (irritation of the heart and lung vagus), bronchial asthma (irritation of the smooth muscle fibers of the bronchial tubes), hyperacidity and increased stomach peristalsis (irritation of the stomach vagus), and still other symptoms.

The above-mentioned authors support their pathogenetic conception of these conditions upon a certain pharmaco-dynamic reaction of the patient in question: all the symptoms of "vagotony," according to them, disappear after large doses of atropin, on the contrary, undergo considerable exaggeration after the administration of pilocarpin, while the so-called adrenalin glycosuria is absent after adrenaline injections. Further investigations must show in how far such a localization of a clinical syndrome can lay claim to proof by means of experimental pharmacology.

Best known and most simply explainable physio-pathologically, is that symptom which is produced by a lesion of the sympathetic in the neck and which we call "*Horner's* symptom-complex." We have already pointed out (see page 146) that through the third ganglion of the cervical gangliated cord, the fibers from the ciliospinal center of the lowest cervical segments of the cord run to the eye: they innervate 1, the superior tarsal muscle; 2, the orbital muscle; 3, the dilator pupillæ; loss of their function hence causes enophthalmus (sinking in of the eyeball), narrowing of the palpebral fissure (sympathetic ptosis) and paralytic myosis (see Fig. 104). Since now, however, the cervical sympathetic also contains vasomotor and sweat secretory fibers for the

corresponding half of the face, there results further from their elimination a vasomotor paralysis in this region; this manifests itself in fresh cases by heat and reddening, later, however, usually by cyanosis and coldness (see Lecture I, page 9), and anidrosis of the same region. Still, in *Horner's* symptom-complex there is quite frequently hyperidrosis of one side of the face, hence irritative and defect symptoms seem combined. As causes of *Horner's* syndrome the following factors acting upon the cervical sympathetic come into consideration: Goiters, wounds (also operative) glandular tumors, for example malignant lymphoma, "cervical ribs," etc. Not rarely, however, the most exact examination does not permit the discovery of such causal factors and we have

Fig. 104.

Horner's Symptom-Complex.

(Enophthalmus, narrowing of the palpebral fissure myosis, in lesion of the cervical sympathetic on the left side.)

then before us a pathological manifestation, harmless in itself, which we, lacking a better explanation, must designate "neurotic." A unilateral symptom of irritation of the cervical sympathetic occurs relatively frequently in tuberculosis of the lung apex, in that upon the side of the pulmonary lesion the pupil, eventually, also, the palpebral fissure, is widened.

As "neuroses" and, indeed, as "vasomotor-trophic neuroses," are described a number of interesting disturbances etiologically unfortunately quite obscure but in any case acting upon the sympathetic distribution, of which we will now consider a few. These are acroparesthesia, angiospastic dysbasia, angiospastic

gangrene, scleroderma, neurotic dropsy, erythromelagia, facial hemiatrophy and hemihypertrophy.

B. Acroparesthesia

Acroparesthesia (the name originated with *Schultze*), readily the most frequent of the vasomotor neuroses, affects in the great majority of cases women, and appears most frequently between the ages of 40 and 55 years. The climacteric appears to create a special predisposition, as does also general nervosity (neurasthenia or hysteria); the frequency of the disease in individuals whose extremities are particularly exposed to wet and cold, for example, washwomen, points to the influence of exogenic factors.

The disease manifests itself in very unpleasant paresthesias, occurring in attacks, sometimes, indeed, increasing to actual pains, which are located mainly in the hands, less frequently in the feet. The causes of these unpleasant sensations (formication, prickling, tickling, stabbing, "going to sleep," etc.) are spasmodic conditions of the peripheral arteries, which are shown by the livid coloration resulting from pallor and cyanosis, which can be perceived in severe cases during the acroparesthetic attack, in the affected parts (while slight attacks present no visible alteration). The attacks occur by preference at night or in the early morning (when the blood pressure is lowest), and, as a rule, bilaterally. After the attack passes away, sometimes reactive reddening of the fingers or toes occurs. In the great majority of cases there are, during the attack, occasionally also interparoxysmally, slight disturbances of sensibility in the affected extremities; they present their maximum in the pulps of the fingers (or toes) and rapidly diminish proximally. *Dejerine*, *Trombert*, and others, have occasionally observed along with this typical condition ("ischemic disturbance of sensation" of *Schlesinger*), radicularly arranged hypesthesias in acromegaly; in spite of many reëxaminations I have not so far been able to confirm these observations.

The diagnosis of the disease is usually exceedingly easy, only one must remember that a number of other affections commence with paresthesias of the fingers or toes, or can do so. So, ergotism, "intermittent limping," "dysbasia angiosclerotica intermittens" (see Lecture XV, page 225), respectively its analogue located in the upper extremities ("dyspraxia angiosclerotica intermittens"), tabes dorsalis, tetany, acromegaly, and finally, as we will soon see, angiospastic gangrene (*Raynaud's* disease which is in so far related to acroparesthesia that it presents a malignant increased degree of intensity of the "vaso-constrictor neurosis of the extremities"). In any case, also the relations between acroparesthesia and vasomotor angina pectoris (*Nothnagel*, *Curshmann*) in which also by preference in the night and morning hours, the same subjective and objective anomalies appear on the fingers, but also with palpitations, precordial anxiety, pain in the region of the heart with radiation into the left arm, also the symptoms of true angina pectoris, are close. In contradistinction to the latter, however, there are absent all symptoms of sclerotic or luetic diseases of the aorta or the coronary vessels; rather does this evidently benign disease affect generally, physically quite healthy per-

sons, usually hysterical young women. Psychical traumata are nearly always discovered in the etiology; *Curshmann* accuses among other things coitus interruptus.

The prognosis of acroparesthesia cannot be laid down generally. There are cases which resist all treatment, though they usually present an alternation of improvements and relapses (such remissions usually occur during the warm part of the year). On the other hand I have seen not a small number of cases pass on to recovery and many permanently improve to an entirely tolerable stage. As to life, the disease is entirely harmless; as to function, not all too disturbing, since it scarcely ever causes actual disability.

From a therapeutic point of view electrical applications are to be recommended in the first place: The faradic brush, faradic hand baths, "four cell baths." Further, a course of quinine from time to time procures for many such patients a permanent or lengthy improvement. At bedtime a capsule of 0.5 quinine sulphate or "chinin-phytin" (combination with phosphorus) is given every night for from 2 to 4 weeks. Between these quinine cures proper, quinine in small doses can be given further, very well in the form of the following pills:

℞ Quinin sulph. ..... 3.0 (grs. 45)
Ext. nucis vom. ..... 1.5 (grs. 22)
Ext. hyoscyami ..... 2.5 (grs. 38)
Ext. valerian ..... 7.0 (grs. 105)
M. Fiat Pil. No. C. S. 2 pills t. i. d. after meals.

For washing, only warm water must be used. At night the patient should wear fur-lined slippers or socks.

C. Angiospastic Dysbasia

The "intermittent limping," occurring from organic disease of the peripheral, also of the spinal vessels, we have already become acquainted with in Lecture XV. There is now, however, a purely vasomotor form of this syndrome to which *Oppenheim* first called attention and which of late has been accurately studied by *Curschmann*. The first author observed a case of intermittent claudication for 15 years, without the severe results of vascular obstruction having developed. On this account there presented itself to him the possibility that permanent spastic conditions of the muscles of the artery wall, though varying in their intensity, can occur, and can produce the intermittent limping. He also expressed the suspicion that a congenital narrowness of the vascular system prepared the way for the occurrence of this disturbance. *Curschmann* saw in people from 18 to 22 years old intermittent limping with all subjective symptoms, also with the absence of pulse in some of the arteries of the foot, and could certainly exclude arteriosclerosis and arthritis. By plethysmographic investigations he was led to assume the possibility of a permanent spasmodic condition of the arteries. I myself have classed as vasomotor neurotic dyspraxia of the upper extremity, forms of writer's cramp which were accompanied by coldness and lividity of the hand, but the cases men-

tioned are on the whole quite infrequent and I would impress it upon you that the diagnosis of a functional, that is, a benign form of intermittent limping or similar functional disturbances of the upper extremities, should in any case only be made after long and thorough observation, and in case of doubt always to suspect an organic vascular lesion and to make this supposition the basis of your treatment.

This last, however, will be in general the same in angiospastic and in angiosclerotic intermittent dysbasia. The use of alcohol is to be limited, tobacco forbidden. Not less important is the absolute prohibition of those perverted therapeutic measures to which such patients only too often turn on their own account or from unwise counsel; cold water procedures, hot foot-baths, excessive gymnastics and massage, forced marches, etc. The same thing applies to the use of rubber stockings and bandages. To be avoided are all substances having decided vasomotor action, strong tea, coffee, spices. Further, care should be taken that the feet and legs are kept evenly warm (proper choice of residence and work-room, foot and leg coverings, bedclothes, occasionally mild rubbing with tepid water). Hot foot-baths are to be avoided; warm foot-baths, on the other hand, to be recommended, as also applications of Fango, peat, *Priessnitz* compresses, etc. *Erb* has quite warmly recommended the galvanic foot-bath; either in the fashion that each foot is placed in a separate tub containing salt water at from 34 to 36° C. on a pole plate and a stabile current of from 12 to 20 milleamperes is conducted through them, first in one direction and then in the other for from 3 to 6 minutes, or so that both feet are placed in a tub containing the cathode while the anode rests over the sciatic in the bend of the knee, or over the plexus on the sacrum.

Of drugs vaso-dilator substances come into consideration in the first place (courses of quinine as in acroparesthesia, also nitrites, nitro-glycerin and diuretin have been recommended); in the second place heart tonics, particularly those from which a better blood supply and favoring of the collateral circulation are to be expected, without action upon the vasomotors; also particularly the strophanthus preparations in contradistinction to digitalis. Simple or carbonated salt baths are to be recommended on account of their vaso-dilator effect. Of great importance, finally, is the regulation of the movements and of the use of the legs. At first, when possible, entire rest and long reclining are to be prescribed. In any event, however, all violent exertion is to be forbidden and walking is to be permitted only to such an extent as does not produce any difficulty. Only when decided improvement has been obtained, the patient, "watch in hand," is allowed to undertake more prolonged exercise.

D. Angiospastic Symmetrical Gangrene, Raynaud's Disease ("Asphyxie Locale Symmetrique")

This very rare disease—I have only seen three cases—is to be considered as the highest stage of intensity of the vaso-constrictor neurosis of the extremities. The occasional, but by no means regularly observed pathologico-anatomical findings (endarteritic and endophlebitic changes) are readily to be

considered as secondary. *Raynaud's* disease, in contradiction to acroparesthesia, shows no special predilection for either sex, and usually occurs in the comparatively young, at between 25 and 35 years of age. *Cassirer*, indeed, saw a nursling affected with symmetrical gangrene. Etiologically, neuropathic predisposition plays in any event the chief rôle. Many cases affect hysterical, psychasthenic, psychotic patients. In one of my cases there was a history of recent, but thoroughly treated syphilis (*Wassermann* negative), and lues is noted in a number of cases of *Raynaud's* disease in the literature. *Nékam* and *Curshmann* observed its family occurrence. As exciting causes, acute infectious diseases, trauma, severe emotions and chilling have been specially mentioned.

The disease begins usually with an attack of "local syncope"; one or several fingers or toes, through spasm of their arterial and venous vessels, become cold, waxy-pale, and so ischemic that deep needle stabs do not draw any blood. Now, however, I must warn you against considering every local syncope of this kind as a forerunner of *Raynaud's* disease. Much more frequently this phenomenon ("dead fingers") is a harmless symptom in neurasthenics and other psychoneurotics, as we will see in Lecture XXVII, also in *Bright's* disease. Characteristic for the *Raynaud* variety of this symptom is, on the one hand, its appearance in symmetrical positions in the upper or lower extremities, on the other, the (often very severe) pains which introduce or accompany it. With continuation, or even increase of these pains, there is added to the stage of local syncope which is of short duration, from minutes to hours, that of "local asphyxia," which lasts considerably longer and can extend over many hours, even several days. On the fingers or toes, and indeed, almost exclusively on the end phalanges, there appears now in spots, at first a bluish, then a continually darker, discoloration, reaching finally slate-gray in color; sometimes, besides this, there are extravasations of blood under the skin and bullous elevations of the epidermis. Now, in the favorable cases, the circulation is established again, when a decided reddening of the fingers appears, in place of the regionary cyanosis ("stage of local reddening"); in bad cases, however, the local asphyxia of the affected parts passes over into gangrene. This can be total, so that entire phalanges become mummified and cast off, or only partial, in which necrosis of circumscribed parts of the skin occurs. Moist gangrene is quite rare. I saw it in the case with luetic previous history mentioned above. In another still rarer atypical form of *Raynaud's* disease, not only the ends of the extremities, but larger or smaller portions of the leg, the buttock, and even the tip of the nose, the lobe of the ear, etc., are affected by the disease.

The relatively mild, that is, the *Raynaud's* paroxysms not coming to the gangrenous stage, can be repeated over years or decades, then at length to lead to mutilations. Without exception the general condition of the patient suffers considerably from the exceedingly painful attack. As a regular accompaniment of the peripheral vascular spasm there is to be mentioned marked hyperesthesia, which makes itself evident during this spasm in the parts affected; of occasional complications, are of interest, the paralysis of the sympathetic of the neck (*Horner's* symptom-complex), further, paroxysmal disturbances of the

sense organs, which may well depend upon vasomotor spasm (transitory deafness, blindness, ageusia). Here belong also the observations of transitory aphasia, hemiglobinuria, etc.

The course can be quite acute, in that, after one or a few attacks the disease comes to an end through casting off of the affected phalanges. In other cases, however, the ever returning paroxysms constitute a life-long scourge. Life itself (if we leave out the rare cases in which sepsis has occurred) is not threatened by angiospastic gangrene.

In differential diagnosis, as I already emphasized, the harmless form of "dead finger" is to be thought of. A benign disease is also presented in the "acrocyanosis chronica anæsthetica" of *Cassirer*. In this affection, an asphyxia of the ends of the extremities develops itself little by little and is accompanied by paresthesias, considerable local dulling of sensibility; sometimes also by trophic disturbances (increase of volume). The distinction of this from arteriosclerotic "spontaneous gangrene" can be difficult when attacks of intermittent limping or intermittent dyskinesia of the upper extremities have preceded this last by some time. Still, the angio-sclerotic dyskinetic paroxysms distinguish themselves from those of *Raynaud's* disease, in that they are dependent upon the use of the affected extremity and disappear upon rest, the accompanying vasomotor skin changes are never very great (a spotted, marbled condition of the integument, but no acro-cyanosis), and the pulse in the peripheral arteries permanently disappears or is greatly reduced. The separation of *Raynaud's* disease from "multiple neurotic gangrene of the skin" in which on different portions of the skin after a painful prodromal stage, or with only burning and prickling paresthesias, superficial necrosis takes place, is not sharp. In making this diagnosis, however, the greatest caution is necessary. The affection attacks very frequently hysterical patients, who as we will see, have an uncommon tendency to self-mutilation; in one of my cases by chemical recognition of silver in the sloughs, I was able to prove that the patient had produced the necrosis with nitrate of silver. Mutilations of syringomyelia and lepra are separated most sharply from *Raynaud's* disease by their painlessness.

The treatment of *Raynaud's* disease is to be carried out after the analogy of that for acroparesthesia and for intermittent limping (see above, pages 374-375), only one will do well to avoid all faradic applications, which here act unfavorably. The always present general nervosity must be combated with the greatest care, according to the principles to be mentioned in Lecture XXVIII; they must, indeed, be considered as the peculiar substratum of the vasomotor neurosis. The severe pains call for resort to the most active antineuralgics. Sometimes morphine injections cannot be avoided.

E. Scleroderma

This disease, which is usually observed in women of middle age and was described for the first time by *Thirial*, is much more frequent than *Raynaud's* disease, but still must be denominated a rare affection. It is characterized by the chronic development of a severe trophic alteration of the skin which only

rarely affects in a diffuse manner the whole integument, usually is limited to definite parts of the face, neck, thorax and upper extremities. The lower half of the trunk and the legs usually escape the disease. Etiologically we know nothing certain; the same points which we brought out in speaking of *Raynaud's* disease, could be repeated here.

The sclerodermatically altered parts of the skin in the fully developed disease, present an exceedingly characteristic appearance: they are thinned, smooth, of a dull, bacon-like polish, tense, cool, hardened and firmly fixed to the subcutaneous tissue. Their color is usually pale, sometimes, however, of a yellowish or brownish pigmentation, also vitiliginous and pigmented spots occur alongside of one another. Also the deeper layers of tissue (subcutaneous fat, muscles, tendons, skeleton of the hand) undergo an increasing atrophy. Move-

Fig. 105.

Sclerodactylism, with Formation of Necroses.

ments of the limbs, opening and closing of the mouth, are more and more hindered by the skin becoming "too tight." True contractures can indeed occur.

So, diffuse scleroderma can give to the patient a mummy-like appearance. As to the circumscribed forms, they furnish a great variety of striking clinical pictures. The alterations usually occur more or less bilaterally and symmetrically. They are found particularly frequently on the fingers; we then speak of "sclerodactylism"; with this there are also atrophic and degenerative processes in the nails, occasionally, indeed, such intense disturbances of nutrition in the end phalanges that actual necroses occur (see Fig. 105). (Such cases have, in my opinion incorrectly, been termed a combination of scleroderma and *Raynaud's* disease; as a diagnostic criterion of the last affection, in any case, the painful, paroxysmal attacks of regionary syncope and asphyxia are indispensable). Exceedingly rare is the "annular sclerodactylism" of *Düring* which, distributed in a circular manner can lead to the strangulation of the phalanges.

The diffuse scleroderma of the trunk and of the face seldom leads to round, but usually to streak-like foci ("sclérodermie en bandes"). Particularly typical are isolated streaks extending from the line of the hair to the eyebrows, reminding one of the scar of a sabre wound ("sclérodermie en coup de sabre," see Fig. 106). Also the mucous membranes can be affected (mouth, nose, pharynx). Particularly interesting, though also very rare, are the observations of certain circumscribed sclerodermas which were exactly limited to the area of a peripheral nerve or to a spinal root zone, or were localized on one side of the body (*Lewin*, *Heller*, *Bonn*, *Bruns*, *Curschmann*, and others).

As preliminary stages of the atrophic changes, a stage of firm œdema and an indurative stage have been described; still, it appears as if this was an artificial generalization of clinical pictures which by no means occur in all cases. No objective disturbances of sensibility worth mentioning occur in scleroderma, on the contrary the intact appreciation of sensation of the greatly altered skin (anatomically it is a "cirrhosis" of the chorium with thickening of the vessel walls) often appears to us paradoxical. Pains, on the other hand, are not rare, particularly in the hands. Though they also sometimes exacerbate, still they have neither the intensity nor the paroxysmal character of those which are peculiar to *Raynaud's* disease.

Fig. 106.

Scleroderma—"En coup de sabre," with *Horner's* Symptom-Complex on the same side.

Scleroderma can be combined with different nervous diseases. Particularly frequently does this appear to be the case with regard to *Basedow's* disease (observations of *Leube*, *Stähelin*, and others) which all the more attracts attention to its eventual connection with internal secretory processes, as *Bruno Bloch* and *Reitmann* in severe scleroderma, have shown, a striking variation of the nitrogen balance between positive and negative values; pathological increase of albumin destruction is, however, as we saw in Lecture XXIII, a typical occurrence in *Basedow's* disease. Rarer is the association with *Addison's* disease or tetany. The combination with *Horner's* symptom-complex is also quite interesting; the patient shown in Fig. 106 showed this last on the side of the scleroderma (in the picture unfortunately obscure). In the most marked case of diffuse scleroderma which I have seen (it affected a 13 year old girl) besides this, in the course of years, a general atrophy of all the muscles of the skeleton developed; also during the progress of the disease ulcers and fistulas, from which carbonate of lime was discharged, broke out in different locations. The patient died of marasmus. The examination by *Dietschy* showed that there was a combination of scleroderma with interstitial polymyositis and calcareous tendinitis.

From a prognostic point of view it is to be remarked that a cure of the changes is only to be thought of in the earliest stages, when no atrophy of the skin has yet occurred; later lesions, however, are to be considered as irreparable. Circumscribed forms in a location which does not cause any disturbance of function and without painful phenomena, usually cause no difficulty worth mentioning and can become stationary. To sclerodactylism, however, all this does not apply and it constitutes a severe disease, sooner or later leading to invalidism. Exceedingly extensive forms of scleroderma are always to be considered as very serious; respiration is impeded, the act of eating is imperfectly performed and the patients die in cachexia, if they are not taken off by intercurrent disease, against which they show themselves little resistant. Many of these patients die of degeneration of the heart muscle; diffuse scleroderma considerably impairs the circulation and so injures the heart.

In differential diagnosis it may be remarked that "glossy skin," as it occurs after a peripheral nerve lesion, should not be confused with scleroderma. The other skin atrophies (the striæ of pregnancy, senile atrophy of the skin, simple and pigmented xeroderma) have no resemblance to the picture of scleroderma, since in them all, the hardening is absent; a similar aspect, however, may be presented by scars (after leg ulcers, burns, lupus, etc.); still here the history will naturally exclude wrong diagnoses.

In treatment, above everything, frequent warm baths (eventually sulphur baths, mud baths, Fango packs) are to be recommended; also the steam bath and the hot air bath ("*Bier's* box") can be tried. Mild massage is nearly always found beneficial. Of internal medication the salicylic preparations appear to exercise a decided effect (sodium salicylate, acetylo-salicylic acid, salol, etc.); also ichthyol capsules (āā 0.25, 1 to 3 capsules t. i. d.) have been recommended. Ichthyol, by the way, comes also into external application, as does thiosinamine plaster. *Hebra* has introduced thiosinamine into the therapy also in the form of injections (every second day 0.5 cc of a 15 per cent. alcoholic solution). These injections are very painful; better tolerated is the nearly related fibrolysin, which *Curschmann* has warmly recommended upon the ground of favorable experiences. Also courses of iodide appear to me often to act favorably. For the sclerodermic pains I recommend the following mixture, which must be shaken up:

℞	Sodii iodid,	
	Lactophenin .....āā	5.0 (gr. 75)
	Codein phosph.	0.2 (gr. 3)
	Spirit vini	20.0 (℥ 5)
	Infus. radic. valerian (10 per cent.) .....ad	150.0 (℥ 5)

M. S. Three tablespoonfuls a day until the pains are relieved, then one or two as needed. Shake before using.

F. Neurotic Dropsy

Exudations of serum occurring from angio- or tropho-neurotic cause we meet with in two different forms, on the one hand as neurotic œdema of the skin, on the other as neurotic dropsy of the joints.

1. *Circumscribed Œdema of the Skin* (*Quincke*).—This affection is relatively rare and occurs in young individuals of nervous predisposition. (Predominance in the male sex is plainly evident.) Partly in connection with exciting emotions, trauma, exposure to cold, partly without recognizable external cause, sometimes hereditarily, circumscribed swellings of the skin which have a doughy feeling and retain the impression of the finger, occur in attacks. Their color is pale or, on the contrary, somewhat reddened, in size they vary from that of a dollar to that of the palm of the hand. Multiplicity of these usually quite indolent, but sometimes itching urticaria-like disease foci, is frequent. They can occur at any point on the surface of the skin, also may extend to the mucous membrane of the mouth, the pharynx and the conjunctiva. The seat of predilection is the face, after this, the backs of the hands. The appearance of the œdema is sometimes accompanied by vomiting and diarrhœa which has been considered as an expression of neurotic-œdematous changes in the intestinal tract; in a case of my observation (which occurred in a previously healthy girl after the railroad accident at Müllheim in Baden) the attacks were at the start accompanied by bloody diarrhœa. *Joseph* has observed hemoglobinuria. After a few hours the circumscribed œdema disappears again, but has a great tendency to recur. Periodic (also menstrual) appearance is sometimes observed. Many patients suffer during their whole lives from this, as a rule, harmless affection, which can only be dangerous when it is localized at the entrance to the larynx. Still, permanent recoveries are not entirely rare. There are certainly close nosological relations between *Quincke's* œdema and chronic recurrent giant urticaria. Many patients suffer simultaneously from *Quincke's* œdema and urticaria; in one case of *Bircher's* an urticarial eruption regularly preceded the attack of œdema and indeed, so that the two affections always appeared at different locations on the skin. Also combination with asthma and migraine attacks occurs. The pathogenesis of the disease is obscure; perhaps it is the result of a regional venous spasm. *Hunziker* as physician to the Basle house of correction, has observed a regular guest of international penal institutions, who (in order to avoid punishment) could produce at will circumscribed œdema of the face, but who would not disclose his method of procedure; *Hunziker* suspected that it was by manual compression of a venous trunk. On the other hand *Stähelin* has shown that gastro-intestinal autointoxication may play a rôle as exciting cause for the appearance of *Quincke's* œdema, as is so often the case in urticaria.

Still more obscure is the pathogenesis of the so-called "chronic form of neurotic œdema." By this is understood hydropic swellings of the skin, usually located on the leg or on the forearm, which appear without any apparent cause, also are observed as a family complaint ("trophœdème familial" of *Meige*) and last for years or for life, without any organic (renal or cardio-

vascular) changes being present. You are warned against confusing traumatic forms with the so-called "hard traumatic œdema." This last, when it appears upon the backs of the hands, is often the result of fraudulent manipulations of skilled simulators, as *Secrétan* and *Haegler* have recognized.

Therapeutically, regular saline purgation, bland and lacto-vegetarian diet, daily cold spongings, rain douches or river baths, are to be advised. Of medications I would recommend long-continued administration of salicylate of quinine (0.25 twice a day) to be tried in every case. Threatened obstruction of the entrance of the larynx, danger of suffocation has on various occasions rendered tracheotomy necessary. After this the application of a permanent tracheotomy tube is recommended.

2. INTERMITTENT DROPSY OF THE JOINTS

Still more rare than *Quincke's* œdema is intermitting dropsy of the joints (*Moore*). This is a periodic swelling of the joints, sometimes recurring with striking chronological regularity, which usually affects one, somewhat more rarely, both knee-joints. In one of my patients the articular dropsy occurred, for example, every 13 days. The patient was, however, during these intervals free, but in them he suffered from cyclic melancholia (also in cases of *Reisinger* and *Morris*, there was a 13-day rhythm). Intermittent dropsy of the joints, apart from a troublesome feeling of tension in the joints, runs its course without important subjective symptoms.

Still, sometimes disagreeable accompanying symptoms have been mentioned (slight fever, palpitation of the heart, vertigo, vomiting, hyperidrosis, polyuria, diarrhœas, migraine, etc.). After from 1 to 3 days the swelling disappears again.

The treatment coincides with that of *Quincke's* œdema; also we can repeat the little that we have said about the etiology of this last here.* A difference consists in the fact that neurotic joint dropsy attacks by preference the female sex, from which it can be concluded that physical overexertion plays no special etiological rôle. The prognosis is worse than in circumscribed œdema of the skin, permanent recovery still rarer.

