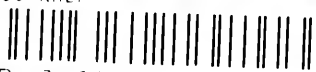


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# PARALYSIS

## AND OTHER DISEASES OF THE

# NERVOUS SYSTEM

IN  
CHILDHOOD AND EARLY LIFE

BY  
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## PREFACE

THE aim of this book is to present in as brief a form as is compatible with completeness an account of the various forms of paralysis and allied disorders, as they are met with in early life. It is hoped that it may be useful to the advanced student, as well as to the practitioner. The absence of references in the text and of any bibliography may disappoint some readers. It was considered desirable, however, not to interrupt the text with references to footnotes, and a bibliography even of those books which have been consulted would have added considerably to the bulk of the volume. The inclusion of a complete bibliography, on the other hand, would have seemed to arrogate to the work a more ambitious character than it aspires to. It is, in effect, an expression of personal opinions, the result of the writer's observations, modified and extended by his knowledge of the views of others, on subjects which have interested him for a good many years, and in regard to which he has had the good fortune to have special opportunities of acquiring experience.

I am very much indebted to writers too numerous to mention individually, but I should like to express my acknowledgments to several of my colleagues and friends.

To Dr. Hughlings Jackson's constant kindness and encouragement I owe more than I can say. I should have liked to dedicate my book to him, had I thought it worthy of such an honour. To Sir William Gowers' kindness and friendly advice I am indebted almost as much as to his teaching, and I have to thank him for the suggestion that I should write this book. I wish I could have brought it up to that high standard which I know he exacts from himself. Dr. James Collier has furnished me with most able and willing help and with numerous photographs, from which the illustrations have been made. Dr. F. E. Batten has also been kind enough to lend me some of his excellent illustrations of the morbid anatomy of infantile paralysis. To Dr. G. A. Sutherland, Dr. Farquhar Buzzard, Mr. Jackson Clarke, Dr. Allen Starr, Dr. Barker, Dr. R. T. Williamson, Dr. J. H. Crocker, and Dr. H. J. F. Simson, I am further indebted for illustrations and for assistance, and I also wish to thank Dr. Grainger Stewart for kindly reading many of the later sheets, and for most valuable help in the preparation of the index.

JAMES TAYLOR.

*Welbeck Street, London, W.*

*March, 1905.*



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AND  
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INTRODUCTION

It may perhaps seem necessary to offer, if not an apology, at least some justification, for a work which deals with nervous conditions as they occur in young subjects. Do such affections, it may be asked, differ so materially from similar or identical diseases affecting adults that it is necessary or desirable to treat of them separately? Undoubtedly there are some points of distinction which justify such separate treatment. Thus, it must be remembered in reference to paralysis, *e.g.*, that such a condition occurring in a child affects structures undeveloped and still growing. Paralysis or disability, no matter what causes it, if it interferes with the nervous mechanism subserving mobility in a child, necessarily leads to interference with the growth of the structures and so produces deformities. Similar paralysis occurring in an adult does not lead to deformity such as is produced in the young. Thus, a child is stricken with hemiplegia—the result of a cerebral condition. Although the brain structures are not nearly so intimately

connected with trophic functions as are the spinal cord cells, yet the paralysis and the consequent immobility of the limbs lead to an interference with their development, so that after some years the difference in the size of the limbs of opposite sides of the body is very striking. Even more marked is the interference with development following acute anterior horn inflammation—so-called infantile paralysis. In spite of its name this disease is met with also in the adult. In him it causes loss of power, with marked muscular wasting. But there is no shortening of the limb, and except, perhaps, for some contraction, the result of unantagonised muscular action, no inconvenience beyond the loss of power. In the child, however, a lesion of similar position and extent leads to marked interference with the growth of the limb or limbs, not only in their circumference, but also in their length, so that if the lower limb be affected, growth may cease to take place and a condition of marked inequality in the length of the limbs naturally results. In other words, a diseased condition which in an adult may only lead to wasting, in a child may produce in addition very striking deformity.

Another point which will at once occur to the reflective is that whilst in the majority of cases of cerebral paralysis in the adult we are dealing with a diseased condition of the nervous system which is not primarily nervous, but is the result of atheroma and other diseases of vessels, in children such atheroma does not occur except in very rare instances; so that other causes have to be invoked—traumatism, acute inflammation, probably of toxic origin—to account for a condition of cerebral paralysis in the child which in the adult is usually the result of degenerative vascular disease. Indeed, it may be stated as a general rule that the cerebral palsies of children as a class are quite different in their etiology from similar conditions arising in adults. And in these diseases as they occur in early life, probably because the structures affected are attacked at a growing stage, there are frequently super-added to the paralysis spontaneous movements of various

kinds—choreiform, ataxic, athetoid—which are distinctly unusual in adults—except in those in whom the paralysis dates from early life.

It must also be remembered that there is a large class of diseases met with in children, the so-called “birth-palsies.” Under this name are included many varieties of paralysis having the one characteristic in common that they date from birth. Some no doubt arise during intra-uterine life from some condition of unknown nature interfering with the development of cerebral structures. Some probably result from maternal conditions, *e.g.* detachments of the placenta. Some are probably to be traced somewhat indefinitely to maternal ill health or injury. A certain number also probably arise as a result of injury during a long and difficult, or at least an abnormal, labour; and the physical condition in these cases may be associated with varying degrees of mental defect.

In another large class of cases there seems to be inherited in certain structures a tendency to early decay and death—what Sir William Gowers has called *abiotrophy*. A child is born healthy and develops naturally and normally for months in certain cases, for years in others, and then a gradual process of decay sets in—sometimes in one set of structures, sometimes in another. Thus, in the so-called amaurotic family idiocy, which Waren Tay was the first to describe, a child, apparently healthy, develops naturally for a few months. It then begins to suffer from weakness in all its limbs and it soon becomes blind. It gradually becomes more and more feeble and dies, as a rule, before it is two years of age. The condition found in the brain is one of almost complete disappearance of all the cerebral cells, but the cause of this remains so far unexplained. In other cases after a lapse of years there develops gradually a spastic condition of limbs, apparently the result of a premature decay of the pyramidal system. Similarly in cases of juvenile general paralysis, we have, as a result apparently of inherited syphilis, degeneration occurring

in the brain and spinal cord. And it is not unlikely that in the condition of varying characteristics which we know under the name of muscular dystrophy, we have a similar inheritance of a tendency to premature decay and death in muscular tissues, for no changes in the nervous system underlying the paralytic conditions due to muscular atrophy and to perversion of muscular nutrition in this disease, have as yet been discovered.

Reference might also be made to various other conditions, if further justification were needed for such a work as this. But it will probably be sufficient, if anyone be still sceptical, if he will look through the table of contents and remark on the number of conditions of paralysis and of nervous disease generally, which, if not peculiar to children, have at least peculiar characteristics as they occur in young people. It has, indeed, seemed to the writer that there is ample justification for writing a book having the aim which this volume has. He would, however, find it much more difficult to offer excuses for the numerous imperfections of which he is conscious, both in the matter and manner of execution of his self-imposed task.



## MENINGITIS

INFLAMMATORY conditions affecting the membranes which cover the brain and the spinal cord may have their seat primarily in the dura mater (pachymeningitis) or in the pia arachnoid (leptomeningitis). Although these conditions are necessarily frequently associated, it is well to make a distinction between them on account of the fact that the one suffers primarily in many conditions, the affection of the other being secondary.

### PACHYMENINGITIS

The cranial dura mater consists of two layers: the outer—which, besides being part of the brain covering, also forms the periosteum for the cranial bones and follows exactly the internal cranial surface, and becomes continuous at the edges of all the foramina of the skull with the external periosteum; the inner layer forms the projecting processes, the falx cerebri, the falx cerebelli, and the tentorium. At each of the cranial foramina two tube-like processes are formed, one within the other. The outer of these is attached to the periosteal layer of the dura mater. At the edge of the foramen the inner is prolonged upon the nerve or vessel entering it. Upon the olfactory nerves these dural tubes are prolonged, even to the terminal ramifications, upon the mucous membrane covering the ethmoid bone; and upon the optic nerves the dural sheath is continued as far as the eyeball, where it joins the sclerotic. The spinal dura mater is not continuous with the cranial membrane, the two structures being attached separately to the edge of the foramen magnum.

and it is quite free from attachments to the vertebral canal, except at the points of exit of the spinal nerves. Where these emerge, the tube-like sheath is prolonged from it upon each spinal root, in part attached to the periosteum of the inter-vertebral foramen, in part prolonged upon the spinal nerve outside the vertebral canal.

Pachymeningitis is, for the most part, the result of morbid processes having their primary seat in the contiguous bone, and is closely analogous to the periostitis which occurs as a result of injury to, and disease of, any bone. Thus pachymeningitis is frequently the result of disease of the cranial bones. It may arise from infective processes occurring in the nasal cavity, the orbit, or the middle ear, and it is usually suppurative. In such conditions it is not uncommon to meet with collections of pus confined between the bone and the dura mater, and if these reach a considerable size the symptoms of intra-cranial abscess may appear. The organisms of suppuration may spread from the dura mater to the leptomeninges, setting up general leptomenigitis; or into the sinuses, producing infective sinus thrombosis.

Pachymeningitis affecting the spinal dura mater is usually tuberculous, and occurs in association with the condition of spinal caries, in which there is also an accumulation of tuberculous granulations and *débris* outside the dura mater.

Pachymeningitis has no special symptomatology apart from that of the antecedent conditions with which it is associated, necrosis of the skull, spinal caries, etc. It is to be noted, however, that tuberculous affection of the dura mater in association with spinal disease may play a very important part in the production of paraplegia, for the compression to which it gives rise, and the obliteration of segmental vessels entering each nerve root which the inflammatory process of the dura mater causes, are apt to produce compression of the spinal cord, associated with interruption of its blood supply; and these combined con-

ditions result in such interference with its functions as to cause paraplegia below the point at which the pressure is exerted. (See "Paraplegia from Spinal Caries.")

#### LEPTOMENINGITIS

The pia arachnoid resembles in many respects the serous membranes lining the body cavities. Its visceral layer, the highly vascular pia mater, follows exactly the external surface of the brain, dipping into the sulci, and projecting into the ventricles along the great transverse fissure, where it forms the *velum interpositum* which carries the choroid plexus of the lateral and third ventricles upon its free edge and its under surface respectively. The pia mater is, in fact, the vascular network formed by the subdivision of the cerebral and cerebellar arteries, held together by delicate connective tissue. From the internal surface of the pia innumerable fine vessels penetrate into the brain substance at right angles to the surface. It is clear, therefore, that inflammatory conditions of the pia mater may cause wide-spread interference with the cerebral circulation. The actual tissue continuity of the pia mater with the superficial layer of the brain substance brings it about that the latter necessarily participates in an inflammatory process invading the former. In other words, leptomeningitis is always associated with some degree of encephalitis. The outer layer of the pia arachnoid is a simple layer of serous epithelium lining the dura mater, whilst the middle layer, to which the term "arachnoid" was originally applied, is a non-vascular membrane projecting over the sulci of the brain substance, and forming also the ligamentum denticulatum, which attaches the spinal cord to the theca between each pair of spinal nerve-roots. The potential space lying between the dura and the arachnoid, and that lying between the pia and the arachnoid, have been called the subdural and subarachnoid spaces respectively. Such a separation does not aid description, and the two are referred to in the following pages collectively as the arachnoid space. This arachnoid space may be compared to any other serous

space—the peritoneum for example—and when the continuity of the arachnoid space of the brain with that of the spinal cord and with the entire ventricular system is considered, the results of microbial infection of the arachnoid space may be clearly conceived. The comparison will serve, further, to emphasise the fact that the usual source of infection is analogous in both: in the peritoneum, the intraperitoneal part of the alimentary canal; in the arachnoid, the naso-pharynx, the Eustachian tube, and the middle ear.

Inflammatory states of the pia arachnoid are all, so far as is known, the result of the growth of micro-organisms which have gained access to the arachnoid space. The affection of the meninges is general, in contradistinction to the local disease, pachymeningitis. When the continuity of the arachnoid space of the brain with that of the spinal cord and with the ventricular system is considered, it will be seen that there is no anatomical limitation to the possible spread of infection over the entire surface of the central nervous system and throughout its central cavity. Such universal affection, indeed, is the rule in suppurative meningitis, if life be not ended before the infection has become general. In epidemic meningitis, also, the affection may be universal. Frequently, however, the morbid process is confined more or less to the base of the brain and to the spinal cord. The relatively greater frequency and greater degree in which the soft membranes at the base of the brain suffer is explained by the fact that the usual paths from the orbit, the nasal cavities, the pharynx, and tympanic cavities, emerge upon some part of the base of the brain. The apposition of the brain to the skull, also, is not so close at the base as elsewhere. This is especially the case in the space round the chiasma and round the brainstem and cisterns of the cerebro-spinal fluid. Through these the relative rate of flow of the fluid is less; and it would thus seem that a micro-organism would have a better prospect of flourishing there than elsewhere, especially since it would, in such a position, escape actual contact with

living tissues. Further, it is probable that the direction of flow of the cerebro-spinal fluid is in the direction of the length of the body, from the vertex to the base of the skull, and from the cervical to the sacral region; and for this reason micro-organisms, even if they arrive at the vertex, tend to be swept down and deposited at the base.

From its position leptomeningitis is termed vertical, basic, posterior basic, ventricular, spinal, cerebro-spinal, and general. The more acute forms of the disease tend rapidly to become general, and while the less acute forms are more or less localised, there is, perhaps, no form of leptomeningitis which may not affect the entire area of the pia-arachnoid. The disease is classified in varieties, according to the pathology, for they are the result of known specific organisms, except in the case of the syphilitic form, the microbial cause of which admits of no reasonable doubt.

Tubercular meningitis	.	.	Koch's bacillus
Simple posterior basic	.	.	Still's diplococcus
Suppurative meningitis	.	.	Staphylostrepto- pneumococci
Epidemic meningitis	.	.	Diplococcus
Syphilitic meningitis.			

## TUBERCULAR MENINGITIS

THE mode of onset of this disease is extremely variable. In children, in whom general emaciation and bodily feebleness have resulted from long-standing tuberculous disease in some other part of the body, the onset of symptoms indicating involvement of the cerebral meninges is rapid. Where, however, the pre-existing tuberculous disease has been local and latent, the illness comes on insidiously. The gradual onset is the more common, and the slight general symptoms which appear during the early days constitute an ill-defined prodromal stage, which may vary in duration from a week to as long as three months. The relation of traumatism to the onset of tuberculous meningitis is an extremely difficult one to discuss. To anyone who has had much experience of the disease in children, the frequency with which the onset of the symptoms is related to some blow or other injury to the head is so striking as to make it impossible to believe that it is always merely a coincidence. There seems to be very little doubt that, in a child already tuberculous, the result of some injury to the head may induce, in some part of the cranial contents, a condition which renders the occurrence of disease—*i.e.* the invasion of organisms at that point—extremely easy. It might be supposed that injury may, in some ill-defined way, lower the resistance of tissues at the point at which it is inflicted, and so render the occurrence of disease at that point comparatively easy.

*Prodromal stage.*—As a rule, the prodromal stage may be ushered in by an appearance of vague and more or less slight illness. General apathy, and neglect of ordinary amusements and play, dulness, fretfulness,

and peevishness, restlessness at night, and grinding of the teeth during sleep, are amongst the symptoms which appear at this time. Vomiting also may occur, without any nausea. Constipation is nearly always present, and the breath has a peculiar factor. Mental perversity may be marked in older children, and if the patient is approaching puberty there may be almost any exhibition of hysteria. The facial aspect is one of illness and of frowning discomfort, there is a disinclination to talk, there may be mistakes in speech, and the articulation may become slow and laboured. Very young children may be speechless for days together. The child also, quite contrary to its usual habits, may pass water in bed; and the association of incontinence of urine or feces with strange behaviour must always be regarded, both in children and in adults, as of very ominous significance.

In the prodromal stage young children ask for nothing and complain of nothing. As age advances, however, headache, generally frontal<sup>6</sup> in situation, becomes a prominent symptom, and is apt to be induced by any mental or physical exertion. Delirium is rare in children, but it increases in frequency as age advances towards puberty. Convulsions may sometimes occur at this period. In explanation of the signs and symptoms which are present in the prodromal stage, it may be mentioned that it has generally been held that the appearance of tubercle precedes the occurrence of inflammation and exudation. During the prodromal stage it is assumed that the meningeal tubercles are developing, but when inflammation makes its appearance the disease definitely declares itself. Doubtless the dissemination of tubercle is in progress during the prodromal stage, for it is evidenced in many cases by the occurrence of palpable glandular enlargement, and an elevated nocturnal temperature. Further, the symptoms of the second, or invasion stage are often only exacerbations of early symptoms, and the two stages overlap so much that it is quite reasonable to suppose that the deposition of tubercle in the meninges

is coincident with the appearance of prodromata, and the occurrence of inflammation synchronous with the onset of severe symptoms.

The most striking termination of the prodromal stage is by the occurrence of convulsions consisting of tonic and clonic spasms, or of tonic spasm only. In many instances, however, a convulsion does not occur, at least at this stage; but the development of the disease is marked by a deepening of the lethargy, an increase of the headache, and more frequent vomiting. From this point it is usual to divide the clinical course into three stages, according to Tanbe, a stage of irritation, relaxation, and paralysis. While such a division is of great practical use for purposes of description, and as an aid to memory, it must be remembered that such a division into stages rarely, if ever, applies accurately to any particular case.

*General aspect.*—The expression of the face tends to lose the frown as the disease advances, and to become vacant, with wide-open eyes and dilated pupils, as if fixed upon some distant object. This has been termed “the far-off look.” There is often some retraction of the angles of the mouth, and there is frequently a bright malar flush. The general surface of the body is dry and harsh, except just before death, when copious sweating often occurs. The *tâche cérébrale* is often marked. The child lies on its side in the “cramped” position, resenting any disturbance, and will often pluck at the bedclothes when they are removed to draw them back (Stocker’s sign). In the later stages the limbs are extended and rigid in most cases, and the head may be somewhat retracted, but the extensor rigidity is never so pronounced as in posterior basic meningitis; the head retraction is never very marked, and generally it “comes and goes.” The abdomen is always markedly retracted. The psychological state at first is one of irritable lethargy, with occasional outbreaks of restlessness. Not only may the patient complain of intense headache, but also frequently of general severe pain in the limbs and joints, such as may suggest the presence of rheumatism.



The single sharp cry, apparently causeless, which has been called the hydrocephalic cry, is of common occurrence in meningitis. It is also frequently heard in other infantile intra-cranial affections.

Ophthalmoscopic examination reveals changes indicative of commencing optic neuritis, which is almost invariably present towards the end of the second week, and may appear in one eye some days before it is noticeable in the other. It rarely reaches a degree comparable with that usually present in cases of cerebral tumour, and the height of the swelling rarely exceeds two dioptries.

The presence of choroidal tubercles is frequently to be observed, and may help in the diagnosis during the prodromal stage. Tubercles of the choroid, however, are signs of general widely-spread tuberculosis, and do not necessarily indicate meningeal tubercle. They commence as minute round dots of pale colour as seen by the ophthalmoscope, which, increasing in size and in number, become raised and white in the centre, and may become confluent at their margins.

*Ocular phenomena.*—These make their appearance towards the end of the first week of the disease. All varieties of varying or persistent strabismus and ptosis are met with; rolling movements of the eyes and independent movements of the eyeballs may occur, but nystagmus is not common. The pupils are generally contracted during the early stages, and may show varying inequality, but in the later stages they are dilated. Oscillating pupil is of common occurrence. Paralysis of the other cranial nerves, such as the facial and fifth, is of rare occurrence.

*Motor symptoms.*—Repeated rhythmic movements are not infrequent, and these are specially noticeable in connection with the mouth, where sucking and champing movements and grinding of the teeth are common. In the limbs, quasi-volitional rhythmic movements may also occur. Coarse tremor upon movement is the rule when the disease becomes established, and there may be then some degree of general rigidity, usually of a varying nature.

There is always partial paralysis of movement throughout the body, but absolute paralysis rarely occurs in any limb, except just before death; yet it may occur in hemiplegic distribution after the occurrence of a convulsion, or without any convulsion, and it may be transient, or permanent, and is usually not severe.

The convulsions occurring in the course of tuberculous meningitis may be generalised or local. The fit that marks the invasion is generally localised and solitary; those occurring in the middle period are apt to be localised, while in the late sub-comatose stage they are again usually general. It is after the occurrence of a convulsion that rigidity of the limbs most often appears, and it increases with the recurrence of the convulsion. The superficial reflexes are generally retained until deep coma comes on. After a convulsion involving the legs, the plantar reflex will usually be found temporarily of the extensor type, and when rigidity comes on the extensor reflex becomes permanent. The deep reflexes are usually increased, but just before death they may be lost. Loss of control over the sphincters is the rule.

General progressive emaciation is invariable, and there is often considerable difficulty in avoiding the occurrence of bedsores.

The pulse is of considerable diagnostic importance, since early in the prodromal stage it shows marked irregularity, and tends to be more rapid than normal. During the second stage it is usually slow, and shows marked increase of tension, while in the final condition of coma it is again soft and tends to be rapid.

The respiration corresponds with the early rapidity; there is sighing breathing, and cerebral grouped breathing, and sometimes true Cheyne Stokes breathing may occur.

For the most part, the temperature of tuberculous meningitis is that of general tuberculosis. Where pyrexia exists previous to the occurrence of the meningitis, the advent of the latter causes a lowering of the temperature, and Barlow considers such lowering of temperature to be of

diagnostic value. The temperature may remain low throughout, or it may become very high for a day or two before death— $106^{\circ}$ – $108^{\circ}$  F. Post-mortem hyperpyrexia is common.

Vomiting, one of the most frequent invasion symptoms, continues at intervals throughout the illness, sometimes incessant, sometimes occasional. It is always of the precipitate causeless type which characterises the vomiting of cerebral disease in children.

Constipation is usually a marked and persistent symptom, but occasionally persistent looseness of the bowels occurs, and this is generally attributable to the presence of tuberculous ulceration of the intestine.

The course is progressive towards a fatal termination, and the end of the second week after marked symptoms have appeared finds the child extremely emaciated and comatose, with optic neuritis. Death occurs quietly from asthenia, sometimes from the embarrassment which accumulation of mucus in the chest places upon the respiration and circulation.

*Etiology—Age.*—Tubercular meningitis, though it may occur at any age, is not frequently met with in infants under six months. It occurs more often in the second six months, and reaches a period of greatest incidence in the second and third years of life. After the fourth year it becomes year by year less frequent until adult life is reached, and a second period of maximum incidence lies between the sixteenth and twenty-fifth years of adult life. The frequency of the disease then rapidly declines from that time till old age is reached, when it is very rarely met with. The sexes are affected equally.

*Heredity.*—While several members of the same family may suffer according to the well-known hereditary tendency of tuberculous disease, yet isolated cases of tuberculous meningitis occur frequently in families in which no history of tuberculous disease is traceable for several generations, and more often, perhaps, than any other tuberculous manifestation. The absence of any cases of

tuberculous disease in a family must not be, for this reason, allowed any weight against the diagnosis of the disease in a doubtful case.

The essential cause of the disease is the entrance of the specific organisms into the system, probably by different channels. A local tuberculous lesion occurs, and subsequently there is generalisation of the disease. No causal connection can be traced between this disease and artificial feeding, or its common result—rickets. As remote causes, any conditions of health, feeding, and sanitation which tend to lower bodily vitality, and therefore resistance to infection, are of importance, as are also the specific fevers, especially measles and whooping-cough, the latter preceding tuberculous meningitis with remarkable frequency.