G. Erythromelalgia

Erythromelalgia, which was first described by *Weir Mitchell*, represents the type of a vaso-dilator neurosis of the extremities. Still, I have seen the disease in a case running a favorable course and of otherwise typical character, appear in the face, which must be denominated "erythroprosopalgia." The very rare disease occurs almost exclusively in grown persons and manifests a certain predilection for the male sex. Those affected are generally persons of neuropathic heredity and as exciting causes, exposure to cold and overexertion have been accused. The upper extremities are much more rarely attacked than the lower, in which, in typical cases, the "erythromelalgic attacks" first manifest themselves by severe neuralgiform pains. Soon there fol-

* No surgical treatment. It cannot aid, but will injure.

lows an intense reddening of the toes, of the feet, sometimes also of a part of the leg, in rare cases indeed, of the whole lower extremity; this reddening, which is usually accompanied by swelling, is sharply separated from the normal parts of the skin. With the appearance of this local dilatation of the vessels, the pain usually somewhat diminishes in intensity. The affected parts of the skin feel hot and sometimes sweat profusely; they are the seat of a decided hyperesthesia. The erythromelalgic attack can last hours, days, or even weeks; in the last case the flaming redness more and more gives way to a cyanotic coloration and the local hyperthermia subsides. In many, perhaps most cases of erythromelalgia, between the individual attacks the integument of the affected parts does not completely return to normal, but there remains a certain degree of vaso-dilatation along with different trophic disturbances which persist interparoxysmally; this is called "chronic erythromelalgia." These chronic cases are prognostically very unfavorable, while true paroxysmal forms can recover. Erythromelalgia, indeed, does not constitute a danger to life. The differential diagnosis from erysipelas, acute gout, inflammatory flat-foot, etc., is often quite difficult. As erythromelia is denominated a painless, vaso-dilator neurosis, which has been observed on the extensor side of the limbs. As treatment during the attack, elevation of the affected extremity with cold compresses, is to be recommended (while a hanging position and warmth considerably increase the distress). Further, naturally, the use of different anti-neuralgics. Electro-therapy is of little or no use. In hopeless cases nerve resection, and, indeed, amputation, have been carried out.

H. Facial Hemi-atrophy and Hemi-hypertrophy

Both these peculiar trophic neuroses, especially the second mentioned, are very rare, and for this reason, particularly, however, on account of their therapeutic intractability, are practically without importance. To complete our discussion though, we will sketch them in their outlines.

Progressive facial hemi-atrophy, or *Romberg's* disease, affects chiefly females in early life and is usually introduced by neuralgiform pains in the affected half of the face which sometimes are also present in the fully developed stage of the affection. Very gradually there develops a strictly unilateral atrophy of the skin and of the subcutaneous connective and fatty tissue, often also of the bones of the face and of a few muscles (the temporals, masseters, tongue muscles). One-half of the face hence sinks in, in a striking manner. As rarities, cases have been described in which the atrophy has extended also to the neck, the thorax and the arm. In the majority of cases the circumscribed atrophy of the face affects the left side. Sensibility remains unaffected in the atrophic region; on the other hand, in it vitiligo, abnormal pigmentation, falling out of the hair, decolorization of the hair, etc., are occasionally observed. Therapeutically, the disease is not to be influenced; it can, however, spontaneously come to a standstill before the deformity has reached a very high degree. In these last cases cosmetic paraffin prosthesis by *Gersuny's* method has sometimes given good results. Pathogenetically *Romberg's* disease is one of the most obscure of the tropho-neuroses. A near relationship to

scleroderma is the more probable, since combinations of both pathological conditions occur.

The contrary condition to facial hemi-atrophy, facial hemi-hypertrophy was first described by *Friedreich* in the year 1862. It is such a rare disease that the last reviewer of the subject, *Wanner*, up to 1908, could find only 29 cases in the literature. It consists in a considerable increase in volume of the soft parts, sometimes also of the bones, on one side of the face, including the forehead and parietal region; the tongue, the ear, the tonsil can take part in the hypertrophy. Anomalies of pigmentation, vasomotor disturbances, unilateral exophthalmus, and other things can complicate the picture. Men and women are found with equal frequency among those affected. Contrary to facial hemi-atrophy, the disease most frequently affects the right side. Neuralgiform symptoms are rarer than in *Romberg's* disease.

I. Herpes Zoster

Herpes zoster ("Gurtelrose," "Zona" of the French authors) is a typical vesicular eruption which in its topographical distribution appears connected

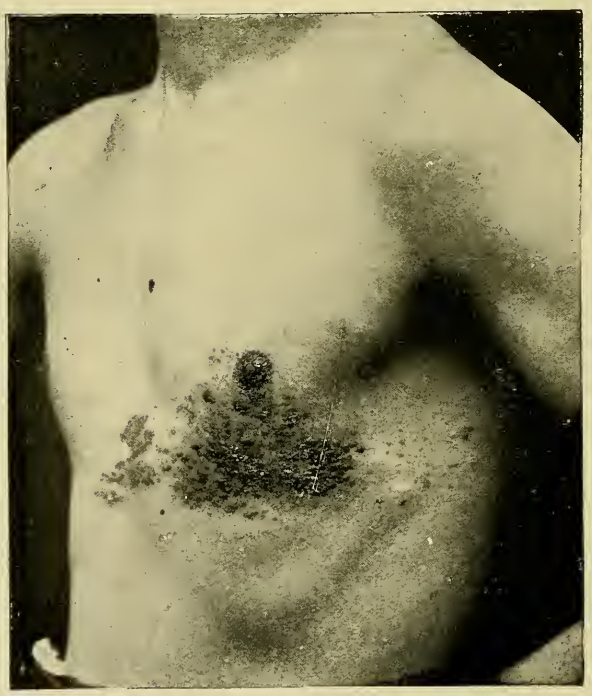


Fig. 107.
Herpes Zoster of the Trunk.

with the segmentary areas, that is, with the regions of distribution of the posterior spinal roots or their cranial homologues. Fig. 107 represents a herpes zoster on the trunk, Fig. 108, one on the face. In the last case I beg you to note that the herpetic area does not correspond exactly with the distribution

of a single branch of the trigeminus, but surrounds the nose more or less concentrically and, hence affects the region of all three divisions. It is indeed, a radicular, not a peripheral-nervous tropho-neurosis. *Bärensprung's* investigations, which have been confirmed by numerous later authors, have shown that the seat of the disease is to be sought in the spinal ganglia, respectively in the homologous structures of the sensory brain nerves (particularly in the *Gasserian* ganglion). It may be an acute inflammation of these (according to *Head* and

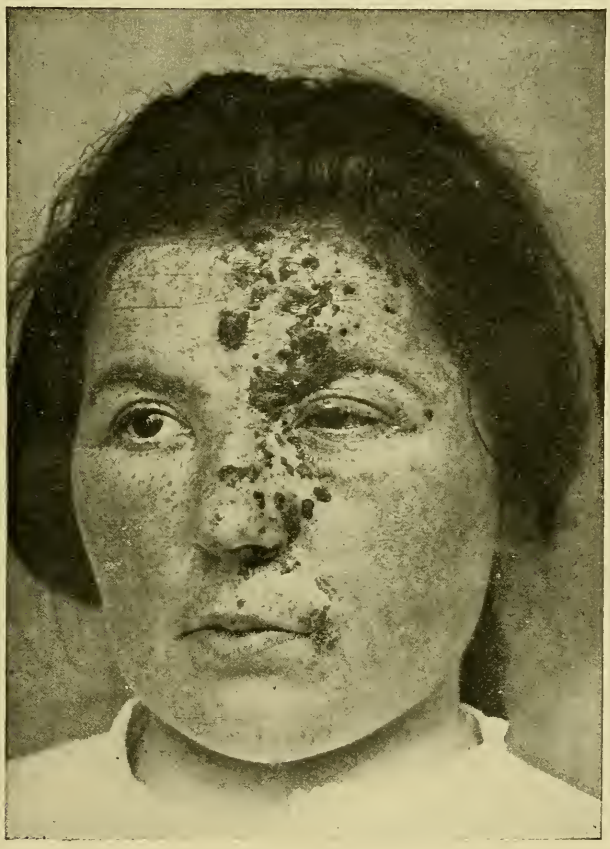


Fig. 108.

Herpes Zoster of the Face (with *Horner's* Symptom-Complex).

Campbell, usually so), but also may be due to tumors, trauma, etc. The physio-pathological basis of the vesicular eruption is indeed obscure to us in spite of this topical diagnostic recognition. In the mechanism concerned, however, connection with the sympathetic comes into question. Of importance in this relation is, for example, the condition that in the patient shown in Fig. 108 a myosis and enophthalmus and the narrowing of the palpebral fissure occurred on the same side as the herpes eruption. Etiologically in the so-called secondary cases, as already said, there have been injuries to the ganglia (fracture of the vertebral column, etc.) or tumors (carcinoma metastases, for example); further, poisoning with carbonic oxide and arsenic should be men-

tioned. With regard to the last, there has been a controversy as to whether the herpes occasionally observed after salvarsan injections is to be considered as a toxic manifestation or as the so-called "Neurorecidiv" of syphilis. In primary or idiopathic herpes zoster, which we will now exclusively consider, there appears to be an acute infectious disease *sui generis*, in which exposure to cold and similar things can only be considered as exciting causes; the actual cause, however, is indeed still unknown to us. For this speak the above-mentioned pathological findings of *Head* and *Campbell*, further the facts that the disease appears to leave behind it an immunity, runs its course with fever, swelling of the glands, and pleocytosis of the cerebrospinal fluid, and last, but not least, its occasionally observed appearance in epidemics.

In typical cases of idiopathic zoster a moderate rise of temperature (to about 39° C. (102.2° F.) with general discomfort, disturbances of digestion, etc., precedes the eruption. With this there is usually very severe burning and boring, sometimes continued, sometimes exacerbating pains in the affected radicular area; these pains can appear simultaneously with the vesicular eruption, or indeed, only after its occurrence. They usually get worse toward evening. The herpes eruption itself is usually introduced by the appearance of red spots upon a normal appearing ground. Soon, however, the epidermis over these spots raises itself to serous vesicles whose contents gradually become cloudy and purulent. If the sensibility of the skin in the neighborhood of the vesicles is tested, sometimes hypesthesia and anesthesia, more rarely hyperesthesia, is found. Sometimes atypical forms of eruption appear: bullous, hemorrhagic, gangrenous, ulcerative herpes; the last two may leave behind scars.

Regional glandular swellings are not rare, also hyperidrosis or anidrosis occur, as do also obstinate neuralgias, which may persist after the herpes has recovered, which takes usually from 2 to 3 weeks, but sometimes considerably longer. Such persistent root neuralgias cloud the prognosis of the in general entirely mild disease, especially in decrepit old people, who form a large contingent of the zoster patients. Serious results may also follow an extension of the herpes zoster to the cornea, when it may lead to ulceration and panophthalmitis. Still, this localization is, fortunately, not frequent.

Therapeutically, we limit ourselves to the administration of antineuralgics (see Lecture III, page 57), and the application of a 2 to 5 per cent. anesthesin ointment, or of a powder of talc, bismuth subnitrate and starch, equal parts. Ulcerated herpes vesicles heal best under 5 to 10 per cent. ointment of balsam of Peru. Ophthalmic herpes should be turned over to the ophthalmologist without delay.

Gentlemen: We will here break off our sketching of the most typical tropho-vasomotor syndromes. I must remark to you, however, that it is just in this class of cases that mixed and transition forms are specially frequent; for instance, not only can the different vaso-constrictor conditions be combined, but also vaso-constrictor and vaso-dilator neuroses have been observed in one and the same patient, for instance, erythromelalgia and *Raynaud's* disease.

It may be also related that many skin diseases by their topographic limitation to definite regions of innervation, symmetric distribution, etc., proclaim

themselves of tropho-neurotic or vaso-neurotic nature. I would recall many vascular nævi, cases of vitiligo, partial albinism, pigmented moles, etc. Also, symmetric lipomatosis or *Madelung's* disease may well belong here (see Fig. 109). As a variety of this last is to be considered the adipositas dolorosa or *Dercum's* disease, which also shows its connection with nervous diseases, particularly by the element of pain. It affects mainly women (often alcoholics)

Fig. 109.
Madelung's Disease.

who, also in general very corpulent, present tumor-like lumps of fat in the subcutaneous tissue of the arm, leg and trunk, but with regular escape of the hands and feet. These lumps of fat are sensitive to pressure, as are also the regional nerve trunks. Also, spontaneous pains may arise in these lipomata. Anesthesia or hyperesthesia of the affected skin zones are occasionally noted. The attempt of a few authors to prove the relationship between *Dercum's* disease and myxœdema must be considered as a failure. Finally, progressive lipodystrophy in which the upper half of the body emaciates to complete loss of the panniculis adiposus while the lower half, on the contrary, shows rather an increased deposition of fat, is to be mentioned.

LECTURE XXVI

Epilepsy

GENTLEMEN: The disease to which to-day's lecture is devoted, must be considered as the first nervous disease of which literary information has been transmitted to us. That it since most remote times has laid claim to the phantasy of physicians and laymen to a high degree and that it, as no other disease, has had the reputation of being of supernatural origin, we need not wonder. Indeed, the "falling sickness" which causes a man apparently in perfect health, to fall unconscious and to go into convulsions, must make upon the masses of the people a particularly alarming, indeed, uncanny impression. Even the "scientific" name epilepsy, which we have inherited from ancient times, indicates nothing but "being seized" or "possessed." You know, indeed, that the disease was considered as a punishment inflicted by the gods, hence the name customary to the Hippocratic body, "ἱερὴ νόσος"=sacred disease. "Morbus sacer" is still occasionally used to-day. Just as respectable is also the age of the designation popular in France, "mal comitial" or "crises comitiales." Namely in ancient Rome if during the Comitia some one had an epileptic fit, on account of this "morbus comitialis," the assembly had to be closed immediately.

We speak to-day expressly of a genuine, essential, or idiopathic epilepsy, since we very properly assign to the epileptiform symptoms as they have been observed in the most varied pathological conditions of the nerve centers—for example, multiple sclerosis, progressive paralysis, cerebral syphilis, cerebral abscess, cerebral tumor, hydrocephalus, infantile cerebral palsy, meningitis, saturnism, alcoholism, morphinism, etc.—a special position as "symptomatic epilepsies." We have, indeed, in the course of these lectures, become acquainted with most of these symptomatic epileptic phenomena and in our further remarks will only consider true epilepsy. This may be defined in the following manner: A chronic, very frequently progressive, disease of the brain, which manifests itself, as a rule, by more or less frequent convulsive attacks accompanied by loss of consciousness (sometimes, however, by paroxysmal disturbances of consciousness without convulsions), and which not rarely can lead to psychical alteration. It appears sometimes even in early childhood, usually, however, between puberty and the 20th year of life. Numerous, however, are the cases of the last category in which we learn in the history that the affected patients as infants have suffered from "tooth spasms," infantile eclampsia, and one cannot escape the impression that these convulsive paroxysms, originally considered as harmless, were really the precursors of epilepsy. The beginning of true epilepsy in the third and fourth decade is less

frequent, in still later years quite rare. An epileptic attack occurring for the first time beyond the 40th year, in the majority of cases is of the symptomatic form (progressive paralysis, brain tumor, brain syphilis, etc.). Both sexes are attacked with about equal frequency.

Symptomatology

We will begin our sketch of the epileptic disease picture with a description of the typical convulsive attack, then will subject to clinical consideration, paroxysms of other sorts and, finally, the permanent interparoxysmal anomalies of epilepsy.

1. The Severe Epileptic Convulsive Attack ("Haut mal," Epilepsia Major, Convulsiva)

The epileptic convulsion can seize the patient without any warning, "like lightning out of a clear sky"; still, in many cases, it begins with prodromal symptoms which we denominate the epileptic "aura" and whose different types we will now enumerate. It occurs not at all infrequently that even before the aura (which is always of very short duration, seconds or minutes) different disturbances of the general condition announce the impending attack. So, many patients complain for several hours or even days previously, of sense of pressure in the head, malaise, abnormal irritability, sexual erethism, sleeplessness, urticaria, diarrhœa, frightful dreams, abnormal appetite, and other anomalies.

The epileptic aura can be sensible, sensory, motor, vasomotor or psychic. The first is the most frequent. The feeling of a cold or warm breeze which passes over the body surface of the patient has given occasion for the choice of the term "aura" (air) which dates back to the time of *Galen*. Further, paresthesias of the ends of the extremities, of the tongue, of the scalp have been described, also a feeling of constriction in the throat, stabbing pain in the head, etc. The sensory aura can affect the various sensory organs, far most frequently it is an optic aura. The patient suddenly perceives dark spots, scotomata, in the visual field, or, on the contrary, flames, lightning, sparks, balls of fire, sees everything in one definite color (almost always red) the so-called "colored vision," or in altered size (macropsia and micropsia), etc. The auditory aura is usually characterized by a roaring, cracking, or hissing in one or both ears, the olfactory, by a penetrating smell, the gustatory by a bitter or astringent taste. In the motor aura there occur, for example, twitching in the facial distribution, choreiform movements of the hands, singultus, yawning spasms, motor aphasic symptoms, purposeless running forward ("aura cursatoria") etc.

As vasomotor aural phenomena, paling, reddening, outbreak of sweat and palpitations occur. Finally, among other things there are designated as "psychic aura," the sudden causeless appearance of a definite memory, often with the plainness of a hallucination, or an abrupt change of mood, for instance, an outbreak of mirth, or feelings of anxiety and imperative ideas. Often the

aura is a complex one made up of psychic, sensory, sensible and motor components in manifold combination, but it usually, in one and the same patient, repeats itself in a thoroughly stereotyped manner, so that he is entirely aware of the threatening attack. Indeed, the aura is so short that in but very few cases the epileptic finds time to take adequate precautions, for example, to prevent falling by lying down.

Whether an aura precedes it or not, the epileptic attack proper begins in the most abrupt and "brutal" manner. In typical cases the patient emits a penetrating cry and falls unconscious, while wounds from striking the head or the limbs or from falling down stairs, etc., upon which the patient is at the time, are nothing rare. Biting the tongue occurs from the sudden bringing together of the teeth, and in one of my cases this led to almost complete hemisection of that organ. Now a general tonic spasm of the muscles appears, the jaws are pressed together, the fists clenched, arms and legs are rigidly extended, the eyes are directed fixedly forward or are drawn up under the fast-closed lids, the pupils are usually narrowed and reactionless; also the inspiratory muscles usually take on a tonic contraction and respiration is suspended. Hence, the sudden pallor of the face, which is usually observed in epileptics at the beginning of the attack passes rather quickly over into a livid, indeed cyanotic color. This "tonic stage" of the severe epileptic attack is of very short duration (about one-half minute) and passes over into the "clonic stage," which usually lasts from several minutes to one-quarter hour. The limbs undergo violent contractions, the body writhes, the eyes roll wildly, the countenance is drawn up into frightful grimaces, the head is thrown in all directions. Also in this stage biting the tongue and other wounds can be produced. There occurs also an excessive secretion of saliva which appears upon the lips as a blood-tinged foam. The pupils are still rigid, but now, however, maximally dilated. The respiration is stertorous and can be heard at a distance; outbreak of sweat and discharge of urine are frequent, rarer, discharge of fæces and ejaculation. The face appears dark red, swollen; the veins of the neck are congested; sometimes there are small extravasations of blood into the conjunctiva, on the neck, on the chest, behind the ears, etc. Now follows, with gradual cessation of the convulsions, passage into a quiet coma, in which the stertorous breathing little by little changes into the normal type of respiration and the cyanosis again gives place to pallor. This last stage of the attack can be of very short duration, but may last from one to two hours; then the patient slowly regains his senses. As a rule, for hours he feels weak, exhausted, depressed, suffers from severe headache, sometimes also from nausea and vomiting. There is complete amnesia for the attack; indeed, the memory of the aura can at times be absent. Sometimes the temperature is somewhat elevated, often there is slight albuminuria which soon disappears. Very rarely transitory paralyses are to be observed in this post-paroxysmal stage, most frequently convergent strabismus. After one or two days they usually disappear. Also there are occasionally temporary pareses of one arm or leg, or of the two extremities on the same side: on two occasions I could elicit *Babinski's* toe phenomenon (once also, ankle clonus). Further, post-paroxysmal transitory aphasia as well as fleeting sensory paralyses (post epileptic

deafness and blindness) need to be mentioned. *Muskens* has found after epileptic attacks cutaneous hyperesthesias of segmental type.

That a true epileptic attack occurs unilaterally ("genuine hemi-epilepsy"), that is, like the *Jacksonian* attacks sketched in Lecture XIX (page 293), is exceedingly rare. In order to include such cases within the limits of the morbus sacer, in spite of the atypical nature of the convulsive attack, one must be able to base his opinion upon a very accurate observation of the course of the disease, the absence of symptoms of a cerebral disease focus and upon a careful study of the history. The occurrence even of a unilateral status epilepticus has been pointed out by *Müller.*

As to the frequency of fits in epileptics, no general statements can be made. They may occur very rarely (once or twice a year), or very frequently (many times a day), and between these two extremes there lie all possible degrees of frequency. In women the menstrual type, in which the paroxyms always fall in the menstrual or in the pre- or post-menstrual period, are relatively frequent. When in the most severe cases of epilepsy, one attack immediately follows another without the patient regaining consciousness between times, this constitutes the so-called "status epilepticus," a form of seizure which may last for hours and is always very dangerous to life. The body temperature usually increases with each successive attack, so that finally hyperpyretic temperatures—over 41° C. (106° F.) can be reached. Also the frequency of the pulse increases in a threatening manner and the action of the heart becomes weaker. When status epilepticus is going to terminate favorably there is gradually a longer interval between the attacks, the convulsions decrease in severity, the pupils begin to react again, the temperature falls, and finally, the patient regains consciousness. In cases terminating fatally on the other hand, there is added to the convulsions a condition of profound collapse in which the pupils remain mydriatic and rigid, all reflexes disappear, breathing becomes continually more superficial, the pulse indeed, slower again but steadily smaller, and finally cessation of heart action and respiration occurs.

In the clinical estimation of epileptic attacks, besides their frequency, the time of day at which the paroxysms occurs is important. There are, besides patients who are only attacked by their convulsions while awake or indifferently by day or by night, also those in whom exclusively nocturnal attacks occur. Such attacks can be easily overlooked when the patient sleeps alone; wetting the bed in grown people, contusions from striking the head against the wall or on the bedstead, also awakening with a feeling of being exhausted and very weak, should always arouse a suspicion of nocturnal epilepsy. The differential diagnosis between the epileptic and the hysterical seizure we will defer until Lecture XXIX.

2. Minor Epileptic Attacks ("Petit mal," Epilepsia Minor, Non-Convulsiva)

At the side of the classical epileptic attack just sketched, to whose most striking criteria the convulsions belong, there are now to be placed other forms of seizure which with equal certainty declare themselves manifestations of the morbus sacer, in which, however, the convulsive factor is either entirely absent or only indicated. Common to all, is the paroxysmally appearing, more or less marked clouding of consciousness, which can increase to complete loss of consciousness. In general, however, their symptomatology varies within rather wide limits. We combine them under the name of "minor" or "non-convulsive epilepsy" and place them as "petit mal" in opposition to "haut mal." Many epileptics suffer from both major and minor seizures, while in others all the paroxysms belong to the same type. Let us now enumerate the most important varieties of non-convulsive epilepsy, proceeding from the more frequent to the less frequent.

A. THE "MOMENTARY ABSENCE"

This peculiar phenomenon may be defined briefly as a sudden and transitory elimination of the higher psychical mechanisms. In the simplest cases the patient ceases suddenly the work which is occupying him, or in the middle of a sentence which he is just speaking, remains quiet a few seconds with fixed look ("as if absent"), then resumes again his occupation or his conversation as if nothing special had happened, and with complete amnesia for the interruption which has occurred, exactly at the point where the "absence" began. Usually, however, this time is filled out with peculiar automatisms varying exceedingly from case to case. The patient, for example, emits grunting or snoring noises, makes movements of chewing, a few twitching movements of the face or of the extremities, claps his hands, bends himself forward as in "salaam spasm" (see above, page 69), speaks senseless words ("epilepsie marmottante"), etc. Rarely the automatisms assume a more complicated form, as was the case in the epileptic President of the Court described by *Trousseau*, who in the middle of a session, rose, muttered something unintelligible, went from the Tribunal into the robing-room, there urinated on the floor, then returned to his post and took up again the proceedings where he had broken off. Occasionally during the absence, the occupation is not really interrupted, but is carried on in a senseless, purely mechanical manner, so, one of my patients, a dactylographer, became aware of her petit mal attacks, by finding that in the middle of the text which she was copying, there was a half line of letters placed together indiscriminately.

B. EPILEPTIC VERTIGO

In epileptic vertigo a feeling of turning or of loss of equilibrium suddenly comes over the patient; he totters, but, as a rule, before falling, regains his equilibrium. These attacks are usually accompanied by pallor, sometimes also

by the escape of a few drops of urine; consciousness is practically never completely lost, although it is markedly clouded (a transitory feeling of confusion, of defective psychic orientation). Epileptic vertigo is hence considered as an "attenuated attack" of the morbus sacer.

C. EPILEPTIC SYNCOPE

Epileptic syncope can be considered as a higher degree of intensity of epileptic vertigo, since it begins, as a rule, with the feeling of apparent movement, tottering, etc., still, here the patient has loss of consciousness and actually falls, but regains consciousness in a short time. Spasms are absent or only slight. Such attacks have been also denominated "apoplectiform."

D. THE "NARCOLEPTIC" ATTACK

In this there is sudden falling asleep in the daytime and the patient cannot be awakened by shaking, etc. After spontaneous awakening the patient often shows a delirious condition; in any case, however, he has no consciousness of having been asleep.

E. THE "PROCURSIVE EPILEPTIC" ATTACK

In the paroxysms which have been denominated "procursive epilepsy" (*Bourneville* and *Ladame*) there is a sudden running forward in which the patient either avoids objects which stand in his way or forcibly pushes them aside. Here also there is lack of memory for the attack, sometimes also for the period immediately preceding it. A still more rare variety is the "retropulsive epilepsy" of *Launois*, in which the patient in his attacks runs backward.

3. Epileptic Equivalents

As equivalents in the broad sense we denominate manifold transitory phenomena, which in the concrete case we consider as of epileptic nature, since they appear to occur in epileptic individuals, or members of epileptic families, in the place of the major or minor attacks proper, run their course usually with more or less clouded consciousness, are generally followed by a condition of decided prostration, and often react in a striking way to bromide treatment (see below). For example, outbreaks of sweat, trigeminus neuralgia, profuse diarrhœas, *Quincke's* œdema (see Lecture XXV), sialorrhœa, vomiting, attacks of migraine, stenocardic attacks, paroxysmal tachycardia, spasm of the glottis, etc. Much more important, however, than these sensory, vasomotor and visceral equivalents (whose inclusion within the limits of epilepsy is through quite a wide extension of hypothesis), are the psychical, for which in general we reserve the term "equivalent" in the narrower sense. *Falret* has divided them according to the intensity of their symptoms into "intellectual grand mal" and "intellectual petit mal," a distinction which is too arbitrary to have been generally received.

There occur, for example, in attacks, hallucinations with conditions of

great anxiety, maniacal deliria, which under the influence of delusions and hallucinations can increase to frenzy and destructive rage, particularly, however, impulsive acts of sometimes quite complicated content. In these different manifestations the patients are in a condition of abnormal, dreamy consciousness ("dämmerzustand" "état second") for which after the paroxysm has passed off, memory is by no means always obliterated (forensically important), almost always, however, markedly cloudy. The impulsive equivalents of epileptics as well as epileptic mania, are responsible for many misdemeanors and crimes. Here belong cases of incendiarism, motiveless homicides, thefts in part miscellaneous, in part confined to certain things, often openly carried out, public exposure of the genitals (exhibitionism), etc. Upon an epileptic basis, dipsomanic attacks may also rest,—sudden impulsion to senseless drinking themselves drunk, in previously abstemious persons. These equivalents are, on the average, of considerably longer duration than epileptic attacks proper; the abnormal condition can last for hours, even days. The last is the rule in the interesting phenomenon which has been denominated "ambulatory automatism," "poriamania," or epileptic "wandertrieb." The patient suddenly leaves without motive his place of residence, and wanders blindly about, during which time as far as buying tickets, paying his way, etc., is concerned, he may conduct himself in an entirely unobtrusive and normal manner. When the equivalent passes away, either there is no recollection of the clouding of consciousness, or the patient has the feeling of waking from a dream whose content he but dimly remembers. A patient of *Legrand du Saulle* gained fame from having been overcome by his poriomania in Havre, and having to his boundless astonishment come to himself again in Bombay.

The epileptic clouding of consciousness ("dämmerzustand") as it serves as the basis of different psychical equivalents, can appear as the forerunner of a convulsive attack, or in connection with such an attack. In the cases in which it does not occur alone in the place of a seizure, we speak of pre-epileptic or post-epileptic clouding of consciousness (delirium), ("preparoxysmal" and "postparoxysmal" or "preconvulsive" and "postconvulsive" would be more correct).

4. The Interparoxysmal Anomalies

When we, as is usually the case in the ambulatory treatment, examine epileptics outside of their attacks, we find occasionally an entirely normal status and must base our estimation of the case entirely upon the history. Nevertheless, a thorough investigation in the majority of cases discloses definite anomalies either in the bodily or in the mental sphere.

Under the first we may mention at the start the so-called stigmata of degeneration which many epileptics (as many constitutional neuropaths in general) present. Most frequent are anomalies of the skull of the most varied sort, for example, marked asymmetry, abnormal shape of the palate (Gothic palate), abnormalities of the teeth, microcephaly, etc. Further, malformations of the ear (adhesion of the lobe, faun-like pointed or protruding, the so-called "jug handle" ears, etc.), and congenital anomalies of the eye (colo-

boma iridis, marked astigmatism, etc.). Much rarer are signs of degeneration on the trunk and on the extremities, as, for example, funnel chest, syndactylism, melanodermia. In the second place I would relate the muscular anomalies, above everything the muscular weakness appearing in contrast to the good morphological development of the muscles, which relatively often appears upon one side. Less frequent are muscle disturbances of atrophic character to which *Onufrowicz* has called attention. So, the occasionally observed scapula alata is to be referred to a paresis of the serratus magnus which may be accompanied by that of the trapezius, the rhomboids and the levator scapulæ. That infantile cerebral palsy is often combined with epilepsy we have already emphasized in Lecture XXII. Also myoclonus epilepsy has already been considered (see page 73). Almost all epileptics show an unusual activity of the tendon reflexes, many are left-handed or ambidextrous.

We find further, occasionally, general reduction of the superficial sensibility as well as the most varied speech defects. Finally, the scars of tongue-bites and other wounds occurring in the attacks should be considered.

Practically much more important, are, however, the (often progressively appearing) permanent psychic and character alterations of epileptics. We will leave out of consideration here those cases in which the combination of congenital idiocy with epilepsy is present (see Lecture XXII, page 336) and confine ourselves to the consideration of such anomalies which must be interpreted as the result of epilepsy, not as a pathological condition coördinate with this last. Many epileptics are excessively quick-tempered and inclined to violence and sudden changes of mood, malignant, cruel, sexually perverse, of brutal egotism. In severe cases, on the other hand, there appears, little by little, a continually plainer reduction of the judgment-forming capacity which increases to actual dementia. In the severest cases the dementia may be complete with loss of all mental capacity and even of speech, uncleanness, refusal of food, etc. "Epileptic paranoia," in which ideas of unseen influence and delusions of persecution, on account of the violence of character of epileptics, may be particularly dangerous, is rare. To these psychical disturbances peculiar to epilepsy are opposed the numerous milder cases in which character and intelligence remain normal during life. It stands beyond question that many eminent personages have been epileptics. I myself know undoubted cases of this sort from my own practice. It must, indeed, be said that in the so-called "pathographies" with retrospective diagnosis, epilepsy has often been diagnosed with unbelievable carelessness, for example, in relation to Napoleon I. That he at the end of the battle of Wagram at a moment when the pursuit of the enemy still imposed upon him important duties, suddenly lay down on the floor and fell into a deep sleep, need not be interpreted as narcolepsy, when one remembers that for over 48 hours in spite of the greatest mental tension and bodily exertion, he had not slept one second.

Etiology and Pathological Anatomy

Gentlemen: That heredity plays a particularly great rôle in the pathogenesis of the disease whose symptomatology I have now brought before you,

there can be no doubt. *Binswanger* has, for example, been able to recognize mental and nervous diseases among the ascendants of from 35 to 40 per cent. of his epileptics, and the heredity can be equally like or unlike. Just as generally recognized is the frequency of epilepsy among the descendants of drinkers, which we can interpret in the sense of injury to the germ ("blastophthoria") through parental alcoholism (compare Lecture VIII, page 127). Further, numerous observations speak for the fact that the changes in the brain which form the basis for genuine epilepsy can be produced, or at least favored, by exogenic factors. This applies particularly in childhood; that "late epilepsy" is so rare depends upon the fact that after its development is completed, the brain has much less tendency to react to pathological alterations by paroxysmal discharges. Among the acute infectious diseases in direct sequence to which, or during whose course the outbreak of epilepsy has often been observed, scarlatina takes readily the first place; next to it are to be mentioned particularly small-pox, influenza, measles and typhoid fever. Among the chronic infectious diseases hereditary syphilis occupies the first place. For late epilepsy, acquired syphilis comes into consideration. Here, however, certain intoxications appear very frequently in the etiology: Saturnism, cocainism, alcoholism (when we speak of the latter we must make it clear to ourselves whether or not it is a dipsomanic form which is to be considered as a symptom of epilepsy, not its cause). Circulatory disturbances also may play a rôle in the etiology of late epilepsy. Often its beginning coincides with the development of severe arteriosclerosis; here, also, belong the not rare observations of epileptic attacks in diseases of the heart and aorta. Finally, it is to be considered as proved that injuries to the head can lead to genuine epilepsy (not only to the *Jacksonian* form depending upon a circumscribed lesion).

A special position in the discussion of epilepsy is assumed by the so-called "reflex epilepsy," whose origin still remains a riddle. Sometimes in previously healthy persons after wounds of the extremities, of the trunk, or of the head, there appear typical epileptic attacks which are introduced by a sensory aura starting in the scar left behind by the trauma. Sometimes indeed, an attack may be produced by pressure upon such a scar (which then is denominated an "epileptogenic zone"). Occasionally diseases of the uterus, of the nose and its accessory cavities, etc., can act in a similar manner, while the convulsions of children having intestinal parasites do not usually appear to be of epileptic character. Further, many cases of hysterical attacks have been incorrectly considered as reflex epilepsy. Nevertheless, there remains a respectable number of these cases which have been regarded as truly epileptic by recognized authorities, as for example, *Oppenheim*. On account of the great rarity of such true reflex epilepsy in spite of the frequency of wounds, we must assume that in the individuals affected, the latent predisposition to epilepsy is anatomically predetermined and the irritation from the scar, etc., only furnishes the exciting cause. Excision of the scar or removal of any other pathological condition frequently does away with the reflex epileptic attacks.

Let us now pass on to the pathological anatomy of the morbus sacer. You have remarked that I have repeatedly spoken of a brain disease, of anatom-

ical alterations, etc. There is for me to-day, as for the great majority of neurologists, no longer the slightest doubt that material alterations of the cerebral cortex lie at the basis of epilepsy, though they are here and there too little intense to be discovered by our histological methods. Epilepsy is no "neurosis," and *Hippocrates* has rightly remarked that its etiology and pathology are in the brain.

Chaslin, in 1889, published the first findings, but in late years these have been extended and systematized by *Alzheimer* through finer methods. In many cases of genuine epilepsy he has shown a "marginal gliosis" of the cerebral cortex with numerical reduction of the medullated fibres and ganglion cells; the latter, besides this, appear atrophic. The vessels are proliferated, show thickened walls, sometimes also lymph sheaths filled with mast cells. In patients who have died in attacks (or in status epilepticus) there are, besides severe acute changes in the ganglion cells and their axis cylinders, also ameboid glia cells loaded with products of disintegration. The last mentioned anomalies indeed are connected with the epileptic attacks, while the marginal gliosis and its accompanying changes can be held responsible for the interparoxysmal clinical pictures, particularly for the dementia and change of character. On the other hand, *Alzheimer* attributes no great importance to the sclerosis of the cornu Ammonis present in 50 to 60 per cent. of these cases, considers it rather only as an occurrence associated with the epileptic degeneration.

What, however, causes the intermittent, sometimes even explosive, onset of convulsive or other paroxysmal phenomena? Where peripheral irritative conditions cannot come into question as in reflex epilepsy, it is natural to consider autotoxic factors and many authors have instituted investigations in this direction, into which, since they concern things still quite hypothetical, we cannot enter further. We need only mention as an example, the work of *Donath*, who asked himself what substances introduced into the circulation of animals are able to set off epileptic paroxysms. He has come to the conclusion that the organic bases resembling ammonia (trimethylamin, cholin, creatinin, guanidin) act as convulsants. He hence believes that in epileptics the occasional accumulation of substances of this sort in the blood irritate the morbidly constituted brain to react by epileptic attacks. That the blood of epileptics during the attack possesses toxic properties appears probable from the investigations of *Ceni* and others, without our knowing how to make much use of this pathogenetic fact.

Prognosis

Epileptic attacks usually only indirectly endanger the life of the patient, that is, in consequence of the injuries which he can sustain by falling; as already said, the status epilepticus, whose prognosis in the individual case I have already discussed, furnishes an exception to this. Though epilepsy in itself does not directly put life into question, still, patients with the severe form seldom live to be old, since they usually have, in general, a reduced resistence to all possible injurious factors. To definite recovery at most 10

to 15 per cent. of the cases are to be brought by proper treatment, still there are along with these, cases in which a very great improvement is to be obtained, so that, for example, the attacks only show themselves occasionally, at intervals of a year or more. The earlier the disease begins and the more frequent the attacks, the worse the prognosis as to recovery and also as to mental integrity. On the other hand, the occurrence of major attacks does not at all cloud the prognosis, in contradistinction to the forms manifesting themselves in minor attacks and equivalents. On the contrary, the last are much more difficult to influence therapeutically and in the long run never leave the psyche intact. Patients with "procursive epilepsy" usually become demented particularly rapidly.

Treatment

In the foreground of the treatment of epilepsy in spite of the manifold other methods of medication continually appearing (most of which fall again into oblivion), stands always the intelligent application of the alkaline bromides which we will hence discuss with a thoroughness corresponding to its importance.

In the following I will not consider the severe cases of epilepsy or those combined with psychoses or idiocy, needing asylum care, but will keep in view those suitable for ambulant treatment or management at home. Patients entering upon treatment are particularly to be impressed with the fact, that under all circumstances the cure must extend long after the cessation of attacks, also that they must not interrupt it even for a few days of their own accord; women, also that bromide medication is in no way incompatible with menstruation and pregnancy. Some person connected with the patient must assume responsibility for the strict carrying out of orders and must keep an accurate account of the number, duration and severity of the attacks.

Among the alkaline bromides, potassium bromide is undoubtedly the most efficient preparation, probably because it is more slowly excreted than sodium bromide and ammonium bromide (the last is in spite of its high content of bromine, of exceedingly low activity; strontium bromide has almost only historical interest). Unfortunately the disagreeable associated effects of bromide medication are much more frequent and intense with potassium bromide than with the other bromine preparations (bromide acne, gastro-intestinal disturbances, general prostration, etc.), which sometimes moves us to abandon the potassium for the sodium salt or for "Erlenmeyer's Mixture"—2 parts each of KBr and NaBr, 1 part of AmBr. The "injurious action of potassium upon the heart muscle" of which we still hear occasionally, need not be brought up against potassium bromide, since it is never observed in practice. To handle a case of genuine epilepsy with organic bromine preparations is an attempt which you will always regret. Next to the choice of a preparation, its dosage is of great importance. Often there is the tendency to make it too low at the start. In general, one should not begin with less than 2.0 KBr a day. Giving a full day's dose at one time (before retiring) is the rule in nocturnal epilepsy; in diurnal forms, however, or in those which do not occur at any definite time of day, you will generally do better (though not always)

if you divide the day's supply into 2 doses of 1.5 each. The patient must take one at bed-time and one at breakfast-time, or—when he tends to have attacks immediately after rising—half an hour before leaving his bed. If this dose proves insufficient we add 1.0 to 1.5 KBr at mid-day. Cases in which daily doses of from 4.0 to 5.0 show no effect, for further increase of the dose (to 6.0 or 8.0 and more) are best brought under hospital treatment or into special institutions for epileptics. Epileptics are almost always more tolerant of bromides than other individuals; particularly so are epileptic children. Since my remarks up to this time—as also the recipes which I will give you later—are adapted to adults, you may note the following figures as representing the usual doses of potassium bromide for young epileptics of slight degree:

	Single Dose.	Dose per Day.
Two first years of life.....	0.1 (gr. 1½)	0.3 (gr. 4½)
3d to 5th year.....	0.2 (gr. 3)	0.5 (gr. 7½)
6th to 11th year.....	0.5 (gr. 7½)	1.0 (gr. 15)
12th to 16th year.....	0.5 (gr. 7½)	2.0 (gr. 30)

Still, when necessary, children can bear much higher doses very well, for example, 2.5 per day in the fourth year.

After the patient has become accustomed to a plainly efficacious initial dose (for this several weeks are needed), for his further management, there is a choice between the following methods:

(*a*) The method of increasing and decreasing doses (*Charcot*). This is especially suited for cases with paroxysms appearing more or less periodically, for example, for the menstrual or pre-menstrual attacks. During the week in which attacks of the last kind are to be expected the patient receives the day's dose proved to be efficient (let us say 4.0); during the following week, 3.0 a day; the next week, 2.0; then for one week again 3.0, and finally at the critical period, again 4.0. After several months we try if 3.5 or 3.0 is not a sufficient maximum dose, and if the result is obtained, in the course of several years continually raising and lowering the dose, we endeavor to dispense with the bromide altogether.

(*b*) The method of gradual disuse. This is particularly suited for cases with irregular, entirely unaccountable attacks and those in which the seizures rapidly follow one another. The ascertained efficient dose is continued for months, then it is attempted every few weeks to reduce it about 0.5 a day. If the case gets worse we return to the next higher dose and endeavor to reduce it again some months later. Even when a gradual withdrawal is carried out without disturbance, we are very cautious about reducing the last dose and wait until no attack has occurred for at least a year. Then we proceed best after the method of *Legrand du Saulle:* For 3 months, 6 "bromide days" a week; for 3 months, 5 "bromide days"; for 3 months, 4 "bromide days," etc.

The most important adjuvant of the alkaline bromide therapy, is the salt-free or salt-poor diet after *Toulouse* and *Richet*. Removal or reduction of the chloride of sodium in the diet increases the pharmacodynamic action of the alkaline bromides so greatly, that we can almost always get along with much

smaller doses and can succeed sometimes with 3.0 to 4.0 KBr in a severe, and even 2.0 in a moderately severe epilepsy. Unfortunately there are plainly individual differences, and here and there no special result can be obtained with the *Toulouse-Richet* method. Since this last not only increases the efficacy of the bromide action, but also the danger of bromism, in practice, only the relative removal of salt comes into consideration (for example, ordinary but unsalted food with 500 grm of salted bread=2.5 NaCl). The technique of a salt-poor diet is considerably simplified by the "Sedobrol" tablets proposed by *Ulrich:* these contain 1.5 bromide of sodium and 0.1 grm chloride of sodium, with fat and extractives of vegetable albumen as a seasoning. Dissolved in hot water they give, depending upon concentration, an almost salt-free, palatable soup or a sauce very suitable for salt-free vegetable food. As to the rest of the food, a predominantly, but not exclusively lacto-vegetarian diet is to be recommended. On the other hand, strongly spiced foods are to be avoided. Alcoholic beverages are to be absolutely forbidden; coffee, tea and tobacco are to be permitted only in very small quantities. Conditions of life as quiet as possible are to be provided for.

If bromide acne appears, compresses of from 10 to 20 per cent. salt solution applied to the diseased portions of the skin act very favorably, as *Ulrich* has shown; of internal medications, *Fowler's* solution acts best. Upon the large acne nodules, pieces of mercurial plaster are placed.

The addition of sodium benzoate (*G. de la Tourette*) or Adonis vernalis (*Bechterew*) appears to increase the efficacy of the alkaline bromides; the first, perhaps, by aiding the absorption from the intestine, the last by improving the brain circulation; both drugs act as diuretics opposing brominism. For severe cases, the combination of bromide and chloral hydrate, which in evening doses of from 0.3 to 1.0 can be given for a long time without any hesitation, has proved particularly satisfactory. Valerian also aids the action of the bromide somewhat. On the other hand I would imperatively warn you against the application of "*Flechig's* Bromide-Opium Cure" outside of a hospital This method, in which the patient within 6 weeks is brought to take 0.9 extract. thebaicum per day, upon the substitution for which of bromide of potassium this last works particularly intensely, has caused some deaths.

Still a few words upon the prescribing of alkaline bromides in epilepsy. For poor people the bromide of potassium can be prescribed as the salt, from which the patient prepares for himself a solution of definite concentration. In general it is convenient to prepare solutions of such strength that a tablespoonful (or a measuring-glass of 15 cc) or a teaspoonful (5 cc) corresponds to 1.0 bromide of potassium. The solutions, on account of the effect on the stomach, should always be taken much diluted. On account of the long duration of the cure, the solutions of the alkaline bromides or the salts are prescribed in large quantities (as long as substances which do not keep well are not mixed with them). It only remains for me to give you the formulæ for several combinations of the alkaline bromides which have proved useful.

℞ Potassi bromid 200.0
Infus. radic valer. (20 per cent.) ad. 1000.0

M. S. One teaspoonful=1.0 grm. KBr.

℞ Potassi bromide ..... 40.0 (10 drachms)
Sodii bromide,
Ammon. bromide,
Sodium benzoat .....āā 12.0 (3 drachms)
Aq. menth. pip. ..... 1000.0 (1 quart)

M. S. One tablespoonful=1.0 grm. bromides. (15 grains)

℞ Potassi bromide,
Sodium bromide .....āā 100.0 (3 oz.)
Ammon. bromide ..... 50.0 (1½ oz.)

M. S. Keep in a well-stoppered bottle.
X grammes dissolved in water 2 to 3 times a day.*

℞ Potassi bromide ..... 20.0 (5 drachms)
Codeini phosphor ..... 0.2 (3 grains)
1% Inf. Adonidis vernalis q. s. ...ad. 300.0 (10 oz.)

M. S. One tablespoonful=1 grm. KBr. (15 grains)

The individual epileptic attack furnishes, as a rule, no indication for therapeutic interference. Only in threatening asphyxia must we proceed to artificial respiration, injections of ether, camphor, etc. In general we content ourselves, however, with placing the patient so that he cannot injure himself during the convulsions, loosening his clothing, and placing a rolled-up handkerchief between his teeth to prevent him biting his tongue.

A cutting short of the attack through certain manipulations which they have come upon empirically occasionally appears possible in certain patients. One can mainly reckon upon this possibility in the so-called "reflex epilepsy" in which, tying a band tightly about the limb wherein the sensory aura appears can prevent the threatening attack. One of my patients whose attacks are preceded by an acoustic aura, can prevent it by inflation of the Eustachian tubes; in a patient of *Heilbronner* singing has had the same effect; two epileptics of *Oppenheim's* have, indeed, found it possible to effect this end by a simple effort of the will.

In status epilepticus active intervention is always in place. Subcutaneous aseptic injections of sterile sodium bromide solution can be recommended. Dosage (according to *Morgan* and *Hodskins*) 100 to 180 cc of a 6 per cent. solution; place of injection, thigh or back. The painful induration which these injections produce, must be treated for several days with cataplasms. Large

* Bromides are very heavy. A teaspoonful equals about 8.0 grm. In order to spare the patient weighing out his dose every time, he can be given a test tube on which is filed a line corresponding to the point to which it must be filled to get a definite quantity of the mixed bromides.

doses of bromide of sodium (15.0) can also be given in a clysma; however, the action when given in this way is naturally slower. For rectal application hence, chloral is better suited (4.0 grm in very dilute solution). A further drug which sometimes is useful in status epilepticus is scopolamin hydrobromide in large, even overmaximum doses (0.001 to 0.002). Also chloroform narcosis is sometimes applied as a last resort.

The question of the operative treatment of epilepsy, in so far as one has in view genuine epilepsy, and does not consider the results in such epileptiform attacks as are produced by local circumscribed disease processes (cysts, meningeal adhesions, exostoses of the skull, etc.), is still under discussion. *Kocher* has expressed himself in favor of the undertaking of a craniectomy through which beneficial relief of pressure is to be effected. Against this theory of relief of pressure is, however, to be opposed the fact that *Tissot* for years has alternately left epileptic patients alone or has subjected them to regular lumbar puncture. He found that lumbar puncture, regularly repeated, even for a long time, exerted a modifying influence neither upon the course nor upon the character of their paroxysms. Also he could find no correlation between the pressure and the quantity of spinal fluid on the one hand, and the convulsive attacks on the other. In favor of partial excisions of the cortex, *S. Auerbach* has expressed himself of late; for "brain massage," *E. Bircher.* I must acknowledge that I have seen so many cases of genuine epilepsy operated upon without result, that I am inclined to refer the favorable changes in the character of the disease which have here and there been observed after operations, to accidental coincidence.

On account of the dangerous character of epileptics who have undergone psychic alteration in any considerable degree, care should of course be taken that they are committed in time to an insane, or epileptic asylum.

LECTURE XXVII

The Psychoneuroses

GENTLEMEN: When in the following lectures I describe neurasthenia and hysteria in particular detail, it is, in the first instance, from thoroughly practical points of view. There can be no doubt that the importance which these two most wide-spread nervous diseases have in the work of the physician, stands in striking contradiction to the neglect under which they suffer in general in clinical instruction. The disadvantage, however, which insufficient acquaintance with these disease conditions can have as a result are numerous and serious. Even among physicians, the view of the laity, which in the symptoms of these diseases sees evidence of an affection of the will, an expression of effeminacy, weakness of character, contrary disposition, is still wide-spread, and so it comes about that such patients, for whose cure the first requisite is that they find understanding, lose all confidence in physicians and in great numbers fall into the nets of quacks and charlatans of every style of practice. Still worse is it indeed, when, as unfortunately so often happens, in the making of the diagnosis neurasthenia or hysteria, wrong is done and, under these false colors, organic nervous diseases sail so long that the time during which therapeutic interference is possible goes by. The treatment of hysterical and neurasthenic conditions is, however, when correctly carried out, on the average a grateful task. Here, the art of the physician may accomplish many striking results, which in a way make up for our depressing helplessness when opposed to so many nervous diseases. But the difficulties in suitable treatment of these diseases also are not to be underestimated. This preconceives not only thorough knowledge of therapeutic resources, but also a never-failing interest in the disease and in the patient. Such prerequisites, however, are absent in only too many physicians; and he who complains about his "tiresome" hysterical and neurasthenic patients cannot reckon upon important therapeutic results. I will attempt to show you that neurasthenia and hysteria are anything but tiresome. Indeed in no branch of medicine is it more brought to our consciousness that our specialty is not only a technical one, but to-day still, does not deny a direct descent from philosophy. The study of hysteria and neurasthenia often brings us into contact with very interesting persons, and when they, in free confession, present us an insight into their psyche, we learn to know mechanisms of the mental processes which are of course pathological, but in general bring to us a rich harvest in knowledge of mankind and psychological understanding. The most satisfactory gain remains, however, I repeat it again, to therapeutic effort when it is granted to us to remodel unfortunate individualities, a burden to themselves and others, into normally feeling human beings.

A. Psychoneuroses and Neuropathic Diathesis

Neurasthenia and hysteria are still much classed in the great group of "neuroses," as which, according to the example of *Cullen*, the Scotch physician (1776), are brought together those nervous diseases which depend, not upon organic alterations of the nerve cells and nerve fibers, but usually upon functional abnormalities, upon an unruly activity of parts of the nervous system. Now, however, of late years it shows itself almost from day to day more plainly how greatly the composition of this group does violence to the facts, and how entirely heterogeneous diseases are brought in such manner under a common classification. The group of the neuroses is shrinking more and more, and this is indeed natural, since their definition is based upon a negative peculiarity, namely the want of an anatomical basis. We should better say, perhaps, upon our lack of knowledge of such. This grouping carries in itself the stamp of the provisory, since with the refinement of our histological technique, and the progress of our etiological investigations, alterations up to this time unsuspected must come to our knowledge and also toxic and infectious factors will be disclosed. As some decades ago, even tabes dorsalis passed for a neurosis, so later tetanus and hydrophobia still figured under this designation, until finally their microbic nature became evident. That acromegaly, *Basedow's* disease, tetany, etc., have as a basis, anomalies of the glandular functions, is a recognition of recent date, which must result in their exclusion from the nosological group "neuroses." As far as epilepsy is concerned, I have laid down to you in the last lecture, that to-day a doubt as to its nature as an organic affection of the brain-cortex is no longer possible, and that we must assume, where no histological changes could be recognized, that it was the fault of the insufficiency of technique; the same conclusion is permitted in chorea.

That on the other hand in neurasthenia and hysteria, as a matter of fact, no anatomical alteration has ever been found, would not suffice in my opinion to prove them the definite survivors from the family of the neuroses; this still could depend upon the weakness of our microscopes and the imperfection of our staining methods. But there are also positive signs common to both affections. To be emphasized above every thing are the transitoriness and changeability, the proteus-like manifoldness in the succession and in the combination of the symptoms, which can make themselves apparent not only from case to case, but also in one and the same patient from day to day, often from hour to hour, while in other nervous diseases, the clinical pictures can repeat themselves from case to case with a stereotyped similarity; for the troubles of neurasthenics and hysterics, if a paradox is permitted to me, the sole rule which prevails is that of irregularity.

Still more important, however, is a peculiarity taking these disease conditions out of the limits of other nervous diseases, I mean the greatly preponderant rôle which in their origin, in the method of appearance of their symptoms, and on account of this also in the curative procedures adapted to combating them, is played by psychic injuries, psychic phenomena and psychic

influences. On this account we accept the denomination "psychoneuroses" which *Paul Dubois* has introduced for these disease conditions. This word, indeed, expresses in appropriate manner that the whole multiplicity of "nervous symptoms," even where they manifest themselves in the bodily sphere, is most closely connected with mental factors; in their occurrence, emotions, hypochondriac fears, exaggerated self-observation, play an important rôle, as do also the suggestive effects of surroundings or the abnormal products of a phantasy directed into false paths.

Welcome to us further is the term psychoneurosis as a common designation for the two disease conditions in question. In these latter, the sentence "natura non facit saltum" (nature does not take a leap), is especially applicable. The cases in which we are in doubt as to whether we shall diagnose hysteria or neurasthenia, in which we must assist ourselves with the designation "hystero-neurasthenia" and where transition and intermediate forms of these diseases are present, are excessively frequent. The boundaries in any case are not to be drawn sharply, and if we do draw them, this is chiefly upon didactic grounds, since without boundary marks to some extent definite, we could not locate ourselves in the confusion of symptomatology.

In tiresome monotony there meets us, over and over again, the commonplace, that the psychoneuroses (or as the popular equivalent has it, "nervosity") are diseases of modern civilization. This is applicable only with limitations, since hysteria, and indeed in its most severe and striking forms, is as old as humanity. This must be plainly apparent to every one who knows how to estimate in their proper nature the hysterical epidemics of ancient times and of the Middle Ages, and *Hippocrates*, even, has furnished good descriptions of hysterical symptoms, which he also recognized as such. It is somewhat different, indeed, with neurasthenia; though there are neurologists who speak of the "discovery" of this disease by the American *George Beard* in the year 1887. Of course this author did not discover neurasthenia, but studied and described it in masterly fashion, recognized it in its nature, and furnished it with a fortunately chosen name. When, however, we look over the medical literature of preceding decades, we find under other designations (such as "nervosism," "nervous erythism," "general hyperesthesia," "cerebro-cardial neuropathy," "spinal irritation") disease pictures which present typical neurasthenic symptom-complexes; particularly, however, is it the diagnosis hypochondria with which since ancient times, neurasthenia has been saddled. With the old authors hypochondria played a preponderant rôle; to-day we make this diagnosis only in rare cases in which there is a mental disease proper, with thoroughly systematized delusional conceptions built up upon fears of disease. The picture, however, which *Molière* in his "Malade Imaginaire" draws of a presumable hypochondriac, is that of nothing else but an excellently observed and, in spite of caricature, unmistakable neurasthenic. The disease which in the seventeenth century under the popular name "Vapors," scourged the Court and noble circles of France, was scarcely anything else; in the letters of *Mme. de Sévigné* and of the *Abbé de Brosse* there are proofs of this. It would lead me too far afield to enter into the documents bearing upon the history of neurasthenia furnished by the ancient writers; for example, the Roman satirists, but

I cannot fail to cite a clinical history taken from the fifth Hippocratic book, "On Epidemics." This was of a patient "Who suffered from bodily relaxation. He could not pass by any pit, neither could he go on a bridge, nor yet was he in a condition to cross over even the slightest trench, although he could walk in the trench itself. Such things happened to him for a very long time." You see here the unmistakable sketch of obsessive ideas which are so frequent in severe neurasthenia. The description of "Gephyrophobia," as we term it to-day, is indeed, classical.