Since tuberculous meningitis is practically always the result of extension of a primary tuberculous focus, some immediate determining cause for dissemination from the focus must exist. Injuries and surgical interference with tuberculous lesions are the presumable determinants in a few cases, but in most cases the agent is as yet quite unknown.

*Morbid anatomy.*—There are three kinds of tuberculous lesions of the meninges which may be met with in tuberculous meningitis: (1) Tuberculous meningitis; (2) meningitic grey granulations; (3) tuberculous tumours. All three forms may exist in the same case, but the third variety never produces symptoms of tuberculous meningitis unless it is accompanied by one of the other lesions.

(1) *Tuberculous meningitis* is characterised by the presence of tuberculous granulations in the meninges, associated with the common products of inflammation—fibrinous and purulent exudation. The superficial tissue of the brain underlying the affected meninges is always involved.

(2) *Grey meningeal granulations* in some cases may be found unassociated with inflammatory exudation.

(3) *Tuberculous tumours* of any size to that of a pigeon's egg may be met with. It is not uncommon to find such a

tumour to be the focus and centre of widely-spread meningitis.

Tuberculous meningitis affects the pia-arachnoid and its processes, the small vessels entering the surface of the brain and the superficial tissues of the brain itself. Occasionally a few tubercles are found upon the dura mater. In nearly all cases the convexity of the brain is least affected or escapes, and the only changes seen in that region are flattening of the convolutions, with pressing together of the latter along the sulci. In the intercranial space, around the optic chiasma, covering the tips of the temporal lobes, along the commencement of the Sylvian fissures, and around the brain-stem, there is inflammatory tissue of a yellow-green colour and of tough consistency. Spreading from the edge of this, in decreasing numbers, grey tubercles are seen in the pia arachnoid, especially in the region of the commencement of the Sylvian fissure. If this yellow tissue be examined by scraping where it is not too dense, or by opening up the Sylvian fissure which it firmly closes, it is seen to consist, for the most part, of grey or white tubercles which have become partly confluent, covered with a fibrino-purulent membrane. So firmly does this substance cling to the brain tissue, that some of the latter comes away in the attempt to remove the affected pia arachnoid.

Tuberculous granulations may be found wherever the pia arachnoid extends (the convexity, as a rule, excepted), predominantly in the depth of the Sylvian fissures, over the gyrus fornicatus, between the cerebrum and cerebellum, and upon the velum interpositum. Except at the base, the tubercles are not generally accompanied by the characteristic tough, greenish-yellow tissue.

The brain as a whole is soft—sometimes diffident—even when no hydrocephalus is present. There are several causes for such softening. Spreading in from the pia arachnoid, the tuberculous process attacks the small vessels running vertically into the cortex, and tubercles arise in their walls sometimes in such numbers that a small

entering vessel, when observed under a low magnification, after the brain tissue has been removed by careful washing, may resemble a bunch of grapes, each grape being a tubercular nodule. Thrombosis is of common occurrence in the fine vessels so involved, and sometimes a similar affection of larger branches of the middle cerebral artery gives rise to definite tracts of yellow softening in the region of the central convolutions. Further, there is actual invasion of the surface of the brain substance by the tubercular process, so that the disease is nowadays correctly termed a meningo-encephalitis. Another important factor in the softening is a widely spread degeneration of those nerve-cells of the brain and cerebellum which are situated near the external or the ventricular surface. Such degeneration of cells involves the degeneration of their nerve-fibre processes in their entire course, and is without doubt in great measure due to the toxic action of some poison arising from the tubercular process. The series of changes which the microscope reveals in the cells of the cortex—for example, in a case of tubercular meningitis—is of the same nature, and involves the cells as widely as do the changes which result from the administration of known poisons.

The occurrence of some degree of thrombosis in the superior longitudinal sinus is usual in this disease, and partial arrest of the central blood-supply of the brain may also occur, by interference with the flow in the veins of Galen which the matting of the velum interpositum brings about. Again, the frequent presence of some degree of hydrocephalus will, from increased intra-cranial pressure, tend to diminish the cerebral circulation. It is obvious, then, that encephalitis, cell degeneration, thrombosis of (1) fine arteries (2) cerebral sinuses, increased venous pressure, and diminished blood supply, constitute sufficient cause for a general soft consistence of the brain upon *post-mortem* examination, and also for the partial or complete abrogation of cerebral function sometimes met with clinically. Circumscribed deposits of tubercle are frequently

found embedded in any part of the cerebrum, cerebellum, or brain-stem. They may be of any size, from a microscopic single giant cell system, to a mass as large as a walnut, and of any age, from a fresh grey tubercle to a caseous mass or a cicatrix with small caseous centre. When such an old tubercular focus is superficial, a local eruption of young tubercles in its immediate neighbourhood may be frequently seen. The ventricles are somewhat dilated, and the fluid which they contain in considerable quantity is turbid, but not purulent or containing flakes of lymph. The turbidity is attributable to the presence of *débris* from breaking down of the living walls of the ventricle. Some excess of the same fluid is found in the arachnoid spaces at the base.

The nature of the meningitic tissue is such that occlusion of the foramina in the roof of the fourth ventricle does not often take place, and it is for this reason in part that marked degrees of hydrocephalus are much less common in tuberculous than in simple meningitis. The short duration of the illness, and the much more destructive nature of the process as regards the ventricle wall, also helps to explain the infrequency of hydrocephalus.

There are two possible causes of the transient and the permanent cranial nerve palsies met with. The one is the implication by adhesions and local interference with the blood supply of the nerves at the base of the brain by the adhesive new tissue. The second is the interference with the function of cranial nerve nuclei by poisons present in the cerebro-spinal fluid.

In a large number of cases the membranes of the spinal cord are affected, and the most common situation of the tubercles is upon the inner surface of the theca, and in the pia covering the lumbar enlargement of the spinal cord.

Co-existent tubercular lesions are found in many organs of the body, and these may be (1) recent deposits of miliary tubercles, and (2) lesions of older date, especially caseous bronchial, mesenteric, or cervical glands, and disease of bone.

*Prognosis.*—Recovery is possible in cases in which the

affection of the meninges is localised, but such cases almost always relapse and succumb. Where the affection of meninges is general, a fatal result invariably ensues.

Where the symptoms of the disease ensue upon those of some chronic form of tuberculous disease, the duration of the illness is short—from three to eight days. Patients in whom the vertex is involved die generally within one week of the appearance of symptoms. Where there has been no previous evidence of tuberculous disease, the duration is usually longer, and may be as long as two months; but as a rule death occurs during the third week.

*Diagnosis.*—The difficulties which attend the determination of the nature of the illness vary according to the stage of the disease. It is relatively simple when the disease is advanced, but it is difficult at the onset. The diseases liable to be mistaken for tuberculous meningitis at the commencement are gastro-intestinal catarrh, and one of the exanthemata or pneumonia. Of the exanthemata, enteric fever is the most difficult to distinguish. In enteric fever the temperature is much higher, and the irritability and resentment of interference are not present. Further, the dorsal decubitus is the rule in enteric fever, and headache is very marked. Widal's test is here of great value. The question as to the other exanthemata is usually settled after a few days by the appearance of the characteristic eruption upon the one hand or of certain signs of meningitis on the other. Where distinctive symptoms of intra-cranial-disease appear, the possibility occurs that some other intra-cranial lesion, such as sinus thrombosis, tumour, abscess, or middle-ear disease may be present. Careful examination of the retinae and of the tympanic membrane is then necessary, together with deliberate comparison of the symptoms of the several conditions upon one of which a determination has to be made. Bacteriological examination of the cerebro-spinal fluid withdrawn by lumbar puncture is of great value in diagnosis (*vide infra*).

*Treatment.*—The prophylactic treatment of the disease is that of tuberculous affections in general. A good milk



supply, sterilisation of the milk, a plentiful supply of good food, especially of assimilable fats, fresh air, sunshine, and perfect sanitation, embody the ideal prophylaxis. The removal of tuberculous glands, etc., and careful attention to bodily nutrition and catarrhal states during convalescence from measles and whooping-cough are all-important. When the disease is once established, the question of treatment resolves itself into the forlorn hope that inoculations of mercury may induce a circumscribed tuberculous meningitis to become stationary, and into the treatment of symptoms. All measures causing discomfort to the patient must be avoided. Ice to the head may be useful in headache and when the temperature is high, but it is not to be employed if it annoy the patient. Sponging frequently quiets the irritability, and chloral and the bromides should be exhibited where convulsion is marked. Feeding must be carefully regulated, and in many cases the nasal tube is required throughout the illness. Constipation is easily remedied. The occurrence of bedsores can always be avoided if sufficient foresight and skilled labour be employed.

Paracentesis and withdrawal of fluid from the lateral and fourth ventricles for relief of possible distension has been followed in a very few cases only with encouraging results.

*Lumbar puncture.*—Drainage of the cerebro-spinal fluid by means of a cannula introduced between the laminae into the lower part of the theca has been extensively practised of recent years. The operation is easily performed without an anaesthetic, and is quite devoid of danger if care be taken that too great a quantity of cerebro-spinal fluid is not allowed to escape rapidly. The procedure is of no curative value, but in certain cases in which indications of greatly increased intra-cranial pressure have been present, marked amelioration of symptoms has followed. Such results are far from constant, yet, considering the simplicity of the operation, there appears to be no objection to its employment for the relief of suffering. Where temporary amelioration of symptoms has followed its employment, the procedure may, with advantage, be

repeated again and again at short intervals. For purposes of diagnosis and prognosis rachicentesis is of more value. Increase of the intra-thecal pressure is at once revealed, and can be measured. In the normal child it is stated to be from 20 to 40 mm. of cerebro-spinal fluid. In tubercular meningitis it has been found as high as 450 mm., and very often as low as 10 mm. The liquid is clear when first withdrawn, in striking distinction to the turbid fluid in cases of purulent and epidemic meningitis. On standing it becomes slightly turbid, and a slight flaky deposit appears. In this deposit, separated by the centrifuge, a few tubercle bacilli may be found. The best method of demonstration of the bacilli, however, is by the injection of the fluid into the subcutaneous tissue of guinea-pigs, when the characteristic lesions of tubercle almost always result.

The following is the method of procedure adopted by Marfan. The usual precautions for the cleansing of the skin and the sterilisation of the instruments having been carried out, a line is marked across the lumbar region joining the highest points of the two iliac crests. This line passes over the spinous process of the fourth lumbar vertebra, which can be felt. The trochar of a small Pravaz syringe is introduced immediately below the fourth lumbar spine and just outside the middle line, the point being directed a little upwards. A trochar of 2 mm. diameter is a convenient size.

The depth to which the trochar should be plunged is from 1.5 to 3 centimetres in the infant, and in the adult from 4 to 6 centimetres. The trochar is withdrawn and the fluid is allowed to escape into a graduated vessel. Not more than 20 cubic centimetres should be allowed to escape at each operation in a child of two years. The puncture is closed by collodion and aseptic gauze on withdrawing the cannula.

## POSTERIOR BASIC MENINGITIS

### SIMPLE OCCLUSIVE LEPTOMENINGITIS, NON-TUBERCULOUS LEPTOMENINGITIS

THE researches of Dr. Gee, Dr. Lees, and Sir Thomas Barlow upon the nature of meningitis affecting the base of the brain in children have succeeded in conclusively proving the not infrequent occurrence of a peculiar form of meningitis, affecting especially the posterior fossa of the skull, in which the *post-mortem* appearances are specific, and in which the clinical picture is so definite that it is not difficult to distinguish this form from tubercular meningitis, with which it was formerly confused. The bacteriological researches of Dr. Still have further proved the existence of a specific micro-organism in the inflammatory exudate in every case he has examined, and there is every likelihood that the disease owes its origin to infection of the meninges with this organism.

*Clinical aspect.*—The onset of the disease is rapid, but the early symptoms are not as a rule alarming. The initial symptoms are head retraction, tonic convulsion, and vomiting, and one of these three symptoms is the first to excite attention in about an equal number of cases. Which-ever symptom may be the first to appear it is usually followed by the other two, and by irritability and languor, within twenty-four hours, and they persist for the most part throughout the course of the disease.

The head retraction is by far the most noticeable and constant symptom in this disease. While it frequently co-exists with tonic spasm, it may be very marked when the other muscles are persistently flaccid. Generally it

involves the extensor muscles of the neck alone and then the deep muscles—complexus, semi-spinalis colli, trachelomastoid, etc.—are affected rather than the superficial muscles (trapezius and splenius). The probable explanation of this condition is that in a position of extreme extension, such as is assumed in posterior basic meningitis, the superficial muscles are by that position placed in a condition of passive insufficiency and cannot act in supporting the



FIG. 1.—Extreme opisthotonos in basic meningitis.

extended position, their points of attachment being too close together. The deep muscles are hard and tender to the touch, and any attempt to overcome the extension is accompanied by great pain. There is never any position of torticollis. In some cases this tonic spasm involves the erector spinæ, partially or completely, and a most severe degree of opisthotonos may result, as, for example, in an infant of nine months in which a bald patch had been worn upon the back of the head by constant friction against the sacrum. This patient recovered completely.

The head retraction persists throughout the course of the illness in fatal cases, and in cases in which recovery occurs it slowly remits and gives place to considerable weakness of the muscles involved. There have been several theories brought forward to explain the appearance of head retraction. In the first place, it was considered to be the result of the irritation of the upper spinal nerve-roots by their involvement in the inflammatory process. Since, however, head retraction occurs when the upper cervical nerve-roots are uninvolved, and since only one set and not all of the muscles supplied by these nerve-roots are affected, while no comparable spasm is ever found in muscles supplied by the oculomotor, facial, hypoglossal, and other cranial nerves when these are involved, this explanation is obviously incorrect.

Again, arguing from Dr. Ferrier's experiment in which stimulation of the inferior vermiform process of the cerebellum by the Faradic current produced temporary head retraction and opisthotonos, it was surmised that irritation of the same structure by the inflammatory process was the essential cause of head retraction in meningitis. If this explanation be correct, the limitation of the rigidity to the neck muscles in many cases requires further explanation, which has not been forthcoming. Further, in a very large series of cases in which surgical procedures upon the inferior surface of the cerebellum and inferior vermiform process were carried out, head retraction was never observed except in a single case in which suppurative meningitis of the posterior fossa supervened.

The probable explanation is, that the meninges of a certain region being inflamed, as a protective act of Nature, that position is assumed in which the inflamed tissues are held most relaxed and kept most at rest. The phenomenon, if this explanation be correct, is strictly comparable to the fixed position met with in diseases of joints and of bones.

Vomiting is usually an early symptom, and may recur frequently throughout the illness.

Convulsions, sometimes repeated, not infrequently usher

in the symptoms, and they are prone to occur in the early stages of the malady. Later on they rarely occur, but in cases of long standing in which hydrocephalus is developing they are again liable to occur and may be of Jacksonian type. Occasionally attacks of tonic spasm (exacerbations of the tonic spasm described later) seem to take the place of clonic spasms in the early stages.

Tonic spasm constitutes one of the most characteristic features of the disease. The most constant manifestation of tonic spasm has been described already as head retraction and opisthotonos. It very commonly affects the limbs and may be present as intermittent spasm or persistent spasm.

Intermittent spasm generally occurs in those cases in which the limbs do not present persistent spasm. It is of the flexor type and affects all the limbs, the child presenting a curious doubled-up appearance. This attitude has been described as that in which the child occupies the least space (cramped position).

Persistent spasm is always of the extensor type and affects the four limbs equally. The upper extremities are strongly adducted, the forearm hyperpronated, and the wrist and fingers clenched. The lower extremities are extended at hip, knee, and ankle, and the toes are strongly flexed; less commonly, the toes are strongly extended, and then, if the spasm has persisted for some time, pes cavus is usually present. A considerable degree of subluxation at the several joints may occur when the persistent spasm is severe.

The face may show signs of the general persistent spasm. The expression is fixed and mask-like, and there is frequently retraction of the angle of the mouth. Nystagmus is the expression of similar spasticity occurring in the ocular mechanism.

Bulging of the fontanelle is not an uncommon early symptom, and it may be followed by considerable distension of the superficial veins of the scalp, sometimes amounting to a "caput medusæ," such as occurs characteristically in hydrocephalus.

*Ocular symptoms.*—Strabismus is commonly met with and is usually of a changing and transient nature. Definite ocular palsies are rare. Marked retraction of the eyelids often occurs, and with it absence of involuntary nictitation is usual. Nystagmus of the spontaneous variety is common, as is also rolling movement of the eyes. These signs must be considered as spastic signs, the ocular analogue of rigidity of limb and trunk muscles.

The pupils usually show no inequality and react normally. They tend to be contracted in the early stages and dilated in the stage of hydrocephalus. Oscillating pupil sometimes occurs. Optic neuritis is quite rare; it occurred in 8 per cent. only of the cases examined by Dr. Lees and Sir Thomas Barlow, and this fact stands out in striking distinction from the frequency of optic neuritis in tubercular meningitis.

Primary optic atrophy may occur when the stage of hydrocephalus is reached.

Transient blindness has been frequently noticed. In such cases the optic discs have been normal and the light reflex has persisted. Moreover, sight has been as a rule completely restored in those cases which recovered. It is far from easy to account adequately for the occurrence of this phenomenon; a temporary inhibition of the function of the lower visual centres by toxic products of the meningitic process has been suggested. Photophobia may be present. In a few cases a rare and destructive inflammatory condition of the eyeball, to which from its ophthalmoscopic appearance the name of "pseudoglioma" has been applied, has been met with.

Champing movements of the mouth and tongue are sometimes very conspicuous, and they may occur not only in sucklings but also in older children. Grinding of the teeth also occurs. Such movements may be met with in all cerebral diseases in childhood. They are common, for example, in diplegia, tumours involving the upper mesencephalon, and in tubercular meningitis.

The psychical state is at first one of irritability alternating

with drowsiness, and there is often much screaming and crying, presumably from acute pain. Subsequently, the condition is one of what may be called shallow coma. In the hydrocephalic stage the child is usually placid and does not cry, nor does it appear to suffer.

Respiration is at first more rapid than normal. Later in the course of the malady "cerebral grouped breathing" occurs. Five or six respirations of equal depth, and not accompanied by flushing or stertor, are followed by a comparatively long pause, at the end of which a deep sighing inspiration commences a second group of respirations, again followed by a pause. This is the form of grouped breathing common to many conditions of cerebral depression (meningitis, tumour, diplegia, hæmorrhage) and has been termed "cyclic" breathing in contradistinction to that form of breathing in which the respiration waxes and wanes between the pauses—Cheyne Stokes respiration. The latter is rarely met with in any morbid state primarily cerebral; it is usually associated with some form of toxæmia.

The pulse rate is either normal or its rapidity is increased, but a slow pulse is unusual.

Vaso-motor phenomena are rare. The *tûche cérébrale* is not often distinct and the herpetic eruptions so common in epidemic meningitis are not met with.

The temperature is moderately raised— $99^{\circ}$ – $102.5^{\circ}$  F.—and is irregular. Terminal hyperpyrexia has been often noticed, but in long-standing cases in which vitality is much reduced, the temperature may be subnormal.

Retraction of the abdomen is as rare in posterior basic meningitis as it is common in the tuberculous variety. The bowels in the later stages of the disease are obstinately confined.

*Paralysis.*—It is usually stated that paralysis is never present in the disease, and it is true that absolute loss of all movement in any part of the body is rare. There is, however, great paralysis in effect; for the child with posterior basic meningitis when general extensor spasm is persistent is much more paralysed than is the dog with his cerebral



hemispheres removed, and perhaps hardly less paralysed than is the cat in which the mesencephalon has been completely transected. Further, the decerebrate extensor rigidity which immediately ensues and persists in the cat upon such transection is probably physiologically identical with persistent extensor spasm. In the child instanced it is doubtful whether any centres situated higher than the mesencephalon are functioning. May not the same factor which causes temporary blindness also act upon the less resistant motor region of the cortex and put this out of action?

General emaciation is invariable, and it may reach an extreme degree even in cases which eventually recover.

*Hydrocephalus.*—For a description of this condition, which often supervenes in cases of posterior basic meningitis, the reader is referred to the special account of this condition.

*Duration.*—The duration of fatal cases usually is from three to fifteen weeks and the average duration eight weeks—that is, about twice as long as the average duration of cases of tuberculous meningitis.

In the series of cases reported by Dr. Lees and Sir Thomas Barlow 16 per cent. recovered completely, 50 per cent. were fatal, while in the remainder recovery was incomplete—hydrocephalus, mental defects, blindness and deafness being among the persistent effects. It is not uncommon for children who have incompletely recovered to die suddenly after a very brief illness, and upon *post-mortem* examination nothing more than slight hydrocephalus may be found.

*Causation.*—The diplococcus of Still occurs in pure growth in the exudation at the base of the brain. It is aerobic and grows readily at a temperature of 39° C. upon most solid media, and especially well upon blood agar. It is very difficult to grow upon blood serum, but it thrives upon broth and milk. The growth upon blood agar is characteristic. It is very rapid and profuse and the superficial growth resembles thick white paint, tending to bead at the margin.

The cocci vary in shape from an almost complete sphere

to a hemisphere, and they have their opposed surfaces flattened or even concave, the two being separated by a narrow, clear space. The diplococci have no distinct capsule and tend to group in pairs, so that an appearance like that of *Micrococcus tetragonus* is produced. They are only rarely found within the cells of the exudate. This organism differs from the pneumococcus in being smaller, in never developing an arrangement like a streptococcus, and in not coagulating milk. It resembles the *Diplococcus intracellularis meningitidis* of Weichselbaum, its much greater vitality, however, excepted. Dr. Still considers that his diplococcus and that of Weichselbaum are two natural varieties of the same organism.

The diplococcus of posterior basic meningitis is pathogenic for guinea-pigs, rabbits, and mice when injected into the peritoneal cavity or into the veins, but no result follows its subcutaneous introduction, thus differing widely from the pneumococcus.

*Path of access.*—In the majority of cases the organism gains access to the posterior fossa of the skull from the naso-pharynx, *viâ* the Eustachian tube, middle ear, and petro-squamosal suture, which in the first three years of life is closed by membrane only. It must be pointed out that micro-organisms may travel along apparently normal mucous membranes, and it is no argument against infection along the path above described that no abnormality of the middle ear should be found *post mortem*. Definite otitis media is found in a few of the cases only. It is most important, however, to bear in mind that in a large number of the cases the symptoms of meningitis definitely follow a catarrh of the naso-pharynx.

Posterior basic meningitis hardly ever follows chronic otitis with bone destruction.

*Etiology.*—The sexes are equally affected. The disease occurs most frequently during the first year of life, and 90 per cent. of the cases occur within the first two years of life. After the third year has passed posterior basic meningitis is rarely met with.

The seasonal prevalence of the disease in London is remarkable. A large majority of the cases occurred in the first four months of the year, and sometimes the number of cases which came under notice within a few days suggest an "epidemic incidence." Heredity seems to play no part in the causation of this malady. Inherited syphilis may be said to act as a predisposing cause only, in that it lowers the general resistance of the child to infection.

*Previous health.*—The disease generally occurs in children who are apparently healthy. Sometimes they are described as having been always weakly, and in others rickets has been present. In a large number of cases the symptoms of meningitis follow immediately upon those of nasal, bronchial, or intestinal catarrh, and in other cases it occurs in the course of those specific fevers which are commonly associated with catarrh, namely, measles, influenza, German measles, and whooping-cough. Otorrhœa, generally of short standing, is not uncommonly present in these cases.