Taken as individual diseases the psychoneuroses need not be designated as the regrettable privilege of our present epoch of culture. Now at present, however, that is for about the last three decades, we have not alone to deal with single cases or with epidemics bounded by time and place, but with a definite nervosity of peoples and races which although not dangerous to life, is nevertheless of great importance, since it injures intensely the capacity for work and productivity of extended strata, of all classes of society. Into the causes of this threatening increase of the psychoneuroses I will not enter at the start. We will reserve the pathogenetic considerations for the next lecture. In order to give you, however, a comparative numerical conception as to the extent of the psychoneuroses, I would refer to the investigations of *Crämer* among the Göttingen student body. By comparison of the whole number of students with the number of those who sought professional advice on account of nervosity, he found the figures of 30 to 40 per cent., and thinks that these figures are certainly not too high, since the students are, with few exceptions, individuals who have as yet been little shaken up by the storms and experiences of life. On the other hand, however, I might point out that among students, other nerve-injuring factors come into action in a relatively high percentage of cases; fear as to examinations, venereal diseases, excesses in drinking, etc. In any case the comparison drawn by *Crämer*, with the frequency of psychoses, is interesting; among 1,000 people there are 2 insane persons needing asylum care, and 2 who can more or less get along in the world.

Shall we class the psychoneuroses with the congenital or with the acquired diseases? This question I can answer you neither in the affirmative nor in the negative. It is true that on the one hand external injurious factors can produce neurasthenia and hysteria; on the other, however, that for many cases the manifestation of an inferiority present in the organism from the start is responsible. Between the congenital, endogenic, and the acquired exogenic factors, there is only a relation of reciprocity. External injurious factors of considerable intensity can make even an individual who possesses from the start a robust nervous system neurasthenic or hysterical. In others, however, with the best will and through the most exact anamnesis, we cannot elicit any adequate external injurious factors; here it is to be assumed (and often to be recognized) that such individuals have brought into the world a nervous system having abnormal lack of resistance, so that they are unable to withstand the demands of life, even on the school-bench, or still earlier, and fall ill under conditions which to one healthy nervously, "ab ovo," would be entirely harmless and irrelevant.

We come here, also, as in so many branches of pathology, upon the con-

ception of "predisposition." In this special case this is denominated "neuropathic diathesis." This conception has been much ridiculed, since it contains in itself much that is vague and unsatisfactory, but even the most exact branch of medicine, bacteriology, has had to recognize that it cannot get along without the conception of predisposition. Only in this way can we understand all the remarkable leaps which an epidemic makes, in that it affects one individual, spares another, although it is known that this last one has been exposed in a high degree to the danger of infection.

Close relationships join the question of neuropathic diathesis with that of "hereditary predisposition." One can well say to-day that the importance of this last is greatly overestimated in the occurrence of the psychoneuroses. It has been implicitly assumed that the healthy person must be free from "hereditary predisposition." This is, however, not so. *J. Koller* has found that among the relations of 370 healthy persons, different neuroses and psychoses were to be found 218 times, which corresponds to a percentage of 59. These figures, high beyond expectation, make for the critical observer the assumption of a sure causal nexus between the psychoneurotic disease of an individual and the neuro- and psycho-pathies occurring in his family, a matter of opinion which must in every individual case be particularly weighed and considered. Also it must continually be emphasized that from an entirely healthy stem neuropathically inclined offspring may be produced. Here, now, in relatively numerous cases the "blastophthoric" injurious factors of which we spoke in Lecture VIII are to be found among the ascendants. In severe cases of neuropathic diathesis somatic signs of congenital inferiority appear to us in the form of the so-called stigmata of degeneration to whose importance *Lombroso*, in his studies on "The Congenital Criminal," has referred. We consider as such stigmata: Marked asymmetry of the skull not to be explained by exogenic factors (for example injury at birth), anomalies of the teeth, great prominence of the upper jaw or of the lower jaw (prognathism, caput progeneum, Gothic palate, cleft palate, harelip), anomalies of the external ear (jughandle ear, adherent lobe, pointed ear), hypospadias, genital infantilism, syndactylism, web fingers, gampsodactylism (inability to extend the little finger), etc.

B. Neurasthenia

Since we have touched upon the most important points which should prepare the way for the understanding of psychoneurotic manifestations in general, we will pass over to the description of those disease pictures which we bring together as neurasthenic; those of hysteria we will reserve until later, since they are mainly more complicated and more difficult to understand. But for the present in the consideration of the neuropathic phenomena we will put aside all theoretical and all pathogenetical considerations and make our description purely clinical.

Symptomatology

The disease manifestations of neurasthenia separate into objective and subjective, of which the last play a specially important rôle. On account of this, the result of the anamnesis is just as instructive as that of the status, and we have to devote particular attention to hearing what the patient has to say. In this, however, a certain routine is an absolute requirement. It indeed is one of the peculiarities of the neurasthenic to enter with the utmost detail into each single symptom of his disease, and he usually makes no difference, or no sufficient difference, between the important and the unimportant. Nothing is more unwise than to cut him off shortly, when his conversation is superfluously long. The neurasthenic takes this very ill, and a physician who is impatient, or more correctly, who gives evidence of his impatience, will rapidly lose confidence and authority with such patients. One must hence learn to curtail the prolixity of his description of his disease, by properly interpolated questions, turning the narrator away from the unimportant and guiding him to the symptoms diagnostically important. A considerable fraction of neurasthenics frequently aid us in taking the anamnesis and compiling the clinical history, in that they hand us written sketches of their disease (in which no symptom is unconsidered or forgotten); "l'homme aux petits papiers" (the man with the little papers) *Charcot* hence has called the neurasthenic.

Let me place before you two examples of such auto-nosographies with the remark that we cannot expect always such short and expressive descriptions:

1. "*Symptoms.*—With ever so slow increase of severe pains in the chest and in the back from the neck to the anus. Burning over the whole body, particularly severe in the legs, a biting feeling over the whole head. Three and one-half to four hours rest at night, then sudden awakening, trembling over the whole body and no more sleep; sweating at night, by day very frequent nervous tremor, anxiety, uncertainty in walking. Attacks of vertigo very frequent."

2. "*Remarks for the Doctor.*—Often headaches and sleepless nights. Short dizziness, a feeling as if the head was drawn backward, pain when there is noise, starting at every noise, for example, the fall of a tablespoon. One morning I felt a stiffness on the left side, felt myself strange in my own house, the people and everything appeared to me different. Little by little the old impressions returned to me, only memory remained poor. Usually the stiffness on the left side repeats itself, indeed it is noticeable also on the right. Sometimes stabbing headache and on strenuous work, sense of nausea. An exceedingly painful uncertainty pursues me the whole day, then I question myself, must I do that, is it I or only a dream, and the interpretation is now obscure in my head. I cannot any longer collect my thoughts."

These two typical personal observations respectively of a patient with principally somatic symptoms and of one with predominantly psychic symptoms ("psychasthenia") show how extremely manifold the symptoms of the psychoneuroses now occupying us can be. It is hence well for us to proceed quite systematically to bring the greatest possible order out of this confusion and to subject the chief phenomena of neurasthenia to a separate considera-

tion. We will suppose a fully developed case of acquired neurasthenia, that is, in which the special congenital degenerative characteristics do not occupy the foreground. The degenerative forms will later be discussed as well as a few semiologically characteristic varieties, such as the so-called "sexual neurasthenia."

Since in the course of taking the anamnesis we become acquainted with the subjective complaints while only examination into the present condition brings the objective symptoms before us, it is justifiable to begin our description with the first group of disease manifestations.

1. PSYCHIC ANOMALIES

a. State of Feeling

The basal symptom in the state of feeling of the neurasthenic is what *George Beard* has characterized as irritable weakness, an abnormal irritability and exhaustibility. The patient is irritable; trifles ("the flies on the wall") disturb him. To disagreeable occurrences, or to actual mental pain he reacts in immoderate manner, after the emotional discharge to sink back into deep apathy. Even agreeable happenings, pleasurable stimuli, produce excessive reaction, and then weakness and exhaustion often with psychical depression. To "sky-high exaltation" succeeds "mortal depression." Still the basal state of feeling upon which such episodes arise is usually a comparatively depressed one, rather morose than sad. This ill humor is in the main to be considered as a secondary symptom corresponding to the uncomfortable general condition; and reflections about the diseased ego play in this a considerable rôle. In any case, it is a quite different symptom from the depression of the melancholiac, since this last in no way arises from reflection, but without any thought as to its cause, it impresses itself upon the patient with overpowering might. Anxiety mainly of a hypochondriacal, sometimes also of a superstitious coloring, as well as defect of energy and inability to form conclusions, are also quite characteristic.

b. Intellectual Capabilities

In the intellectual sphere on the other hand, no important disturbances are noticeable except for the abnormally rapidly occurring and the abnormally disagreeably perceived fatigability, which gives to the mental work of the highly endowed and talented persons, who form no small proportion of patients with acquired neurasthenia, an aphoristic and broken-off character. In any event the patients often complain of "reduction of their mental force," that the ability to receive and combine impressions and the memory have suffered. A more exact investigation, however, makes it plain to us that these functions are in fact uninfluenced, and that only the continued occupation of the patient with the observation and explanation of his troubles prevents him from turning his attention sufficiently to things of the surrounding world. This "narrowing of the field of consciousness," brings it about that a neurasthenic must read the same sentence several times before he understands it, that

names, words, dates, easily escape him (since he has imperfectly noted them), that he is unable to concentrate his thoughts strictly upon an intellectual task.

2. HEADACHES

More than half of all neurasthenic patients complain of headache. If we require them, however, which we should not neglect in any case, to give an accurate description of this trouble, we will usually find that there are not actual pains, but only abnormal, more or less unpleasant sensations. Mainly we have to do with what *Charcot* called "le casque neurasthenique" (neurasthenic helmet), a feeling of a weight pressing upon the whole skull. Frequently the pressure is located only in the frontal region, more rarely at the back of the neck. Other patients complain of a feeling that the brain is threatening to burst the skull, or, on the contrary, that it is too small, that the head is empty. Often it is only a "dullness in the head." Of true severe headaches only such neurasthenics who live in terror of suffering from a severe organic brain disease, as brain tumor or brain syphilis, complain usually. Nevertheless, such head troubles increased in consequence of intense fixation of the attention and autosuggestive factors to considerable intensity, are to be recognized as to their neurasthenic nature before exact investigation, by the fact that upon the mental abstraction of the patient they disappear.

3. SLEEPLESSNESS

This, also, constitutes an exceedingly frequent complaint of neurasthenics; still, their expression "sleeplessness" is never to be taken literally, even although the patients expressly insist that they cannot sleep at all, "hear every quarter hour strike." In such cases one has to do, as a rule, only with an insufficient depth of sleep, with a condition of half sleep, which we, with the Hungarian author *Lechner*, who has studied the disturbances of sleep particularly thoroughly, will designate "Dysnystaxis." Other patients suffer from difficulty in falling asleep ("Dyskoimesis"), or from too early awakening ("Dysphylaxia") although sleep is deep enough. Distressing dreams, nightmare, are not rare. The patients very often, indeed, complain of a sudden starting which frightens them just as they are going to sleep. Even when the duration and the depth of sleep apparently present no particular anomalies, the refreshing action of sleep is absent and we hear then that the patients "arise more tired than they were when they went to bed." Especially trying is a drowsiness during the day which is in great contrast with the abnormal wakefulness at night. "Night is the enemy of the nervous," correctly says *Hermann Oppenheim*, who has subjected the genesis of the neurasthenic disturbances of sleep to a keen analysis. With the abandonment of activity and all external interests, the attention of the neurasthenic may be turned in a higher degree to his bodily processes, so that there occurs a hyperesthesia toward sensations which the healthy person does not perceive at all, for instance, his own heart-beat, the peristalsis of his intestines. These continued sense perceptions naturally oppose going to sleep, for which the elimination

as far as possible of the irritants reaching the sensorium furnishes the best conditions. Through a specially exaggerated "self-perception" every organic feeling can take on an actually painful coloring, on account of which we speak of " hypnalgias" and "nyctalgias." The healthy person also for all that, can provoke such phenomena in himself by way of experiment. Attempt during the stillness and darkness of the night when your attention cannot be distracted by external perceptions to fix this in the greatest concentration possible upon a definite part of your body, for example, upon one heel; you will in a few minutes perceive an unpleasant sensation in this region. The contact with the sheet will be first found noticed as an unpleasant tickling, then as itching and burning, and in case you do not allow your attention to be distracted in some other direction, finally as actual pain. Upon the basis of this personal experiment which can be repeated without difficulty, I cannot agree with *Oppenheim* when he writes that the interpretation of hypnalgias and nyctalgias gives particular difficulty.

4. IRRITATIVE SYMPTOMS ON THE PART OF THE SENSE ORGANS

Exaggerated feeling of self furnishes us the key to the understanding of a number of subjective ear and eye symptoms which occur in a large number of neurasthenics and in which we are justified in assuming a morbid irritability of the sense nerves in question, a reduction of their threshold of irritability. The following irritative symptoms are to be observed: Hyperesthesia to light, blinded feeling, spots before the eyes, hyperesthesia for noise, so that such patients, for example, perceive the whistle of a locomotive as actual pain and anxiously avoid one. Further, tinnitus aurium, sometimes rhythmic, synchronous with the pulse, and in this case dependent upon the perception of the noise of the circulation within the skull, to which the cochlear nerve with normal threshold of irritability would not react. The entoptic perception of neurasthenics, denominated "musci volitantes," has nothing to do with similar phenomena in opacities of the vitreous body (indeed, ophthalmoscopic examination shows here that all the media of the eye are clear), and is probably produced by the circulation in the retina or by the corpuscular elements of the tear secretion. Acuteness of vision and hearing remains uninfluenced in all the phenomena mentioned. The single defect symptom which neurasthenia may produce in the visual apparatus is usually to be referred to the abnormal fatigability of the internal recti and of the ciliary muscle; these are the so-called "asthenopic" troubles which make themselves apparent after a more or less short time, upon fixation and accommodation for near objects.

5. VERTIGO

Only in quite rare cases do true attacks of vertigo with disturbances of equilibrium occur (for example having a rotatory vertiginous character), which can be considered as an irritative symptom on the part of a pathologically irritable vestibular apparatus, which answers in this manner to sudden

variations in the blood distribution in the internal ear. When, in spite of this, our neurasthenic patients complain of vertigo with excessive frequency, this comes from the fact that under this term, the public understands very different things. A momentary fit of weakness, the feeling that it is suddenly black before the eyes, as if the legs were sinking into the floor, or as if they were taken away from the body, all these abnormal sensations generally come under this designation. If a neurasthenic has once experienced a subjective difficulty of this sort the fear of "vertigo" usually makes itself felt in very marked form and the reoccurrence of such symptoms naturally increases considerably this fear, a painful, vicious circle. Fortunately in very few cases does an actual "permanent vertigo" appear.

6. PARESTHESIAS

Abnormal sensations occur in neurasthenics not only in the regions of the skull and the nerves of special sense, but also in those of the skin, the mucous membranes, muscles, bones, tendons and joints, in varied and manifold forms. If they are also of unpleasant character and indeed actually painful, so is there here also, as we emphasized under the "Headache" and "Sleeplessness" of neurasthenics, very frequently an unconscious tendency to exaggeration. Not at all infrequently such patients complain of their "terrible" pains not ceasing a moment, but their healthy appearance, their expression, their abstractibility, the fact that they can exercise their calling undisturbed, etc., form such a contrast to the patients who suffer from the lancinating pains of tabes or trigeminal neuralgia, that we are in a position without difficulty to reduce their description to its proper proportion. The most frequent seat of such painful paresthesias is the back; we speak then of spinal irritation, of rachialgia. But also in the muscles of the limbs are located various sensations which probably correspond to those which the normal person perceives after intense exertion as the well-known "fatigue pain." In rare cases "dysbasia neurasthenica intermittens," which should not be confounded with the angiogenic forms of intermittent limping, can so occur (see above, pages 225 and 374). Here, probably, the two components of "irritative weakness" (morbid increase of motor fatigability on the one hand and of sensory irritability on the other) play an important rôle. As paresthesia of the bones is to be interpreted the so-called "anxietas tibiarum," a dull, difficultly definable feeling in the bones of the leg—whose parallel occurring in healthy people is the well-known sensation designated by the expression, "the fright has got into my legs"—which can be experienced continuously by the neurasthenic. Neurasthenic arthralgias have led to confusion with rheumatic joint diseases. In contradistinction to these last, however, they increase in intensity during rest, diminish on movement, a criterion which applies to many neurasthenic pains, as *Kollorits* has expressly emphasized. Pruritus, itching in the skin and mucous membranes forms, finally, a frequent complaint of neurasthenics. It is almost never general, as are often the parasitic, the senile, and the forms occurring in diabetics and arteriosclerotics; it is rather, almost always, localized upon definite areas. Itching of the pharyngeal mucous membrane leads to dry, nervous "irritative cough," pruri-

tus of the urethral or rectal mucous membrane to distressing tenesmus of these excretory passages; pruritus of the genitals may be the starting-point for masturbatory practices. It is self-evident that the diagnosis of nervous pruritus should only be made, when an exact investigation can exclude the other disease conditions above mentioned.

When we turn our attention to the subjective symptoms of neurasthenia, we are first expressly reminded of the polymorphism of the neurasthenic disease picture. This brings it about, that we need not expect in every neurasthenic to find represented each of the four groups of symptoms into which, upon didactic grounds, it is well to arrange these anomalies. Still, almost never will a neurasthenic fail to show any objective symptoms at all, so that their diagnostic importance is not to be underestimated.

7. INCREASED IRRITABILITY OF THE NEURO-MUSCULAR APPARATUS

This anomaly expresses itself particularly in a general increase of the bone and tendon reflexes, which indeed in general remains below that which we find in organic affections, for example, in the spastic symptom-complex, but still is occasionally so intense that a doubt may arise in this direction. In such cases, the fact that functional increase of reflexes is never accompanied by *Babinski's*, *Oppenheim's*, or the *Mendel-Bechterew* phenomenon, associated movements, etc., is of great importance. Further, permanent hypertonia of the muscles is absent. The so-called "functional" or "pseudo" ankle clonus could at most give occasion for confusion. Still, there are never here the thoroughly rhythmic, even contractions which can be provoked as long as desired, which characterize true organic ankle clonus, but irregular movements of varying amplitude, which, besides, are soon exhausted. This is shown most plainly in graphic reproductions of true and false clonus upon the rotating drum (clonograph), still, an examiner of any experience can always dispense with this complicated method of investigation. A reduction of the tendon reflexes can sometimes be found in such neurasthenics who, in consequence of marked nervous dyspepsia, are in a condition of considerable muscular emaciation.

The absence of a reflex phenomenon normally constant, particularly also of the patellar reflex, in a neurasthenic, indicates without exception a complicating organic anomaly. Also the skin reflexes, namely the plantar reflex, are in neurasthenics sometimes exaggeratedly lively.

Frequently an increased mechanical muscle irritability is to be found, so that even slight blows with the percussion hammer upon the extensor side of the forearm set up a powerful motor effect at the wrist and in the individual fingers. Also in the quadriceps and in the pectoralis major is this mechanical hyper-excitability of the muscles often very clearly to be recognized. The "idiomuscular contraction" is sometimes very plainly visible. On the other hand, mechanical overirritability of the nerve trunks is much rarer. It is most frequently demonstrable upon the ulnar nerve behind the elbow,

in that by rolling this nerve under the palpating finger a definite contraction of all the muscles supplied by it can be produced.

8. TREMOR AND SIMILAR PHENOMENA

In the great majority of neurasthenics, when we request the patient to close the eyes firmly we can notice a marked tremor in the orbicularis palpebrarum. The slightest degrees of this phenomenon are, as you should note, presented sometimes by an entirely healthy person, for instance, in a condition of fatigue, after long reading; in the neurasthenic, however, this may increase to a blinking, which is increased upon attempt to keep the eyelids as still as possible. Less frequent are tremor of the outstretched tongue, and that of the fingers which is usually very fine and vibrating. *Quinquaud's* symptom, already mentioned in Lecture IV, page 63, is by no means infrequent in neurasthenics, while the phenomena of fibrillary contractions and myokymia described in the same lecture are only very exceptionally observed here, and hence should naturally only be considered as neurasthenic, after exclusion of an organic substratum. In functional cases they occur most frequently in the first interosseus of the hand.

9. CARDIO-VASCULAR AND RESPIRATORY DISTURBANCES

The cardiac disturbances in many neurasthenics to such an extent occupy the foreground in the symptomatology that these patients frequently turn at first, not to the neurologist, but, thinking that they have heart disease, to the internist. So, *Gerhardt* affirmed that over half of the patients who came to consult him about heart disease actually suffered from "neurasthenia cordis." On the other hand, you are expressly warned against making this diagnosis too precipitately, and I greatly recommend to you to carefully examine the heart, the vessels, the urine, etc., even in "plain neurasthenias," in order not later to have to reproach yourself for having overlooked a myodegeneration of the heart, a contracted kidney, a coronary sclerosis, etc. The most important objective symptom of the nervous heart is the quickening of the pulse, which may be permanent—analogous to that which we have learned to know in *Basedow's* disease,—but which shows itself usually only on occasion, by preference at night, but also during the day particularly with psychic disturbances. In exaggerated cases there is true paroxysmal tachycardia, with abrupt rise in the number of heart beats to 160 to 200 per minute.

A diagnostically valuable symptom is, further, the increase of lability and variability of the pulse-beat. By lability I understand the difference between the number of beats under different conditions, for instance on change of position (lying, sitting, standing), on effort (for example, mounting upon a chair 3 to 6 times, rapidly one after another, which corresponds to a work of 100 to 200 meter-kilogrammes)—as variability, the difference between the pulse-rate on different days (measured as near as possible under identical conditions). In healthy people pulse lability and variability are slight; in cardio-vasomotor neurasthenia, on the contrary, exceedingly great. The same

thing applies to the lability and variability of the blood-pressure, as I have been able to determine by measurements with *Gärtner's* tonometer, which for the rest in such forms is on the average abnormally high. While, for example, I found in healthy persons an average of 95 mm with the *Gärtner* apparatus, among 27 neurasthenics with heart and vascular symptoms I found the blood-pressure only 7 times under 100, 8 times between 100 and 115, over this 12 times. The maximum was 160. Only in two cases were the figures abnormally low, 75 and 60 mm. To be placed parallel to this is the great rarity of a functional nervous bradycardia. Where this is present in neurasthenics, it can often be found that there is a combination with nicotinism.

Irregularities of the heart action also belong to the rare objective symptoms of neurasthenia. Among them, true "pulsus inæqualis" (allorhythmia) upon a purely functional basis, that is, if an affection of the heart muscle can be certainly excluded, may be counted a rarity; somewhat more frequently the heart neurosis is accompanied by the missing of individual contractions, extrasystoles or arhythmia. In no way unusual, however, is the abnormal response of the heart to the respiratory phases, in that on inspiration an alteration of the pulse in the sense of "pulsus respiratione intermittens" makes itself more or less apparent.

That upon a purely nervous basis, variations in the size of the heart, the so-called "acute dilatation of the heart" can occur, I do not consider proved; also, in spite of the favorite diagnosis "nervous heart weakness," I have always considered the finding of symptoms of stasis (engorged liver, congestion of the kidney, cyanosis of the lips, œdema, etc.), even when these were only of slight degree, as proof of an organic heart disease, and this has always proved correct. On the other hand, in neurasthenics "cardioptosis" is a strikingly frequent symptom; if such patients are placed upon the left side the apex-beat moves across the mammillary line outward, and this "movable heart" is also to be recognized by percussion. Further the second sound over the aorta and the pulmonary valves is often abnormally loud. According to *Oppenheim*, upon excitement, besides increased rapidity of the pulse, a temporary systolic blowing may appear. I have never been able to confirm this.

The objective heart anomalies just enumerated are practically always accompanied by subjective troubles, among which palpitation, the feeling of painful thumping of the heart, occupies the first place. Of other sensations, precordial pressure, which may increase to actual "pseudo-angina pectoris," comes into consideration. Between these last, in which psychogenic components in the sense of "exaggerated perception" which we have considered in the preceding lecture must play the chief rôle, and which also has properly been designated "precordial anxiety," and the already mentioned "angina pectoris vaso-motoria," the boundaries are not always to be sharply drawn. An original symptom of cardiac neurasthenia, which *Trommer* has described, you can here and there confirm. This is a soiled area on the shirt-bosom of male patients, corresponding to the position of the heart, which has been produced by their constantly placing the hand upon the heart region on account of the disagreeable sensations there localized.

Of vasomotor disturbances there are most frequently to be found in neuras-

thenics "rushes of blood," that is, congestions of the head with great local feeling of heat; the ears and cheeks feel hot and sometimes the cervical vessels beat with exaggerated intensity. With the hyperemia of the head is contrasted often the coldness and lividity of the hands and feet, also a rapid alternation of reddening and pallor is peculiar to many patients. A cooperation of the psyche manifests itself in many cases in characteristic manner; his tendency to frequent and motiveless blushing is well known to the patient; he fears it ("ereuthophobia") and this preoccupation is to blame that blushing actually occurs regularly upon meeting unknown persons, in any sort of situation not entirely common. Also circumscribed and migrating erythematous spots ("fliegende röten") are a frequent symptom in neurasthenia. Dermographism and factitious urticaria occur in the same manner, as has been described for *Basedow's* disease. But many neurasthenics also suffer from spontaneous urticaria, and are further, as we already emphasized in the beginning lecture, predisposed to different vasomotor neuroses.

Much more rare than the vasomotor-cardiac, are respiratory symptoms in neurasthenics. In spite of the certain relations of bronchial asthma to nervous influences there are actually among the great horde of neurasthenics strikingly few typical asthmatics. What is designated by such patients as asthma is generally only a "nervous tachypnœa" sometimes running parallel with the tachycardia.

10. ANOMALIES OF SECRETION

The secretory anomalies of neurasthenia are somewhat less frequent than the cardio-vasomotor. Most widespread is readily the tendency to excessive outbreaks of sweat, general or localized (forehead, hands, feet); in the second place are to be mentioned anomalies of the gastric secretion, among which hyperacidity is the rule, only exceptionally hypacidity, or even anacidity. The occurrence of an acid gastrorrhœa, in attacks, is called "paroxysmal gastroxynsis." Increased salivation and polyuria are rare, exceedingly frequent, on the contrary, the symptom denominated, little justifiably, "phosphaturia," a precipitate of the earthy phosphates (calcium and magnesium) soluble in acids, present in freshly passed urine, or occurring when it is warmed. Without a trace of proof this precipitate has been improperly interpreted as a sign of increased decomposition of phosphorus containing nerve substance (lecithin, protagon). This view, however, may be considered as aside from the subject, so long as it is not supported by exact experiments on metabolism carried out under all precautions upon such phosphaturic neurasthenics, and there are no such at present. With the simple determination of phosphates in the urine nothing can be decided; satisfactory conclusions can only be expected from the exact determination of the phosphorus intake and its excretion by the neurasthenic during a given time. At the present time I consider it probable that simple alteration of the conditions of solubility in the urine are to be held responsible for the so-called neurasthenic phosphaturia. These are perhaps due to abnormalities in the production of acids in the body. Finally, is to be mentioned the "mucous colic" or "pseudo-

membranous colitis," in which, in attacks with severe abdominal pain, tube-like formations of mucus and fibrin are discharged. This disease, a secretory neurosis of the colon, is observed particularly in psychoneurotic patients (mainly females); the popular designation, "nerve mucus," customary in many places, shows sharp observation on the part of the laity.

LECTURE XXVIII

The Psychoneuroses

B. Neurasthenia (Continued)

GENTLEMEN: We have now brought to an end the analytical consideration of the manifold subjective and objective semiology of acquired neurasthenia, begun in the last lecture, and from now on we will direct our attention more to the grouping of symptoms than to single symptoms. It is self-evident, and I have sufficiently emphasized it, that all these disease symptoms cannot be present in every neurasthenic; rather must an exceedingly great variety of clinical pictures arise, according to the special prominence of this or that anomaly and the absence or recession of other disturbances. The necessity for systematization has given rise to the separation of the disease according to such so-called "localizations" of the neurasthenias into special varieties.

So, many neurologists speak of cerebral neurasthenia or encephalasthenia when the troubles of the patient consist chiefly in pressure in the head, tinnitus aurium, vertigo, disturbances of sleep, irritability, etc. As hyperalgetic neurasthenia has been denominated, that form in which pain in the back and disagreeable and painful sensations in the limbs constitute the chief complaint. Such disease pictures, however, in which palpitation of the heart, congestions, tachycardia, fleeting erythema, dermographism, etc., predominate, are classed as cardio-vasomotor neurasthenia. These three varieties, however, show so many transition forms and combinations that their separation must be considered quite an artificial one. A much more independent position must, on the contrary, be assigned to those varieties of neurasthenia which are denominated nervous dyspepsia and sexual neurasthenia, hence you shall have a connected exposition of these before we turn to the etiology and pathogenesis of neurasthenic conditions.

NERVOUS DYSPEPSIA

Here disturbances on the part of the digestive organs to such an extent occupy the foreground that it is the stomach specialists, and not the neurologists, who have contributed mainly to our knowledge of this exceedingly widespread form of disease.

The average picture of nervous dyspepsia, which naturally presents the most varied degrees of intensity, is about the following: The patients complain of a frequent burdensome feeling of pressure that either comes on after meals or is independent of taking food. The qualitative character of the food

is in remarkable manner often a matter of entire indifference, or, indeed, there is the paradoxical phenomenon that heavy, fat, acid foods, etc., are best borne; hence, sometimes the peculiar "régimes" which many such patients lay down for themselves, and to which they hold fast with great pedantry and obstinacy.

Precipitate change of the disease picture is a frequent occurrence; often we seek in vain for an adequate cause for the beginning of the dyspeptic trouble, very frequently, however, the influence of psychic factors is unmistakable; an anger, an excitement, sometimes a ridiculously slight annoyance, has "fallen on the stomach" of the patient, and only with the passing away of the mental annoyance the troublesome symptoms disappear. Frequent accompanying symptoms of stomach pressure are sour or stale eructations, anomalies of the stomach chemism (usually in the sense of a hyperacidity (see above, page 416), heartburn, regurgitation). The appetite is usually capricious, anorexia and buliminia can alternate with one another. Quite characteristic is a painfully perceived feeling of hunger which usually occurs on an empty stomach (particularly at night or upon awakening in the morning), and has been called by *Boas* "gastralgokenosis." The tongue often looks quite normal; usually, however, it is slightly coated; also the intestinal functions are in very many cases disturbed; there is most frequently atonic constipation (*Kussmaul's* "torpor peristalticus"), sometimes spastic constipation; that for the rest the combination of atony in the proximal and spasm in the distal divisions of the colon occurs, *Stierlin* has of late recognized roentgenologically. Among the spastically constipated dyspeptics, the disposition to occasional paroxysms of mucous colic is most widespread. More frequent, indeed, than the last so exceedingly characteristic disturbance, there are in nervous dyspepsia, in consequence of peristaltic intestinal unrest, diarrhœas. Such "nervous diarrhœas" are often abruptly set up by an emotion, a fright, etc. Very frequent is distressing pressure to empty the bowels, which usually occurs at an inconvenient time (in the theater, for example).

A by result of the intestinal atony, found particularly distressing by the patient, is, finally, flatulence; distention making itself apparent during the night, is often designated as the direct cause of sleep disturbances. In spite of these manifold digestive disturbances the condition of nutrition of the patient in nervous dyspepsia does not usually suffer to an appreciable degree. There are found among them, indeed, people of blooming appearance to whom may be applicable the exclamation of the visitor of *Beard's*, who, after a look at his waiting-room filled with neurasthenics, cried out, "Your patients are, indeed, regular giants."

It is otherwise, however, with the (in general, rare) severe cases of nervous dyspepsia, since here there may be extreme emaciation, indeed, real marasmus. In such cases the gastric troubles have taken on a chronic character, not remitting to any appreciable extent. Loss of appetite predominates. Along with the slight desire for nourishment the continued anxiety as to supposed dietary errors leads the patient to limit his bill of fare more and more, and to undertake actual fast cures in which he is often not aware of the insufficiency of his nutrition. In one of my patients, for example, who, although reduced

to a skeleton, affirmed that he abundantly nourished himself, it appeared from the investigation of his metabolism by *Jaquet* that he, in his hypochondriacal anxiety about abdominal distress, had actually reduced his ration to 1,350 calories a day (an average of 3 days). (As a comparison it may be recalled that in a woman lying in hysterical sleep, the daily minimum needed was reckoned by *Sondén* and *Tigerstedt* at 1,680 calories, while *Voit* gives the normal food value for adults at moderate labor at about 2,750 calories.) The loss of strength in severe cases of nervous dyspepsia can be increased in that there is not only eructation and regurgitation, as in the slighter forms, but there may be actual vomiting. Since, further, the feeling of pressure in the stomach sometimes increases to more or less severe pain, the differential diagnosis of such cases from ulcer or carcinoma of the stomach is often only possible after long observation with the aid of Roentgenological, chemical and microscopical examination of the stomach motility and stomach contents.

SEXUAL NEURASTHENIA

To the preponderant rôle which sexuality plays in the life of the individual and its manifold relationships to the most varied expressions of the psyche, corresponds the great extent of those forms of neurasthenia in which the whole disease picture revolves about disturbances of the sexual function. If the symptoms here are exclusively those to which organic basis is to be denied, nevertheless their appearance in material diseases of the male or female sexual organs (in the sense of a simple combination) is a very frequent occurrence. This is explainable, as we will assume in advance of our discussion of the etiology and pathogenesis of neurasthenia, in unconstrained manner from the fact that diseases of the genital organs, particularly venereal affections, to a special degree favor hypochondriacal self-observation and brooding. When discussing the treatment of neurasthenia I will shortly impress upon you the necessity of educating the patients to look upon their troubles with as much equanimity as possible, even to disregard them. Allow me to warn you against too close inquiry as to sexual anomalies which may furnish still more encouragement to reflection on things which without this already dominate too much the thoughts of the patient. Here the necessity of finding out the cause, with the conscientious physician, must give way to the principle "non nocere" (do no harm).

For the rest, the patients almost always acquire such confidence in a tactful, reserved medical adviser that they spread out before him voluntarily a sufficiently detailed sketch of the disturbances in question. These last can be quite satisfactorily arranged under *Beard's* definition of "irritable weakness." Indeed, abnormal sexual irritability and exhaustability can sometimes be differentiated as a first and second disease stage; more frequently, however, symptoms of both categories occur actually along with one another. In our consideration we will for clearness hold fast to this division.

The increased irritability first manifests itself in that the sexuality makes itself evident in a manner inadequate to the constitution of the patient. Erotic thoughts press upon the psyche in such predominant fashion as can often

find no satisfaction within the sphere of the attainable, and develop into sexual phantasies and waking dreams. Upon the same basis arise, also, the most varied sexual perversions. As physical expressions of the hyperaphrodisia are to be mentioned, in men excessive and continued erections which, particularly at night, reach distressing intensity and may lead to an agrypnia which, on account of the ever-present danger of masturbation, is particularly exhausting. Still more serious than this last is psychic "onanism," orgasm intentionally provoked by means of erotic phantasies. To be mentioned further are the excessively frequent (sometimes, indeed, nightly or several times in a night) nocturnal pollutions. If these are to be regarded as a morbid manifestation, when they occur in periods of sexual abstinence, only from excessive frequency and from the psychic and physical depression which follows them, diurnal pollutions are to be considered absolutely and without exception as a pathological occurrence. They occur, nevertheless, only in the most severe forms of sexual neurasthenia. From the increased sexual irritability, the carrying out of the sexual act is usually interfered with, often impossible; namely there is premature ejaculation. As irritative symptoms in sexual neurasthenia in females are to be mentioned: vaginismus (intercourse prevented), pruritus of the genitals (a starting-point for masturbatory practices) and pollutions, discharges of mucus from the glands about the vaginal entrance (*Bartholin's* glands, etc.) occurring with orgasm, in half-sleeping conditions.

As a symptom of weakness is to be mentioned the great exhaustion which even with active sexual desire follows coitus as well as pollutions. Sexual weakness in the male occurs most markedly, however, in the form of impotence which here, in contradistinction to the organic form (about which we learned in connection with tabes, for example), is to be denominated psychic. If here, insufficient response of the nerves of the sexual organs to erotic stimuli is often at fault (anaphrodisia), as a rule, the chief blame is to be ascribed to fear of failure, lack of self-confidence, etc., for this chief complaint of male sexual neurasthenics.

Still higher grades of sexual weakness are characterized by the substitution for pollutions of the discharge of semen while awake and without orgasm. This spermatorrhœa occurs upon urination and defecation (micturition and defecation spermatorrhœa). In women sexual weakness manifests itself in absolute frigidity.

The near relationships between the genital and the uropoetic apparatus, renders it easily understandable that the majority of sexual neurasthenics also complain of urinary troubles, namely of desire to urinate at inconvenient times, pollakiuria, dysuria and strangury. Particularly characteristic is the symptom which has been called "urinary stuttering." The patient during micturition must repeatedly stop, and so empties his bladder in small quantities at a time.

Finally, without exception, general neurasthenic symptoms manifest themselves in disturbing fashion, often favored by the fear of threatened disease of the spinal cord. The belief on the part of the public that sexual neurasthenia is the precursor of "consumption of the spinal cord" (tabes) maintains itself with ineradicable obstinacy, since it is intentionally supported by the

quacks and charlatans. The patients show a more or less severe depression, occasionally increasing to suicidal impulsion, are misanthropic, unable to apply themselves to any work, lose all interest, complain of pain in the back, headache, etc.

Pathogenesis and Etiology

Since now we have obtained a sufficient insight into the semiology of neurasthenic conditions, having used as examples the ordinary acquired neurasthenia as well as two of its varieties practically important, nervous dyspepsia and sexual neurasthenia, we must enter more fully into the question of the nature and the occurrence of such and similar symptom-complexes.

Here I would refer to my remarks already made in Lecture XXVII, on the question as to whether the psychoneuroses are to be counted among the hereditary or the acquired diseases. We saw there, that between the endogenic and the exogenic factors forming the basis of the individual case there exists a reciprocal relation, so that on the one hand under the influence of powerful external factors, even a normally constituted nervous system is affected, on the other, a neuropathic disposition brought into the world can lead to the development of pathological conditions even from the ordinary stresses of life.

For proper estimation of the individual neurasthenic disease picture, it is now of prime importance to take into consideration whether an endogenic or degenerative factor is present or absent. It is not the least service of *Charcot* that he first perceived the necessity for separating what he called true, and what hereditary, neurasthenia. In order to assign to both etiological varieties the same rank within the limits of *Beard's* disease, indeed, we had better strike out the epithet "true" and use for it "acquired" or "accidental" neurasthenia, in contradistinction to "congenital" or "constitutional neurasthenia." That these two varieties prognostically are to be estimated quite dissimilarly, and therapeutically are to be managed in an entirely different way, does not need to be specially emphasized. That, however, this principle of classification is a correct one, appears from the circumstances that in their symptomatology, also, as you will see directly, the accidental and the constitutional cases show differences, whose importance leaves far behind all that formerly justified the attempt at a topographico-symptomatological classification (see above, page 418).

Let us turn next to the etiology of the accidental form, which includes always recognizable and well-characterized influences, often affects, acutely, a previously entirely normal organism, and in which neuropathic heredity plays no rôle worth speaking of (see page 407). When we ask the patient himself what causes he holds responsible for his nervosity, we will hear him complain either of strokes of fate suffered, of agitations, distress and care, mental overexertion, business troubles,—he accuses himself of sexual excesses, or finally, he dates his nervous weakness back to a physical disease of acute or chronic course, from which he has suffered. When we now search through this rather varied list of causal factors, after a common criterion, it may be seductive to regard the factor of exhaustion usually as such. So *Möbius* would consider acquired neurasthenia as a variety of chronic exhaustion, its individual symp-

toms as potentiated exaggerated symptoms of the physiological fatigue process, developed into a permanent condition. This pathogenetic conception, however, does decided violence to the facts and hence, has even in neurological circles, been able to establish no support. Of course, there is a chronic fatigue, an exhaustion become a permanent condition ("defatigatio"); we observe it after long lying in bed with fevers, in the undernourished, among the women of the proletariat affected by hard work and numerous labors, after exhausting maneuvers, after excessive feats of strength in various sports, but we miss in this the most important physical and psychic stigmata of *Beard's* psychoneurosis, and the picture of simple fatigue is rather that of a very general relaxation and prostration, which usually disappears leaving no symptoms, upon rest and full nourishment, without medical aid.

For the development of an acquired neurasthenia, something more is needed than for that of a chronic exhaustion, not the excess of stimuli that press upon the nervous system, but their qualitative nature gives the result its marked emotional coloring.

An example: A business man attributes his severe neurasthenia to the heaping up of the duties of his calling; and still he admits that some years back he passed over a period of much more severe, even excessive exertion, almost like child's play. The explanation is furnished us by the fact that he at that time was in a position of less responsibility, but in the meantime had moved up to a post of heavy responsibility. The robust nervous system over which the affect-free exertion passed without trace, gave way under that marked by strong affect. Also in the wide-spread "examination neurasthenia" the anxious tension, not the forced work, is chiefly to be blamed.

Now there are indeed persons enough who under such circumstances do not fall victims to neurasthenia, and there is no doubt that certain "temperaments" are more exposed to the acquisition of a neurasthenia than others. The person conscientious from the start, strict with himself, may be considered as predisposed; the one of sentimental nature and soft heart, not less so; none more so than the ambitious person whose mental work can never develop without affect, but through stimulated expectation, or painful undeception experiences a continuous emotional coloring.

So we understand also, why neurasthenia grew first in North America to be a disease of the masses; in the last decades, however, it has overflown into the civilized countries of the Old World: parallel with the haste and unrest which modern methods of communication and the acquisitions of technology in all branches of human activity have liberated, in all nations the tendency to continually more complicated methods of life has developed. The movements of trade, the relations between production and consumption, have become continually more complicated, so that to speak with the national economist *Bücher*, "The existence and work of every individual is interlaced more and more with the existence and work of many others." In all departments of business life, we see more and more dependence upon numerous unaccountable factors enter, a feeling of uncertainty which is continually increased by the constantly more acute competition. Along with these specifically increased cares of occupation, those conducting different enterprises bear more and more the burden of

crushing responsibility. The measure of responsibility has, however, also increased for the subordinates in industrial and commercial enterprises. The middle class is engaged in a hard and desperate struggle upon two fronts. The psyche of the working man is in continued tension through the sharpening of the social contrasts, through class struggle, through the danger to life of the mechanical occupations. Also for him the danger of crises is a continually threatening one; the hours of labor are shorter, but the work has become more intensive and intellectual. In short, if the "American nervousness" of *Beard* has also mastered modern Europe, this has its basis in those phenomena which have been designated as the "Americanization" of our social conditions.

Also in the cases where, following bodily diseases, a symptomatologically typical neurasthenia (not as more frequently a simple exhaustion) is acquired, the affective factor is never absent. No wonder that among these diseases, just those which give occasion for distressing self-reproaches, or to anxious fears for the future, far excell all others in importance and frequency; namely, the venereal affections. Also in the sexual neurasthenia caused through excesses or errors, there manifest themselves, as the most important etiological agents, remorse, shame, but particularly the fear of evil results. That this fear is encouraged by quacks and medical persons fallen into devious ways, by brochures, prospectuses, etc., which have as their end to draw into their nets as many victims as possible, I have already emphasized. One of the most favored tricks of these people consists in representing such normal things as the occasional occurrence of pollution in sexual abstinence, and one testicle hanging lower than the other, as the consequences of youthful sins, and the precursors of incurable marasmus, and in suggesting in this manner to the readers of their literature, sexual neurasthenic troubles, for which then they assert that they possess the only cure. For the origin of severe dyspepsia sometimes improper medical treatment is at fault, as *Dejerine* has forcibly emphasized quite recently. As a prophylactic, one should refrain in neurasthenics from bringing into action for slight digestive disturbances, which, however, are greatly exaggerated by the patient, the imposing apparatus of complicated bills of fare, frequent washing out of the stomach, water cures, etc., but should on the contrary seek to represent to the patient his troubles as unimportant and fleeting symptoms and should avoid overtreatment.

Special importance in this era of social legislation has been acquired by "accident neurasthenia," which often in little satisfactory manner has been brought together with accident hysteria, the fright neuroses, commotion psychoses, etc., as "traumatic neurosis." Accident neurasthenia develops from the abnormal mental condition into which the injured person falls as a consequence of the accident, especially when this has affected the "noble parts" (the head, spine, region of the heart). The fear of permanent injury to health or economically, especially about the effect upon his ability to work and earn money, puts the patient into a state of chronic emotional unrest which in spite of undisturbed recovery from the material results of the accident, may develop into the severest neurasthenia. A particularly injurious influence is exercised here by preoccupation with regard to the question of damages, which the compensation and accident insurance laws of the different civilized states cause

the injured person. Fear that he will receive too little indemnity keeps him in continued tension; it more and more impresses itself upon his mind that high damages are due him, and there is anxiety lest this be cut down or withheld from him, either carelessly or malignantly, or that his troubles will be estimated too lightly.

The system of paying a monthly allowance customary in Germany, for instance, greatly encourages the formation of such "avaricious concepts," and indeed the continued "struggle for pension" furnishes ever fresh material for accident neurasthenia. Much more rational is our Swiss system of cash settlement, in which the injured person after being paid the sum awarded by agreement or through a decision of Court, is as greatly interested on the ground of health, as financially in regaining as soon as possible his full capacity to work. In what manner the estimation of what compensation should be paid to the traumatic neurasthenic is to be estimated, cannot be rigidly laid down. A first principle is, however, that a *permanent* impairment of working capacity must not be assumed upon the basis of a traumatic neurasthenia (naturally the most exact observation must have permitted the exclusion of symptoms organically caused). As an expert, I take the stand that according to the severity of the neurasthenia present (as to which the objective symptoms should be decisive), it must be considered that the disabilities should be estimated as likely to extend over from six months to at most three years. My after-histories, in agreement with those of *Nägeli* and other observers, prove the correctness of this view.

The closing portion of one of my reports, for example, reads as follows:

"The cure of the troubles is dependent upon the earliest removal possible of the question of compensation. As long as this is pending, in spite of all the efforts of the physician, the subject will be exposed to emotional disturbances which will furnish conditions favoring the continuation of the traumatic neurosis present. If, however, Mr. X realizes that the question of compensation is definitely settled, his troubles will gradually disappear and he will regain his ability to work.

"In consideration 1, of the kind and manner and of the intensity of the present troubles of the subject (vertigo, headache, hypochondriacal depression); 2, of his occupation as a carpenter (which requires frequent bending over and in which vertigo is very disturbing); and 3, of experiences acquired in similar cases, I would recommend to the Court a compensation upon the basis of the assumption of a diminution of earning capacity from 25 to 33 per cent. during 2 years. Though the reduction of working capacity is greater at present, it is nevertheless to be assumed that after definite adjudication, this will diminish progressively and at latest will have disappeared at the end of the second year."

Quite otherwise than in the acquired forms are the etiologico-pathogenetic relationships in constitutional neurasthenias. The degenerative factor manifests itself equally from the anamnesis and from the semiology, now in unmistakable intensity, now in more obscure forms, sometimes indeed, only as a suggestion. For neuropathic heredity, in contradistinction to the acquired cases, we seldom seek in vain, also it is usually furnished by numerous and severe

forms of disease in the ascendants or in the collateral branches of the family. These disease forms belong only in the minority of the cases, under the division of similar heredity. More frequently (and in the more severe cases) there are in the parents, brothers and sisters, not neurasthenic symptom-complexes, but more severe neuropathies, hysteria, psychoses, epilepsy, so that the "degenerative neurasthenic" in contradistinction to the victims of a reinforced pathological heredity (see Lecture VII, page 197) often appears to us as the relatively least injured member of an in general much more severely degenerated family. Often, also, separated still further from homomorphous heredity, we find common among the progenitors, affections injurious to the germ in general, as alcoholism, syphilis, tuberculosis, more rarely diabetes, perhaps also the uric acid diathesis, if we follow the statements of French and English authors, since with us the rarity of true gout does not permit the formation of a definite opinion in this direction. Finally, consanguinity of the parents is to be considered here.

There are now, however, also neurasthenias of the most severe degenerative character in which we can find no hereditary factor. In general we must expect to form a decisive conclusion as to the constitutional factors from the study of the patient himself. And hence we will take up now the previously unconsidered clinical peculiarities of these degenerative cases.

CONSTITUTIONAL NEURASTHENIAS

The three chief criteria of the cases falling under this head are: 1, the occasional presence of somatic stigmata of degeneration; 2, beginning of the disease manifestations in early or earliest childhood; 3, certain anomalies to be considered as psychic stigmata of degeneration.

The first point we have considered at length in our introduction to psychoneuroses (Lecture XXVII), and only need to refer to it to-day to refresh your memory. Only it must be expressly emphasized that such morphological anomalies, which are entirely foreign to acquired neurasthenia, are not very frequent also in the constitutional form.

Much more frequently, however, in any event in the majority of the cases, there is a history of the so-called congenital neurasthenia or that of childhood. We find that already in school, or even before this, in the nursery, the patient is characterized by abnormal irritability, motiveless outbursts of anger; that he now misanthropically and discontentedly isolates himself from contemporaries, again manifests a restless motor impulsion, that he suffers from stuttering, or for years, often until puberty, from nocturnal enuresis; that he has masturbated long before the age of puberty. Important, also, is the history that the patient as a child suffered from night terrors, those peculiar attacks of frightened waking out of sleep with momentary confusion and loud screaming, which even then, when they are referred to nasal polypi, adenoid vegetations of the pharynx or to intestinal worms, indicate an abnormal reflex excitability of the brain. Milder fright conditions are more frequent; fear of darkness, of the whistle of a locomotive, of being alone;

further peculiar habits, eating paper, forming senseless words, crying out on certain occasions, grimacing during serious situations, etc.

Over these manifold expressions, which it is often not at all easy to discover anamnestically, the psychic stigmata which so often as phobias and imperative conceptions dominate the clinical picture of constitutional neurasthenia in adults, cast their shadow.

As best known and perhaps most frequent paradigm of the phobias, *Westphal's* "agoraphobia" (fear of open spaces) is to be mentioned. This is an overpowering feeling of anxiety which always affects the patient when he has to traverse an open space. A feeling of anxiety, which expresses itself in the countenance, is accompanied by pallor and palpitation of the heart, the breaking out of a cold sweat, and the arrest of the salivary secretion. A feeling of anxiety in which the person affected has the sensation that he is about to sink through the ground, that his legs are paralyzed. A feeling of anxiety that in severe cases actually compels the individual to turn about, to creep along the houses, or to solicit the attendance of a passer-by, although he is entirely aware of the lack of motive and absurdity of his phobia. Endless is the list of other phobias which we meet as a frequent or occasional occurrence, in the picture of constitutional neurasthenia, and to which an endless number of foreign words have been applied; the gephyrophobia already described in the works of *Hippocrates*, that is the fear of passing over a bridge; siderodromophobia, the fear of traveling on a railroad train; claustrophobia, the fear of being pent up in a narrow space. Excessively variable are the special manifestations of the so-called "situation anxiety," which, for example, affects the barber when he is about to shave, the preacher when he is about to mount the pulpit, etc.; all manifestations which are strictly separable from the vague anxiety of the accidental neurasthenic.

Closely related to the conditions of anxiety are imperative conceptions (obsessions), from which the constitutional neurasthenics so often suffer. "Conceptions which do not arise by way of associations," so *Oppenheim* defines them, "but appear spontaneously and forcibly press into the circle of ideas, so that they cannot be banished from them although the individual considers them something foreign, not belonging to the mental ego." The most frequent paradigm of these disturbances is the "Folie du doute" ("doubting mania") studied by *Griesinger*, *Legrand du Saulle*, and *Falret* long before *Beard*.

This consists in questions which continually and without motive press upon the mind, as "Why have I said this and not that?" or doubts which cause even the most indifferent acts to take on a painful character and are often combined with anxious concepts, "Would it be better to take this journey or not?" "May I not be the victim of a railroad accident?" or "May not somebody break into the house during my absence?" "Did I really put a stamp on the letter just mailed?" "Have I not forgotten to shut the door of my house?" "Have I not forgotten to turn out the gas?" Such doubts appear compulsorily upon the most indifferent occasions (they can occur as episodes even within physiological limits) and are, besides, first disproportionately strong, and, second, so obstinate that they do not allow the patient to rest, although

(and partly because) there is complete insight for the senselessness and pathological character of these manifestations, a phenomenon which is easily differentiated from the general lack of decision of the accidentally neurasthenic.

More severe symptoms are arithmomania, the compulsion to count the windows of a house, the lanterns in a street, the flowers in the pattern of a carpet, onomatomania, the compulsion to call to mind certain forgotten proper names, the "délire du toucher," the obsession that a defilement or infection must be connected with every touching of a strange person or object, etc.

As to the psychical mechanism of the peculiar phenomena of the phobias and obsessions, we can with good conscience repeat "Ignoramus," since the attempt of *Sigmund Freud* to explain imperative conceptions as in general symbolical from certain "displaced" psychic complexes of erotic nature cannot be brought into accord with the facts; also I consider the sharp separation between an anxiety neurosis and a compulsion neurosis undertaken by him as impracticable.

Much more does it recommend itself to denominate both forms with their transitions just as frequently found, with *Janet* and *Raymond*, as "Psychasthenia." Now "psychic powerlessness" (and this is indeed caused by the giving way without resistance under hyper-quantivalent ideas in spite of complete insight), impresses a characteristic stamp upon the whole clinical picture. This does not indeed indicate that somatic signs of disease are absent—still they play a much smaller rôle than in acquired neurasthenia. It seems to me deserving of emphasis that pressure in the head and sleeplessness never reach a very high degree, indeed, can be entirely absent. Among the objective symptoms which we mentioned for the acquired form, only the increase of reflexes and the cardio-vascular lability usually manifest themselves with marked intensity, sometimes also vibrating tremor. Of other somatic phenomena the not altogether rare occurrence of "tics" (see Lecture IV, pages 67 and 72), indeed deserves mention.

The conceptions psychasthenia and constitutional neurasthenia would agree with one another if it were not that a considerable contingent of cases which in their history prove themselves certainly constitutional, are entirely wanting in the phobic and obsessive element, and approach the picture of accidental neurasthenia. Indeed, they then usually bear a chronic character, the time of their development cannot be precisely determined and they have a less stormy symptomatology. In spite of this, just these cases are to be considered much more serious and therapeutically much less influenceable—which of course applies also to the psychasthenias.