*Injury.*—In quite a number of cases the history of a fall or a blow upon the head, often of a comparatively trifling nature, which immediately preceded the onset of symptoms, is forthcoming. Such histories are met with too often to admit of the relationship of injury to symptoms being always one of coincidence alone. A similar history is often obtained in cases of tubercular meningitis and also in cases of cerebral tumour. There can be little doubt that such injuries act as immediate exciting causes of the appearance of symptoms.

*Morbid anatomy.*—The initial seat of the inflammation is most commonly the pia arachnoid overlying the fourth ventricle; less commonly it is the pia mater, which, passing below the splenium callosi, enters the great transverse fissure of the brain as the velum interpositum, and when commencing in the latter situation it is usually much less extensive than when starting over the fourth ventricle. The pia arachnoid, at first thickened and injected, becomes soon covered with a yellowish-green coating of pure lymph, which as the inflammation spreads comes to cover the

medulla, and extending forwards along the base of the brain, fills up the hollows and invests the issuing nerves as far forward as the optic chiasma. Generally there is some affection of the membranes covering the tips of the temporal lobes upon their lower and internal surfaces. The pia upon the lower surface of the cerebellum is generally affected, and that upon the upper surface is almost always involved to a greater or less extent. Extending downwards from the medulla, the meningitis may involve any extent of the spinal cord. Sometimes the spinal cord is quite free, and in some cases when the lower part of the spinal cord and roots of the cauda equina are found covered with yellow-green lymph, the dorsal region being free, it is probable that this lymph is simply carried down into the lower theca by the current of cerebro-spinal fluid, and adheres to the spinal cord and nerve-roots. For in such cases the spinal pia to which the lymph is adherent may not show any signs of inflammation. The inflammation does not extend beyond the limits here given, never reaching the anterior part of the base or on to the vertex of the brain. Reaching the ependyma of the fourth ventricle through the posterior foramina, and the lateral and third ventricles by the great transverse fissure and the velum interpositum, the inflammatory process is said to spread throughout the ventricular system. Finally, condensation of the deposited lymph and cicatrisation of the affected pia arachnoid occur which leads to closure of the foramina of Majendie and Luschka in the roof of the fourth ventricle, through which the cerebro-spinal fluid leaves the ventricular system. The ventricular meningitis may lead by adhesion to complete obliteration of the cavity of the fourth ventricle. More frequently the aqueduct of Sylvius becomes blocked or obliterated. Further, the involvement of the velum interpositum may cause strangling of the veins of Galen. There are, then, all the factors present for the production of hydrocephalus, which condition supervenes in proportion to the severity and extent of the meningitic process and its effect in obliterating the usual channels of outflow for the cerebro-

spinal fluid and also in proportion to the prolongation of life after the disease is manifest. (See Hydrocephalus.)

There is usually considerable excess of fluid in the sub-arachnoid space at the base of the brain. The fluid is turbid yellowish-green in colour and contains flakes of purulent lymph. It is not usual for dense cicatrization to occur on the exudate investing the cranial nerves, and it is for this reason, perhaps, that cranial nerve palsies are rare in this disease as compared with the tuberculous varieties. Except in cases very early fatal the usual signs of raised intra-cranial pressure are present. The convolutions of the vertex are flattened and dry upon the surface and the theca spinalis is almost always distended in cases of any standing. Local annular obliteration of the thecal space by adhesions between the spinal cord and its membranes has also been met with.

The other organs of the body present no special peculiarity beyond extreme emaciation, except that in some cases periarticular swellings have been noticed from which, in several cases Dr. Still obtained pure cultures of the specific diplococcus.

*Diagnosis.*—In the earliest stages of posterior basic meningitis the diagnosis may be difficult. A distinction has to be made between this disease and other acute diseases the onset of which is marked by rapid rise of temperature, headache, vomiting, and convulsions. The disease cannot be distinguished from tuberculous meningitis at this stage, and it is liable to be mistaken for pneumonia, typhoid fever, and acute miliary tuberculosis.

Pneumonia is especially deceptive in that transient head retraction may occur as a symptom. Careful observation of the respiration frequency, and of the pulse-respiration ratio should lead to the avoidance of error.

If in a given case the head become strongly retracted within the first few days, the condition is probably one of otitis media or of posterior basic meningitis, and examination of the tympanic membranes will here aid the diagnosis. It should be remembered that head retraction

which is variable is rarely due to posterior basic meningitis.

At the end of the first week the question "Meningitis or not?" will become settled, and having regard to the wide difference in the prognosis that the determination whether the meningitis is simple or tuberculous entails, this question must be carefully considered. The distinction can only be made by a careful consideration of the collective symptoms and signs of the case, and not by the presence or absence of any one or two symptoms. The chief points of difference are here given in the form of a table.

	<i>Posterior basic.</i>	<i>Tuberculous.</i>
Age. . . . .	Under two years.	{ Usually over one year, often over two.
Preceding condition . . . . .	Catarrh . . . . .	—
Head retraction . . . . .	Prominent symptom . . . . .	Not marked.
Persistent tonic spasm . . . . .	Usual . . . . .	Not usual.
Nystagmus . . . . .	More marked . . . . .	Often absent.
Amaurosis . . . . .	Occurrence . . . . .	Non-occurrence.
Optic neuritis . . . . .	Rare . . . . .	Common.
Bradycardia . . . . .	Rare . . . . .	Common.
Abdominal retraction . . . . .	Never marked . . . . .	Very marked.
Hydrocephalus . . . . .	Common . . . . .	Not common.
Duration . . . . .	Longer . . . . .	Shorter.
Result . . . . .	16 per cent. recover . . . . .	Always fatal.

*Prognosis.*—In these cases the outlook is decidedly more hopeful than in cases of tuberculous meningitis. In children of over a year the prognosis is better than in infants. Unfortunately, cases very frequently come under observation in so advanced a stage of the disease as to give little hope that treatment will materially assist the healing process.

Amaurosis usually disappears completely.

It must be remembered that in cases apparently recovered a considerable degree of mental deficiency may result, and also that when fair recovery seems attained sudden and unexpected death may occur, and an unsuspected condition of ventricular distension may be found at the autopsy.

*Treatment.*—In view of the difficulty of diagnosis between

this disease in its early stages and otitis media, it seems advisable that puncture of the tympanic membrane should in all cases be performed, especially as this operation has definitely improved the general condition in cases of posterior basic meningitis, and is quite without danger when performed by skilled hands.

Vigorous use of mercury and iodide of potassium in large doses (one to three grains every two hours, even in a young infant) should be always employed.

The greatest care in the feeding and nursing of these children is necessary. Drainage of the lateral, or of the fourth ventricle in cases in which hydrocephalus has developed has not been followed by any satisfactory results at present.

#### SIMPLE VERTICAL MENINGITIS OF CHILDREN

Under this name Dr. Lees and Sir Thomas Barlow describe a form of meningitis affecting chiefly the convexity of the brain upon its anterior aspect. The posterior fossa is rarely affected. Following a stage of hyperæmia and œdema of the pia arachnoid a suppurative exudation beneath the arachnoid occurs, covering the convolutions of the affected area with a uniform yellow sheet. Inflammatory conditions of other viscera often co-exist, such as pneumonia, empyema, and pericarditis.

These cases present considerable difficulty in diagnosis. Retraction of the head and ocular symptoms are usually absent. Vomiting is less frequent than in posterior basic meningitis; convulsions and pyrexia are the rule, but in some cases they are entirely absent. The duration of the disease is generally not more than a week, and the result is always death. These authors do not attribute the disease to the organism of posterior basic meningitis, and they consider the path of infection to be from the nasal cavities to the anterior fossa of the skull. There does not seem to be sufficient reason for separating this form of meningitis from the varieties of acute suppurative meningitis to be subsequently described, and it appears to resemble clini-

cally the cases referred to later on as acute meningitis occurring in adynamic subjects and presumably due to pneumococcus infection. The term "simple" is perhaps confusing when applied to an acute suppurative process with a highly probable specific microbic origin.

#### ACUTE SUPPURATIVE MENINGITIS

This condition is met with much more frequently in childhood than in adult life. The majority of infantile cases occur in children during the first two years of life, when acute meningitis is stated to be of more common occurrence than tuberculous meningitis. It is very rare between the ages of 2 and 14 years, and at the latter period it attains its more frequent adult incidence.

Hereditary influences appear to have no relation to its occurrence. The causal factors are many, and they may be conveniently divided into three groups: (1) Certain special causes; (2) Extension to the meninges of microorganisms from areas of disease in their immediate neighbourhood; (3) Infection of the meninges as a part of a general septicæmia.

(1) *Special causes.*—These are comparatively rare and unimportant, insolation and injuries to the head being alone worthy of mention.

(2) *Extension from infective disease of neighbouring structures.*—This group comprises by far the most important causes of acute meningitis, and in comparison cases of this disease arising from other causes are rare. Septic affections of the cranial bones and of the face and scalp (wounds, furuncles, anthrax, cellulitis, impetigo) may give rise to acute meningitis, and in these conditions the infection of the meninges is preceded by a septic thrombosis of the veins draining the infected area and opening into the intracranial blood sinuses, the path of infection being the thrombosed veins.

Much more frequently the disease results from suppuration occurring in the nasal cavities, the orbit, and the



middle ear, the last being by far the most important. Sometimes the infection spreads to the meninges by vascular migration, sometimes along small veins joining the meningeal circulation. More often destruction of the bone separating the dura mater from the infected cavity allows a direct spreading of the infection into the meninges. In rare cases acute meningitis may arise from the rupture into the arachnoid space or ventricles of a cerebral or cerebellar abscess, itself the result of nasal, orbital or mastoid disease.

(3) *General diseases.*—There is, perhaps, no general infective disease that may not be complicated by acute meningitis either directly or through the advent of one of the conditions which determine meningitis.

In the first case, it is only necessary for an organism of the disease or one of secondary infection to gain access to the meninges by the blood-stream. In the second case, a suppurative otitis, for example, may arise under the influence of the primary infection, and from this focus of disease the meninges may be affected.

Among general diseases, those most frequently causing meningitis are scarlet fever, measles, smallpox, rheumatic fever, enteric fever, influenza, all kinds of pyæmic and puerperal infections, and especially pneumonia; for it has been conclusively proved that suppurative meningitis may result from infection of the meninges with the pneumococcus alone (Netter, Laveran, Leyden, and others). The meningeal infection may be pure or mixed. When pure the pneumococcus is generally the organism present; more rarely it is a streptococcus. More commonly several kinds of organisms are found in the meninges, the pneumococcus, staphylococci, streptococci and the *Bacillus coli communis* being those usually present.

*Morbid anatomy.*—The affection of the meninges may be partial or it may be general, and may involve the whole surface of the brain and spinal cord and the central ventricular system. The pia mater is especially affected, and in the early stages it is markedly injected; afterwards

there is a small-celled exudation both into the pia mater and into the substance of the underlying nervous tissues. Later, a sero-fibrinous, or more often a purulent, exudation occurs beneath the pia mater, and this collects especially along the vessels; it extends to the outer surface of the pia, filling the subarachnoid spaces; the ventricles also may be filled with pus. More often the ventricles contain an excess of clear fluid. The hemispheres of the cerebrum and cerebellum are not infrequently adherent to the dura over a limited area, which is deeply infiltrated with pus, the surrounding tissue being much softened in cases in which infection has proceeded directly from diseases of the cranial bones. As a rule, the convexity of the cerebrum is predominantly affected, the base being affected to a less degree; acute meningitis confined to the latter situation is rare.

*Clinical aspect.*—The onset of symptoms is sudden, and no prodromal signs are manifest other than those of the antecedent condition, when such is present. A severe rigor followed by high pyrexia and general or partial convulsion generally occurs. Intense headache, photophobia, and irritability are followed by delirium, often of a violent character, soon lapsing into coma. Vomiting and constipation are almost always present. The fontanelle when still present becomes markedly prominent. Coma usually makes its appearance upon the third day and persists till death, the advent of which is rarely later than the eighth day. During the comatose period certain signs make their appearance, which are common to many forms of meningitis—general extensor rigidity, head retraction, trismus, champing of the jaws, grinding of the teeth, irregular movements of the eyes, and strabismus are common symptoms. The pulse is rapid throughout. The respiratory movements are rapid, and in the final stages are apt to be irregular and cyclic. Vomiting and convulsions usually occur at intervals until the last. Sometimes periods of delirium alternate with periods of coma from the second day onwards. Optic neuritis is rarely observed, since in

most cases life is not sufficiently prolonged to allow of its development. Death occurs either during an attack of convulsions or from respiratory failure.

From the above brief description of the clinical aspect of acute meningitis it will be seen that a stage of irritative phenomena, followed by a stage of paralytic phenomena, is very striking. The two stages, however, merge very gradually the one into the other.

Special mention must be made of cases in which acute meningitis supervenes upon a grave adynamic bodily condition, for then the severe initial symptoms of acute meningitis are prone to be absent, and the patient may lapse into coma and die without any marked indication of meningitis.

The *prognosis* in this disease admits of no discussion except, perhaps, in cases in which immediately after the appearance of symptoms the obvious cause of the meningitis can be removed by surgical interference—for instance, mastoid disease. Such cases have recovered, but it is not easy to distinguish between the symptoms of severe disease of the middle ear and those of commencing meningitis, and it is only fair to acknowledge that a case which recovers under these circumstances had probably not progressed to actual meningitis.

The *diagnosis* presents little difficulty except in adynamic patients. The sudden onset of convulsion and delirium with a rigor and high pyrexia are characteristic. Further, in many cases a local cause indicating the nature of the disease, such as ozæna, orbital cellulitis, or otitis media, can be found on examination.

*Treatment.*—Any removable local cause should be at once attended to, otherwise the treatment of the disease is palliative only. The pyrexia should be treated with sponging, and ice to the head is advisable if it relieves the suffering. Bromides and chloral should be freely administered, and violent convulsions may be treated by inhalation of small quantities of chloroform. Lumbar puncture may be employed to relieve intense pain.

## EPIDEMIC CEREBRO-SPINAL MENINGITIS

### CEREBRO-SPINAL FEVER, MALIGNANT PURPURIC FEVER

THE description of this condition is usually included in that of the specific fevers. The close-parallelism, however, of this form with other forms of meningitis, both in clinical features, anatomical characteristics, and undoubted micro-organismal origin, demands its description both as a specific fever and as a variety of meningitis. A consideration of such a disease as epidemic meningitis, linking, as it were, the specific fevers as a class with conditions of meningitis as a class, serves greatly to advance our knowledge of the pathology and results of microbic infection. Since modern methods of bacteriological research have been available, the epidemics which have occurred have been few and small, and therefore our knowledge of the nature of the specific organism is scanty.

The disease was first recognised at the commencement of the nineteenth century, during the course of which two great outbreaks occurred in Europe and three in the United States. The epidemic outbreaks here lasted from three to twenty-five years, apparently becoming shorter as sanitation improved. Great Britain has suffered but little, and of recent years only small outbreaks have occurred. These have been in the following localities: Dublin, 1885-1886; Birmingham, 1876; Galston, near Kilmarnock, 1884; and at certain villages in the eastern counties, 1890.

*Etiology.*—The outbreaks have occurred during a period of mild weather, following cold, and usually in the spring. Very cold winters seemed to prevent the occurrence of the disease, which is unknown in Arctic climates. Other cli-

matic conditions seem to have no influence upon its incidence. It has no predilection for sex, but it affects chiefly children and young adults.

The manner in which the disease appears and spreads is peculiar. It tends to break out simultaneously, in disconnected areas, and does not spread either by contiguity or upon lines of traffic, neither can its origin be traced to food or water supply.

*Micro-organism.*—In almost all the cases which have been sufficiently worked out a peculiar micro-organism has been found in the cerebro-spinal fluid and meninges, considered by many competent observers to be identical with the *Diplococcus lanceolatus* of pneumonia. Other observers regard it as similar to, but not identical with that organism. The organism found in epidemic meningitis differs in many minor points from the pneumococcus, and at the present time it is difficult to be sure of the meaning of such minor differences, whether they indicate specifically distinct organisms or natural variations in virulence and mode of growth, of one species.

*Morbid anatomy.*—In epidemic meningitis there is an acute, widely-spread inflammatory condition of the pia arachnoid of both brain and spinal cord, followed after a few days by purulent effusion into the arachnoid space. Except for the general tendency to the occurrence of hæmorrhages, the *post-mortem* appearances are similar to those of suppurative meningitis.

*Symptoms and course.*—These are liable to great variations. Speaking generally, it may be said that the onset is acute, with a rigor, vomiting, high fever, depression, severe headache, delirium, and retraction of the head. Preceding the onset for a day or two, there may be rigors, malaise, and headache. The pain generally spreads down the back and to the limbs, and extreme restlessness ensues, to be followed by delirium and coma. Cutaneous eruptions, erythema, herpes, and petechial spots are common. The eyes are almost always suffused in the early stages of the disease. In the late stages destructive panophthalmitis and otitis media are common.

The duration of the illness varies greatly, from ten days to several weeks, and almost at any period recovery may set in or death may occur.

As in typical specific fevers, an abortive form, from which recovery takes place, and two malignant forms, adynamic and hæmorrhagic, occur; the first of the malignant forms characterised by sudden onset, early collapse, and severe cardio-vascular depression, the second by the early appearance of cutaneous and other hæmorrhages; both of these forms are invariably fatal. An intermittent form with some superficial resemblance to malaria is also described.

Other nervous symptoms are met with similar to those of the other forms of meningitis, but they are, as a rule, more severe.

Herpes facialis and cutaneous hæmorrhage are almost constant features of the disease. The most likely but not invariable sequelæ in cases that recover are, severe blindness, deafness, hydrocephalus, and a form of arthritis resembling pyæmic arthritis.

Recovery may take place at any stage of the disease. In the early stages it may be complete, but in the later stages some permanent disability of sight, hearing, mobility, or sensibility persists. In the late stages recovery leaves the subject hopelessly crippled with paralysis and joint disease, and generally both blind and deaf. Convalescence is generally tedious.

The *prognosis* is worse in the young and in the old; middle-aged people recover more frequently. It is influenced by the character of the prevailing epidemic, whether mild or severe, and depends largely upon the symptoms present in a given case; and as the most unfavourable symptoms there may be mentioned, absence of prodromal signs, sudden and severe onset, early coma, deep and prolonged coma, symptoms indicating widely spread involvement of both brain and spinal cord, pulmonary complications, and severe purpura.

The *diagnosis* is aided by the knowledge of a prevalent

epidemic and the distinction from other specific fevers is simple on account of the early appearance of conclusive signs of meningitis, of herpes, and of purpura.

The *treatment* that is described for other forms of meningitis applies to the epidemic variety. Pyrexia generally requires special treatment. All measures tending to produce depression and collapse must be avoided.

## SYPHILITIC MENINGITIS

It is highly probable that affections of the meninges referable to a luetic cause are not of themselves productive of symptoms in any way comparable with those making up the clinical pictures of the several forms of meningitis already described. In children who are the subjects of congenital or acquired syphilis pathological conditions of the soft meninges are not rarely met with *post mortem*. In some of these cases the meningeal affection is associated with widely spread affection of the central nervous system and symptoms arising from the former condition are consequently indistinguishable from those of the more important central nervous lesions. This condition is invariably present in adolescent general paralysis of the insane.

In some cases of acute cerebral syphilis examples of which have occurred in children nearing the age of puberty as the result of acquired syphilis, gummatous inflammation of the meninges or the presumable result of such a condition, local thickening of the meninges associated with local perivascular thickening of the vessels, has been found. The symptoms in such cases resemble those of acute cerebral tumour, and are similar to the clinical picture met with in the acute cerebral syphilis of adults.

Local meningeal thickening and adherence to the surface of the brain are frequently met with *post mortem* in the subjects of congenital lues in whom no symptoms of meningeal affection have been apparent. The soft meninges covering the vertex of the brain are most often affected, but those of the base and of the spinal cord may be involved, and both the ependyma of the choroid plexuses and of the ventricles may be affected. The pia arachnoid may



lose its usual translucency and present general opacity. It may also adhere locally to the dura or to the cerebral cortex. Local patches of thickening are of a dead white colour and vary in size from that of a lentil upwards. Such patches appear to have their starting point around a small blood-vessel as a perivascular thickening which spreads into the meninges. In these vessels endarteritis and general periarteritis may be met with. Disease of the larger blood-vessels is usually conspicuous by its absence.

Thickening and scarring of the ependyma is very marked

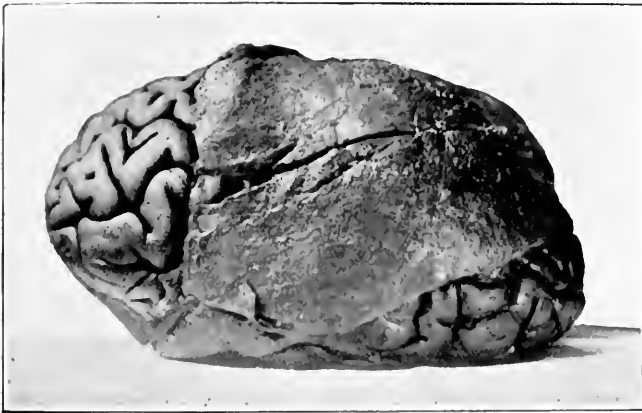


FIG. 2.—Gummatous meningitis, the result of acquired syphilis in a young adult.

in some cases. It may be distributed in patches or may be general, and in the lateral ventricles the surface may present a markedly rugose appearance. When the ependymal affection is extensive there is always some degree of hydrocephalus and in rare cases the latter may be extreme. It may be said without hesitation that syphilitic disease is the cause of a considerable number of cases of so-called idiopathic hydrocephalus. Thus in two cases cicatrization, presumably syphilitic, had obliterated the cavity of the fourth ventricle below the auditory striae, and in another case the Sylvian aqueduct was closed below the posterior commissure. In neither of these cases had there

been symptoms of meningitis, and both of them presented symptoms similar to those met with in acute cases of cerebral tumour and a rapidly fatal result from hydrocephalus occurred. The spinal meninges seem to be rarely affected in these conditions, and when so affected no symptoms referable to such affection seem to occur.

## HÆMORRHAGIC PACHYMENINGITIS

### HÆMATOMA OF THE DURA MATER

This condition is met with occasionally in children during the first three years of life, but after this period it seems not to occur. It is met with almost always as a complication of certain specific fevers and a few other diseases in subjects of poor vitality. Smallpox and typhoid fever are the most important of these, but the condition has been also found in pneumonia, tuberculosis, gastro-enteritis, and eczema. Hæmorrhagic pachymeningitis also sometimes occurs in those diseases characterised by the frequent occurrence of hæmorrhage, such as scurvy rickets, as was first pointed out by Sutherland. The disease is local in its distribution upon the inner surface of the dura mater, and it is confined to the convexity of the brain, the region of the middle meningeal arterial supply and the neighbourhood of the superior longitudinal sinus being the places where it most commonly occurs. The morbid process consists of two stages—(1) pachymeningitis, (2) hæmorrhage into the affected area of the dura.

The process commences with a separation of the endothelial lining and some of the subjacent tissue of the dura mater over a certain area subsequent to a small rounded infiltration of these tissues, and the condition may be with advantage compared to the formation of the blister which occurs in pemphigus upon the skin and mucous membranes. Upon the inner surface of the walls of the cavity thus formed layers of fibrinous deposit mixed with small round cells are deposited in the form of lamellæ which soon are found to contain numerous blood-vessels of large calibre, but having only capillary walls, and these walls are always found in a state of more or less advanced

fatty degeneration. Thus the common occurrence of considerable hæmorrhage between the lamellæ and the formation of subdural hæmatoma is easily explained.