Differential Diagnosis

Now before we pass over to the so important treatment of neurasthenic conditions, it must not be neglected to point out the, unfortunately all too frequent, serious diagnostic and practical misconceptions which arise from the extension of the conception neurasthenia, to conditions which have with it nothing in common but a more or less extended semiological analogy. We should hold to the rule, only to make the diagnosis neurasthenia, by exclusion.

At first it must be considered that incipient phthisis, diabetes mellitus, chronic alcoholism, nicotinism, morphinism and cocainism may produce similar symptoms. The same remark applies to certain organic brain diseases whose "pseudo-neurasthenic" initial stage we have already described at length: dementia paralytica, and cerebral arteriosclerosis. Dementia præcox should also be thought of here; indeed, for example, it has been shown by *Mlle. Pascal* that of 75 patients sent to the Insane Asylum, Ville-Evrard, with dementia præcox (of both the simple as well of the paranoid, hebephrenic and catatonic varieties) not less than 32 had first been wrongly diagnosed and treated for neurasthenia, an error which may have serious results for the patient, his family and society. Against such serious errors only careful psychological investigation into the mental condition of the patient will protect us. It will be noticed that in the neurastheniform prodromal stage of dementia præcox—in contradistinction to true neurasthenia—emotional indifference is the basal element of the psychic syndrome,—the hypochondriac ideas are characterized by their variability and their trifling character, concern themselves preponderantly about the physical condition of the health and are emitted with little affective accompaniment (emotional dulling). Notice also, peculiarities, odd demeanor, motiveless laughing; negativism in the form of systematic opposition, and a hard-headedness alternating with childish docility and suggestibility; "psychographic disturbances," an excess of pretentious word-forms used by preference, bombastic and inflated style further appear. Quite as frequently the confusion of neurasthenia with cyclothymia, that is, with the "formes frustes" of circular melancholia occurs. This happens in patients usually of the female sex, who at more or less regular periods—for example, every Spring or every Fall, become relaxed, without energy, and inclined to tears, complain of dyspeptic troubles, pressure in the head and similar things, and usually suffer from marked agrypnia. As definite indications of their relationship to melancholia, besides the periodicity to be mentioned are, the regular occurrence of self-reproach and depressive ideas ("I cannot preside over my house, am a burden to my family! Oh! what a lazy and unenergetic person I am!" etc.), and that in the great majority of cases there is an alternation of morning exacerbations and evening remissions. That the formes frustes of *Basedow's* disease usually sail under the colors of neurasthenia cannot be denied; also one must be on his guard against confusing this last with beginning *Addison's* disease.

Treatment

The interest which of late has been specially directed to the psychoneuroses in general and to neurasthenia in particular, has fortunately redounded also to the advantage of the therapeutic side of the problem. Proceeding to discuss these matters I shall chiefly keep in mind those methods which can be carried out outside of a hospital or sanitarium; since most of you will meet your neurasthenics in private practice.

In every rational treatment of neurasthenic conditions the leading position must be assigned to those measures for which the word "psychotherapy" has been coined. That this expression has become quite a commonplace, must

be admitted. Nevertheless, it has one great advantage in that it shows us in striking manner, how great a change our therapeutic measures for psychoneurotic affections have experienced of late. Here the service of the path-finding work of *Paul Dubois* cannot be enough emphasized. Although even before him, consciously or unconsciously, every really good physician practiced mental treatment, and *Ottomar Rosenbach* is to be considered as his predecessor in this method of cure (the so-called "dialectic psychotherapy"), it is only through the efforts of *Dubois* that the recognition of how predominant a position is to be attributed to psychic influences in the circle of therapeutic agents has become generally realized, and how astonishing results it is permitted to obtain by its systematic application. Further, he has made it clear to us that psychotherapy has its technique and its rules, like all other methods, and that to be applied intelligently and systematically it presumes in the physician a certain makeup besides practice and experience. These requirements can never be replaced by theoretical study, also you should not expect from me more than some suggestions and indications toward developing your own psychotherapeutic system. According to the personality of the therapeutist his methods will have to bring into play the factors of educational influence, explanation of facts and authoritative intervention in quite varied combinations, to estimate whose relative value would be a sterile pedantry. Rather are they to be judged by the effects produced. That, however, the personality of the patient must have a decisive influence upon the choice of the psychotherapeutic method, is self-apparent; every word that the physician addresses to a neurasthenic exercises upon him a psychic action, and hence must be adapted to his peculiarities.

So, there are individuals who are entirely competent to realize that they must suffer throughout life from certain troubles, if it is only permitted to the physician to convince them that these symptoms do not place in question either their lives or their working capacity; other neurasthenics, on the contrary, cannot bring themselves into such a condition of stoicism, and by the direct communication of such a prognosis are reduced to despair and driven deeper into their psychoneurosis. It is not given to every one to react to the appeal to duty, altruistic feeling, etc., in the sense of correcting the egocentric view of the world peculiar to many neurasthenics. Occasionally we appeal directly to self-love—so doing must of course be carefully considered in advance—"If you do not carry out my recommendations and put aside the injurious factors pointed out to you, you are liable to become still worse." In short, a great measure of diplomatic skill is necessary, in properly estimating how the patient is best to be reached, corresponding to his psychological individuality. This insight into the nature of the patient we must endeavor to obtain, particularly from the manner in which he describes his troubles to us. Since this is usually done with extraordinary detail and verbosity, we have, as a rule, only too much time to become acquainted with the mentality of the individual. Naturally, as I have already emphasized in the last lecture, it must be our endeavor not to allow the history to be extended indefinitely, since we cannot devote all our time to a single patient. But we need not cut off a neurasthenic by saying to him, "What you wish to tell me further is unim-

portant; I know enough," since by so doing we would irreparably lose his confidence. On this account one must learn, by skillful introduction of questions, to lead the patient away from unimportant matters and to keep his descriptions within reasonable bounds. The neurasthenic must, however, gain the impression from the conduct of the physician while taking the history, that his complaints are sufficiently considered and are not taken "with a shrug of the shoulders." Also the physical examination must be careful and thorough, since in this way we procure a basis for convincing strength in the reassuring advice which is to be imparted to the patient.

Removal of the hypochondriacal ideas is important above everything else. To this end I do not hesitate, in the case of intelligent and educated patients, to support my word by clinical proof and explain to them, in a simple but scientific way, why their heart troubles are to be considered functional and cannot be organic, in what way their abnormal sensations are produced; with the hemoglobinometer I demonstrate that the suspected anemia is not present; by reading selected paragraphs from *Beard*, *Dubois*, etc., I show that phenomena which greatly frighten the patient do not represent anything but typical neurasthenic symptoms; that phobias and obsessions are not at all precursors of mental darkening, that many views spread through inferior sexual literature which hold the sexual neurasthenic under their ban, are fabulous, etc. A wide and thankful field for explanation and reassurance. In this short discussion of the nature and symptoms of neurasthenia, I would only advise you to carefully avoid one thing, the word "imagination," which almost always makes such patients restive, while "autosuggestion" can perhaps be risked.

With explanation to the patient of the nature and genesis of his troubles, the matter is indeed finished in only very few cases. Usually teaching self-control, consideration of the subjective disease symptoms as a "negligible quantity," training into a rational method of occupation, correction of exaggerated sensitiveness to the annoyances of life, etc., requires quite high qualities and psychotherapeutic skill on the part of the physician. It is usually specially difficult to talk a psychically impotent person back into his lost self-confidence, by far most troublesome, however (unfortunately often also most thankless), to influence anxious and imperative conditions through logic. Here, as there, it is in general to be recommended, at first to advise the patient as far as possible to avoid for a long time the "critical situations" (for example, cohabitation, crossing bridges, etc. Through this, the memory of former ill results and attacks of anxiety becomes gradually less marked, and the inhibitions which oppose themselves to rational psychotherapy are gradually reduced. Further, a great number of phobias and obsessions prove themselves accessible to psychic influence only after there has been a change of surroundings as radical as possible; hence, treatment in sanitariums and asylums comes frequently into question in such cases. For the rest, however, I certainly agree with *Edinger*, when he warns against being too ready to order institution treatment. The slighter cases of neurasthenia are really often better suited when the changes in surroundings are procured in a less radical manner, as in the form of a sojourn in the country, or a sea voyage, provided that, while so doing, enough new impressions are furnished for the mind to keep

it from the observation of self. Even the prescription of such a cure acts often encouragingly upon patients who had feared to be sent to a sanitarium. A disadvantage of a sanitarium is often that the neurasthenic patients mutually lament about their troubles to one another, which hardly acts in the sense of an "abstraction." In neurasthenic conditions shall psychotherapy push entirely into the background other methods of treatment (formerly incorrectly occupying the foreground), or has each of these its justification along with the other? In this question I would occupy an eclectic stand-point and would epitomize the matter as follows:

That the psychic abnormalities of the neurasthenic cannot be directly influenced by dietetic rules or by prescriptions is self-evident, and here psychotherapy in the direction which was sketched above has uncontested results. Nevertheless, in the state of mind of the accidental neurasthenic the primary is not always easily to be differentiated from the secondary, namely what is irritable weakness of the brain functions from what is the natural result of a frequently quite painful feeling of general loss of strength and prostration. The last, however, can be influenced by somatic measures of treatment. To the subjective disturbances in whose estimation the secondary ideogenic element is difficult to differentiate from the results of primary injury, belong headache and pain in the back, loss of sleep, loss of appetite, vertigo, etc. Here, also, psychotherapy belongs in the foreground; as further curative factors, however, physical and dietetic methods can be applied, and properly applied, they give excellent results. Only in the second place, however, should treatment of these subjective disturbances by drugs come into application, since in no other instance is there greater danger of the establishment by the superficial therapeutist of a "therapy of little symptoms." The following principles should prevail: "Limitation of the indications as much as possible—the eventual application of drugs is permissible only as a therapeutic compromise and by reason of opportunity, and in any case as rarely and as temporarily as possible." This applies namely, to the different "headache remedies" from the group of the antipyretics and for hypnotics proper.

The better one understands how to carry on the general treatment of neurasthenia, the more rarely will he have to resort to remedies of this class, if only temporarily, it should always be considered as a forced position. Not only does the objection to this convenient method of treatment with symptomatic drugs rest upon the danger of habit formation, but also in that it directly opposes a rational psychotherapy. Everything is in bringing the patient primarily to the conviction that all his individual complaints are not diseases in themselves, but represent the outcome of a unique abnormalization of the whole nervous system. If, however, we force these by too active therapeutic consideration into the foreground of the treatment, we make him doubtful as to our assurances that his "everlasting pain and misery so thousandfold" is to be cured from one point. It is somewhat different, however, with the manifold somatic visceral disturbances, which we have enumerated as objective symptoms of neurasthenia. These disease manifestations (for example, abnormal variability of the pulse and of the blood pressure, factitious urticaria, hyperidrosis, fine tremor, tachycardia, hyperacidity, etc.) do not

permit themselves to be absolutely subjected to a primordial mental alteration as in hysteria, in which the symptoms manifesting themselves in the territory of somatic or sympathetic nerves are brought to pass in a roundabout way through the conception. Rather is it here plainly the coördination of irritable weakness both in the psychic and in the physical sphere, so that in neurasthenia we can recognize a general neurosis in the fullest sense of the word. The somatic visceral disturbances of neurasthenia have hence a claim to a treatment to a certain degree autonomous, to be begun along with psychotherapy and based upon a rational empirical basis. The disappearance of these symptoms will be an indication of its efficacy, that is, a reduction of the reflex and mechanical irritability, and return to normal secretory conditions, the quieting of the heart, etc., and since we can accomplish these actually by physical, dietetic and medicinal agents, I cannot decline these methods. Presuming that with the removal of the causal factors, an accidental neurasthenia can be cured, a constitutional, decidedly improved, by psychotherapy alone, nevertheless, in my experience our task is greatly lightened in most cases by a combined procedure.

No drug is so often given to the neurasthenic as the alkaline bromides; to many physicians inclined to routine, ordering this drug follows almost as a matter of course the diagnosis neurasthenia. There is, however, no point in administering bromides to relaxed neurasthenics without any marked symptoms of excitation. At most a short course of bromide (with daily doses up to about 3 grms of KBr) can be occasionally recommended for such patients in whom irritative symptoms dominate the clinical picture, while weakness occupies the background; for example, in threatened predominance of conditions of anxiety, in disturbance of the always labile psychic equilibrium from some annoyance small or great. Very dilute solutions are best borne (10 grammes (℥iiss.) of the salt in 150 cc (℥v) of aq. menth. pip., of which a tablespoonful can be taken in a glass of water). If to this mixture 0.3 to 0.4 (gr iv to vi) codein phosphate is added, the sedative action is increased.

A further quite useful sedative is Indian hemp, in the prolonged administration of which very small doses suffice. It can also very well be combined with tonic remedies; for example, I have recommended the following combination under the name of pilulæ cannabinæ compositæ:

℞ Quinini sulphat ..... 1.0 (gr. xv)
Acid. arseniosi ..... 0.06—0.1 (gr. 1/10 to 1½)
Extract. cannabis ind. ..... 0.45 (gr. 6¾)
Extr. et pulv. rad. valerian q. s. ut f. pil. xxx.

S. One pill every evening.

A course of arsenic is sometimes above everything a curative factor in thin, chronically fatigued neurasthenics having little appetite. Large doses are practically never desirable; as a rule, in long administration a daily dose of 0.002 to 0.003 (gr 1/30 to 1/20) of arsenious acid or 0.2 to 0.3 (♏iii to ♏v) of Fowler's solution, is sufficient. The last, combined with tinc. nucis vom., is usually well borne. More convenient are the Asiastic pills, of which one can

be given daily after the evening meal (Acid. arsenios, 0.2; pip. nigri, 5.0; sach. alb., rad. alth. āā. 3.0, M. f. pil. No. C.). Where exceptionally a very sensitive stomach refuses the arsenic preparations, I administer them subcutaneously in the form of sodium cacodylate in (sterile ampoules of 0.05). Where there is definite anemia, iron is called for. Very popular are *Erb's* "Tonic Pills": Ferr. lactat., extract. cinchon., aquos, āā 4.0 (3i). Extract. nucis. vom., 1.0 (gr. xv); extract. gentian. q. s. ut f. pil. No. C. S. 2 pills t. i. d. after meals.

Where iron does not appear indicated, cinchona and nux vomica are ordered in the form of drops (tinct. nuc. vom., 5.0; tinct. cinchon. comp., 10.0; M. S. 30 drops t. i. d. before meals, in water). This prescription is useful in intestinal atony and other dyspeptic troubles.

A drug in which tonic and sedative properties are united is valerian, an indication for which is furnished particularly by the cardio-vascular disturbances. A systematic course of the infusion, prepared in the cold, has proved most efficient. Directions for its use will be found under the treatment of *Basedow's* disease (see Lecture XXIII, page 353). Where the valerian tea is refused, the extract can be substituted for it, for example: ℞. Extract. valerian. 10.0 (3iiss); extract. hyoscyam., zinc. oxid. pur., āā 5.0 (gr lxxv). M. Fiat Pil. No. C. S. 1 pill three or four times a day. Phosphoric medication, when it is indicated, namely in thin but full blooded neurasthenics, is to be carried out as recommended under *Basedow's* disease (under treatment with sodium phosphate, or calcium glycero-phosphate, there is often satisfactory gain in weight and improvement in the general condition), further, in neuroses of the stomach and, finally, in nervous irritative conditions of the sense organs, particularly in tinnitus aurium.

Also with regard to one further point we can refer to what was said under *Basedow's* disease in Lecture XXIII. The ovo-lacto-vegetarian régime can act exceedingly favorably in neurasthenics with marked circulatory lability; quite as satisfactory is sometimes the effect of such change of diet in nervous dyspepsia, in which the gastric anomalies of secretion are favorably influenced by the relative lack of irritation of the mild meatless, or meat-poor diet. Finally, this is the most harmless method of combating the chronic intestinal sluggishness so frequent in nervous people, particularly when an increase in the content of cellulose is provided for by the free use of fruit, Graham bread, etc., and in obstinate cases, ½ liter of warm physiological salt solution is given in the morning on an empty stomach. The intestinal fermentation and the distention by which the atonic intestine of the neurasthenic is so often troubled, are decidedly reduced. Where there is anemia, naturally in the regulation of diet its content in iron should be taken into consideration (spinach, carrots, yolk of egg, oats, asparagus, strawberries, Graham bread). In certain cases, particularly in cardio-vasomotor neurasthenia, alcohol is best entirely forbidden; under all conditions, however, it is only to be permitted in small quantities and well diluted. Strong coffee is to be forbidden, at most, that well diluted with milk is permitted. Tea appears to be less injurious, but also is to be permitted only in small amounts. With regard to tobacco, it is to be forbidden in the cardiac and vasomotor forms of neurasthenia; otherwise, however, we

should, as a rule, be satisfied with restricting its use within moderate bounds. Indeed, we should consider the agreeable feeling which it procures for the patient as an ally, from a psychotherapeutic standpoint, since it raises his spirits.

A simple dietetic prescription which well suits most patients with sense of pressure in the head and dizziness, is to allow them to take some slight refreshment every hour in the day so that the stomach is almost never quite empty.* It is astonishing how rapidly almost every one becomes accustomed to this régime and how advantageous it is found. In cases in which the head troubles mentioned above, together with weakness and prostration, make themselves apparent immediately after rising, breakfast had better be taken in bed about one hour before getting up. This simple dietary measure can be denominated "fractional overnutrition"; in ambulatory treatment and in slight cases, it can accomplish what is sought in severe cases by the "*Weir Mitchell* rest cure," which in any case can only rarely be carried out at home, and for which in general a sojourn in a hospital is necessary.† The marked taking on of flesh, which is attained in such cures by the gradual training to continually more frequent and continually more nutritious meals and aided by the large amount of physical rest, often acts as a marked sedative to the whole nervous system.

A certain comfortable laziness gradually comes over those subjected to this "cure," and the proverb "Plenus venter non studet libenter" gradually finds application in the matter of constantly studying himself and his woes by such a patient. Besides this, the isolation with which the *Weir Mitchell* cure is united, gradually prepares the way for psychotherapy.

The physical curative measures which may be of value to the neurasthenic can be only briefly mentioned. With warm full baths and half-baths, air-baths, hot foot-baths, alternating foot-baths and cool spongings, we can succeed wherever baths are found advantageous and we do not need complicated hydrotherapeutic apparatus. The foot-baths are particularly appropriate in combating the congestions and tinnitus aurium, the full-baths and half-baths for quieting the patient at night, the rubbings and the air-baths for general tonic treatment. With cold-water procedures and sun-baths for which the laity have such a great fondness we are more likely to do harm than to aid. Also the wide-spread idea that the practice of different athletic sports is a panacea for nervosity must be decidedly opposed. It is well to lay down for the patients, who seek recovery in the country, rational directions with regard to walks, periods of rest, reclining in the open air, etc., as otherwise we risk having them come back in a seriously exhausted condition. Finally, I would recommend to you to take to heart the reminder of *Beard:* "Every case of neurasthenia is a study in itself; no two cases are just alike. If two cases are treated alike from start to finish it is probable that one of them has been wrongly treated."

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* For example, milk, cakes, figs, prunes, chocolate tablets, etc.

† *Doctor Mitchell* always preferred carrying out the "rest cure" in special private houses, away from the hospital.—*Translator.*

LECTURE XXIX

The Psychoneuroses

C. Hysteria

In contradistinction to neurasthenia, which, as we saw, has only been isolated nosologically and named in recent times, in its sister neurosis hysteria, we have to do with a very ancient disease conception. That the striking symptoms of the disease were brought into connection with supposed disturbances of the functions of the uterus, has, as is known, found expression in the name for the affection (ὑστέρα = the womb). *Hippocrates* regarded the hysterical phenomena to a certain extent as abstinence symptoms on the part of the organ withdrawn from its natural function; the disease appeared most frequently in old maids or in women early widowed, to whom the advice to have relations with men as soon as possible must be given; "since if they become pregnant, they will be cured." Particularly fantastic is the pathogenetic conception attributed to *Timæus,* who imagined that the unsatisfied uterus wandered restlessly about, like a rutting animal ("animal liberorum procreandorum appetens"="an animal desirous of begetting children"), throughout the body, and in this way set up the hysterical symptoms. That this grotesque view has been retained to this day among the country people of our neighboring Alsace, deserves mention: we occasionally hear from a peasant woman troubled about her daughter, the expression "her womb is trying to get out of her throat."

Further, you know indeed, that among the laity of all classes of society (naturally also in romance literature), to-day still, the conceptions "hysterical" and "man-crazy" are nearly synonymous. No wonder then, that even from the chain of thought of physicians, the teachings of *Hippocrates* only disappeared in the course of the eighteenth century. For these the equally incorrect view that hysteria and hypochondria (this last term we must to-day replace by "neurasthenia") were the same disease was substituted; only the first was the special privilege of the female, the last, that of the male sex. It is not the least service of *Charcot* that he made clear that hysteria and neurasthenia are different conditions and that each may affect both sexes. As to hysteria indeed, its great preponderance in the female sex was recognized by *Charcot,* and statistics of later authors give this as 85 to 90 per cent., but the determining factor for this predisposition of women is, as our later discussion of the nature and causes of hysteria will show, not the female genital organs, but the female psyche, in which, in comparison to the male, a preponderance of phantasy and conceptional life, as well as diminution in the power of judgment and of critical inspection, are evident Also, men who become hysterical are throughout of the so-called "feminine natures" (which, as is

understood, is meant only in the psychic sense and may be combined with the most virile physical make-up). In analogous manner the predisposition in childhood as well as that of certain peoples (South Europeans, Slavs, Jews) is to be explained.

Only as a curiosity does the theory of *W. A. Freund* introduced not so long ago, according to which a causal connection between hysterical phenomena and gynecological diseases—for example, contraction of the broad ligaments—exists (in the sense of a reflex neurosis) need mention. This development of a one-sided specialistic view has unfortunately led to the removal of the pelvic organs in hysterical women, by which the psychoneurosis was naturally not extirpated. To-day, fortunately, the operative era in the treatment of hysteria can be considered as definitely closed.

Symptomatology

As we did in the case of neurasthenia, in hysteria we will first describe the different disease manifestations, mainly in their most important clinical features, only after this to enter upon the more difficult task of an etiological pathogenetic consideration. The symptomatological description of hysteria is made easier for us from the fact that a portion of the symptoms is common to both psychoneuroses; this concerns namely, such phenomena in which exaggerated self-observation plays a mediating rôle as we have sketched it in Lecture XXVII. Still, the intensity of these disturbances, corresponding to the incomparably greater auto-suggestibility of hysterics, is usually much more marked; and further from their increased auto-suggestibility there are also qualitative differences: since only upon a basis of hysteria, never upon that of neurasthenia, can paralyses, contractures and anesthesias (of sensible or sensory nature) occur. Special diagnostic importance is finally to be attached to the "hysterical character" and to the special psychic disturbances of hysterics. These anomalies manifest, however, such intimate connections with the nature of the disease that we will first study them along with the pathogenetic relations. To bring some order into the striking multiplicity of the hysterical phenomena, it is advisable to subject the permanent symptoms, (stigmata) the hysterical attacks, paralyses and contractures, to a separate description.

1. PERMANENT SYMPTOMS ("HYSTERICAL STIGMATA")

a. Sensible and Sensory Symptoms

The majority of hysterics complain of pain of one sort or another. Individually very variable in localization, they usually differentiate themselves from pains as they occur in "hyper-algesic neurasthenia" through their greater intensity, or let us say rather, through the greater activity of the expressions of pain (complaining, crying, etc.). Particularly frequent are hysterical pains in the back, also hysterical headaches, which last, as a rule, are not described as pressure in the head only, but as a throbbing, burning or boring pain, which is felt by the majority of patients superficially, that is, in the scalp, and not

inside the skull. Usually there is with this, marked hyperesthesia of the hairy region, pressure or even touching the scalp, a slight pulling on the hair, produce lively expressions of pain; percussion is described as "actual torture." Very frequently headache and hyperesthesia center themselves in circumscribed regions over the top of the skull (see Fig. 110); the simile of a nail driven into one of these areas, occasionally used by hysterical patients, has led to the denomination of this symptom "Clavus hystericus." There is also a hysterical pain in the face which is differentiated from neuralgic prosopalgia, above everything by the continuity and diffuse nature of the pain and in contradistinction to that, is very often bilateral. The greatest intensity of pain is usually located in the temporal region; in many cases the pain occurs day after day at some definite time—habit pains (*Brissaud*). Analogues of the clavus hystericus, that is circumscribed sensitiveness to pressure, often also spontaneously painful areas, the so-called "topalgias," occur on certain portions of the trunk (Fig. 110) with a certain predilection. By the *Charcot* school so much industry was expended upon the study of these "mammary," "sternal," "ovarian" points, etc., that one cannot escape the impression that just this searching for hyperesthesia over these "classical" areas, then as to-day, frequently unintentionally suggested to the patient the phenomena sought. Nevertheless, just the possibility of conjuring up in this fashion hyperesthesias and pains, is a clinically very useful symptom of hysteria; further, the examination of healthy persons shows that the apex of the skull, as well as the other areas of predilection for topalgias often are somewhat more sensitive to pressure than the rest of the body surface, so that perhaps the physiologically caused local predisposition may also come into play. That the so-called "ovarian point" has nothing to do with the ovaries is shown by the fact that this symptom also occurs in male hysterics. Since occasionally by the production of pain on pressure over the spots of circumscribed hyperesthesia, hysterical attacks may be set up, on the other hand, however, those already in progress may be cut short; such areas have been denominated "hysterogenic" and "hysterofrenatory" zones.

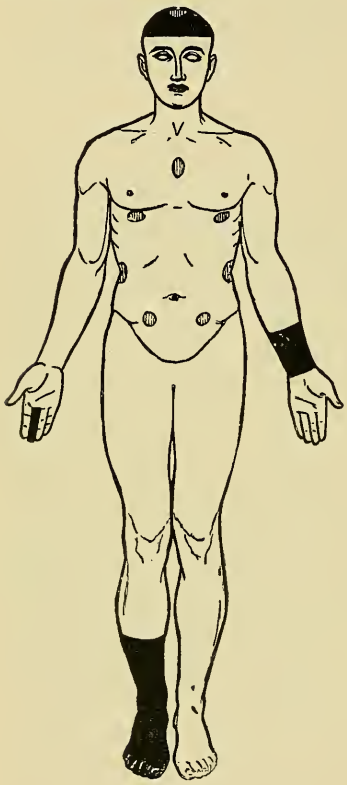

Fig. 110.

Typical Localizations of Hysterical Topalgias (Shaded) and Anesthesias (Black).

Diagnostic difficulties are presented sometimes by hysterical arthralgias, as they may localize themselves in joints to which the attention of the patient has been directed by some sort of material anomaly, for instance, genu

valgum, crackling of the joint surfaces, etc. Often, indeed, the surgical or roentgenological findings are entirely normal, or the hysterical nature of the pains is clear from their localization at an anatomically indifferent point of the joint region. For the rest, however, the diagnosis can only be made after long observation. Of important indication is particularly the disappearance of joint pains when the attention is distracted—which also applies to the other hysterical "psychalgias." These last are, however, not always of such circumscribed nature as in the examples already mentioned. They can affect the whole body (pantalgia), or more rarely, one entire half of the body (hemialgia). When the patient on account of hysterical pains which occur or are increased by every movement, is condemned to the most absolute helplessness, we speak with *Möbius* of "Akinesia algera"; where every touch of the skin calls forth pain, with *Pitres*, of "Haphalgesia."

Of greater diagnostic importance than pains and hyperesthesias are the hypesthesias and anesthesias of the hysterical. Indeed, as already emphasized, just through the occurrence of such disturbances which are foreign to neurasthenia, hysteria assumes a special clinical position in the circle of psychoneurotic conditions.

What I remarked when speaking of hysterogenic zones, about the suggestive production of hysterical symptoms by the physician applies here also and there is no doubt that hysterical anesthesias can be provoked by the examiner in the patient intentionally or not, that further, by repeated examinations (which hence are to be avoided as far as possible as soon as the hysterical nature of the disease is certain) they usually increase in intensity and extent. On this account never test the sensibility in a patient suspected to be hysterical in the way of asking him, "Do you feel the prick equally plainly upon both sides?" or "Is there any place where you do not feel the prick?" This is as if you had in mind directing his suggestibility toward the test. If you wish to rule this out as far as possible I would advise you to avoid all questioning during the examination, since the patient (not before examined) usually spontaneously remarks: "In this location I have not felt the prick," or when upon application of a strong faradic current to the right hand he expresses pain, to the left, however, he remains passive, then you know at least that you have not exercised any foreign suggestion and only autosuggestion remains in question.

That suggestion in general often plays the most important rôle in the production of hysterical anesthesias is proved by the fact that such disturbances of sensibility can be cured by suggestion, and indeed, may be changed topographically thereby (which is called transference). Why, however, the suggestibility of the hysteric manifests itself in circumscribed loss, or reduction of sensation (and besides this in quite characteristic distribution) remains a puzzle. While the generally diffused conception of motor paralysis permits us, in almost every hysterically paralyzed patient, the presumption that he has already seen paralyzed persons, or at least has heard of them, with regard to the anesthesias, similar relations are not to be assumed, hence observations of suddenly appearing hysterical hemianesthesias as have been published by

Stierlin, myself and other authors, in previously healthy individuals, as a result of severe fright (railroad accidents, earthquakes, etc.), compel us to assume that some organic regularities in the apparatus of apperception supply a ready-made pattern through which the psychogenic disturbance receives its clinical stamp.

The hysterical disturbances of sensibility vary greatly in intensity from case to case and there are all possible transitions from slight hypesthesia to complete anesthesia. Still, in contradistinction to the anesthesias caused by organic nervous diseases, the paradoxical fact is noticeable that the movements of extremities which upon testing sensibility show themselves totally insensible, occur in entirely normal, coördinated and undisturbed fashion, that with an anesthetic hand the patient can write, play the piano, etc. Also the tendon reflexes of the region are usually not diminished, indeed, they are often increased as in neurasthenia, only the mucous membrane reflexes—pharyngeal reflex, conjunctival reflex—are in hysterics relatively often very weak, or indeed, absent. Sometimes upon pinching or pricking the anesthetic cheek, dilatation of the pupil occurs. Sometimes the different qualities of sensation are affected to a different degree, only pain sense may be noticeably disturbed. An important characteristic of the hysterical disturbances of sensibility, their suggestive influenceability ("Pithiatism" according to *Babinski*) naturally can only be tested with considerable reserve. Attempts at "transference" for example, in which through any hocus-pocus, an anesthesia is transferred from one side to the other, should be directly forbidden, since they distinctly increase the morbid suggestibility. As to the topography of hysterical disturbances of sensibility, we can distinguish 1, universal anesthesia (excessively rare); 2, hemianesthesia; 3, circular or geometrical anesthesias; 4, insular or disseminated anesthesias. Typical for hysterical hemianesthesia, which usually affects also the mucous membranes, is in contradistinction to the organic form, its sharp limitation to the middle line. Also, circular anesthesias are separated from the normally feeling skin areas, by very sharp lines which usually run at right angles to the long axis of the extremity—so-called "amputation lines" (see Fig. 110); we find instead of peripheral or radicular areas, cap-like, sleeve-like, sock-like, glove-like, finger-like, anesthesias. Of organic diseases only lepra sometimes gives similar pictures. That between the different topographical types of hysterical anesthesia there are transitions I need not specially emphasize. Experience teaches that hysterical anesthesias more frequently occur upon the left side. Perhaps this coincides with that, as *van Biervliet* has shown, in healthy people there is usually a slight difference in the sensibility of the skin unfavorable to the left side, when tested by delicate methods.

Very frequent is the combination of hysterical disturbances of general sensibility, with those of the sense organs in which there are naturally phenomena analogous in principal. Among these stands first in importance, concentric limitation of the visual field, in confirming which with the perimeter, we notice in many cases that the visual field, in the course of the investigation, becomes constantly smaller; also that the mutual relations of the color fields have experienced a change from the normal. Namely, in healthy people, the

visual field for green is smallest, somewhat larger is that for red, still larger the area for yellow, particularly, however, that for blue. In hysterical dyschromatopsia (we speak of *Förster's* type of alteration) on the other hand, the limits for red more or less overlap those for blue. In spite of the most pronounced hysterical narrowing of the visual field the patients can usually move about with entire safety without striking against anything and have no suspicion of the anomaly appearing upon perimetric examination. Occasionally, however, there is a hysterical amblyopia proper in which, besides the visual field, acuteness of vision, color sense and light sense are found diminished. Hysterical blindness is, on the other hand, quite rare; like hysterical amblyopia, it occurs with intact light reflex of the pupils. A very great rarity is monocular polyopia. The hysterical disturbances of vision occur, as a rule, upon both sides, still, they are generally most marked upon the side on which general sensibility is most affected. Next to vision, taste is most frequently affected in hysteria; now it is perversions of taste in which bitter or otherwise disagreeable tasting substances are found agreeable, and eaten by preference (hysterical "longings," "pica") again, ageusia for single or for all qualities of taste. This ageusia can occur uni- or bi-laterally. Less frequent is hysterical anosmia, indifferently unilateral or bilateral, still rarer hysterical dullness of hearing which, as a rule, occurring unilaterally, can sometimes increase to complete deafness. The different tests for hearing (see Lecture II) give the same results as in nerve deafness ("Rinne"), positive, etc., but the subjective ear noises, so frequent in organic diseases of the auditory nerve apparatus, are almost always absent; further, the patient, deaf on one side, usually in conversation conducts himself like a healthy person, while in unilateral organic hypacusis of any considerable degree, we nearly always notice that the patient attempts to turn his better ear toward us. Sometimes hysterical deafness is an elective one, in that, for example, whispers are heard while there is complete deafness for the ticking of a watch, etc.

b. Vasomotor Symptoms

If we disregard the not infrequent combination of hysteria with different vasomotor neuroses (see Lecture XXV), as well as the disturbances of vascular innervation frequent also in neurasthenia (dermographism,* factitious urticaria, fleeting erythema, "dead finger," etc.), the vasomotor anomalies occurring in the region of hysterical anesthesias are especially to be mentioned. The absence of bleeding on deep stabbing with a needle has a peculiar importance in the history of civilization. The famous "Malleus maleficarum" was accounted one of the surest "witch signs" and it has hence contributed to the delivery of innumerable victims to the stake.

Sometimes there appears also about the stab a circumscribed œdema.

Further, in hysteria there is a not entirely rare, firm swelling, not pitting

* In hysterics sometimes more complicated forms of dermographism occur; for example, in one of my patients, upon stroking the skin with the finger, there appeared one red between two white streaks.

upon pressure, combined with intense cyanosis and coldness of the skin over the anesthetic or paralyzed hand which has been called "blue œdema." The circumstance that in persons especially susceptible to hypnosis it has been possible to suggest the appearance of skin vesicles and sugillations, suggest the possibility that trophic skin disturbances and extravasations of blood may occur upon the basis of hysteria. Still, it is certain that the impressive majority (if not indeed all) the cases described as "hysterical pemphigus," "hysterical ulceration," "hysterical hemorrhages," etc., have been produced by self-injury, a point to which, when speaking of the hysterical psyche, we will come back again. Also most cases of the so-called "hysterical fever" are to be referred to simulation (warming or shaking the thermometer), further, diagnostic errors—failure to recognize a latent tuberculous focus, etc.—often occur. On the other hand there are known a number of undoubted cases, in which the temperatures were taken by the physician himself in the rectum or vagina and the eventuality of an organic cause could be excluded. These are chiefly of temperature disturbances which occurred in hysterical seizures or at the time of the menses, some reaching quite high degrees (up to 42° C.=106° F.), but were not accompanied either by corresponding alterations of pulse and respiration, or by the urinary changes characteristic for fever. On this account the name "hysterical hyperthermia" is decidedly to be preferred. Palpitation of the heart and rapidity of the pulse occur in hysteria much less frequently than in neurasthenia.

c. Motor Symptoms

Hysterical tremor can be unilateral or bilateral, and may affect the upper or the lower limbs. In *Basle* as the "shaking disease" it excited great wonder some years ago: A hysterical school-girl had in this way exerted so striking an effect upon the phantasy of her school-mates that the tremor gradually spread from child to child. In this manner two hysterical school epidemics occurred, which were only stamped out by strictly enforced isolation. For the rest, we observe hysterical tremor with special frequency among "traumatic hysterics." It is usually in a great measure dependent upon attention, much more so than is the case in the tremor of neurasthenics or even in tremor organically caused. Only in a minority of cases is it the fine vibrating tremor mentioned in Lecture XXVII, usually it is of greater amplitude and moderate rapidity (5 to 7 oscillations a second): Its character can, however, be subject to frequent change ("polymorphous tremor"). Further, occasionally the most varied dyskinesias can be "imitated" by hysteria, in which long observation, careful taking of the history and accurate study of the suggestive influenceability is needed to determine the hysterical nature of the disease. Here belong among other things the disease pictures which we have already mentioned in Lecture V as "Chorea Major" and "*Bergerons*' Electric Chorea."

The frequent coincidence of hysteria and local spasms as well as tics has been alluded to when speaking of the dyskinesias; I would mention particularly,

œsophagism, pharyngism,* yawning, sneezing, snoring spasms, facial tic, etc. (see pages 67 and 72).

Anesthetic limbs upon being tested with the dynamometer usually show a diminution of the gross strength with very rapid fatigability, more rarely, what has been described by *Charcot* as the "contracture diathesis." Massage, faradization or binding the affected extremity has as its result a contracture which only ceases again after some seconds or minutes.

d. Visceral Disturbances

On the part of the digestive organs practically the most important, since in severe cases they lead to marked inanition and cachexia, are hysterical anorexia and hysterical vomiting. In the last, the food is usually vomited immediately after taking; also independent of taking food, there is bringing up of masses of mucus. Very often in hysteria, in contradistinction to neurasthenia, vomiting is the only anomaly on the part of the stomach and the feeling of pressure in the epigastrium, eructations and the other symptoms of nervous dyspepsia are absent. That only an exact examination of the stomach with exclusion of an organic affection permits us to diagnose the vomiting as hysterical is self-evident. Very typical of hysteria are the cases in which patients regularly react with vomiting to some definite situation (*e.g.*, coitus), upon seeing certain colors, hearing certain noises, etc., or where they are able to vomit "at command." Meteorism, the distention of the intestines by gas, reaches on an average much higher degrees than in neurasthenics; in hysterics, as a rule, the swallowing of air (aerophagia) plays a part in this. As curiosities may be mentioned here the cases of "hysterical pregnancy," in which women not pregnant but filled with longing for a child, have shown along with a meteoristic distention of the abdomen, cessation of the menses, swelling of the breasts, the appearance of colostrum, morning sickness, etc., in short, the most varied signs of pregnancy, and only upon the supposed beginning of labor, was the actual nature of the case made evident. Obstinate constipation is particularly frequent in hysterics; also the mucous colic already mentioned is often a symptom of hysteria. On the part of the uropoetic apparatus the most important hysterical anomalies are oliguria which usually (to a certain extent vicariously), goes hand in hand with hysterical vomiting,† and polyuria (up to 12 liters a day and more); due to polydipsia. Particularly frequent are genital disturbances; almost all female hysterics suffer from nervous symptoms at menstruation; sexual frigidity is frequent, with which are contrasted an exalted psychic eroticism and a resulting "desire for excitement." On the other hand sexual hyperesthesia occurs frequently. It leads often to a vaginismus hindering cohabitation, or the introitus directly represents a hysterogenic

* Globus hystericus, with which we will become acquainted as an initial symptom of hysterical convulsive attacks, which, however, occurs in many patients without relation to such attacks, is probably a variety of pharyngism specially colored by accompanying paresthesias.

† In the vomited matter, even in cases in which it could not be assumed that the patient had swallowed urine, urea has been frequently recognized.

zone. Many hysterical men, in spite of erotic monomania, are impotent or of feeble potence. This explains in part the sexual aberrations in such individuals, while in other perverse hysterics the "psychopathia sexualis" (homosexuality, sadism, fetishism, etc.) must be considered as a primary psychic stigma of degeneration.

2. HYSTERICAL SEIZURES

When in the 70's and 80's of the last century through the brilliant descriptions of *Charcot* and his pupil *Richet*, the interest of the whole medical world in hysteria was aroused anew, for a long time the study of the so-called attacks of "grand hysteria" occupied everywhere the foreground. To this circumstance it is proper to attribute the fact that at that period these "classical" paroxysms were much more frequently observed than now. At that time the great rôle which suggestion, imitation and other psychogenic factors play in the disease picture of hysteria, in the great enthusiasm for the work of symptomatological detail, was entirely overlooked, so that patients with hysterical attacks were placed in a common ward with other psychoneurotics, were demonstrated clinically as "show cases," and the physicians discussed in the presence of their patients with imprudent naïveté the picture of these paroxysms, which besides this, through newspaper and literary elaborations, though distorted, reached a more extended audience. To-day, however, such cases when they occur are isolated as far as possible; in the interest of the patient we avoid clinical demonstration, and otherwise withhold all concern in the attack which may act suggestively. So it has come about that the "grand hysterical attack," as *Charcot* and *Richet* described it, now occurs quite rarely; while in Charcot's time, at the *Salpêtrière*, day by day such paroxysms occurred in different patients. During my stay at this giant hospital in 1904-1905, I was not able to study one single case of major hysteria, and still, if I may so express it, they had "learned" their attacks under *Charcot*.

When now I lay before you the theatrical picture of this most peculiar manifestation of hysteria, I would not have the differentiation of the four stages of the "grand attack" understood in the schematic sense which *Charcot's* school gave to it. The symptomatology of the attack, indeed, is subject to considerable variation from patient to patient; nevertheless, there come under observation from time to time cases which with some good will can be denominated typical and regular. In such a case there appear at first certain disturbances, which in analogy with the epileptic paroxysms, may be called the "hysterical aura": particularly typical is the feeling of a ball arising from the abdomen or from the epigastrium up into the throat ("globus hystericus"), combined with great anxiety, palpitation of the heart, tinnitus aurium, dizziness, darkening of the visual field, etc. Then begins the first stage of the attack proper, the epileptiform, characterized by general tonic and clonic convulsions. As a difference from the true epileptic attacks, the patients do not usually fall down, but sink down and do not suffer any injuries; also the piercing initial scream of the epileptic is wanting and the face does not become cyanotic. After some time the epileptiform stage passes over into that of "contortions and grand movements" or of "clownism"; the patient makes

faces, throws the arms about, throws herself around; like the "bridge" customary in wrestling, the body rests only upon the occiput and the heels ("arc de cercle"), flings her legs about, stamps, rolls about her long axis, produces by tension of the neck muscles the "hysterical swollen neck" ("Blähals"), makes movements with the pelvis, etc. There follows now the period of "passionate attitudes" in which, apparently under the influence of dream-like alterations of consciousness, the patient mimics certain emotions, for example, fear, erotic rapture, religious ecstasy, deep grief, etc. Sometimes these conditions find expression simultaneously also by screaming, moaning, declarations of love, lascivious grunting, singing, praying, etc. Without sharp delimitation this phase of the grand attack passes over into the "terminal delirium," in which the mimicry ceases and there is chiefly vocal expression of hallucinatory perceptions (*e.g.*, visions of animals). A "major" hysterical seizure lasts on the average from a quarter to half an hour. After the attack, the patient sometimes remains for a time in a cataleptic condition, usually, however, she immediately gets up, and in spite of the great motor efforts which she has made, shows no fatigue worth mentioning. Sometimes, however, the attack leaves behind hysterical paralyses or contractures, later to be described.

Much more frequent than the major attacks, are the so-called minor attacks, whose manifestations may be quite various. Usually they are epileptiform, more rarely only the phase of contortions is assumed ("demoniacal attacks"). Other seizures occur only as loss of consciousness with slight muscular tension ("syncopal attacks"), or a sudden falling asleep ("lethargic attacks"), or as catalepsy, in which the patient with consciousness fully retained, passes into a condition of complete immobility. Not at all infrequently we observe the so-called "affect and respiratory spasms," that is, laughing spasm, weeping spasm, screaming spasm, tachynöic attacks (in which the frequency of respiration may increase to 180 per minute—figures which in general only occur in hysteria, and which hence may be considered as pathognomonic); further, attacks of "hysterical barking cough," particularly unpleasant for the surrounders. As "psychic equivalents" of the hysterical convulsive attack, may be mentioned attacks of sleep-walking (somnambulism), *Ganser's* symptom ("symptom of approximate answers"), that is, incorrect answers to the simplest questions, although the patient is apparently clear mentally,* "ecmnesia," by which expression is meant psychic paroxysms in which the patients reproduce events of their childhood in word and gesture.

In the epileptiform attacks under certain conditions a differentiation from the morbus sacer may be quite difficult, usually, however, on the basis of the criteria which I have collected in the following table, the differential diagnosis can be made without great difficulty.

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* Characteristic of *Ganser's* symptom is the fact that while the answers of the patient seem incorrect and inappropriate, they nevertheless have a certain connection with the matter in hand.—(*Translator.*)

	Hysteria	Epilepsy
Etiology	Often after some definite causation (rage, fright, etc., provocable by suggestion)	Usually without apparent causation; never provocable by suggestion
Sensory aura	Usually absent	Often present
Initial cry	Usually absent	Often present
Fall	Gradual sinking down; the patient almost never suffers injury	Sudden falling; injuries frequent
Biting the tongue	Very rare	Very frequent
Color of the face	Usually not markedly altered	Initial paling, then marked cyanosis
Discharge of urine	Almost never	Very frequent
Pupils	Only in very rare cases reactionless	Dilated and rigid
Consciousness	Not entirely lost; the patient reacts to tickling the mucous membrane of the nose, sprinkling with cold water, etc.	Totally lost
Duration	Often one half hour to an hour	Apart from status epilepticus, only minutes
Terminal sleep	Almost always absent, sometimes sudden coming to	Present
Possibility of being cut short	Frequently can be (hystero-frenatory zones, suggestive influence, external irritants, etc.)	Absent

3. HYSTERICAL PARALYSES

Hysterical paralyses usually appear suddenly either in connection with an attack of hysterical convulsions, or independent of this, in consequence of an emotion—fright, anxiety, rage, grief, etc. Also the physical traumata with which hysterical paralyses are relatively frequently connected, act only through the medium of the affect accompanying them, but have on the other hand in so far pathogenetic importance, since they usually determine the localization of the paralyses, in that this affects the part exposed to violence. The trauma can be very slight, even purely imaginary: so, in a case of my observation a blow directed at the patient was caught upon his open umbrella and did not touch the paralyzed arm at all. After explanation to the patient of the unreality of the supposed trauma, the severe paralysis rapidly recovered.

Sometimes after the trauma a certain time elapses before the paralysis appears: in such cases the conception that a paralysis may result gains the upper hand only after the accident and leads by autosuggestion to the elimination of the innervations in question. This method in which the patient "hatches out" his paralysis is not at all infrequent in "accident hysteria" (which also may manifest itself by anesthesia, spasms, contractures, etc.). The disease producing action of definite nosological conceptional complexes is here favored by the factors which we have already considered under accident neurasthenia

(page 424, etc.). We hence speak also of "Pension hysteria." Hysterical paralyses are usually flaccid, more rarely spastic, and belong now to the monoplegic, again to the hemiplegic or paraplegic type. They may represent transitory, fleeting phenomena (this applies particularly to post-paroxysmal paralyses which *Gendrin* hence considers as exhaustion paralyses), on the other hand, however, are marked by great obstinacy. The combination with hypesthesias or anesthesias of corresponding topography is very frequent. Hysterical paralyses when they persist very long can lead to a definite reduction in size of the affected limbs, still, this is only an atrophy from disuse which is never accompanied by reaction of degeneration or change of the superficial outlines.

The reflexes are never lost in the flaccid form; in the spastic form they are usually somewhat exaggerated, but not greatly so. The phenomena of *Babinski*, *Oppenheim* and *Mendel-Bechterew* are always absent as is also true ankle clonus, while we occasionally find the "pseudo-ankle clonus" already described. *Oppenheim* has further pointed out that the examiner who holds a hysterically paralyzed extremity for some time in his hand sometimes plainly perceives occasional innervation impulses in its muscles and that such a limb, when passively held in a certain position, then suddenly let loose, is able to maintain for some time a position, which is only explainable by the activity of the otherwise paralyzed muscles. Hysterical hemiplegia almost always avoids the facial and hypo-glossus. The gait differs markedly from that of cerebral hemiplegia, in that the paralyzed leg is not circumducted, but is simply dragged along, the sole or the heel scraping the ground. If the patient uses a crutch, he particularly does not bring his leg into contact with the floor. In hysterical paraplegias the functions of bladder and rectum are almost always intact; in all hysterical paralyses tropho-vasomotor disturbances are much rarer and in every case less intense than in the organic. Finally, it is pathogenetically as well as diagnostically important that only the conscious voluntary movements are suspended. In alcoholic and chloroform intoxications, on the contrary, there is lively gesticulation.

A very characteristic syndrome is the so-called "astasia-abasia," in which the patient in bed can execute every movement with normal strength and completeness, but, on the contrary, upon attempting to walk or to stand, simply sinks down (different from this symptom-complex first described by *Jaccoud*, is *Mingazzini's* "Stasobasophobia," an emotionally caused psychasthenic disturbance of gait nearly related to agoraphobia).

A frequent form of hysterical paralysis is hysterical aphonia, usually the immediate effect of a fright, a particularly obstinate symptom, tending to recur. The patients speak only in whispers; in it, as laryngoscopic examination shows, the vocal chords are not at all, or only imperfectly approximated. Coughing and clearing the throat usually occur noisily, however. Rare, but almost pathognomonic of hysteria is suddenly appearing mutism, in which, in contradistinction to aphasia, no sound or portion of a word can be produced, while mimic movements remain quite active. Excessively rare is hysterical ptosis, which, according to *Oppenheim*, is to be distinguished from that organically caused, by the fact that the contraction of the frontal muscle observed in this last is absent; it should not be confused with hysterical pseudo-ptosis

which occurs from spasmodic closure of the eyes, also introduces us to the hysterical contractures.

4. HYSTERICAL CONTRACTURES

In these contractures there is permanent fixation of a limb in a definite position by the tonic contraction of certain muscles. They develop in contradistinction to the hysterical paralyses by degrees in the majority of cases, though usually in connection with the exciting causes mentioned under those paralyses. Upon distraction of the attention, the contracture diminishes (in sleep it usually disappears), while, on the contrary, the manipulation by the examiner of the contractured limb plainly increases the muscular tension. In very long duration of the hysterical contracture there appear, however, as in mechanically caused fixation in a definite position, material alterations in the muscles, the ligaments, etc., which are responsible for the fact that sometimes even upon cure of the psychogenic anomaly a complete restitutio ad integrum is not attainable. In contradistinction to the contracture after organic diseases of the pyramidal tracts, in hysterical contractures (they also assume by preference in the arm, the type of "flexion," in the leg that of "extension"), the reflexes are not exaggerated and the other accompanying phenomena of the spastic symptom-complex are absent. Also upon passive shortening of the distance between the origin and the insertion of the contracted muscles a relaxation does not occur. Along with the contractures of the extremities and the pseudo-ptosis already mentioned as relatively frequent in hysteria, there are to be named contractures of the neck muscles (see in Lecture IV, under Tonic Local Spasms, Torticollis, Retrocollis, etc.) and glosso-labial hemispasm which draws the lips and tongue laterally, in a grotesque fashion.

Psychology and Pathogenesis

Gentlemen! Many physicians, when asked their position upon the question as to the nature of this peculiar psychoneurosis whose semiological study we have now finished, confess still to-day the standpoint of *Laségue*, "The definition of hysteria has never been given and never will be." This convenient axiom we can only accept in the sense that it is not possible to include the varied symptomatology of this "protean neurosis," the manifoldness of its special etiological factors and the numerous variations in its course, within a short formula. It is, however, possible to arrive at a pathogenetic conception of what authenticates itself as hysterical if one attempts to view the tangle of clinical details from a psychological point of view.

If we start out with the description of the psychic stigmata of hysteria, whose discussion we have intentionally put off until this time, we can distinguish with *Crämer* four groups of phenomena which naturally, like the somatic stigmata, need not all be present in every individual case, of which, however, one or more occur in definite or discrete manner in most hysterics. These are: 1, Excitability under affect; 2, ideas of being wronged; 3, inability to accurately reproduce impressions; 4, striking variations of mood. In general the hys-

terical individual is moved with extraordinary intensity by affects of agreeable or disagreeable tone, and reacts to these emotions in a specially unrestrained manner. In general a morbid egocentrism manifests itself in the tendency to interpret occurrences in their relation to his own person and in so doing by preference to assume the rôle of a martyr; in him in general the retouching of the objective facts through subjective factors, as well as the description of external happenings, particularly in that of his own sufferings, reaches the highest degree, even "pathological untruthfulness;" finally, in general, the hysterical are entirely irresponsible in their disposition.

It is a question now if this "hysterical character" furnishes a comprehensive and satisfactory explanation for the fact that just in such individuals autosuggestibility (whose importance for the occurrence of hysterical phenomena can be experimentally recognized) is increased to an actual disease condition. This question is to be answered in the negative upon the ground that many healthy people, but particularly many psychasthenics and neurasthenics, present one or another of these stigmata, or indeed, all of them, in marked development, and even then, do not show specific hysterical symptoms if they experience severe psychic traumata.

Further, *Dubois* has pointed out in a brilliant manner, that autosuggestibility, in conjunction with the constitutional inferiorities expressing themselves in the hysterical character, may, indeed, explain the appearance of inadequate somatic reactions to emotions, not, however, the following important criteria: "The persistence during weeks, months, years, indeed during the whole life, of an array of functional disturbances which appear in the absence of every primary material injury and which in consequence must be referred to psychic causes." This characteristic fixation of the somatic phenomena arising from emotions (actual or suggested), *Dubois* attributes to a psychic anomaly which is to be considered the most important factor in the specific hysterical diathesis, "sense impressionability."

Now the Bern neurologist defines "sense impressionability," "The ability to impress upon the sensations arising from affects, the stamp of reality;" you see in how satisfactory an agreement what I remarked to you about the rôle of phantasy as a predisposing factor for hysterical affects, stands with the deductions of *Dubois*. That, also, in the stigma—the inability to accurately reproduce impressions—which can increase to "mythomania," "phantasy," reaches clinical expression, has scarcely escaped you.

Though I confess myself without reserve an adherent of the teachings of *Dubois* as to the pathogenesis of hysterical conditions, I cannot leave unmentioned the much-discussed views of the Vienna neurologist *Freud*. He assumes that an affect when it does not lead to an adequate reaction, is transposed into somatic disturbances. The hysterical disease manifestations are based upon such a "conversion." And, indeed, the disease-producing "complex" is regularly to be sought in the sexual experiences of the earliest childhood, which had exercised a powerful impression upon the childish psyche, but from natural reasons is concealed and not "reacted out." In that, little by little this complex disappears from consciousness, that is, is "supplanted" by hysterical phenomena (which, all the same, are symptomatologically in accord with the

sexual trauma in childhood, for example, a hysterical attack with a parental coitus witnessed), from a therapeutic point of view it is necessary through "psycho-analysis" to bring up the "pent in affect" from the subconsciousness into consciousness and rid the patient of it.