The disease may exist in the form of a single patch of variable size, which may be unilateral or may extend across the falx and become bilateral. Sometimes there are two or more distinct patches. The extravasations of blood occurring in these areas of disease vary from the size of a lentil to that of a large mushroom, and when small they may be multiple. When examined soon after the occurrence of hæmorrhage the hæmatoma may contain liquid blood. In later stages a red clot or a laminated clot may be present, and long after the extravasation a cyst with laminated walls and containing yellowish-brown fluid is found.

*Symptomatology.*—In young cachectic children the condition occurs without symptoms, and is revealed only at the necropsy. In other cases the first symptoms to arise are those of raised intra-cranial pressure with repeated and rapidly fatal convulsions, and presumably their appearance is coincident with the occurrence of hæmorrhage; while in a few cases lassitude, severe localised headache, vomiting, myosis, and slight convulsions precede by some days or even weeks the onset of severe pressure symptoms.

This disease in children has progressed to a rapidly fatal result in those cases in which the diagnosis has been possible. But several instances of serous subdural cysts, presumably the final result of hæmorrhagic pachymeningitis, have been found in children dying from other diseases at long periods after the occurrence of cerebral symptoms. The diagnosis of the condition is often impossible, and where symptoms indicating some intra-cranial disturbance are present it is difficult, sinus thrombosis, acute meningitis, and cerebral tumour being the conditions with which it is liable to be confounded. From sinus thrombosis the differential diagnosis cannot be made except where some obvious cause for sinus thrombosis, such as ear disease, is present. The characteristic signs of meningitis, strabismus,

retraction of the abdomen, and rapid wasting are not present in hemorrhagic pachymeningitis. Cerebral tumours have a more gradual course, and are accompanied usually by optic neuritis and by signs of localising value, phenomena which do not occur in association with pachymeningitis. The course of the disease after diagnosis becomes relatively possible, is very rapid in children.

The pachymeningitis which occurs in juvenile general paralysis will be referred to in the section dealing with that disease.

## PARALYSIS OF CEREBRAL ORIGIN

ENCEPHALITIS, INFANTILE HEMIPLEGIA, PORENCEPHALY, CEREBRAL AGENESIA, CEREBRAL DIPLEGIA

CEREBRAL paralysis in childhood may be the result of the incidence of very varied lesions upon the tissues of the brain. A general description of the nature of the initial processes of those diseases that have been actually demonstrated and those which are theoretically probable is of service in facilitating the understanding of the several clinical types, and aids in the comprehension of the many individual cases met with which are apparently intermediate forms between two or more of the definite clinical entities described in the following pages.

Bearing in mind that the tissues of the brain consist of (1) the essential nerve-cells and their processes, the dendrites and the nerve-fibres, (2) the supporting neuroglia, and (3) very numerous capillaries, vessels, and lymphatics, it is in the first place possible to conceive of an inflammatory lesion local in origin and either limited in distribution or widely spread. The incidence of this inflammatory lesion is upon all the elements of the brain tissue simultaneously. It is not primarily incident upon the vessels, although these show the earliest changes that can be recognised by the microscope, and throughout the course of the inflammatory process the vascular changes—changes in the vessel-wall and its immediate neighbourhood—minute areas of thrombosis and minute hæmorrhages may be by far the most noticeable histological alterations. Nor is the inflammatory process primarily incident upon the neuroglia. It must be remembered that the neuroglia

does not show such marked changes as the result of inflammation as do the connective tissues generally. Further, the essential nerve-cells are not the elements first affected, but the changes that are noticeable in these suggest in the first place the effects of poisons acting upon the cells and in the later stages the effects of interference with their blood supply.

As to the essential nature of this inflammatory process, there is little doubt that it is in all cases the result of the advent of micro-organisms to the brain tissue. The organisms that produce non-suppurative inflammation in the brain are probably of a special kind, and our knowledge of them is scanty. When the common organisms of suppuration gain access to the brain tissue an inflammatory process such as is above described arises and the result is either abscess formation or suppurative meningo-encephalitis. In the other case simple encephalitis ensues and may result either in recovery, partial or complete, cicatrization, porencephaly, cyst formation, or in death. Of such a nature are cases of acute encephalitis and many cases of infantile hemiplegia.

The most commonly occurring vascular lesion is thrombosis, which is usually arterial, often venous, and sometimes both arterial and venous. Such thrombosis is due sometimes to primary changes in the vessel walls; more often it is the result of certain blood states and of marasmus.

Embolism is an occasional but not a common cause of cerebral lesions in fetal life and during the first six years of childhood.

Hæmorrhage as a primary condition is of rare occurrence. The majority of cases of cerebral hæmorrhage in children are, as in adults, the result of a rupture of a vessel in a small focus of previously occurring thrombosis, and are comparable as regards cause with that pulmonary hæmorrhage which occurs in phthisis. Such hæmorrhage in children is usually the result of syphilitic vascular disease.

Meningeal hæmorrhage has been proved to be of frequent occurrence during birth, but any case in which symptoms

have undoubtedly arisen therefrom is as yet lacking in the records of the subject.

Among vascular lesions may be also placed the occasional results of instrumental delivery and dystocia, which amount in nearly all cases to actual tissue disruption with consequent hemorrhage and softening. In such vascular lesions many cases of infantile hemiplegia and monoplegia and cases of bilateral spastic hemiplegia have their origin.

The result of vascular blocking is peculiar when this occurs in childhood. Softening in the form met with in adults rarely occurs. Atrophy without death of the supporting tissue is the rule, and the evascularised convolutions, still retaining their form and colour, shrink and become hard, and the sulci are widened. Microscopic examination reveals absence of the nerve elements and relative increase of the glial tissue.

Sometimes complete absorption of the damaged tissue occurs, and the area of such absorption may be so exactly limited from the surrounding healthy brain tissue that an excavation may result, clearly cut as with a cheese-scoop. The condition is known as porencephaly.

Of lesions primarily affecting the neuroglia we know nothing. The commonly occurring condition of atrophic sclerosis (causing cerebral diplegia) has been attributed by Richardière to primary overgrowth of the neuroglia, but recent investigations do not support the view.

Primary change in the nerve-cells of the central nervous system is the initial lesion in that large group of maladies included under the term "cerebral diplegia." Seeking further for the presumable causes of such cell degeneration, we find that toxic influences probably stand first, while in those cases which are congenital, maternal morbid states are in all likelihood the potent cause.

In other cases hereditary tendencies and parental diseases and post-toxic states impart to the nerve-cells a condition of diminished potentiality in the tenure of life, and after a period of apparently normal vitality the limit of life of such cells is prematurely reached and degeneration



sets in. This condition has been termed by Sir William Gowers *abiotrophy*, and it may make its appearance at any period of existence even when it is inherited, and the life of the cells may cease at any period from early foetal life till middle age. In the majority of cases abiotrophy is confined chiefly to one physiological system of cells and generally to those concerned with the higher functions of the nervous system—such, for example, as the pyramidal cells. Often, however, the cells of more than one system are involved and every degree of involvement of the several systems exists until a class is reached in which “all the cells of the nervous system are affected,” as is the case in amaurotic family idiocy.

The appearance of the convolutions of the cerebral and cerebellar cortex where such cell-atrophy has occurred is remarkable. The sulci are widened, the convolutions stand away from one another and are unduly firm, while frequently the cerebral convolutions present a faceted appearance comparable with the surface of hammered silver. Sometimes they present a worm-eaten appearance. The term “walnut” type aptly describes the appearance of the brain as a whole. The arrangement of the convolutions varies according to the period at which atrophy takes place. If this occur during foetal life, the development of the part ceases or is retarded and a primitive form of sulcine arrangement is met with; while if the atrophy be of post-natal onset, a normal form of arrangement will be found.

*Agnesia*.—According to the period in the development of the nervous system at which a lesion occurs, whatever its nature, an arrest or diminution of the development of the parts concerned occurs. For example, if at the third month of foetal life a lesion of the central cortex occurs, the corresponding pyramidal tract never develops or never reaches a considerable degree of development. If the lesion occur before the complete development of that tract, further development of that tract ceases. Not only do the elements directly concerned in the lesion suffer arrest of development, but also elements in close physiological

connection with them fail to develop to the normal extent. Thus, damage to one side of the cerebrum, if extensive, entails arrest of development of the opposite half of the cerebellum.

The cerebral palsies of childhood fall into two chief groups : (1) those in which the paralysis is confined to one side of the body—hemiplegic forms—and (2) those in which the whole body is more or less affected—diplegic forms. The hemiplegic forms and the diplegic forms are for the most part distinct pathological and clinical entities, the former being mostly diseases of post-natal onset and due to gross lesions of the brain, the latter, on the other hand, being often of congenital origin and due to widely spread cell-death in the cerebral cortex unassociated with any gross anatomical lesion. In certain cases, however, these groups come into close resemblance clinically ; for example, cases of infantile hemiplegia, arising in children who were born healthy and remained so usually for years, are for the most occasioned by a unilateral focus of encephalitis or by a unilateral vascular lesion, but in some cases symmetrical and bilateral lesions of a similar nature occur, and the clinical picture produced is with difficulty distinguished from that of cerebral diplegia. Conversely, the paralysis in a case of cerebral diplegia may be much more marked upon one side of the body than upon the other, and if marked amelioration in the symptoms take place, the resulting condition, years after the onset, may be one of unilateral paralysis. It will be of advantage in this place to set forth briefly the reasons for considering hemiplegic forms and diplegic forms as distinct diseases :—

The mode of onset and the course of the disease are different in the two forms.

The hemiplegic forms are characterised by a preponderance of paralysis over rigidity upon the side affected, whereas in the diplegic forms there is a preponderance of rigidity or of perverse movement (athetosis, choreiform movements, or myoclonic movements) over paralysis.

Cerebral diplegia frequently occurs as a familial

disease, several children of the same mother being affected, but hemiplegic forms are never met with in these families, nor does infantile hemiplegia occur as a familial disease.

The anatomical characteristics found *post mortem* differ in the two groups, gross lesions being found in the hemiplegic group and widely spread cortical cell death in the diplegic cases.

According to the position of the lesion in infantile hemiplegia and according to the major incidence of the cell-death in cerebral diplegia, the clinical aspect of the disease may vary, and consequently in connection with both infantile hemiplegia and cerebral diplegia forms departing widely from the usual clinical type may occur. A focus of encephalitis causing permanent damage may occur either in the frontal, central, parietal, occipital, or temporal region of the hemisphere. Directly after the occurrence of the lesion in all probability there will be hemiplegia, wherever the situation of the lesion; but when time has elapsed it is only when the lesion involves the central convolutions that hemiplegia will remain. If the lesion be in the pre-frontal region defective intelligence and asymmetrical cranium, and if the lesion be upon the left side backwardness in speech, will be the most marked symptoms, possibly associated with slight weakness of the lower face as the only remaining link binding this variety of case to the ordinary form of infantile hemiplegia, though the two are pathologically the same diseases. Similarly, if the lesion be situated in the parietal or temporal lobes, mental deficiency is the only lasting result. Such lesions seem to be very rare in the posterior part of the hemisphere, but cases are on record of hemianopia associated with hemiplegia, or of hemianopia alone clinically comparable with cases of infantile hemiplegia. And there is little doubt that similar lesions of an acute character occur in the cerebellum and give rise to ataxic symptoms and others showing interference with the functions of that structure.

Among cases of cerebral diplegia a similar difficulty exists in that according to the situation of the major incidence and the degree of the cell atrophy and in accordance with the physiological function of the groups of cells that may be affected, widely different clinical phenomena may result from one and the same morbid process. As examples, the whole cerebral cortex may be equally and severely affected when the most complete diplegia and mental deficiency result; or the central regions may be predominantly involved and then a severe form of diplegia results—the mental faculties remaining relatively intact. Again, the frontal and occipital regions are often affected out of all proportion to the central region, and the clinical picture is then one of idiocy, with slight generalised rigidity. Again, the atrophy of the cortical cells may be almost entirely limited to the paracentral and marginal convolutions, and a condition of bilateral spastic paralysis of the legs resembling paraplegia of spinal origin results. Further, the destruction of cells in the cortex may involve certain cells only, and the clinical result is, not actual paralysis, but a condition of more or less marked spasticity associated with perverse movements—athetosis, choreiform movements or myoclonus.

The varieties of infantile cerebral palsy that may be distinguished clinically are as follows:—

Hemiplegic forms, resulting from encephalitis, vascular lesions, and injury . . . . .	{ Acute encephalitis, infantile hemiplegia, infantile mono- plegia, idiocy, with cranial asymmetry, choreiform hemi- paresis, hemiathetosis.
Diplegic forms, resulting from death of cortical cells . . . . .	
	{ Generalised rigidity, idiocy with general rigidity, paraplegic rigidity, spastic facial diplegia, amaurotic family idiocy, bi- lateral athetosis, choreiform diplegia, epileptiform myo- clonus.

## INFANTILE HEMIPLEGIA

THE majority of cases of infantile hemiplegia are examples of a disease peculiar to children, and *sui generis*, to which no comparable disease occurs in adults. The majority of cases of the disease may be likened to the anterior poliomyelitis of children, in that the two diseases are the results of acute inflammatory lesions occurring in the grey matter of the cerebral and cerebellar cortex and in the grey matter of the spinal cord respectively. Beyond doubt both anterior poliomyelitis and encephalitis are the result of infective processes, but of the nature of the organisms responsible we know nothing. It has been suggested that these two diseases are the result of one pathological process, having its incidence at one time upon the cerebral cortex, at another upon the grey matter of the spinal cord. This is not improbable; for although affection of the brain and of the spinal cord has not been found to concur in one case, as might be expected were the conditions the result of cerebral and spinal foci of the same disease, yet in one instance of simultaneous onset in two members of a family one was affected with the spinal disease, the other with the cerebral.

The clinical picture of the commencement of encephalitis or infantile hemiplegia has no parallel in the hemiplegia of adults. It must be summed up as follows:

A child hitherto healthy and without hereditary stigmata suddenly becomes ill without any apparent cause, between the ages of one month and six years. The early symptoms are severe, and consist of fever, convulsion, often vomiting and coma. These symptoms persist from twenty-four hours to a week; and towards the end of the period, as the

general condition improves, hemiplegia is noticed to come on, first in the face, then in the arm, and lastly in the leg. The hemiplegia becomes severe and the limbs remain flaccid for some days, after which the flaccidity begins to lessen and the limbs become spastic. In some cases the paralysis disappears completely. In most cases a greater or less degree of paralysis or perversity of movement remains.

In another class of cases the hemiplegia becomes manifest in the course of certain known specific fevers, such as measles or scarlet fever or diphtheria. It seems probable that the majority of such cases are the result of encephalitis in relation to which the specific fever stands as a predisposing cause only. Some cases, however, are certainly the result of thrombosis of the cerebral vessels, and this is especially prone to occur when the subject of the specific fever is marantic. Others are the result of embolism from endocarditis, which has arisen as a complication of the specific fever.

In a small group of cases severe injuries to the head occurring at birth, not necessarily from instrumental delivery, or injuries from a blow or fall on the head after birth, are responsible, the lesion in these cases being one of bruising or actual rupture of the brain tissue.

Again, in whooping-cough or during convulsion rupture of a cerebral vessel may occur during the paroxysm, and hæmorrhage may be responsible for the production of infantile hemiplegia.

Lastly, meningeal hæmorrhage occurring at birth has been a commonly attributed factor.

*Etiology—Age.*—The onset of the disease occurs within the first three years of life in two thirds of all cases. A few cases are of pre-natal origin. In others the paralysis is first noticed shortly after birth. In many of these cases it is difficult to determine from the history whether the child presented any noticeable disability at birth or not. After the sixth year the disease is rare. It may be remarked with justice that many of the cases of hemiplegia occurring

after the sixth year are due, not to encephalitis, but to vascular lesions, embolism, and thrombosis. Of 594 cases collected by Freund the age distribution was as follows: Congenital, 76 cases; 1st year, 162; 2nd year, 139; 3rd year, 81; 4th year, 36; 5th year, 26; 6th to 10th year, 75 cases.

*Sex.*—It would appear that the sexes are equally affected, but it has been held by some authorities that males suffer more frequently than females.

*Heredity* does not seem to play an important part in the etiology of the disease. A history of neuropathic heredity is almost always wanting. More important, however, are alcoholism and syphilis in the parents. While manifestations of syphilis are rare in children who are the subjects of the disease, a history of parental syphilis is very common. Its connection with the disease is probably in some cases more than that of a predisposing cause which lowers the vitality and therefore the resistance of the child to infective processes, for occasionally choroidal atrophy of syphilitic origin is present.

If cases of infantile hemiplegia be divided into three groups according to the time of occurrence of the lesion, namely (1) those of prenatal origin, (2) those occurring during the process of birth, (3) those occurring after birth, such a classification will separate the cases etiologically; for the pre-natally occurring cases will be of thrombotic origin, and will have as their most frequent cause endarteritis referable to maternal syphilis, those occurring at birth will be due to injury during parturition, while the post-natal cases are referred to encephalitis.

*Antecedent infective diseases.*—In about one third of all cases of infantile hemiplegia the disease appears in the course of an attack of one of the definitely known specific fevers. By far the most important of such fevers are scarlet fever and measles, the other fevers which are sometimes complicated by the occurrence of hemiplegia being smallpox, rotheln, whooping-cough, diphtheria, dysentery, pneumonia, typhus, typhoid, mumps, malaria, chorea, and endocarditis.

The clinical picture of the condition occurring in association with these acute diseases is exactly similar to that of infantile hemiplegia resulting from primary acute encephalitis. In a number of cases, and especially in those associated with scarlet fever, endocarditis is also present, and an embolic lesion has been found *post mortem*, while in marasmic subjects a thrombosis of cerebral veins has occasionally been found. In the majority of cases, however, where opportunity has been afforded for the examination of the brain soon after the onset of the disease, a condition of encephalitis has been met with, similar to that found in cases in which the malady has been primary and unassociated with a specific fever. It seems highly probable that this encephalitis is due to a secondary infection of the brain tissue with organisms of a similar nature to those responsible for primary encephalitis, and that the specific fever acts as a predisposing cause by lowering the vitality and the resistance of tissues to infection.

When encephalitis occurs primarily there seems good reason to suspect that it is the result of the invasion of the brain tissue by organisms of a specific nature; those organisms have not been isolated or cultivated. Yet the sudden onset of the malady in healthy children, with accompanying symptoms such as are usual in specific infective diseases, its short course and rapid amelioration, and the inflammatory nature of the lesion leave no alternative but to consider this disease as of specific micro-organismal origin in the large majority of post-natally occurring cases.

It may here be remarked that acute encephalitis has frequently been described apart from the subject of infantile hemiplegia, but without sufficient warrant. The most severe cases, in which death takes place within a fortnight of the onset, and where perhaps the patient has lain in a condition of more or less profound coma during the second week of the illness, may never present demonstrable hemiplegia, which would be evident in a less severe case. And on the other hand, less severe cases, in which



hemiplegia becomes apparent, are not often fatal. It is for these reasons that the conditions of acute encephalitis and infantile hemiplegia have been described separately, as if they stood in no relation the one to the other.

There has been much discussion as to the connection of hereditary syphilis with infantile hemiplegia. Many of the congenital cases are the result of maternal syphilis, and the cerebral lesion has been proved to be syphilitic disease and thrombosis of the middle cerebral arteries, or of the veins entering the superior longitudinal sinus. A similar condition of thrombosis has been met with in a few cases of post-natal onset which presented distinctive signs of hereditary lues. But, as has been mentioned above, encephalitis and infantile hemiplegia are only rarely found in the subjects of congenital syphilis. Their incidence is upon previously healthy children. It has been pointed out, however, by Freud that a large proportion of the parents of the patients have suffered with syphilis, though their children have shown no taint. Such a fact cannot be used as a sound basis of argument in favour of a syphilitic origin for encephalitis, and it may be stated confidently that while syphilitic vascular disease is responsible for many congenitally determined cases and for a few cases occurring after birth, infantile hemiplegia resulting from encephalitis has no causal connection with syphilis. This fact stands out in striking distinction to the important relation of syphilis to the diplegic forms of infantile cerebral palsy.

In quite a large number of cases a history of shock or fright or a blow upon the head has immediately preceded the onset of symptoms. The connection is too striking to be one of coincidence only, and it is probable that these events stand in the relation of immediate exciting causes of the appearance of symptoms. A similar relation has been noted in connection with the occurrence of intracranial tumour and tuberculous meningitis.

*Clinical aspect.*—In two thirds of all cases of infantile hemiplegia the disease is ushered in by convulsion, which may be frequently repeated during a period of from twelve

to forty-eight hours. Following the initial convulsion, pyrexia, often reaching a high degree, and vomiting frequently occur. Coma may come on after a single initial convulsion, when the patient gradually becomes drowsy and sinks into unconsciousness. More often it appears after several convulsive attacks have occurred. In rare cases convulsions may be absent, and drowsiness, gradually deepening into coma and associated with pyrexia, are the only initial manifestations of the disease.

In a great many cases the convulsions are confined to that side upon which the hemiplegia subsequently appears; they are of the usual epileptiform type—a tonic stage being followed by a clonic stage—ending in exhaustion, with coma and stertor. The duration of the convulsion varies from a single attack lasting a few minutes to a rapid sequence of convulsions lasting twenty-four hours, without return to consciousness between the attacks. Sometimes the child dies in this condition, which may be termed a *status epilepticus*.

The time at which the hemiplegia becomes apparent and the relation of its appearance to the occurrence of convulsion vary in different cases. It may be immediately apparent and severe in degree directly after the occurrence of the initial convulsion or convulsions, upon the first or second day of the illness. In other cases the initial convulsions are followed by a slight hemiparesis, which becomes more marked after the occurrence of another attack, and is profound only after repeated batches of convulsions. More commonly the convulsions which occur during the first three days of the disease leave no sign of paralysis, but the paralysis appears towards the end of a week, either suddenly after a further attack of convulsion, or gradually as the patient is recovering from the comatose state.

The hemiplegia occurs in an almost equal number of cases upon the right and upon the left side. Of 692 cases collected by Freud 374 were right-sided, and 318 cases were left-sided.

The paralysis is at its onset a flaccid one, and in almost

every case it involves the whole of one side of the body to a greater or smaller extent. An initial monoplegia is of extreme rarity; and though where considerable amelioration has taken place severe paralysis may be confined to one limb, yet the face and trunk and the other limb of the same side in most cases show some defect of motility. The paralysis reaches the height of its intensity between the end of the first week and the end of the second week. Subsequently it lessens, in some cases rapidly disappearing, either completely, or leaving only some slight disability in using the hand. In other cases the hemiplegia remains unaltered. Between these two extremes every degree exists. No trace of the paralysis may be noticeable three months after the onset in a very favourable case.

The distribution of the paralysis differs somewhat from the hemiplegia of adults.

The face and the trunk do not show much loss of voluntary movement. The arm is affected to a greater extent than is the leg, and the peripheral segments of the limb are much more severely paralysed than are the proximal joints.

The *face* upon the paralysed side is weak, and there is generally slow movement and over-action in volitional movements. It is stiff and somewhat expressionless and the palpebral aperture is generally wider than upon the sound side. It is in mimetic and emotional movements that the facial paralysis shows itself especially. When the child laughs and cries the affected side of the face is conspicuously paralysed. Such paralysis of emotional movements with retention of volitional movements occurring in adults has been associated by von Bechterew with deeply seated lesions in the region of the thalamus; but no such connection exists in infantile hemiplegia, for loss of emotional movements with preservation of volitional movement has been repeatedly observed in cases of infantile hemiplegia when the necropsy has shown that the lesion was confined to the cortex of the motor region.

Associated with the facial paresis choreiform and athetotic movements are often found.

Sometimes both sides of the face show evidences of spastic paresis, perhaps with choreiform or athetotic movements, notwithstanding the fact that the lesion is strictly confined to one cerebral hemisphere. This condition is probably the result of the innervation of the muscles subserving the bilaterally associated movements of the face from each cerebral hemisphere.

In some few cases the facial paresis does not clear up to the same extent as does the palsy of the limbs, and it may remain as almost the sole sign of an antecedent infantile hemiplegia.

The *upper extremity* is almost always more severely paralysed than the lower extremity, and the palsy is progressively more marked towards the periphery of the limb. There is always associated vaso-motor palsy, the limb presenting a characteristic red-blue appearance.

Stunting of growth is the rule, and choreiform and athetotic movements are of more common occurrence than in the leg or face. Contracture develops early and the limb assumes a characteristic attitude. The upper arm is adducted and the elbow is thrown slightly across the body and is pressed into the trunk. The forearm is flexed upon the upper arm at about a right angle and is semi-pronated; the hand is flexed at the wrist and deviates to the ulnar side. The thumb is flexed into the palm and the fingers are closed over it.

The *leg* is for the most part less affected than the arm, and it is most paralysed in the periphery. In the rest position it will be found flexed and rotated in at the hip and slightly flexed at the knee. The foot is dropped, and either presents a well-marked pes cavus, with the toes extended at right angles to the line of the metatarsus, or there is a position of equino-varus with the toes flexed. The leg presents the same stunting of growth and vaso-motor palsy as does the arm. Choreiform movements and athetosis are much less common than in the arm.

In many cases the opposite leg shows slight signs of

spasticity in that an increase of the reflexes and slight contracture or pes cavus may exist. Sometimes the opposite leg is affected to such an extent as to make progression almost impossible.

The gait resembles that of adult hemiplegia. The limb is swung round at the hip, and is often brought across the body too much and the knee may be raised too high. Since the limb is almost always shorter upon the side of the hemiplegia, the patient walks upon the toes of that foot and frequently there is compensatory tilting of the pelvis and lateral spinal curvature.

The contractures which occur when paralysis persists in marked degree come on early and are severe. The stage is soon reached at which they cannot be overcome by force even during profound anaesthesia. Subsequently, under the influence of growth, unalterable distortions of bones and joints occur.

The amount of volitional movement that can be accomplished in such spastic and contracted limbs is sometimes truly remarkable. A leg that appears practically useless when the patient is lying in bed, intensely rigid and contracted in the hemiplegic position, may be quite a serviceable member. The gradual assumption of the hemiplegic position directly it ceases to be used is also remarkable. The deep reflexes may be diminished or lost in the first few days following the onset of the illness; after this they are almost invariably increased. Foot clonus is sometimes, but not always, to be obtained. The plantar reflex upon the hemiplegic side is nearly always of the extensor type, and if there be marked implication of the opposite leg, the plantar reflex on that side may also be of the extensor type. In some of the most severe cases, however, when the foot is severely paralysed and very rigid, a flexor type of plantar reflex upon the paralysed side is met with. It is curious to note two conditions with which the presence of a flexor plantar reflex upon the paralysed side may be associated. The one is in cases in which the hemiplegia is of congenital origin or

has occurred in the first year of life ; the other is when marked athetosis of the leg is present.

The sphincters are not affected after the severe symptoms of the onset have passed off.

Hemianopia is not an uncommon symptom, but it is easily overlooked on account of the difficulty of testing the visual fields in young children. It may occur as a transient symptom, to disappear rapidly when the paralysis begins to improve : and in this case its presence does not indicate a direct implication of the occipital lobe in the lesion, but it is the result of a temporary disturbance of function of the whole cerebral hemisphere, which may take place on the occurrence of an acute lesion anywhere within the hemisphere. In other cases the hemianopia is permanent and this is evidence that there is permanent damage to the visual cortex, and when this occurs the child usually comes to hold the head rotated to the right or left, according to the side upon which the visual field is lost.

The best method of determining the presence of hemianopia is to move an object which the child has seen and desires to possess across the visual field. The expression of doubt and bewilderment when the longed-for object disappears into the blind half of the field is very striking.

Hemianæsthesia occurs about as frequently and in the same form as in the hemiplegia of adults. In children as in the latter it is often slight and soon lessens or disappears. In younger children the difficulty of testing the tactile sensibility offers an almost insuperable obstacle to investigation.

Interference with speech is of very common occurrence and it may take several forms, the deductions as to localisation of the lesion and prognosis being different in each case. The following are the chief varieties :

1. Aphasia may occur when the speech centres are involved in the lesion, but it is different from the aphasia of adults, for it is the result of a lesion in undeveloped structures, and it is never permanent. It is commonly associated with right hemiplegia, but it is much more

commonly associated with left hemiplegia in children than in adults. It would appear that the speech centres in the left cerebral hemisphere have not acquired such a dominant position over those of the right cerebral hemispheres in children, and that a lesion of these centres in the right hemisphere is sufficient to produce a severe temporary aphasia. Out of 324 cases of hemiplegia associated with aphasia in children 200 were cases of right hemiplegia and 124 were cases of left hemiplegia. Doubtless a few of the cases of left hemiplegia occurred in children hereditarily left-handed.

2. Aphasia may occur as a rapidly evanescent phenomenon from a lesion of either hemisphere not involving the speech centres. It is here one of the phenomena of the widely-spread disturbances of cerebral function which are liable to occur temporarily as a result of a local cerebral lesion, whatever be the situation of the latter.

3. Children who have recently acquired speech may lose that faculty as the result of the general depression produced by any acute illness, such as a specific fever or an attack of encephalitis.

4. When an attack of infantile hemiplegia occurs before speech is learnt the child is backward in learning to speak, and this deficiency is the more marked when the speech centres (especially those of the left hemisphere) are involved in the lesion and most marked when there is associated mental reduction.

Aphasia occurring in children is always of a temporary nature provided that fair intelligence remain and provided bilateral lesions of the speech centres do not exist.

*Post-hemiplegic disorders of movement.*—Perverse movements of the face and limbs of the hemiplegic side are very common sequelae in cases of infantile hemiplegia.

They fall into two chief classes: (1) Defects accompanying voluntary movements and (2) spontaneous involuntary movements. The defects accompanying voluntary movements do not stand in direct relation to the degree of paralysis present, but they appear as a rule only when consider-

able voluntary movement is possible. The most important of these disorders are rigidity, ataxy, and choreiform movements. Notwithstanding that voluntary movements are possible, they are executed slowly and stiffly from the persistent spasm of the muscles. This stiffness of movements is more marked after the muscles have been at rest some time. To a certain extent it wears off with prolonged exertion, to again reappear in its full intensity after rest. Again, ataxy is a common post-hemiplegic trouble, showing itself in unsteadiness, uncertainty, and clumsiness of movement. The probable explanation of such ataxy is that as a result of a partial destruction of the cerebral centres concerned in a movement, there is unequal distribution of innervation to the muscles. Further, in many cases the attempt to perform a certain movement is accompanied by shock-like jerking movements, "choreiform movements," which interfere greatly with accomplishment of the desired act.

Such disorders of voluntary movement are much more frequent in the upper than in the lower extremity. Ataxy and choreiform movements are always most marked in the hand and in the performance of the finer co-ordinate movements; choreic and athetotic movements are also met with in the face.

Spontaneous involuntary movements are of three kinds: (1) sharp shock-like or twitching movements—choreiform movements; (2) slow lasting rhythmic movements which may be likened to the movement of the tentacles of an octopus—athetosis; and (3) almost every variety of fine or coarse tremor.

(1) Choreiform movements make their appearance in cases in which a moderate degree of volitional movement is possible, and spastic contracture is not severe. The stage of the malady at which they appear varies in different cases. Commonly they appear some time after the onset of the hemiplegia when paralysis and spasticity are abating. In other cases they appear coincidentally with spasticity at the end of the initial flaccid stage of the palsy; when this is so, it sometimes happens that they are developed in the arm



alone, while spasticity appears in the leg alone. Such spontaneous movements are of serious prognostic importance, for they tend to become progressively more apparent and to render the effectual use of the limb impossible.

Under the name of choreiform movements are included movements which present almost every gradation between the shock-like movements of myoclonus, the movements seen in Sydenham's chorea, and the slow tentacle-like



FIG. 3.—To illustrate position of upper limb in athetosis. From a photograph by Dr. James Collier.

movements of athetosis. The short shock-like movements generally affect the proximal joints of a limb, while choreiform and athetotic movements are seen chiefly in the periphery of the limb.

(2) Athetosis occurs more frequently than does choreiform involuntary movement; sometimes the two occur in the same case, athetosis presenting in the leg, while choreiform movements are seen in the arm. Unlike the choreiform movements, athetosis often shows itself in the periphery of a limb which is much paralysed and very rigid. Such

rigid athetosis is usually confined to the forearm and hand, less frequently to the leg and foot. It does not become noticeable until long after the onset of the hemiplegia, and is for the most part a persistent symptom. Further, its appearance usually denotes that no further recovery of voluntary power will take place in the affected limb.

Just as a condition of choreiform hemiparesis may develop upon the clearing up of an initial hemiplegia, so may



FIG. 4.—Another position of upper limb in athetosis. From a photograph by Dr. James Collier.

a condition of generally distributed hemiathetosis develop in the absence of any great degree of paralysis or rigidity, and in such cases volitional movement is accompanied by much athetotic movement.

The nature of these movements is in all cases the same. The movement is slow, rhythmic, intermittent, and uncontrollable, and is not dependent upon reflex causes. In the face it takes the form of a series of grimaces affecting the lower part of the face rather than the upper.

(3) Tremor of almost every variety may be met with in

the hemiplegic limbs. It is chiefly seen in the hand, and when paralysis is incomplete. It may occur during rest, but more often accompanies or follows exertion. Simple tremor, either fine or coarse, is common. Sometimes rhythmic tremor resembling that met with in paralysis agitans occurs, and intention tremor comparable with the jerking often seen in cases of disseminated sclerosis may occur. It is not possible to make a clear distinction between the latter and the ataxic movements which have been already described in connection with the disorders of voluntary motion in infantile hemiplegia.

*Trophic disturbance and arrest of development.*—In every case of infantile hemiplegia in which symptoms are permanent, trophic disturbances occur affecting the growth and development of the soft parts, and also of the bones and joints upon the affected side. The hemiatrophy thus arising is of a double nature. In the first place, atrophy of the muscles, often of considerable degree, rapidly follows the onset of the disease, and continues to progress until voluntary power is regained, if that event takes place. In the second place, the cerebral lesion, in accordance with its persistence and the amount of destruction of cerebral tissue entailed, causes partial arrest of development upon the opposite side of the body.

Deformity of the skull is often the result of the adaptation of the brain-case to the loss of cerebral substance at the site of the lesion. Sometimes the flattening of the skull over the damaged region of the brain is very conspicuous. This is not infrequently the case when porencephaly is present, and sometimes the bone may be actually absent over the porencephalic defect.

The contour of the skull does not always, however, follow that of the brain when the latter is locally atrophied, and in cases in which very great atrophy of part of one hemisphere exists the skull may show no striking asymmetry, since fluid accumulating between the membranes may compensate for the loss of brain tissue. Careful craniometric observations will show in nearly every case of in-

faulile hemiplegia that the skull case is smaller in the region of the affected hemisphere. It should be borne in mind that an extensive lesion of one cerebral hemisphere occurring in early childhood entails arrested development of the opposite half of the cerebellum, and that corresponding flattening of the skull in the latter situation is likely to occur. The face participates markedly in this hemiatrophy, in some cases being smaller than upon the sound side, and both the eye and ear may be distinctly smaller upon the affected side. The half of the trunk is often smaller upon the hemiplegic side, and a comparison between the scapulæ or the halves of the pelvis shows a striking inequality. There is frequently a scoliosis, with the concavity to the affected side, due in part to the hemiatrophy, and also to the non-use of the affected side. The mamma and testicle are sometimes found strikingly smaller than on the sound side. The limbs show varying degrees of muscular atrophy and smallness and shortness of the bones, together with the results of growth in bones and joints which are subject to the constant strain of the contracted muscles. In marked cases the limbs are cold and blue from deficient vascular innervation.

In a few cases fat hypertrophy is met with similar to that which is often met with in a limb paralysed from infantile poliomyelitis—the limbs appear of normal size, but the place of the muscular tissue has been taken by fat. When athetosis and chorea are present (especially in the hemiparetic form) there may be actual hypertrophy of the muscles. Changes in the skin of the periphery and in the nails are commonly met with, and are probably referable rather to the presence of vascular paralysis than to a cerebral trophic influence.

The temperature upon the paralysed side is always lower than that upon the unaffected side.

The trophic disturbances which occur in infantile hemiplegia cannot be considered as atrophy from disuse. They are almost certainly the direct result of the lack of cerebral innervation. The atrophy affects, not only the

motor structures, but also the bones, connective tissue, and glands; moreover, the degree of atrophy is never in proportion to the degree of paralysis, the most marked hemiatrophy being not infrequently met with in cases in which the paresis is quite slight. Neither has the atrophy any strict relation to the clinical type of the disease, whether spastic, choreiform, or athetotic, and it would be more correct to speak of it as arrested development.

*Epileptic attacks.*—About one half of all cases of infantile hemiplegia are the subjects of recurring epileptic attacks. Such attacks commence in a large number of the cases long (one to ten years) after the onset of the hemiplegia, and are repeated. In a few cases the attacks recur from the time of the onset. Very remarkable are the occasional occurrence of unilateral convulsions prior to the onset of the hemiplegia, and they occur upon the side which is subsequently hemiplegic.

Epileptic attacks following infantile hemiplegia are not so severe as those of idiopathic epilepsy. They are always unilateral in commencement, and often strictly confined to the paralysed side. A peripheral sensory warning is usually present. The initial cry is rare and consciousness is often retained in the fit or if lost it is lost late. Post-epileptic coma and automatism are rare, but occasionally occur.

Mental deficiency is met with in all degrees, from slight loss of intellectual acuity to complete idiocy. The impairment of the intellectual faculties is in relation to the position and extent of the cerebral cortex which is involved in the lesion, and is especially met with when the pre-frontal and temporal lobes are affected.

*Morbid anatomy.*—The condition of the brain in a large majority of the cases of infantile hemiplegia, in which opportunity for *post-mortem* examination has been afforded, has been investigated at a period long after the time of occurrence of the primary lesion. The anatomical changes that have been found are, in the order of frequency of their occurrence:

- (1) Circumscribed atrophic sclerosis.

(2) Cyst formation.

(3) Superficial shrunken patches resembling wet wash-leather. In the two latter conditions there is always associated some degree of atrophic sclerosis of the convolutions in their immediate neighbourhood.

(4) Porencephaly.

From the character of such final lesions alone it is often impossible to determine what has been the nature of the original process of which the lesion in its final form is the result. It has been already stated that the tissues of the infantile brain tend to undergo a process of sclerosis and atrophy, with increase of the glial elements, as the result of defective blood-supply or inflammatory processes, rather than a process of softening, as is the rule in the brains of adults similarly affected. Consequently a process of acute encephalitis, as the inflammatory process lessens and ceases, is followed by increase of the glial structures, and entails death of the nerve elements wholly or in part, the final result being atrophic sclerosis.

Again, as the result of vascular obliteration, either by embolism or thrombosis, the nerve elements perish, but the neuroglia retains its vitality in most cases. Fresh blood-vessels are developed and a condition of atrophic sclerosis again results. So also, as the result of the bruising of cerebral tissue that may occur during the process of birth, atrophic sclerosis may arise.

On the other hand, an intense inflammatory process such as acute encephalitis may cause actual death of all the elements, including the glia, and considerable extravasation of blood may occur into the inflamed tissue, with the result that a cyst is formed. Such a cyst was found in one case twenty-one days after the onset of acute encephalitis. Though atrophy and sclerosis are the usual results of vascular obliteration, yet it sometimes happens that softening and cyst formation take place. This is especially the case when a central vessel not supplying the cortex is obstructed. Cerebral hæmorrhage frequently ends in cyst formation, and if it be borne in mind that such hæmorrhage

commonly occurs in an area of antecedent thrombosis, the presence of atrophic sclerosis in the neighbourhood of the cyst will be intelligible.

From the above it is clear that a condition of atrophic sclerosis may have its origin in acute encephalitis, or in embolism, thrombosis (arterial or venous), or injury, and further, that cyst formation may similarly be the result of encephalitis, embolism, thrombosis, or hæmorrhage.

The superficial area of softening—the wet wash-leather patch (*plaque jaune*) is invariably the result of vascular obliteration.

The origin of porencephaly is somewhat difficult to determine. It has been proved to be the result of embolism in some cases, and it has been produced experimentally in animals by ligation of cerebral vessels. It is certain that many cases are the results of local cerebral lesions occurring in early foetal life, for other developmental peculiarities of the brain often co-exist. All cases of porencephaly are not, however, congenital, and it seems highly probable that the post-natal cases have their origin in vascular obliteration.

*The initial pathological process.*—During the last few years a considerable number of cases have been placed on record in which an anatomical investigation was made soon after the onset of the disease. The conditions found have been, according to their order of frequency :

1. Acute encephalitis.
2. Thrombosis, arterial or venous.
3. Hæmorrhage.
4. Embolism.

The lesion of acute encephalitis a few days after its occurrence presents the following appearances. Over a part of one hemisphere the convolutions present a dark, somewhat plum-coloured appearance, and where this affected part joins the non-affected part of the hemisphere this colour fades gradually into that of the normal cerebral tissue. The convolutions are swollen and slightly softer than normal. The dura mater and arachnoid are normal over the affected area, but small spots of sub-pial hæmor-

rhage are frequently seen. Upon section the same purple colour is seen extending through the grey matter and affecting the underlying white matter to a variable extent, but a focus of encephalitis does not as a rule spread deeply into the white matter; its major incidence is upon the cortex. Upon the surfaces of the sections many minute hæmorrhages are seen, and dark streaks, which are minute vessels in a state of thrombosis. Sometimes a portion of the affected area may be almost diffuent, or a cavity may be present beneath the cortex containing softened brain tissue and extravasated blood.

Microscopical examination shows a condition typical of inflammation; there is marked perivascular infiltration of round cells. Many of the finer vessels are thrombosed and multitudinous small foci of hæmorrhage are seen. The nerve-cells are swollen and distorted, the tigroid substance is seen in various stages of fragmentation and disappearance and the nuclei are eccentric and have often disappeared.

The *post-mortem* appearances of recent thrombosis, embolism, and hæmorrhage do not need a separate description in this place. Thrombosis of the cerebral veins is met with chiefly in the superficial veins, which pour their contents into the superior longitudinal sinus. It may or may not be associated with thrombosis of the sinus itself.

Arterial disease in children is often confined to the smaller vessels, and care must be taken that a judgment be not formed as to the presence or absence of arterial disease from the examination of the vessels at the base of the brain; for these may be absolutely free from disease even to the third or fourth order of their branching when the most extensive disease of the walls exists beyond. Attention may be again drawn to the dependence of cerebral hæmorrhage in many cases upon local degeneration of the walls of a vessel as it passes through a small focus of softening due to antecedent thrombosis.

As the result of the processes that have been above described, tissue absorption, sclerosis, cyst formation occur,



together with interference with the further development of those parts of the central nervous system which are in close physiological connection with the damaged area of the hemisphere. Consequently as the final condition of the affected cerebral hemisphere there is found either local atrophic sclerosis, cyst formation, patches—the result of softening or porencephaly, while not unfrequently a combination of these lesions co-exists.

#### ATROPHIC SCLEROSIS

In this condition the brain as a whole is of less than the normal size and the affected cerebral hemisphere is smaller than the other. Over a certain area of this hemisphere the surface is shrunken and the convolutions are small and separated by wide sulci. These gyri are hard to the touch and are of a tough leather consistency when cut.

The surface of the affected convolutions may present a faceted or worm-eaten appearance. The apex of each gyrus as seen in cross section is sharp and the shape of the gyrus may be compared to that of a knife rest. There may be some thickening of the membranes over this area and the sinking of the surface of the hemisphere is compensated for by an accumulation of cerebro-spinal fluid between the membranes. At the head of the atrophic area the shrunken gyri merge gradually into the surrounding convolutions. The corresponding portion of the corona radiata and internal capsule are reduced in size and there is usually some compensatory enlargement of the lateral ventricle in the neighbourhood of the lesion. Sometimes the superficial yellow-brown patches of a consistence like that of wet wash-leather already referred to, exist in the atrophic region, and small multiple cysts may be found in the substance of the atrophied tissue, and these are, in all probability, produced by enlargement of the lymph spaces compensatory to the shrinking of the tissues. The arrangement of the atrophied convolutions varies according to the period at which the initial lesion occurs. If a very primitive arrangement

of the gyri obtain in the atrophied area it is certain that the lesion occurred in early foetal life, whereas if the shrunken gyri do not differ in arrangement from the normal the lesion did not occur until that period of post-natal life at which the brain reached its complete external development. Owing to the effect of the shrinking of the atrophic patch upon the surrounding convolutions it is usual for the sulci separating the latter to run a somewhat straight course into the hollow and present a radiate (cart-wheel) arrangement.

Microscopically the atrophic convolutions show a partial or complete absence of the nerve elements according to the severity of the atrophy, these elements gradually reappearing at the edge of the affected area. The glial tissue is much increased both relatively and absolutely. The blood-vessels are numerous, but they are of finer calibre than those of the unaffected convolutions. Their walls do not present pathological changes, and it is probable that they are for the most part vessels which have been formed subsequently to the occurrence of the lesion.

*Cyst formation.*—Cysts and patches of softening are usually associated with some degree of atrophic sclerosis. Cysts as a rule extend deeply into the centrum ovale and do not reach the surface of the hemisphere, but sometimes their chief extent is superficial and they may be confined externally by the pia mater alone. Such cysts present a smooth lining membrane and their contents consist of a yellowish-green fluid.

#### PORENCEPHALY

This term implies an absence of the whole thickness of the brain tissue forming the walls of the lateral ventricle over a particular area. There are three varieties of such defects :

(1) A cavity may exist in the thickness of the cerebral hemisphere bounded externally by the pia mater and internally by the ependyma of the ventricle. It contains a

fluid comparable with the fluid of the serous spaces. There is no distinction of this form of cavity from a cyst except its greater size.

(2) The cavity is bounded externally by the pia arachnoid, but internally it communicates freely with the ventricle.

(3) The cavity communicates freely both with the ventricular cavity and with the sub-arachnoid space.

The two latter varieties of porencephaly are not infrequently associated with the presence of other developmental defects in the central nervous system, such as syringomyelia, microgyria, and spina bifida. Sometimes a condition of hydrocephalus co-exists.

The condition of the pyramidal tracts and of the other fibre systems leaving the cortex cerebri, such as the fronto-tectal and fronto-pontine tracts and temporo-pontine tracts, etc., in cases of infantile hemiplegia, varies according to the period of fetal or post-natal life at which the lesion occurs, and according to the position, extent, and severity of the lesion. A lesion occurring before the development of these tracts entails their non-development. Subsequently occurring lesions are associated with varying degrees of arrest of development in its several stages and secondary degeneration of these tracts. Incomplete lesions in which some of the nerve elements retain their vitality in the affected area of the cortex, cause arrest of development and degeneration in those fibres whose cells of origin have been destroyed. An extensive lesion of the central region of one hemisphere occurring in early childhood is associated with interference with the development of the crus cerebri, the pons, and the medulla upon the same side and of the cerebellum upon the opposite side.