That in spite of the very great exaggeration of the frequency of an infantile sexual etiology, the extensive lack of criticism in the development of the doctrine of conversion, and the regrettable growth which the therapeutic application of psychoanalysis has attained, *Freud's* views contain a useful principle, is to be acknowledged without dispute. So he has pointed out effectively, not indeed for the first time, but most clearly, that every hysteria is a traumatic one, and that it is regularly psychic factors which bring the dormant psychoneurosis to an outbreak. These are mainly acute and brutal psychic traumata, and not, as a rule (as is generally the case in acquired neurasthenia) a more chronic emotional unrest, the storm of affective irritations, small in themselves, but always protracted in action, which supporting themselves upon former ones not yet run out, undergo summation and accumulation. It is also true, that not rarely the symptomatology of the concrete hysterical attack is influenced by the initial trauma. I have already mentioned this for accident hysteria. As to sexual traumatic hysteria, a like relationship is sometimes found, so three cases of contracture of the adductors of the thigh which I observed in females arose from attempts at criminal assault, on which occasion these muscles were put into a position to do honor to their ancient name "custodes virginitatis." Let it be remarked in passing, no "psycho-analysis" is needed to uncover sexual etiological factors (or to suggest them); they are naturally not subconscious and "displaced," but usually they are intentionally concealed from the physician until he has inspired a sufficient confidence by tactful demeanor. That then talking it over with the psychotherapeutist and his soothing reassurance can only act advantageously and can assist recovery, is evident to any one without his having to confess adherence to the dogma of the "hemming in of affects." The Psalmist wrote about "sin," "Should I conceal it, my limbs would give way," and also the ordinance of auricular confession bespeaks eminent psychological intuition. Still one thing more: it is shown that many cases of hysteria can be dated back to initial traumata of sexual or erotic nature; this corresponds to the powerful rôle which the sexual factor plays in the life of the individual. Only one must not think that the sexual traumata must always occur in early childhood, and must be connected with actual irritation of the genitals, as *Freud* has apodictically affirmed. Rather do sentimental-erotic misfortunes, as, for example, a broken engagement in a young girl, play a very great rôle. Also in the frequent hysteria of childless women, the insufficient potence of the husband is but rarely to be held responsible; much more frequent is the collapse of a long-cherished hope of offspring; the same thing applies to the hysteria of old maids, which often breaks out at the time when the last illusion in regard to the longed-for "settlement" is dissipated.

Dubois has said of the female hysteric, that she is a comedienne, but does not know it, that she is acting, and honestly believes in the truth of her

rôles. This may in general be entirely correct, but does not apply to certain cases in which the desire for commiseration, the "pose," the coquetting with illness, lead to refined "pathomimicry" which sometimes does not draw back from self-mutilation. Fig. 111 represents the hand of an hysterical woman who was detected as she was producing her "hysterical pemphigus" with hot sealing wax. Still more monstrous was the case of a young girl who had produced in herself a "puzzling trophoneurosis" by about 150 cuts reaching down to the fascia on the left arm and both legs and in whom my diagnosis of self-injury nearly brought me into an unpleasant situation for suspecting the "brave daughter" of such a thing. Fortunately the discovery of a pair of bloody scissors concealed between the mattresses rehabilitated me, and the patient then furnished an interesting written account, from which it appeared that after an exciting family scene, while in her bath, she accidentally noticed the anesthesia of her left arm, and then, partly in order to make herself interesting and partly to mystify the family doctor, had begun the automutilation. In another girl, when at different points in her skin needles appeared, I received the confession that she had swallowed them in order to provoke a hemorrhage from the stomach. In a case of *Dieulafoy's* the patient allowed it to go so far that the extremities which he had intentionally cauterized with lye were amputated. Opposed to these rare extremes, forcing up the thermometer, the production of blood-spitting by wounding the gums, putting albumen in the urine, etc., represent the frequent "formes frustes" of pathomimicry. From the fact that self-injury, swallowing foreign bodies, etc., occasionally result fatally, the otherwise good prognosis of hysteria as regards life becomes more serious in such cases. The same remark applies to the sometimes theatrically staged suicidal attempts, which occasionally succeed better than was intended. True, intentionally carried out suicide is exceedingly rare in hysterics.

Fig. 111.
Hysterical Self-Injury. Pseudo Pemphigus.

But one word on the so-called "hysterical psychoses." I am not willing to accept without reserve the view of *Aschaffenburg*, according to which there is no definite clinical form of hysterical insanity, but rather that usually a

mixture of more or less numerous and striking hysterical features with the most varied psychoses (for example, with manic depressive insanity) can occur.

Treatment

That from what has been said above, the rational treatment of hysteria can only be a psychic one is apparent without further remark. While here, in general, the same underlying principles apply as were laid down for the psychic treatment of neurasthenia, in the previous lecture, on account of the preponderant rôle which in the clinical picture of hysteria is to be assigned to autosuggestive factors, some modifications are desirable.

While, for example, the neurasthenic is favorably influenced psychically by frequent and accurate examination into his condition, since he thereby becomes assured that the physician is carrying out his task particularly thoroughly, and on this account is inclined to put more and more faith in the assurance that there is no organic disease, and his troubles are bagatelles, in hysterics it is well to avoid frequent examinations. For example, if by an exact examination carried out under all precautions, an anesthesia is recognized as certainly hysterical, it is wisest to completely ignore it thereafter until taking the condition of the patient upon discharge. In severe cases of hysteria, isolation from the home surroundings and bed treatment, not ambulatory, is absolutely required. In hysterics much reduced in nutrition this can be combined with the rest cure after *Weir Mitchell*. Anorexic hysterics must have the psychical genesis of their repugnance to taking food explained to them and after this must, without severity but with unyielding persistence, be trained to take an increasing quantity of nourishment. One should refrain as far as possible from using the œsophageal tube. While a few patients have a wholesome horror of this instrument and so may be persuaded to take a sufficient quantity of food, on the other hand the patient finds pleasure in his rôle of martyr, and there is difficulty in laying the ghost that has been called up. Also astasia-abasia, aphonia, contractures, etc., must be cured by irresistible reassurance and patient explanation of their psychogenic nature by methodical re-education of the will. Seizures are disregarded as far as possible; also the family and surrounders must learn to not make much of these, but to content themselves, after sprinkling the patient with some cold water, to leave him alone as much as possible until he comes to himself. In paralyses, aphonia, etc., in fresh still untreated cases, the so-called "surprise method" is often found a success. In this, by the application of some striking procedure previously unknown to the patient (the application of static electricity, endo-laryngeal faradization, hypnosis, etc.), a sudden cure of the symptom (naturally not of the disease) bordering on the marvelous may be sometimes obtained. Against this method no objection can be made, only if we explain afterward to the patient the psychotherapeutic nature of the cure, that the procedure is only active suggestively, so proving to him directly the autosuggestive nature of his disease. For the rest, the object of our treatment must be not only the explanation to the patient of the nature of his troubles, but also of their cause; he must learn to combat his "sense impressionability," his phantasy, his egocentrism,

his impulsiveness, his exaggerated emotivity, as well as to oppose the psychic traumata which has set up his hysteria, with objectivity and stoicism, after he liberates himself from the pressure of painful memories by their communication to a sympathetic counselor. All these things allow themselves to be put into words in a very condensed and plausible manner, but in practice are the most difficult and trying tasks which come to the psychotherapeutist.

LECTURE XXX

Migraine

In the course of these lectures we have become acquainted with different varieties of headache as a symptom occurring in a large number of nervous diseases: so in arteriosclerosis and syphilis of the brain, in internal hemorrhagic pachymeningitis, in the different leptomeningitides, in brain tumors, brain abscesses, hydrocephalus, in progressive paralysis, after epileptic attacks, hysteria, etc. The occurrence of headache (cephalalgia, cephalæa) extends, however, far beyond the boundaries of neurology. I would remind you of those forms which are known to you as an accompaniment of fevers, as the expression of slight uremia, in consequence of chronic nephritis, as an initial symptom of syphilitic infection; further, of the locally caused headache in diseases of the ear, the eye, the frontal sinus and antrum of *Highmore*, of the bones of the skull (gummata, tumors, caries, periostitis), of the neck and frontal muscles, and the cephalæa of chronic lead, alcohol, nicotine and many other intoxications, of those which are complained of during the incubation stage of infectious diseases, of the transitory headache with which even the healthy person becomes acquainted, after mental or physical overexertion, excesses in drinking, sleepless nights, sojourn in hot and poorly ventilated rooms, etc. To-day, however, there remains for us the description of a form of headache which even in the second century after Christ was described as a disease *sui generis* (by *Galen* and *Aretæus*, of Cappadocia), and still preserves its nosological autonomy. This is hemicrania or migraine. Just as in Lecture XXVI, we kept in mind only genuine, not symptomatic epilepsy, here also we will disregard those cases in which migraine-like attacks occur during the development of a brain tumor, as symptoms of multiple sclerosis, or progressive paralysis; also those observations very interesting in themselves, where in epileptics migraine attacks alternated with convulsive seizures, so that their conception as "epileptic equivalents" has impressed itself. Such cases, like the not very rare occurrence of members of one and the same family, afflicted part with epilepsy and part with hemicrania, suggests the thought of a certain nosological relationship between the two affections (to which besides the occurrence in paroxysms is common), while on the other hand, opposed to such a conception, is the fact that epilepsy usually manifests progressive tendencies; migraine, on the contrary, shows practically always with the advance of age, a reduction in the frequency and intensity of the attacks and indeed, in many cases finally spontaneously disappears. Also in migraine patients degenerative symptoms are, as a rule, absent.

It is an exceedingly frequent nervous disease, particularly among the upper

classes of society, which affects women in a quite preponderant manner; in my material at the most 20 per cent. of the hemicranics were of the male sex. Like heredity, it is often recognizable. That the arthritic diathesis, which is much supported by the French and English, plays any rôle worth speaking of in the etiology of migraine, is very questionable; in any case, the notorious rarity of true gout in our neighborhood stands in striking contrast to the extent of migraine. The combination with psychoses, neurasthenia and hysteria is quite frequent; that with vasomotor neuroses not rare. Very questionable is the frequently affirmed causal relationship of migraine to gynecological, rhinological, or stomach diseases.

The disease appears usually at the time of puberty, still, its beginning at an even earlier age, for example, 6 or 7 years, is in no way rare. Quite 90 per cent. of cases set in before the 20th year of life; an occurrence of attacks of migraine for the first time in persons between 40 and 50 years of age is always suggestive of the symptomatic form. Relatively frequently the attacks cease during the 6th decade, after it has decreased in severity and frequency from the 40th year; in women the recovery sometimes coincides exactly with the menopause. On the other hand, there are persons who are still subject to migraine attacks even at an advanced age.

Symptomatology

In the preponderant majority of hemicranics—if we disregard the accompanying psychoneurotic conditions—the migraine attacks occurring at more or less regular intervals (it can show a regular or an irregular type), represent the whole clinical picture and in the intervals there are no anomalies to be observed. A minority of patients, on the other hand, show also interparoxysmal symptoms. We will now, to gain a comprehensive view, first sketch the usual clinical picture of migraine attacks, then consider the different varieties of the paroxysms, and finally the interparoxysmal phenomena.

a. The Regular Migraine Attack, Hemicrania Simplex

For the occurrence of a migraine attack, there are either no provocative agents to be found, or certain exciting causes can with more or less probability be held responsible; this most certainly applies to the menstruation, since many patients are exclusively and regularly affected by hemicrania at the time of their catamenia; for the rest, the patients frequently give as exciting factors overexertion, psychical excitement, coitus, excessive use of alcohol, too short or too long sleep, indigestion, hunger, the occurrence of sultry weather, etc. The time of predilection for the outbreak of the attack is the early forenoon, still it can occur at any hour. Occasionally it strikes the patient like lightning out of a clear sky, though he has gone to bed with a clear head and feeling particularly well, he wakes in the morning with severe migraine; usually, however, characteristic premonitory symptoms precede its appearance. *Grasset*, *Rauzier*, and others have differentiated two types of these prodromi, the "excited type" in which there comes over the patient a peculiar

motor unrest along with psychic irritability, and sometimes also sexual excitement, and the "depressed type" which is characterized by the preponderance of a sad or morose mood with a feeling of depression, by drowsiness and yawning. When the attack is coming on, the patients usually look pale and prostrated; often they complain of pressure in the epigastrium, precordial pressure, urinary irritability, shivering, dullness in the head; when the migraine comes on in the morning, during the preceding night, sleep is often restless and accompanied by bad, in part stereotyped, dreams. An actual "aura" occurs only in a few severe forms of migraine, for example, in the form of paresthesia of the tongue, or of one hand, tinnitus aurium, spots before the eyes, etc.

The headache is, as the name hemicrania expresses, at the start one-sided or predominating upon one side, but not at all infrequently in the course of the attack affects both sides of the head with equal intensity. It is usually frontal or temporal, more rarely occipital, but can also radiate into the neck, shoulder and arm. Its severity usually increases little by little to remain at its maximum several hours, and then just as gradually to disappear again; when the migraine has so far decreased that the patient can go to sleep he usually awakes with his head quite free again, rarer is a "critical" discontinuance of the pain while the patient is awake. Its severity is quite different from case to case, sometimes also in the same patient from attack to attack. With the mildest migraine the patient can still go about his business, the more severe forms compel him to lie down in a darkened room as free from noise as possible (since light and noise increase his trouble); the most severe cases, however, increase to actual torture which forces from the patient groans and lamentations. The character of the pain is varied; usually it is described as hammering or boring, sometimes as cutting or burning. As to its topography, it is by no means always stereotyped; even in the unilateral type, sometimes the right, again the left, now more the forehead, now more the temporal region can be affected, etc.

The migraine headache is always accompanied by nausea, which usually increases to retching, often to actual vomiting; this last is independent of taking nourishment. It can occur upon an empty stomach (for example, immediately after awakening), with the bringing up of gastric juice or bile, or there are indeed, abortive vomiting movements, a particularly painful condition. The loss of appetite is complete during the attack.

The face is usually pale and sunken in (the so-called "white" or "angiospastic migraine"), more rarely reddened and swollen ("red" or "angio-paralytic migraine"). Sometimes these disturbances of vascular innervation are accompanied also by other sympathetic phenomena; so the physiologist *Du Bois-Reymond* noticed in his own migraine attacks, besides pallor of the face, redness and heat of the ear, hyperemia of the conjunctiva and narrowing of the palpebral fissure. *Flatau* found, at the height of the hemicranic paroxysm, hardening of the temporal arteries, myosis or mydriasis, swelling of the upper lids, etc. *Oppenheim* and others have described increased discharge of mucus from the nose, *Tissot*, *Labarraque* and others, small extravasations of blood in the face, in the nose, in the retina. *Curschmann* noticed "vasomotor angina

pectoris," *Berger*, diarrhœas, *Calmeil*, polyuria and pollakiuria, I myself circumscribed œdema of the face, "dead fingers," and flow of tears, as accompaniment of the hemicranic attacks. The pulse is sometimes slowed, indeed to 40 a minute.

Möbius has pointed out the relatively frequent occurrence of sensitiveness to pressure of the *Valleix's* points of the trigeminus and occipitalis (see Lecture III, pages 48-49. Also a hyperesthesia of the scalp and the skin of the face appears during the migraine attacks, further, paresthesias of the lips, the tongue and the hand.

The attacks just sketched, which in a few patients show themselves only at long intervals (about two or three times in the year), in others once or twice a week, upon the average, however, have a tendency to appear about every three or four weeks, can in severe cases succeed one another in such a manner that there is a status hemicranicus (or, according to *Oppenheim*, a "permanent hemicrania"). In this after the migraine is apparently on the decrease, the headache increases again to maximum height, and this repeats itself for several days. From this condition *Flatau* with propriety wishes to see separated "continued hemicrania," that is, cases in which the migraine instead of the average duration of the attack of about 12 hours, persists for days in unaltered intensity, and then slowly disappears.

b. Particular Varieties of Migraine

Among the varieties of migraine, on account of their frequency, the abortive or rudimentary attacks deserve to be mentioned in the first place; in these, after more or less definite development of the prodromal symptoms, there is a slight pressure in the head with some nausea, but after a short time the patient feels entirely well again.

Of greater symptomatic interest are the migraine forms which occur with cerebral irritative or defect symptoms; so, at the height of the attack, there are not so very rarely more or less outspoken aphasic disturbances (usually of *Broca's*, more rarely of *Wernicke's* type); further *Féré* and others have pointed out fleeting hemipareses on the opposite side to the maximum of pain; *Liveing*, to objective disturbances of sensibility of cerebral topography; *Oppenheim*, to transitory typical cerebellar ataxia, with severe vertigo (cerebellar hemicrania); *Flatau*, to clonic contractions on one side of the face, etc.

As "ophthalmic migraine" *Charcot* and *Féré* have isolated a variety of hemicrania, in whose clinical picture certain visual disturbances appear particularly prominently. One of the most frequent forms of course of this eye migraine, is the following: The patient has suddenly a peculiar light perception, in that either flames, sparks or lightning, move before his eyes, or with darkening of the central part of his visual field, in its periphery bright serrations, which now separate from, again approach one another, now rotate like a cog-wheel, disappear, again glitter with all colors of the rainbow, but do not prevent the perception of surrounding objects. We speak of "scintillating scotoma" or also of "teichopsia" (τεῖχος =rampart) since the serrated figures remind us of the plan of a citadel after *Vauban*.

The light phenomena, which usually occupy only one-half of the visual field, disappear after some minutes, usually to make place for a transitory hemianopsia, eventually, also, for a temporary amaurosis, to which then, the usually specially severe unilateral pain, the nausea, the prostration, in short, the ordinary migraine symptoms, succeed. The psychiatrist *Jolly* and the astronomer *Airy* have furnished very good descriptions and pictures of their own attacks of ophthalmic migraine and scintillating scotoma. Ophthalmic migraine can also begin without the entoptic phenomena described, with simple temporary hemianopsia or amaurosis;* further, the ocular symptoms instead of affecting as is usual both eyes, exceptionally affect only one; finally, there is sometimes an intense sensitiveness to pressure of the one eye ("iritic migraine" of *Piorry*). Combination of ophthalmic hemicrania with the focal symptoms already enumerated (transitory aphasia, facial hemispasm, etc.), is relatively frequent. *Antonelli* and *Siegrist* have been able to recognize ophthalmoscopically during the attack, a spasm of the retinal vessels.

As ophthalmic migraine affects the visual organ itself, ophthalmoplegic migraine (denominated by *Möbius* "periodic oculomotor paralysis") involves its muscles. The attack begins like simple hemicrania, which, however, as a rule, is characterized by great severity and long duration (up to 14 days); then, however, it passes over into an oculomotor paralysis (usually total, but irregularly distributed among the different muscles). This last is homolateral with the headache and usually lasts for several weeks, while the pain generally disappears with the appearance of the ophthalmoplegia. The trochlearis and abducens are only rarely affected.

As analogue of ophthalmoplegic migraine, *Flatau* has isolated "facioplegic" (better, "prosoplegic") migraine, a form unknown to me personally, in any event very rare. It leaves behind a facial paralysis of peripheral type which recovers only after several weeks.

We will close the enumeration of the atypical forms of migraine with the mention of "olfactory migraine," "gustatory migraine," and "otic migraine." These are attacks which are accompanied by disturbances of smell, taste or hearing, or are followed by such. Such observations are also to be considered as great rarities.

Entirely outside of the limits of hemicrania are to be placed forms of headache which have been described by *Hartenberg*, *Peritz*, and others, as "migraine of the arthritic," myalgic migraine, etc. As a matter of fact, these cases belong to an interesting rheumatic affection which has been isolated by *Henschen*, *Norström*, *S. Auerbach*, *Edinger*, and others, as "nodular" or "indurative" headache. In this, the pain radiates from the neck and forehead muscles unilaterally or bilaterally over the scalp, and in the trapezius, rhomboideus, splenii, frontalis and other muscles there are to be recognized upon palpation, partly diffuse, partly circumscribed swellings. Now the swelling is yielding and elastic, again firm and hard. The first corresponds more to the

* One of my patients, a Polytechnic student, while in school, noticed the onset of hemianopsia, in that suddenly, instead of two blackboards. he only saw that to the left; immediately afterward his right hand and right side of his tongue "went to sleep," and pain in the left side of the forehead appeared.

subacute, the last more to the chronic cases. Almost always there are refrigeratory influences, sometimes of local nature (in women washing the hair in winter) to be discovered. *His* has also pointed out the presence of uricemia. Under the influence of fatigue and dragging upon the affected muscles, of local refrigeration, and getting wet, this nodular or rheumatic cephalæa usually exacerbates. Besides muscular nodules and indurations, enlarged lymph glands, and infiltration of the skin at the back of the neck may sometimes be found. Reflexly (perhaps also by irritation of the sympathetic in the neck), the severity of the pain can lead to nausea, rarely to vomiting. In pieces removed from such "headache nodules" I have not been able to find anything abnormal microscopically; there is also no infiltration or true induration, but as *A. Müller* thinks, the result of a localized hypertonia. Local applications of heat combined with massage and percutaneous or internal salicylic treatment, is if persisted in long enough, of sovereign efficacy in this "nodular headache."

c. Interparoxysmal Symptoms

Expressly to have pointed out the greatly neglected interparoxysmal symptoms in migraine patients, is a special service of *E. Flatau.* Among these are on the one hand interesting, those syndromes which occur themselves in the form of attacks and so justify their consideration as "migraine equivalents"; on the other those which are characterized by their constancy. Of migraine equivalents there may be mentioned as examples: Attacks of vasomotor angina pectoris, *Quincke's* œdema, bronchial asthma, neuralgias (for example, *Morton's* metatarsalgia), rotary vertigo, tinnitus aurium, psychic depression, gastralgia, yawning and sneezing spasms. Of the permanent symptoms which usually affect older persons, most frequently described is the persistence of hemianopsias, or minor defects in the visual field after repeated occurrence of ophthalmic migraine. *Meige* saw develop in an aged migraine patient a hemiparesis of the face with unilateral œdema. Exceptionally ophthalmoplegic migraine can leave behind permanent paralysis of the oculomotorius or of some of its branches. Finally, I would mention that two of my migraine patients had permanent bradycardia.

Pathogenesis

As to the basis of migraine, a number of theories have been proposed, into all of which I naturally cannot go. The most modern, supported by *S. Auerbach*, is based upon *Reichardt's* theory of "swelling of the brain" already touched upon in Lecture XIX. *Auerbach* thinks that the symptom-complex migraine is explained with least constraint, if we assume that the "hemicranic predisposition" depends upon a lack of relation between the space within the skull and the volume of the brain, and that the attacks are set up by exciting causes which are calculated to increase this lack of correspondence by way of the vasomotor system. In my view, this theory, seductive from many points of contact between the semiology of the hemicranic paroxysm and that of the

brain pressure syndrome,* is not sufficiently well founded for my acceptance of it. Rather do I confess myself, with the great majority of neurologists, of the opinion that the migraine attacks depend upon vascular spasms in the brain, that is, the hemicrania represents a vaso-constrictor neurosis; no other theory can explain so satisfactorily the fleeting focal symptoms (hemipareses, aphasia, hemianopsia, etc.), the accompanying paresthesias; the combination with vasomotor angina pectoris, "dead fingers," etc. Also, the ophthalmoscopic findings of *Siegrist* and *Antonelli* may be referred to. Further there are observations in which thromboses of the brain vessels and foci of softening, occurred after unusually severe attacks of migraine. The attempt of *Du Bois-Reymond* and *Möllendorf* to differentiate a vaso-constriction and a vaso-dilatation migraine is, on the other hand, to be considered a failure; from the investigation of vasomotor neuroses we know so much about sharply localized vascular spasms, that it does not do to deduce from the different condition of the arteries of the integument in "white migraine" and "red migraine," that the condition of the brain vessels must coincide with that of the superficial vessels. That autointoxications, hormonological anomalies, etc., are to be held responsible for the periodic setting up of migraine attacks, is often assumed; but the nature of the substances coming into question is still entirely obscure.

Prognosis and Treatment

In genuine migraine, fatal complications, as we have indicated, are most extreme rarities, so that the prognosis as to life is in general quite favorable. As to recovery, a somewhat reserved standpoint is proper, since not all cases recover in later life, as is the rule. Namely, ophthalmic and ophthalmoplegic migraine may be characterized by great obstinacy. Fortunately, however, from the point of view of treatment, we do not stand helpless against migraine, rather in the great majority of cases are we able to favorably influence its course.

As a general rule applicable to all migraine patients, it may be stated that a bland, predominantly lacto-vegetarian diet, with frequent taking of small portions of nourishment between the principal meals (see Lecture XXVIII, page 434) and abstention from alcoholic beverages, exercises a favorable influence upon the number and intensity of the paroxysms. A distribution of work and rest as rational as possible, avoidance of all excesses, treatment of constipation, regular exercise in the open air, generous vacations spent when possible in the mountains, are further important prophylactic measures.

Under medicinal treatment are to be mentioned in the first place, occasional courses of bromides recommended by *Charcot, Gilles de la Tourette, Möbius, S. Auerbach* and others, in which several times a year for several weeks at a time, potassium bromide raised to a dose of from 3.0 to 6.0 grammes a day and then gradually reduced, is given. More efficient than the simple alkaline bromide appears to me the following combination recommended by *Mendel:* for 20 days at a time the patient takes every morning in a large

* Headache, vomiting, slow pulse.

cup of hot orange flower tea, a powder of the following composition: ℞ Sodii bromide, 2.5 (gr. 35); sodii salicylat, 0.25 (gr. 4); aconitini, 0.0001 (1/10 mg. = 1/600 grain). For prolonged treatment, the extract of cannabis indica is especially suitable; of this 0.015 (gr. 1/5) is given, best every evening for several months. The pil. cannabinæ comp. which I have recommended, has also been found useful by *Curschmann* and others. When necessary, the dose of hashish can be doubled or tripled without hesitation. The administration of ext. cannabis ind. in a maximum dose to cut short the attack (℞ Pastæ guaranæ, 0.25 (gr. iv); caffein. citrat, 0.05 to 0.1 (gr. 3/4 to 1½); extract cannabis ind., 0.1 (gr. 1½ in capsule), however, sometimes produces unpleasant symptoms. Here the different anti-neuralgics (see pages 57 and 59) are to be preferred, particularly pyramidon, migränin, trigemin, aspirin, or the so-called "mixed powders"; among the last I would mention specially the standard recipe of *Krafft-Ebing:* ℞ Past. guaran., 0.2 (gr. 3); caffein citrat., 0.06 (gr. 9/10); codein phosph., 0.02 (gr. 1/3); phenacetin, 0.5 (gr. 7½); further, the equally efficient *Edinger's* formula: Past. guaran., 0.3 (gr. 4½); antipyrin, 0.5 (gr. 7½); caffein. citrat. 0.02 (gr. 1/3); 2 powders at an interval of an hour as needed. Since the patients are only too much inclined to misuse "headache powders," the taking of them except for actual severe pain must be firmly opposed. Alternation between the several drugs, in order to prevent habit formation and the patient's raising the dose on his own responsibility, is greatly to be recommended. Only in very rare cases is it necessary to use the certainly acting morphine, for particularly severe attacks (status hemicranicus). Of course the patient should never be given the syringe and the solution; do not be afraid, however, to give a large dose; from less than 0.015 (gr. 1/4) a prompt action is not to be expected. We need not order the patient during the attack to lie quiet with loosened clothing in a darkened room from which all noises are excluded; he does this himself when it is necessary. Also, the patients know better than the physician whether cold or warm compresses to the head suit them best. As a prophylactic against the paroxysms, however, I recommend daily application of very hot compresses to the forehead for a quarter of an hour after going to bed. This is an adequate substitute for the hot forehead douches recommended by *Carron de la Carrière* (which are in practice quite complicated). From electrotherapy (only transverse galvanization through the head comes into question), on the other hand, not much is to be expected. In order to prevent the "vomiting on an empty stomach," the patient should be urged to take some food during the attack; milk, to which strong black tea has been added, is usually best tolerated.

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