*Course of the disease.*—In a small proportion of cases the patient does not survive the initial manifestations of the disease, the widely spread interference with cerebral function caused by a severe and extensive lesion being incompatible with life. Under such circumstances death takes place usually in the second or third week of the disease. In

certain of the cases which arise in the course of a specific fever death may occur from the cerebral condition or from some other complication of the specific fever. Apart from these occurrences infantile hemiplegia has little tendency to destroy life.

In many cases improvement begins to take place within a few days and the hemiplegia rapidly disappears, no sign perhaps remaining six weeks after the onset. Such cases may never present any subsequent defect referable to the disease, but there is always the liability in the first place that the temporary interference with the function of one cerebral hemisphere may result in some imperfection in the development of the opposite half of the body—shortness of the arm and leg; and in the second place there is always a tendency to the occurrence and recurrence of epilepsy.

In a large majority of cases the initial flaccid hemiplegia gives place to a slowly improving spastic hemiplegia. The amelioration is more marked in the face, trunk, and proximal joints than in the distal joints and usually more marked in the leg than in the arm. With increasing recovery of power, perversity of movement, stiffness and slowness, ataxy, choreic movement, and tremor become apparent and interfere seriously with the necessary re-education of the limbs.

Spontaneous movements come on in most cases within a year of the onset—athetosis in a much paralysed limb, choreiform movements in a slightly paralysed limb. With the onset of athetosis improvement ceases in the region in which the rhythmic movement is manifest.

As the growth of the body proceeds the retarded development of the affected side commences to show itself. While it is usually most marked in those parts which are most paralysed, it may be very apparent when the paralysis is slight. Otherwise the degree of the retardation of development that will result finally depends upon the age at which the disease is manifest; the younger the child the greater will be the inequality between the two sides of the body when the age of puberty is reached, other circumstances being equal.

The subjects of infantile hemiplegia are very prone to the occurrence and recurrence of epilepsy, nor does this tendency disappear, though no attack may have occurred, till several years after the age of puberty is reached.

The majority of the cases in which paralysis persists show some degree of mental deficiency, but this is regressive except when severe in degree, especially if careful education be afforded.

Cases of hemichorea and hemiathetosis show this difference from cases of infantile hemiplegia, that they tend to become progressive until the patient is completely incapacitated upon the affected side. This rule is not invariable, however, for some cases improve remarkably.

*Diagnosis.*—The onset of acute encephalitis with convulsions, vomiting, and fever resembles closely the mode of onset of many of the specific fevers and of meningitis. From the specific fevers it may be distinguished by the prolonged coma, by the absence of characteristic signs of the specific fevers, and by the appearance of hemiplegia.

The onset of encephalitis is more acute than the onset of tuberculous meningitis and the prodromata of the latter disease are absent. From acute meningitis, infective sinus thrombosis, and cerebral abscess, the distinction may be very difficult in the absence of any discoverable local cause for the latter conditions.

The onset of acute encephalitis closely resembles also a severe attack of simple infantile convulsions. The presence of rickets, of peripheral irritation, or some adequate cause for convulsion, together with the severity of the coma and the results of the treatment usual in infantile convulsion, should serve to prevent errors of diagnosis.

When the hemiplegia is established, there is little difficulty in diagnosis, the history and non-progressive nature of the malady at once separating the disease from the other common form of hemiplegia in children, namely, that resulting from cerebral tumour. With hemiplegic chorea infantile hemiplegia can hardly be confused.

The disease has been confounded with infantile paralysis

of unilateral distribution ; the rigidity, the presence of perverse or spontaneous movements, preservation of electrical excitability in the affected muscles, and retention of the deep reflexes are the obvious distinguishing signs.

Choreiform hemiparesis and hemiathetosis do not resemble any other conditions and can be at once recognised by the nature of the spontaneous movements.

*Prognosis.*—It has been already stated that infantile hemiplegia has little tendency to destroy life except at the onset of those cases which are due to acute encephalitis, when a fatal result occasionally occurs. The subjects of infantile hemiplegia in whom the mental state is much reduced are rather more prone to the occurrence of intercurrent maladies than are normal children. The prognosis as regards bodily capacity cannot be made until some time after the onset of the hemiplegia, when the progress of improvement has been observed. In many cases the hemiplegia disappears completely within three months and the prognosis as regards bodily capacity is good, but these reservations must always be made: (1) That although the paralysis has disappeared, some impairment of development of the formerly affected side of the body may possibly result; (2) that epilepsy is very likely to come on at some period before the age of twenty years; (3) that the mental acuity may be somewhat below the normal.

When paralysis has persisted months or years after the onset in acquired cases or after birth in congenital cases, the prognosis depends upon the nature of the paralysis, the presence or absence of chorea and athetosis, the age of the patient, and lastly upon the mental state.

If the paralysis be nowhere complete, if the age of the patient be not advanced and if the mental state be good, great improvement will certainly take place if appropriate and prolonged physical training can be administered. Marked mental deficiency renders physical training impossible, and as age advances the capacity for increasing function of the maimed nervous system lessens with each year. Even when paralysis is complete in the hand or foot several

years must elapse before a certain statement can be made that improvement will not take place and some useful voluntary power be regained. The presence of athetosis in a severely paralysed extremity is of certain unfavourable prognosis.

The presence of perversity of volitional movement is not necessarily unfavourable, but it renders the training more difficult and improvement is slower.

Choreiform and athetotic hemiparesis are nearly always of unfavourable prognosis. Slight conditions of mental reduction are susceptible of great improvement under the influence of careful training. Severe degrees of mental impairment render the outlook as regards capacity hopeless. The presence of epilepsy does not influence the improvement that may occur in physical and mental capacity. The attacks are not as a rule refractory to the ordinary treatment of epilepsy.

*Treatment.*—We know of no measures that avail to prevent the occurrence or lessen the severity of the hemiplegia in cases of encephalitis. Doubtless the damage to the cerebrum has happened as soon as a diagnosis is possible, and the treatment to be adopted in the early days is that suitable for any acute febrile disturbance.

When the paralysis has developed, treatment is to be directed to the prevention of rigidity, the regaining of voluntary control, and the improvement of the mental acuity. There is perhaps no disease which demands greater patience and persistency in the carrying out of suitable treatment, and there are few diseases in which more brilliant results are produced from apparently hopeless cases, by pertinacity in treatment, than infantile hemiplegia.

On the other hand, it is a disease in which treatment is for the most part neglected, especially in the early years of the disease, when good results are more readily and quickly obtained. From the first, regular massage and passive movement of the affected limbs should be employed, and bearing in mind the tendency to vaso-motor palsy, the affected limbs should be clothed in woollen garments. As

soon as some voluntary power is regained it should be encouraged as far as possible. It is often necessary to tie up the sound arm in order to compel the child to make what use he can of the crippled limb. He must also be regularly assisted in re-learning to walk. Subsequently, as power and movement increase, gymnastic exercises of every kind and Fraenkel's exercises should be employed. Rigid apparatus for the prevention of contracture and for the alteration of deformity are harmful, for they increase the weight of the limb, and interfere with movement, which is the remedy by which paralysis is to be combated. Similarly a heavy boot should never be worn upon the affected side. The mother of a patient, for reasons of economy, usually loads the paralysed foot with a large hob-nailed boot—large because it is more easily put on to the pes cavus, hob-nailed because "he wears the boot out so quickly at the toe."

In regular cases, where there is much contracture and deformity, tenotomy is of great service, and it should be soon followed by passive movements. It need hardly be said that tenotomy is useless unless some voluntary power be present in the muscles, the tendons of which are to be divided.

Of great importance is attention to the mental training of the children, for they are frequently backward, and careful and individual teaching is necessary.

Recurring convulsions should be treated as "idiopathic epilepsy."



## CEREBRAL DIPLEGIA

The various diseases which are grouped together under this title are the results of widely-spread changes in the nerve-cells of the cortex of both cerebral hemispheres. Presumably such changes may be caused by harmful or traumatic influences exerted upon the elements of the cerebral cortex at any epoch during fœtal life, during the process of birth, or during post-natal existence. Some hereditary taint or toxic influence may deprive the cortical elements of their developmental capacity or of their capacity for maintaining a long tenure of life (abiotrophy). In the latter case the symptoms resulting from the degeneration become apparent without any visible causal antecedent at some time remote from that at which the essential cause of the degeneration was in action. Hence it is probable that in certain cases of post-natal cerebral diplegia the disease is not acquired, but is in reality the result of an inherited defect. In the familial varieties of the disease this is of frequent occurrence. For example, when several children of the same mother are affected, it sometimes happens that while some of them are born paralysed, others develop paralysis soon after birth, while others may show no defect until comparatively late in childhood. In such a condition one cannot but refer all the cases to one and the same cause, and that a congenital one.

The varieties of cerebral diplegia present so widely different clinical aspects that some justification is needed for the plan of grouping together such apparently dissimilar conditions as paraplegic rigidity, bilateral athetosis, and amaurotic family idiocy. The explanation is that, according to the incidence and extent and degree of the

degeneration upon the cells of the cortex, the resulting clinical picture varies greatly. Thus the cell affection may be confined almost entirely to those elements of the pyramidal system which complete their development latest. For this reason these elements seem especially prone to suffer from harmful influences occurring during the last two months of foetal life. The resulting condition is paraplegic rigidity, or Little's disease, cerebral spastic paralysis. Again, the whole pyramidal system may be extensively affected, and the result is "generalised rigidity." If the cell affection be partial in the motor region of the cortex, chronic and athetotic diplegia are produced. A predominant incidence of the lesion upon the frontal and occipital lobes results in various combinations of idiocy and spastic paralysis. Lastly, the affection of the nerve-cells may not be confined to the cortex, but may affect all the cells of the central nervous system, as is the case in amaurotic family idiocy.

The nature of the lesion in cerebral diplegia explains the occurrence of almost every transitional form between the clinical types of the disease as tabulated below. Every variety of combination of the symptoms—rigidity, athetosis, and mental reduction—are met with.

*Clinical aspect.*—In the majority of cases of generalised and paraplegic rigidity, and in athetotic and choreic diplegia the symptoms are noticed soon after birth. In severe cases the nurse in washing the child is often the first to notice the stiffness of the limbs or the regular assumption of a curious bodily attitude. In less severe cases the abnormalities are not obtrusive until the child begins to get about, when perverse movement, rigidity, and *pes cavus* are often the first symptoms that excite the attention of the parents. In other cases backwardness in learning to walk and talk and mental deficiency first suggest that something is wrong with the child.

In cases in which the symptoms arise after birth these may appear during convalescence from an acute disease, and are then usually preceded by convulsions, and the first noticeable symptom is rigidity. In other cases an acute

febrile disturbance associated with convulsions precedes the appearance of symptoms. Sometimes repeated convulsions without febrile disturbance occur, after which the symptoms of diplegia gradually appear.

In other cases the onset is accompanied neither by febrile symptoms nor by convulsion, but the disease comes on insidiously, as do such diseases as myopathy and Friedreich's disease, suggesting an abiotrophic pathology. Such an onset is the rule after the sixth year of life.

*Age.*—In the majority of cases the disease is apparent at birth or as soon after birth as the infant commences to perform such movements as render clumsiness and rigidity observable. In less severe cases of paraplegic rigidity the defect may not be noticed until the child commences to walk. In the family types of the disease, however, the child is healthy until some time after birth. In amaurotic idiocy the disease makes its appearance during the first year, usually between the third and the sixth month. In other varieties of familial diplegia the symptoms appear later in childhood, usually between the sixth year and puberty. Following acute diseases cerebral diplegia occurs most frequently between the second and the fifth year, but it may show itself at any time until the age of puberty is reached.

*Sex.*—The sexes are usually equally affected. It has been stated that in cases resulting from birth injuries males are more liable to be affected than are females, on account of the greater size of the fœtal head in males. This was strikingly illustrated in a series of cases seen by the writer, but several authorities deny its importance.

The clinical picture of the several forms of cerebral diplegia presents a combination in varying degrees of certain characteristic symptoms always bilaterally distributed, though often more severe on one side of the body than on the other. These symptoms are: muscular rigidity, paresis, perverse movement, contractures, and increased deep reflexes. Other important and characteristic symptoms are mental deficiency, optic atrophy, ocular palsy, and bulbar paresis.

The clinical aspect of amaurotic idiocy and epileptiform myoclonus differs greatly from that of the more usual types, and will subsequently be described apart.

*Generalised rigidity.*—As its name implies, the chief characteristic of this type of diplegia is a condition of muscular rigidity associated with weakness, affecting to a greater or less extent all the muscles of the body. Except in the most severe cases in which the weakness amounts to



FIG. 5.—Cross-legged progression in cerebral diplegia. From a photograph by Dr. Collier.

complete paralysis there is more rigidity than weakness and in some cases it is astonishing that there should be so much power in the presence of such a degree of rigidity. The lower extremities are generally most affected, the upper to a less degree, and the facial region still less. Paroxysmal exacerbations of the rigidity have been noticed by some writers. Movement is slow and clumsy, and often athetotic and choreic movements are present in the limbs. If the patient is able to walk, the gait is digitigrade from contracture of the calf muscles; the knees are slightly

flexed from contracture of the hamstrings; the thighs are rotated inwards and the knees pressed close together, rubbing against one another with each projection of the limb. More severe adductor spasm gives rise commonly to the "cross-legged" progression.

The assumption of the sitting position may be impossible in marked cases, when such a patient is placed in a chair. Owing to the rigidity preventing the necessary flexion at the junction of the lower extremities with the trunk, in the most severe cases the patient lies in a log-like fashion, unable to turn or move in bed, and then the rigidity may reach such a degree that he may be lifted by the heels in one rigid piece, the weight of the body being supported on the back of the head. In this stage, owing to muscular contractures, certain characteristic attitudes are assumed which will be described later on. The knee-jerks are exaggerated, ankle clonus may be present, but is not invariable, and the plantar response is extensor in type.

*Mental symptoms.*—Every degree of mental defect is met with in these cases, from precocity and slight mental dullness to complete amentia. As a rule the average degree of mental impairment is more profound than in the other types of diplegia, but it does not always correspond with the severity of the rigidity and of the paralysis, for the most profound mental deficiency may exist in cases in which the rigidity and paralysis are quite slight. This depends upon the greater affection of the pre-frontal lobes. The amount of mental impairment cannot be estimated from the facial expression, as conditions of facial over-action and rigidity, perverse mimetic or athetoid movements of the face may lead to an opinion that there is much greater mental impairment than exists in reality. The mental state is, as a rule, placid. Where the affection is slight the patients are occasionally irritable, mischievous, and passionate.

*Speech defects.*—In congenital cases, and in cases occurring before the eighteenth month, speech may never be acquired, or may be much delayed, and if acquired it is

often imperfect. Articulation is defective, and slowness of speech, with great facial over-action, slurring, and stammering, often occur. When the disease arises after speech is acquired, it is generally completely lost, at least temporarily, if the child be younger than six years of age, and if the case be at all severe. The loss of speech may be quite gradual, and where some intelligence persists the understanding of words may remain in patients who can utter no articulate sound.

Under this heading may be mentioned the curious and characteristic manner in which children affected with severe generalised rigidity cry. They give vent to a series of hoarse prolonged roars very unlike the screaming, sobbing, and broken respiration of a normal child.

Convulsive attacks are very common in cases of generalised rigidity. In about one half of all cases general convulsions associated with loss of consciousness occur at intervals, and in one eighth of the cases epilepsy becomes established. The occurrence of such attacks bears no relation to the severity of the other symptoms.

*Special senses.*—Primary optic atrophy occurs in a small number of cases, and it is bilateral, rarely unilateral. It may produce complete blindness, and has been shown to result from a primary degeneration of the nerve elements of the retina similar to that occurring in the cerebral cortex.

*Cranial nerves.*—Inequality of the pupils and slowness of the light reaction are not uncommon, and oscillating pupil is often met with. Spontaneous nystagmus of very wide range is common in cases in which the face is markedly affected. It is to be regarded as the spastic ocular sign analogous to the rigidity elsewhere, and produced in the same manner. Convergent strabismus occurs in about 30 per cent. of the cases. More rarely divergent strabismus is present, and in a few cases paralysis of the third or of the sixth nerve is found.

In nearly all cases the face shows signs of paresis and rigidity. With slight involvement of the face this may be shown by a general loss of emotional expression, slight

retraction of the eyelids, and some retraction of the angles of the mouth. With more severe involvement of the face, an unvarying mask-like expression, with wide palpebral apertures and a large open mouth, are present. Slobbering is common. In some cases involuntary facial over-action is present and gives rise to varying grimaces on attempts to move the face or on speaking. Athetoid movements of the face are uncommon. The tongue is often very large and the hard palate very much arched and the presence of adenoid vegetations in the pharynx is frequently to be observed. Difficulty of swallowing is present in the large majority of cases, though it may be slight. On watching the act of deglutition in these cases the difficulty seems due to rigidity of the muscular apparatus rather than to paralysis. Cases with severe difficulty in swallowing have been described by Oppenheim under the name of "infantile pseudo-bulbar paralysis." There may be difficulty with protrusion of the tongue, and this organ usually shares in the facial athetosis, if this is present, and laryngeal difficulties—dysphonia, stridor, etc.—have been noticed.

*Conformation of the skull.*—In congenital cases some degree of microcephaly is generally present. Asymmetry and flattening of the region of the central convolutions are common, and in cases of slight generalised rigidity with profound mental changes the frontal region may be small and flat proportionately to the rest of the skull. In a case of slight generalised rigidity under the writer's care, in which speech was retarded till the ninth year, great flattening of the left frontal region and of the right cerebellar region was present.

*Attitude.*—In the more severe cases characteristic attitudes are assumed. The head may be rigidly retracted, but more commonly the chin is pressed down upon the chest, and torticollis has been noticed. The spinal column generally shows kyphosis, sometimes lordosis, and often scoliosis. The cases presenting lordosis usually have head retraction, and those with kyphosis a flexed position of the head.

Several characteristic attitudes have been described and

named, and these occur with sufficient frequency to deserve special note.

(1) *The cramped attitude.*—The head and trunk are bent forward in kyphosis. The elbows are pressed into the sides and the forearms crossed on the chest in contracture. There is contracture at the hip and knee joints. The position may be described as that in which the patient occupies the least possible space.

(2) *The attitude of adoration.*—The head is retracted and the eyes are staring. There is lordosis. The elbows are pressed into the sides and the hands are held supinated below the face. The legs are rigidly extended.

(3) *The extension-supination position.*—The limbs are rigidly extended and the forearms are fully supinated. More rarely extreme pronation is associated with the extended position of the limbs.

*Pes cavus* is present in most of the cases, but some cases show a condition of *talipes equino-varus*. *Genu recurvatum* sometimes occurs, and this is usually the case when paralysis is complete and rigidity extreme.

The mode of progression in these cases, where any is possible, is characteristic, and has been compared with that of the tardigrade animals. The "cross-legged" progression is so well known as to need no description here. Where the legs are so severely affected as to render walking impossible, while fair power remains in the upper limbs, the patient may crawl along the ground with the upper limbs dragging the useless lower limbs and trunk.

*Muscles.*—In some cases a curious hardness and resistant feeling of the affected muscles is present resembling a similar condition in pseudo-hypertrophic paralysis. If athetosis is associated with the rigidity, there may be actual hypertrophy of the muscles.

In late stages of severe generalised rigidity the muscles are always wasted, and both the myotatic irritability and electrical excitability may disappear. Fibrillary twitchings of the muscles are common, especially in cases approaching puberty.



*Perverse movements.*—Under this heading must be grouped the very constant maladroitness of movement, the facial overaction and grimacing in speech, and in mimetic expression, choreic movements, athetotic movement, and intention tremor. Athetotic movements sometimes are present in the face, choreic movements rarely, if ever. Not rarely choreic or athetotic movements are present in the upper extremities, with slight rigidity, while severe rigidity affects the lower limbs.

Choreic movements may develop *pari passu* with the rigidity, or generalised rigidity may clear up for the most part, leaving a choreic diplegia, but athetotic movements always appear at a later date than rigidity.

Tremor precisely resembling that seen in disseminated sclerosis is sometimes seen in limbs in which the rigidity is slight, and it is likely that some of the cases described as those of disseminated sclerosis in children are in reality cases of diplegia.

Common sensation and the muscular sense are unimpaired in the cases in which their accurate investigation is possible. In the severe cases there is very great pain on passive flexion of the rigid joints, probably owing to the formation of rest adhesions. Trophic changes are confined to a stunting of growth, but this is not nearly so well marked in the diplegias as in infantile hemiplegia and in bilateral spastic hemiplegia. In diplegia the bilaterality of the distribution renders it difficult to estimate. The sphincters are unaffected. The deep reflexes are exaggerated, but are difficult to obtain in limbs which are very rigid, and for this reason foot clonus is not generally obtained. Of the superficial reflexes, those of the trunk may be absent in marked cases. The plantar reflexes have nearly always shown an extensor response.

*Atypical mental forms.*—These cases are placed under a special heading because of their peculiar clinical aspect. They present slight symptoms of generalised rigidity with profound mental changes which generally amount to complete idiocy, and in these cases the incidence

of the cortical lesion is chiefly upon the frontal convolutions.

*Paraplegic rigidity—Little's disease.*—This form of diplegia is always of pre-natal origin, and is the variety most frequently associated with premature or difficult labour.

The clinical aspect differs from that of generalised rigidity only in that the rigidity and paresis are confined almost entirely to the lower extremities. According to the severity of the affection walking is impossible or progression is tardigrade or of a "crossed-legged" type (see Fig. 5). Adductor and extensor spasm of the legs and the presence of pes cavus is the rule and there is sometimes hyper-extension of the knees. Frequently slight defects of mobility exist in the upper extremities and face, and from this condition every gradation exists between paraplegic and general rigidity. In one third of the cases convulsions occur and convergent strabismus is also present. Mental defects, difficulty of speech, and dysphagia are of frequent occurrence, but usually these are not so marked as in general rigidity, and are often conspicuously absent.

*Bilateral athetosis and choreic diplegia.*—These may be described as conditions of slight generalised rigidity in which uncontrollable spontaneous movements and great irregularity of volitional movement occur. The rigidity and paresis may be so unobtrusive and the spontaneous movements so incessant and universal, that the clinical aspect may show little to connect these conditions with the other forms of cerebral diplegia.

Bilateral athetosis and choreic diplegia are only separated as types by the form and extent of the spontaneous movements, and both forms of movement may occur in the same subject. The movements in bilateral athetosis are not so slow or so like tentacle movements as are the athetotic movements of infantile hemiplegia, and may be described as having partly the character of the squirming movements of chorea and partly the character of the rhythmic tentacle movement of athetosis. Not only do the move-

ments occur spontaneously, but all voluntary movements may present the same perversity.

The movements of bilateral athetosis may affect all the voluntary muscles of the body. Sometimes the movements may be confined to the lower extremities or to the upper extremities. Not rarely the movements are first noticed in one limb and spread to the other limb of the same side and then to the opposite limbs. As a rule the movements are more marked in the distal than in the proximal joints of the limb. The muscles of the trunk and face are frequently affected, and in the latter region the movements take the form of spontaneous grimacing and great facial overaction and grimacing upon voluntary movement. The tongue when at rest in the mouth and when protruded may show obvious athetosis, and when the affection is severe great inconvenience may be experienced in taking food. The movements in bilateral athetosis are as a rule increased by volitional movement and they cease during sleep, but some exceptions to the latter rule have been recorded.

The movements of choreic diplegia are not sharply distinguished from those of bilateral athetosis, and range between the squirming movements of true chorea and the sharp shock-like movements of myoclonus. Marked unsteadiness on voluntary movement is associated. Such choreic movements are seen chiefly in the limbs and at the proximal joints rather than at the distal joints. They are, perhaps, never seen in the face.

The same mental changes, affection of speech, nystagmus, optic atrophy, and dysphagia as have been described in "generalised rigidity" occur in bilateral athetosis and in choreic diplegia. A few points, however, demand special notice. Even in the most severe cases of bilateral athetosis the mental changes may be very slight. Convulsions are common in choreic diplegia, but are very rare in cases of bilateral athetosis. In both types intention tremor is not infrequently noticed. In athetosis actual hypertrophy of the muscles is sometimes seen.

*Course of the disease.*—In cases in which the condition is

present at birth the course of the disease varies greatly. In many cases of generalised rigidity and most cases of paraplegic rigidity the disease is not progressive, and many of the cases show a tendency to a slow amelioration of the rigidity and an increase of voluntary power and control of the muscles, especially under the influence of careful physical training. It may be said that all cases of paraplegic rigidity tend to improve, if the mental acuity is not seriously reduced, and if appropriate physical training be employed early in the case. On the other hand, many cases of generalised rigidity show a tendency to become progressively worse, the rigidity increases and voluntary power decreases, the patient becomes bedridden, and much reduced mentally. Progressive emaciation comes on, and death occurs either from exhaustion or from broncho-pneumonia, which is often brought about by the passage of food into the trachea, owing to the great difficulty in feeding the child which pharyngeal rigidity and paresis entail. It is usual for cases of a progressive nature to end fatally before the end of the fourth year of life.

Bilateral athetosis and choreiform diplegia, as a rule, present a slowly progressive course, but there is no tendency towards a fatal result.

Speaking generally of cases of diplegia present at birth which survive the first six years of life, it may be said that in proportion to the degree of physical and mental disablement the tenure of life is short, and the majority of those afflicted do not reach far into the third decade of life. Cases of diplegia, the onset of which is after birth, most often show a progressive course to a fatal termination within three years of the onset. Sometimes remarkable and lasting improvement takes place, and this occurs chiefly in those cases in which there has been an acute onset and rapid development of paralysis.

*Etiology.*—In rather more than one third of all the cases of cerebral diplegia there are no clinical conditions, either in the child itself or in its parents, with which the onset of the trouble can be associated. In the remaining

two thirds, however, some condition has been present, either in the life of the child or in its family history, which can be brought into causal relation with the disease. Amongst these etiological factors is, first, heredity. In the cases in which heredity is an important factor it is usually found that more than one member of the family suffers; and sometimes there is a history that a similar condition has been present in preceding generations. Another point in reference to this condition is, that in cases in which it plays a part the onset of the disability is usually about the age of puberty. The clinical form which the disease assumes is that in which rigidity, especially of the lower limbs, is a marked symptom. Associated with this rigidity there is an increase of the reflexes, indicating disease in the pyramidal system. There is sometimes, also, nystagmus present. There may also be a slight degree of optic atrophy, and, in some cases, articulatory impairment is distinct. It will thus be seen that the condition, in some of its clinical features, closely resembles that present in disseminated sclerosis; but the fact that it sometimes affects more than one member of a family, that it is of more gradual onset than disseminated sclerosis usually is, and that the sphincter affection and tremor are both absent, would be sufficient to separate the cases from those of that disease. In the amaurotic family idiocy of Sachs the hereditary influence seems to be very strong, several cases frequently occurring in the same family. And the racial factor is of the utmost importance, for, so far, no case has been recorded which did not occur in a child of the Jewish race. The second factor of very definite etiological importance is illness on the part of the mother during pregnancy. Sometimes such illness is of a vague and indeterminate character, associated with obstinate and troublesome sickness and vomiting. Occasionally the illness has been of a somewhat acute character, associated with a high temperature. Traumatic influences have also been present so frequently as not to allow of their being regarded as mere coincidences. Blows, especi-

ally abdominal injuries, and falls—sometimes violent and resulting in concussion—seem to have been very frequent precursors of the condition. In many cases, also, eclampsia of a very severe and prolonged character has been present before the birth of the child, and in some instances the mother has been actually mentally deranged. A third factor of importance is difficulty or abnormality in the labour. Frequently the labour has been a prolonged and difficult one, has been effected by instruments, and the child has been born asphyxiated. Such difficulty in labour is much more apt to be present in primiparæ, and in a large proportion of the cases of which the writer has notes the conditions seemed to be more effective in male children than in female, the result, apparently, of the larger size of the head of the male child. Malpresentation, also, is important in this relation as naturally causing greater difficulty, a longer duration of the labour, and a greater likelihood of injury. In a large number of cases investigated by Dr. Herbert Spencer, in which the children were born dead, hæmorrhages were present on the surface of the brain, or at the base of the brain, or over the cerebellum, and even in the spinal cord. And it is to be presumed that in many cases in which there were difficulties similar to those resulting in still-birth, but in which the child survived, there may have been similar lesions of the nervous system. Such conditions would, of course, according to their position and extent, give rise to varied forms of paralysis. Another condition of labour which has been occasionally present in cases of diplegia is the precipitate labour; and in this we must assume either that the pressure was exerted so quickly upon the child's head, or that the conditions under which it was born were such, as probably to produce some injury to the brain. A fourth factor of much etiological importance is acute infectious disease and fevers occurring in the early extra-uterine life of the child. These seem to be effective in inducing a condition of cells which leads them to undergo a slow, but steady, degeneration. Of course it is quite possible that

the cells which suffer in this way in the child are originally endowed with a somewhat uncertain tenure of life, and that the acute infectious disease, or fever, is merely the proverbial "last straw." Certain vascular lesions, also, would seem to be effective in producing diplegia, especially lesions occurring in surface veins. These, of course, will tend mostly to occur in marasmic children. Epilepsy, also, is a condition which at all events is associated with the onset of diplegia, and may be present before the diplegia asserts itself. Under such circumstances one must assume that the epilepsy and the diplegia are both results of the same cause, the epilepsy resulting first of all from the smaller lesion, leading to instability of the cells, and consequent discharge; the diplegia resulting from the further progress of the disease leading to abolition of function of those cells, and possibly also of others in their vicinity.

*Morbid anatomy.*—In the large majority of cases of cerebral diplegia, whatever the clinical type, the *post-mortem* condition of the cerebral hemispheres is one of widely-spread atrophic sclerosis. The more severe the case and the longer life has been preserved, the greater is the degree of sclerosis and atrophy in the affected regions. The atrophic sclerosis may affect the whole surface of the hemispheres equally, but more often certain regions are profoundly affected while others escape relatively or completely, but the distribution is practically always symmetrical upon the two hemispheres. In severe generalised rigidity with idiocy, the whole of the hemispheres may be affected. Where motor symptoms are slight and mental deterioration profound, the frontal and occipital regions are predominantly affected. In paraplegic rigidity the atrophy is confined to the paracentral lobes and neighbouring convolutions upon the convexity of each hemisphere, a lozenge-shaped depression of remarkable appearance occupying the summit of the cerebral vertex. In bilateral athetosis and choreiform diplegia the atrophy is of the central region, and is not so marked as in the other forms of the disease.

In atrophic sclerosis the hard and soft membranes are found to be normal, but there is always some excess of subarachnoid fluid compensatory to the shrinking which takes place in the volume of the brain from the atrophy. Further, the blood-vessels are almost always found to be healthy. In the atrophied region the convolutions feel unduly hard to the touch, they stand away from one another, and are smaller and more yellow in colour than are normal convolutions, while the sulci separating them are widened. The convolutions do not preserve their even breadth as do normal convolutions, but are very narrow in one place, at another relatively very broad. Their surfaces often present a worm-eaten and faceted appearance. Each convolution is ridge-like and does not preserve its level, now sinking and now rising. This irregular form of the convolutions, with the wide sulci separating them, gives the surface of the brain a characteristic appearance like that of a walnut-kernel (Figs. 6 and 7).

On section the consistence of the cortex is tough, sometimes gritty, and occasionally many minute cysts are present. The general arrangement of the convolution does not usually depart from the normal, but sometimes a primitive arrangement of gyri obtains, and is indicative of interference with the development of the brain in early foetal life.

The centrum ovale is shrunken in proportion to the degree of cortical atrophy. Sometimes in rapidly progressive cases it may present a translucent, almost jelly-like, appearance from the disappearance of the fibres—the neuroglia only being left. The basal ganglia are usually small, and in the pre-natal cases in which the pyramidal tracts are not developed the appearance of the ventral aspect of the crura cerebri, pons, and medulla is correspondingly modified. In some cases atrophic sclerosis is also present in the cerebellar cortex.

Microscopical examination of the atrophic cortex shows degeneration and disappearance of the nerve-cells, and in a severe case of general rigidity there may be hardly a cell



to be found in the cortex of the central region. No change is to be seen in the blood-vessels, which are numerous, but of reduced size in the diseased region. The neuroglial



FIG. 6.—Brain from case of atrophic sclerosis, showing contraction and faceting of convolutions and widening of sulci, "walnut type." From photograph by Dr. Collier.

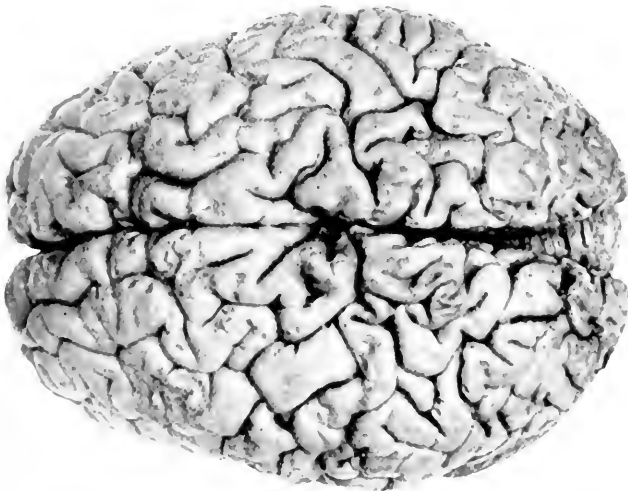


FIG. 7.—Another aspect of the same brain as Fig. 6

elements are greatly increased, and many spider cells are visible. The projection systems corresponding with the affected regions of the cortex are found either not developed



FIG. 8.—Section of normal cerebral cortex.

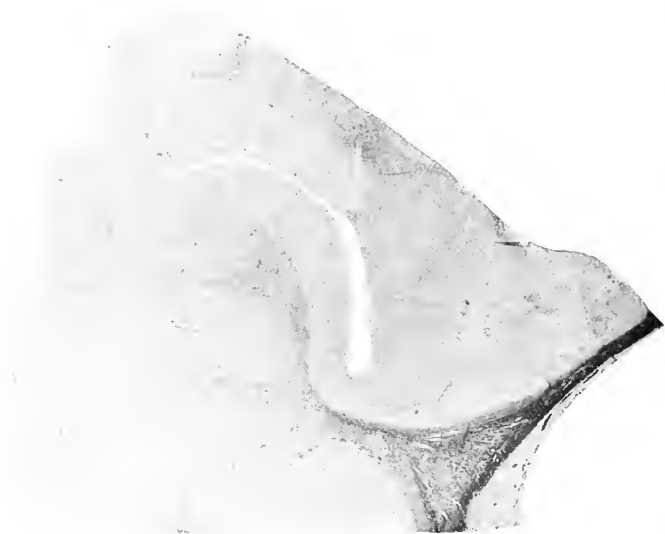


FIG. 9.—Section of cortex in atrophic sclerosis, prepared in the same way as the section in Fig. 8, showing disappearance of cells and fibres.

or are degenerated, according as the death of the nerve-cells took place before or after the time of development of these fibres. For example, the pyramidal tract in pre-natal cases may be found undeveloped, partially developed, or incompletely developed, while in post-natal cases it is found degenerated.

With the exception of the abnormalities of the pyramidal tract, the brain-stem and spinal cord present no peculiarities, but in cases in which life has been preserved for some time in a condition of extreme emaciation widely spread degenerative changes may be found. In a few cases of cerebral diplegia, bilateral and symmetrical porencephalic defects are found, in others, symmetrical cysts or symmetrical softenings. Such conditions are met with chiefly in the cases of diplegia which arise after birth.

Of the essential pathology of atrophic sclerosis we know little, but it is certain that the cell degeneration is a primary one, and that it is not secondary to an overgrowth of the neuroglia strangling the nerve elements. The neuroglial change follows and does not precede the cell degeneration. There is strong presumptive evidence that the death of the cells is occasioned by toxic agents which especially affect the physiological group of elements of the cerebral cortex. The immediate effects of the presence of such toxic agents may be the degeneration of the cells, or as a result of the action of the toxins, the tenure of vitality in the cells may be reduced so that after a period of apparently normal life they undergo degeneration at some subsequent period when the toxic agent is no longer exerting its effect.

In a minority of cases, and among the post-natal cases chiefly, atrophic sclerosis has been proved to result from symmetrical disease of the blood-vessels supplying the cerebral cortex. Thrombosis of the cerebral veins entering the superior longitudinal sinus, thrombosis of both Sylvian arteries, and symmetrical embolism of the latter vessels, have been found occasionally. In these somewhat rare cases the signs of the original vascular disease remain and

indicate the nature of the original process of which atrophic sclerosis is the result.

FIG. 10.

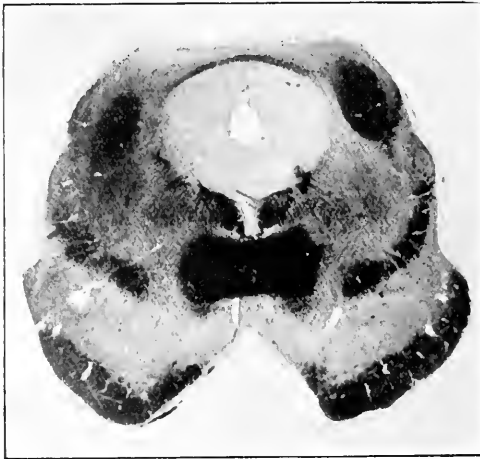


FIG. 11.



Porencephalic defects are always the result of vascular obliteration, either by embolism or thrombosis, but the eon-

dition determining the complete absorption of the cerebral tissue which occurs is quite unknown.

In explanation of the essential symptoms of diplegia—rigidity preponderating over paralysis, perversity of volitional movement and the occurrence of spontaneous movements—stands the important fact that the disease is one affecting chiefly the cerebral cortex at the period of life before the preponderating and almost exclusive government and control of voluntary movement by the highest centres is attained. In the lower animals the function of the cerebral cortex in the initiation of voluntary movements and in

FIG. 12.



FIG. 13.



Figs. 10, 11, 12, and 13, showing absence of pyramidal system in case of atrophic sclerosis. From photographs and preparations by Dr. Collier.

the control of the lower centres is far less important than in man; while the importance of those structures corresponding with the optic thalamus basal ganglia and mid-brain is much greater, and in these animals the whole cerebral cortex may be removed without producing paralysis of movement. In the human adult the supremacy of the cerebral cortex in initiation and control of voluntary movements has become complete, and the functions of the basal ganglia, etc., are entirely subsidiary, so that in the absence of the cerebral cortex the latter are incapable of initiating any voluntary movement. The child at birth, however, resembles the lower animals in this respect, that the functions and supremacy of the cerebral cortex have not

been attained and the lower centres still possess the function of initiating movements, though perhaps only of a primitive character. Consequently, if the cerebral cortex be put out of action at this period of life, voluntary movement of a slow and clumsy nature is still possible.

FIG. 14.



FIG. 15.

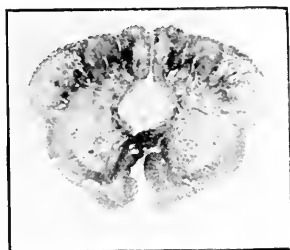


FIG. 16.



Figs. 14, 15, 16.—To illustrate very defective development of pyramidal tracts in a case of diplegia. From preparations by Dr. Collier.

Rigidity is in part due to the uncontrolled action of the centres in the spinal cord, but it seems to be chiefly the result of the absence of the influence of the cerebral cortex upon certain centres situated in the region of the cerebellum and pons. If, for example, in the cat the mesencephalon be cut across above the pons and cerebellum there ensues at

once an intense extensor rigidity of all four limbs—"decerebrate rigidity." This decerebrate rigidity is immediately abolished by the hemisection of the spinal cord in the cervical region or of the medulla oblongata, upon the side of the hemisection, and it must, therefore, be due to influences proceeding from the ponto-cerebellar region. The similarity of the decerebrate rigidity of animals after mesencephalic transection and the rigidity of cerebral diplegia is very striking, and probably the phenomena in the two conditions are the result of decerebration.

Perversity of movement is the expression of the disordered action of a nervous system, the correlation in action of the several elements of which is disturbed by the impairment of the functions of certain elements. The normal condition of these is necessary for the performance of perfect movement.

Spontaneous movements of a choreic, athetotic, or myoclonic nature appear to be associated with a partial destruction of the cells of the central region of the cerebral cortex. Convulsive phenomena are due, as in other examples of organic epilepsy, to the instability of the remaining elements of the cerebral cortex which partial damage and interference with associational connections entail. It is probable that some of these phenomena are the result of the action of toxic substances produced during the degeneration of the nerve elements upon the surrounding nerve elements.

The explanation of the strabismus and ocular paralysis so frequently met with in cerebral diplegia is unsatisfactory. Affection of the nerve-cells of the oculo-motor nuclei has been suggested, but no change in these structures has been demonstrated histologically, and it is unlikely. The strabismus is not usually the result of paralysis of a muscle, but of an affection of movement, and is, therefore, most likely due to a cortical affection.

*Diagnosis.*—The diagnosis of the condition when the disease dates from the earliest infancy, and when the symptoms are marked, presents little difficulty. Paraplegic

rigidity may possibly be confounded with other varieties of paraplegia and especially with that resulting from spinal caries. The absence of spinal pain and deformity and the frequently associated fits in the cerebral disease should help to distinguish the two conditions. Pain and deformity of the spinal column may, however, be completely absent in spinal caries; and if a case of paraplegic rigidity progress, there is great probability that it is of spinal origin and the appearance of anæsthesia should be carefully watched for.

Certain cases of pontine tumour in which headache, vomiting, and optic neuritis are absent may almost exactly simulate generalised rigidity of post-natal and insidious onset; the occurrence of hydrocephalus or the loss of the ocular associated movements of lateral deviation from involvement of the sixth nuclei should suggest a gross pontine lesion.

Usually the presence of headache, vomiting, and optic neuritis serve to prevent confusion in diagnosis between the results of the presence of cerebral tumour, hydrocephalus, and meningitis and diplegia.

Much greater difficulty arises in those cases in which the rigidity and paresis are slight and perverse movements or volitional tremor are present. Such cases may closely simulate disseminated sclerosis and Friedreich's disease, and several cases are on record in which the clinical aspects and the pathological investigations have proved the existence of the lesions characteristic of both diseases. Many of the cases which have been recorded as examples of disseminated sclerosis in children have been subsequently shown by *post-mortem* examination to present a brain lesion similar to that above described as characteristic of diplegia. The possibility of the presence of cerebral diplegia should always, therefore, be entertained when symptoms suggesting disseminated sclerosis are present in a child, and a careful survey of the history and symptoms made before a decision is arrived at, since the prognosis in disseminated sclerosis is that the disease will necessarily progress, whereas in those cases of dip-



legia which simulate the former disease a tendency for the condition to remain stationary or to improve is the rule. Friedreich's disease is characterised by the usual absence of the knee-jerks and the clumsy ataxy, but sometimes the distinction from diplegia is exceedingly difficult.

Juvenile general paralysis of the insane is another condition which may give rise to difficulty in diagnosis. The distinguishing points of this disease are the Argyll-Robertson pupil, the ataxy, and the frequent absence of the knee-jerks, and, not infrequently, evidence of congenital syphilis.

In the final stage of wasting and flaccidity there may be some resemblance to anterior poliomyelitis, but in diplegia an antecedent history of spasticity and incomplete paralysis is always obtainable.

The *prognosis* as regards life depends upon the severity of the case and upon whether the symptoms show a tendency to become progressively worse; it is also influenced by the age of the patient. In cases of paraplegic rigidity, in slight cases of general rigidity, and in choreic and athetotic diplegia, there is little tendency to the destruction of life, either directly or from a greater susceptibility to the occurrence of intercurrent diseases, but the patients are usually short lived.

The majority of the cases of severe generalised rigidity occurring in early infancy do not survive the fifth year, marasmus and broncho-pneumonia being the common cause of death. If a patient survive the sixth year, life is usually prolonged till after puberty. In progressive cases the patients do not usually survive for more than two years after the symptoms commence to progress. Post-natal cases of acute onset rarely remain stationary; they show a tendency to either rapid amelioration or rapid progression to a fatal result. The prognosis in those which improve can only be judged from the rate of improvement and nature of the malady in a given state. When the onset is insidious, in later childhood, a very slowly progressive course not tending to end life is the rule.

The prognosis as regards mental and physical capacity is hopeless in severe cases, in cases showing a progressive tendency, and in most cases of bilateral athetosis and choreic diplegia. In pre-natal cases of slight generalised and paralytic rigidity, improvement almost always takes place, and the degree of such improvement as can be expected depends largely upon the mental condition, the capacity for learning, and the possibility of bestowing upon the patient the necessary skill in the laborious task of mental and physical training. Under the most advantageous circumstances the prognosis is that, though the child will never completely lose his defects of movement, he may be able to earn a livelihood. Even under these circumstances the probability of the occurrence of epilepsy is great. In post-natal cases of acute onset, where rapid amelioration takes place, almost all the symptoms may disappear.

*Treatment.*—We know of no treatment that is of avail either in prophylaxis or in arresting the course of the disease when it is of a progressive nature. But much may be done in slighter cases in the way of improving the mental condition by careful training, and by measures for increasing voluntary power and control by regular physical training, such as have been described under Infantile Hemiplegia. Passive movement from the earliest days is most useful in preventing deformities of the limbs, and if, from neglect, these have occurred, tenotomy, followed by passive movement, has proved of great service.

## AMAUROTIC FAMILY IDIOCY

This disease, to which Sachs gave the above name, is also known as "arrested cerebral development," "family form of idiocy," "infantile cerebral degeneration" and "agenesis corticalis." It is a rare disease, probably always fatal and characterised by the gradual onset of mental and physical weakness, coming on after a period of normal development, and, by the presence of symmetrical changes at the *macula lutea*, leading to complete blindness. The condition was first described by Mr. Waren Tay in 1881. Since then Sachs, Peterson, and Hirsch have written on the subject and made autopsies of cases, and a very important contribution to the study and elucidation of the nature of the disease has been made by Kingdon and Risien Russell in this country. Kingdon was fortunate enough to observe clinically three cases, and to examine the *post-mortem* condition in conjunction with Risien Russell. These three cases all belonged to a family of seven, five of whom suffered from the disease.

*Symptoms.*—A child who becomes the subject of this disease is born apparently healthy and remains so until the age of a few months—three or four—is reached. In exceptional cases a more advanced age is attained before any abnormality is noticed. One of the earliest signs is a weakness of the neck muscles, leading to inability on the part of the child to keep its head up. This weakness spreads by degrees until the muscles of the trunk and limbs are affected in a similar way, and the condition of the child is one of great weakness and almost universal flaccid paralysis. As time goes on the condition of flaccidity is replaced by stiffness and spasticity, the muscles waste, and the head becomes retracted. Simultaneously with the onset of

physical weakness the mental condition begins to deteriorate. The child ceases to take notice of things and becomes dull and lethargic. It is less troublesome than ordinary children of its age, but occasionally it screams loudly and persistently. At an early stage of its physical and mental deterioration its sight is obviously affected. It is indeed difficult to say whether its early apathy and inattention may not be due to the impairment of its vision, which rapidly proceeds to complete blindness. An examination of the eyes reveals the condition which is characteristic of the disease. At first no obvious change may be noticed in the optic discs. Occasionally some pallor may be present, and Waren Tay has observed early in the disease a slight degree of optic neuritis. But the macular changes are striking. The macula itself is represented by a dark cherry-red point which is surrounded on all sides by a large area, about twice the size of the disc or even more, very pale, with soft woolly-looking edges, its longer diameter being horizontal. This condition of the fundus remains the same throughout the illness, except that the disc becomes more and more pale and finally completely atrophic. The ocular muscles are not affected, although a squint may be occasionally present, and nystagmoid movements are not uncommon. Slow rolling movements of the eyeballs may also occur, usually after the patient is blind.

As regards the muscular condition of the limbs and trunk, that, as already stated, is in the early stage one of flaccidity. The knee-reflexes are active. Later, spasticity becomes distinct, and there are frequent jerkings of the arms as well as of the legs. There is marked wasting of the muscles, but the electrical reactions remain unchanged. One curious phenomenon is present, viz. the very marked quite sharp jerk of all the limbs, and also of the head and eyelids, which takes place when any sharp sound is made. A tap on the iron bed-frame with a key or penknife is followed at once by such a jerk, apparently the result of the oversensitiveness of hearing. Convulsions are exceedingly rare in this disease—indeed, it is probable that they do not occur

at all as part of it. The mental and physical condition gradually becomes weaker, and death occurs from gradual exhaustion. Occasionally the end is unexpectedly sudden, before the wasting has proceeded to an extreme degree. There may have been some difficulty in swallowing before death, but as a rule the organic reflexes remain unaffected, so far as one is able to judge in such young patients, and there is no evidence of any disease of the thoracic or abdominal viscera.

*Etiology and pathology.*—One very striking etiological factor has been present in almost every case in which it has been inquired for, viz. that these children are the offspring of Jewish parents. No signs of syphilis are present either in the parents or children as a rule, and that disease does not probably constitute one of the causal factors of the condition. The fact that it is so frequently present in more than one member of a family would seem to place it amongst those diseases in which there is an inheritance of a nervous system prone to early decay and death. Whether there is further some toxic substance present we cannot say. The majority at least of the patients are breast-fed, and it is possible, as Russell and Kingdon suggest, that there may be in the child's food something which determines the degeneration. It is probably not to be regarded as a condition simply of arrested development. It would seem as if the nervous system were normally developed at first, and went on growing naturally. The subsequent history of the cases would seem to suggest that development becomes arrested by some influence which determines rapid and complete decay, especially in the pyramidal system, and the morbid anatomy of the condition would seem to bear out such a view.

*Morbid anatomy.*—There are no naked-eye changes of significance in the nervous system except that occasionally abnormal fissuration of the brain indicating defective development has been described. As a rule, however, the sulci and convolutions are normal in arrangement, although the former may be deeper and more open than usual, due to a wasting of the latter. There are marked degenerative

changes in the neurons of the cerebral cortex, especially the pyramidal cortex, the cells being seen in all stages of degeneration, and their processes similarly affected. The direct and crossed pyramidal tracts in the cord are also intensely degenerated, but it is difficult to say whether this follows, or precedes, or occurs simultaneously with, the degeneration of the pyramidal cortex. There is no overgrowth of the neuroglia, no real sclerosis, and the walls of the blood vessels are normal. The optic nerves are atrophied, and the ganglion cells of the retina have also been found degenerated. Treacher Collins has found a thickening of the retina in the macular region, the result apparently of a spacing out of the outer molecular layer as if by œdema.

*Diagnosis.*—When the changes in the fundus are once seen the diagnosis is easy. They are quite characteristic, and, so far as we know at present, pathognomonic. The physical and mental conditions are similar to those met with in other diplegic conditions, but the association of these with visual defects and changes in the fundus of the nature described are quite conclusive.

*Prognosis.*—The prognosis is essentially bad. The majority of the cases that have been described have been known to end fatally, usually within the first two years of life. One case has lived to four, and it is possible that life may be prolonged to even a later age. But this is scarcely likely if the view of its pathology which has been given above be the correct one.

*Treatment* has been quite inefficacious. Mercury, iodide of potassium, strychnia, and various other drugs have been tried. Kingdon has also tried cerebrine and pituitary gland, but without effect. The same observer has also tried the administration of iodide of potassium during pregnancy. He tried it in two cases. One child escaped, the other suffered from the disease. As already stated, Kingdon and Russell suggest that children in a family in which this disease has appeared should be hand-fed rather than breast-fed, on the hypothesis that the mother,

although healthy herself, may possibly communicate some toxin to her child. It is a suggestion which, in such a hopeless disease, is certainly worth a trial.

A condition of symmetrical changes at the macula occurring in two members of the same family associated with mental defect has been described by Dr. F. E. Batten. But the changes in the fundus were rather pigmentary, and were closely similar to those previously described by Dr. Rayner Batten in two brothers, in whom, however, the mental condition was normal. It is possible that these changes are related to syphilis. They have not the racial affinity of the cases of amaurotic family idiocy.

## SINUS THROMBOSIS

FROM certain peculiarities of anatomical structure and arrangements, it would seem that the venous sinuses of the dura mater are particularly prone to the clotting of blood within them. The lumen of the sinuses is irrégular and generally triangular in shape; their walls, embedded in the substance of the dura mater, contain no muscular tissue, and their calibre does not vary; so that in conditions of feebleness of the circulation or diminution of the volume of blood, such as occurs, for instance, in cases of prolonged diarrhœa, the blood flow through them becomes very slow. The fact also that trabeculæ occur crossing the lumina of the sinuses predisposes to thrombosis. The superior longitudinal sinus seems to be in a position of peculiar disadvantage as regards thrombosis, for the cerebral veins opening into it ascend from the surface of the brain and enter the sinus in a forward direction, the current of blood in them being thus opposed to the direction of flow in the sinus.

Thrombosis of the cerebral sinuses early attracted attention and was first described by Abercrombie in 1818. An excellent memoir was published by Tonnèlé in 1829.

*Etiology.*—The condition may result from a general condition of malnutrition and prostration and is then called “marantic thrombosis” or primary thrombosis of the sinuses; or it may occur from the extension of infection from other regions to the sinus—secondary or infective thrombosis.

### PRIMARY THROMBOSIS

This occurs more frequently in the superior longitudinal



sinus than in the others. It is more common in the first year of life than in the whole remainder of life. Among adults the old and feeble are liable to be affected. The common causes of this condition in infants are: (1) Gastro-enteritis where there is much exhaustion, especially if there is prolonged diarrhœa; (2) bronchial diseases. It is questionable whether the thrombosis in these cases may not be of an infectious nature. Bouehut states that chronic bronchial infection is by far the most common cause of sinus thrombosis in infants; (3) the conditions of exhaustion met with in tuberculous disease and in congenital syphilis; (4) it may follow certain acute diseases, such as measles, scarlatina, influenza, typhoid fever, and diphtheria.

Under all these conditions, thrombosis of veins may be met with in any part of the body, and in most cases of marantic thrombosis of the sinuses thrombosis of other veins of the body is met with.

#### SECONDARY OR INFECTIOUS THROMBOSIS

*Causation.*—The essential cause is the advent of micro-organisms to the sinuses. The infection is often a mixed one and several varieties of the germs of suppuration are met with; *e.g.* *streplococcus*, *Bacillus coli*, and the *pneumococcus* have all been found in the infected sinuses.

The modes in which infection may take place are as follows:

(1) The sinus may become infected as part of a general pyæmia.

(2) Infection may spread directly through the wall of the sinus from a focus of local disease—nearly always an extra-dural abscess immediately in contact with the affected sinus.

(3) In the great majority of cases the infection spreads along the veins opening into the sinus from a local focus of disease in a contiguous region of the skull by a process of infectious thrombosis spreading along these veins. The source of infection is often disease of the middle ear and

mastoid cells. It is important to note that an intra-cranial affection following otitis is more commonly meningitis in children, more frequently sinus thrombosis in adults. Sometimes the infection spreads from cellulitis of the face or orbit along the facial and angular veins to the ophthalmic veins which open into the cavernous sinus. Again, infection of the cellular tissues of the bucco-pharyngeal cavities may cause thrombosis of the lateral sinus, since the pharyngeal and pterygoid veins communicate with it. Another frequent cause is local cranial osteitis, either from injury, osteomyelitis, tubercular or syphilitic caries, or from suppuration of the scalp.

The position of the thrombosis depends upon the nature of the process, marantic or infectious, and if infectious it is determined by the situation of the focus of infection. In simple thrombosis the superior longitudinal sinus is nearly always the one first affected. In the septicæmic form the superior longitudinal and lateral sinus are prone to be the first involved.

When infection is local the sinus thrombosed will be that into which the veins draining the infected part open.

*Morbid anatomy.*—The affected sinus is bulged and distended. To the touch it feels like a hard cord, and as if injected with a solid mass. The wall of the sinus is thickened and the intima may be rugose; more often it has disappeared. The lumen is at first completely obliterated by a clot, which is more or less adherent to the walls. Later on, as the clot tends to contract, it may be attached to the wall in certain places only, and a lumen may in part be restored. The clot in early stages is red and friable, but as time goes on it becomes reddish-yellow and tough, then yellow, and finally grey. In the infectious forms the clot may very quickly break down into pus and general pyæmia result. The veins opening into an affected sinus are, as a rule, extensively thrombosed, and the wall of the affected sinus may show changes analogous to periarteritis and endarteritis, even when it is not thrombosed.

Except in cases in which the clot is very small, the ob-

literation of a sinus produces evident intra-cranial effects. Taking, for example, thrombosis of the superior longitudinal sinus, there is marked congestion of the cerebral convolutions of the convexity with, perhaps, a cord-like condition of clot-distended veins, and there is nearly always bloody serum in the arachnoid space with small surface hæmorrhages, and occasionally larger meningeal hæmorrhages. The pia arachnoid is œdematous, as is also the general lining membrane of the ventricles.

There is generally some increase of the intra-ventricular fluid. In later stages extensive softening of the hemispheres may occur.

Upon microscopic examination multitudinous capillary hæmorrhages are seen. It is difficult to determine correctly whether a healing process may occur in this disease. Some circulation may be kept up when the superior longitudinal sinus is obliterated with a limited clot by a communication which exists between this vessel and the lateral sinus. Sir William Gowers has reported a case in which bilateral atrophy of the paracentral and neighbouring convolutions was undoubtedly the result of a thrombosis of the superior longitudinal sinus a considerable time previous to death. In Marfan's case a child survived a thrombosis of the lateral and cavernous sinns of one side. Death occurred from broncho-pneumonia nine months later, when these sinuses were found reduced to fibrous cords.

*Clinical aspect.*—The symptoms of sinus thrombosis are somewhat variable and the diagnosis may be very difficult. Sometimes thrombosis may occur without giving rise to any appreciable symptoms: (1) In newly-born children whose cerebral functions have scarcely come into action, and (2) in cachectic infants in whom any symptoms that may arise from sinus thrombosis are concealed by those of the preceding malady, and lastly (3) in certain cases in which obliteration is not complete or in which an anatomical peculiarity in the establishment of a vessel permits collateral circulation to occur.

The clinical phenomena associated with thrombosis of the

cerebral sinuses may be conveniently divided into three groups:

(1) Cerebral symptoms indicating some intra-cranial disturbance.

(2) Local signs revealed by external examination of the head.

(3) General signs of some bodily condition with which sinus thrombosis is likely to be associated.

(1) *Cerebral signs*.—The symptoms of the intra-cranial lesion present no specific peculiarity. In very young children they simply consist of vomiting, convulsion, and coma, rapidly ending in death. In older children the initial symptoms are headache and restlessness followed by vomiting, delirium, convulsions, and coma. Rigidity of the muscles of the neck, trismus, inequality of the pupils, strabismus, nystagmus, and irregularity of the pulse and respiration are often present.

(2) The *local signs* of sinus thrombosis differ with the situation and extent of the thrombosis. When present they are often pathognomonic, but, unfortunately, they are so often completely absent that they cannot be considered as negative signs for purposes of diagnosis.

When the superior longitudinal sinus is affected there may be some cyanosis and œdema in the forehead region. The veins entering at the anterior fontanelle, the angular vein, and the temporal veins may be distended and in rare cases thrombosed.

If the lateral sinus is affected the superficial cranial veins of that side may be empty in contradistinction to those of the other side of the head, which may be distended. The clot may extend into the internal jugular vein and cause pain and stiffness upon that side of the neck. Sometimes the jugular vein may be felt beneath the anterior border of the sternomastoid as a solid cord very tender on pressure. The deep glands of the neck may be enlarged and the deep cervical connective tissue œdematous. Sometimes thrombosis of the mastoid emissary vein which opens into the lateral sinus occurs, and a circumscribed patch of hard

œdema which is very tender upon palpation appears immediately behind the mastoid process. From extension of the thrombosis to the veins of the tympanum, facial and acoustic, paralysis of the same side may occur.

When the cavernous sinus is affected, there is usually slight proptosis of that side with œdema of the conjunctiva and of the upper part of the face. Amblyopia is the rule, and ophthalmoscopic examination reveals swelling of the disc or perhaps thrombosis of the central vein, with multiple retinal hæmorrhages. Paralysis of the ocular muscles and trophic destruction of the eye on the affected side may also occur.

When considering this local symptomatology it is necessary to remember that thrombosis of the cerebral sinuses in most cases advances from sinus to sinus. It is quite exceptional to find *post mortem* the thrombus confined to one sinus.

(3) *General signs.*—The symptoms of these maladies, which are the usual antecedents of sinus thrombosis, are of great importance. A condition of marked emaciation, with persistent diarrhœa associated with a subnormal temperature, suggests marantic thrombosis. Broncho-pneumonia in a fuller subject, suppurative disease of the middle ear, orbit, nasal cavities, or cranial bones accompanied by pyæmia suggest infective thrombosis.

Severe pyrexia, rigors, embolic phenomena, and enlargement of joints suggest pyæmic thrombosis.

*Diagnosis.*—The indications for diagnosis, where this is possible, are embodied in the preceding account of the disease aspect. In cases consequent upon middle-ear disease there is some possibility that an early diagnosis may give a chance of saving life, but there are few indications to aid one in distinguishing whether cerebral symptoms suddenly arising in the course of chronic otitis are due to meningitis, cerebral abscess, or sinus thrombosis in the absence of any localising sign such as thrombosis of the jugular vein.

If the onset of cerebral symptoms is accompanied by rigors and high pyrexia, with wide oscillations, the presence of thrombosis of the lateral sinus may be considered highly probable.

*Prognosis.*—The disease as a rule is rapidly fatal. It is probable that some subjects of non-infective disease survive the thrombosis. When it is local or incomplete, atrophic sclerosis of the brain may occur in those areas from which the veins open into the affected sinus. Cases of infective thrombosis following ear disease have been saved by early surgical interference with ligature of the jugular vein and of the affected sinus on either side of the thrombosed area, the sinuss being incised and the clot removed. Where pyæmia is present, either as the cause or as the result of infective thrombosis, the result is always fatal.

*Treatment.*—Beyond vigorous prophylactic treatment and the palliative treatment of symptoms, surgical interference in cases of local infective origin is alone of any value.

## INTRA=CRANIAL ABSCESS

LOCAL collections of pus within the skull fall into three groups, according to their situation: (1) Extra-dural—situated between the inner surface of the skull and the dura mater; (2) subdural—situated between the dura mater and the surface of the brain and confined in this position by adhesions which have formed between these structures. Such abscesses almost always involve the brain tissue to a greater or less extent; (3) encephalic abscess—situated beneath the surface of the brain.

In the first two groups, extra-dural and subdural, the abscesses are acute and are due always to local infection from injury, wounds, or local disease of the skull. In the third group the abscesses are usually chronic in nature and may result from chronic infective disease of the middle ear or nasal cavities, by indirect infection, or they occur in association with suppurative diseases of the thoracic and abdominal viscera (tuberculous disease excepted) or rarely as complications in certain infective diseases.

A fourth variety of encephalic abscess occurs in connection with conditions of pyæmia and ulcerative endocarditis. The lesions of the brain are usually small and multiple and do not give rise to symptoms apart from those of the general condition.

*Etiology: age and sex.*—Among adults, males are more often the subjects of intra-cranial abscess of any variety than are females, in the proportion of rather more than two to one. It hardly ever occurs in infants before the end of the first year, and is more common between the ages of 7 years and puberty than during the first six years of life; acute cerebral abscess of traumatic origin occurs much more fre-

quently in children than in adults. Encephalic abscesses occurring in association with distant suppuration or in the course of pyæmia are extremely rare in children, while in association with specific fevers they are more frequently met with than in adults.

Abscesses, the result of disease of the middle ear and nasal cavities, occur in much the same proportion as in adults (the first year of life and old age being excepted), but the situation of the abscess shows a marked difference. In adults encephalic abscess resulting from ear disease is found three times as often in the temporal lobe as in the cerebellum, but before puberty it is found ten times more often in the temporal lobe than in the cerebellum.

*Organisms.*—The germs commonly found in intra-cranial abscesses are staphylococcus, streptococcus, and pneumococcus. More rarely the *Bacillus coli* is present and in very rare cases *Oidium albicans* has occurred in pure culture.

*Causation.*—The most important cause of acute cerebral abscess is injury to the head. It may follow simple contusion unassociated with a wound of the skin or fracture of the skull. It must be presumed in these cases that the injury causes by bruising a “locus minoris resistentiæ” within the skull, of which organisms present in the blood take immediate advantage. More frequently the injury has caused a wound, which suppurates, and perhaps in addition there has been a fracture of the skull, or a fracture of the base of the skull has opened a way from the nasal or tympanic cavities into the subdural space. Again, punctured wounds and bullet wounds extending through the skull into the brain are common causes of abscess. When the condition follows a wound of the scalp, associated or not with fracture of the skull, it is quite common for the indications of the presence of abscess to make their appearance after the wound has healed, sometimes even after the wound has cicatrised.

Local disease of the bone, osteomyelitis, syphilitic and tuberculous disease, are also prone to cause abscess—the latter when secondarily infected with the germs of suppura-



tion. Chronic disease of the nasal cavities, of the orbit, and of the tympanic cavity may cause acute extra-dural or subdural abscess, either by direct extension or as one of the results of venous thrombosis.

The causes of encephalic abscess may be *local* or *remote*, and the former are by far the most important. These *local causes* are chronic disease of (1) bones of the middle ear and (2) bones of the nasal cavities. Long-standing disease of the middle ear is the cause of more than one half of all cases of intra-cranial abscess. During its acute stages otitis media seems never to cause intra-cranial suppuration, but when it has become chronic, and especially when discharge has ceased and the disease has apparently become obsolete, it is prone to be followed by a dangerous intra-cranial extension of infection—either meningitis, sinus thrombosis, or abscess.

It has been already stated that tympanic disease may by direct extension cause an extra-dural or a subdural abscess, but these results are not common. Generally the abscess is encephalic, and it is situated either in the temporal lobe or in the lateral lobe of the cerebellum of the same side. It is said that when the carious process affects the superior surface of the temporal bone the resulting abscess is in the temporal lobe, while, when the posterior wall of the tympanic and mastoid cells is diseased, the lateral lobe of the cerebellum is the seat of suppuration. The relatively much greater frequency of temporal abscess in children suggests that this is not an invariable rule. It must be remembered, however, that chronic otitis in children is more often followed by sinus thrombosis than by abscess, while in adults abscess occurs more often than does sinus thrombosis.

In what manner does the infection from the diseased middle ear reach a deep situation in the closely adjacent temporal lobe and lateral cerebellar hemisphere? In the majority of cases the dura mater is intact, and the meninges and surface of the brain appear normal, and there can be, therefore, no direct extension of disease such as is the rule in traumatic abscess. Neither is there any connection

between the vascular supply of the infecting and infected region except through the general circulation. In a few recorded cases the abscess has been situated on the opposite side of the brain to the ear disease, but such cases are so exceptional as not to interfere with the correlation in position of diseased bone and abscess. Similarly in those cases resulting from disease of the nasal cavities, the abscess is situated invariably in the frontal lobe near to the orbital surface. The question cannot be answered at present. It has been suggested that the organisms travel along lymphatics, but it is difficult to bring forward any anatomical facts in support of this statement.

Chronic disease of the nasal cavities, ozæna, syphilitic and tuberculous diseases, sometimes cause encephalic abscess. These are always situated in the lower part of the frontal lobes and generally upon the side of more advanced bone disease.

*Remote causes.*—These are somewhat rare ; there are three groups :

(1) Suppuration of thoracic and abdominal viscera or of bone.

(2) Pyæmia and ulcerative endocarditis.

(3) Certain other diseases.

The occurrence of a single encephalic abscess in association with cases of empyema, abscess of the lung, and bronchiectasis where no signs of pyæmia were present, excited the attention of observers many years ago and the term "pulmonal abscess" of the brain came into use. More recently such abscesses have been reported also in connection with suppurative peritonitis, post-peritoneal abscess, and chronic suppuration of bone. It is remarkable that suppuration arising in these regions primarily from tuberculous disease is never associated with brain abscess and also that pulmonal abscess has always been found upon the same side of the brain as was the lesion of the thorax. A few only of such cases have been reported in children and these have occurred in subjects nearing the age of puberty.

Pyæmic abscesses of the brain are always multiple and

sometimes very numerous. They rarely reach a size larger than a small nut, and they are especially prone to occur in the neighbourhood of the basal ganglia. These occurrences are very rare in the pyæmia of children.

Certain other diseases are occasionally complicated by the occurrence of encephalic abscess, such as enteric fever, typhus, erysipelas, and influenza.

*Morbid anatomy.*—Extra-dural and subdural abscess are invariably situated in the immediate vicinity of the antecedent lesion. An extra-dural abscess may reach a very considerable size by separating the dura mater from the skull; if it be in contact with the wall of a sinus, septic thrombosis of that sinus usually results. Such an abscess may burst externally or into the arachnoid space or into a blood sinus.

A subdural abscess is confined between meningeal adhesions between the pia and dura mater. When it is in contact with the brain, the surface of the latter is found softened or it may have disappeared, the abscess cavity extending more or less deeply into the brain tissue, which is extensively softened. Sometimes when there is no actual excavation of the brain substance, the latter is found to be absolutely riddled with pus over a considerable area. It is rare for this variety of abscess to have a definite capsule on the cerebral side. In either variety there may be a sinus leading to the surface of the skull, the nose, or the middle ear.

Encephalic abscesses (those of pyæmic origin excepted) are rarely, if ever, found in the grey matter, but their starting-point is most often in the subcortical white matter of the cerebral and cerebellar hemispheres. In one half of all cases the abscess is situated in the temporal lobe and in about one third in the lateral lobe of the cerebellum. The remainder are divided between the parietal lobe, the pons Varolii, the frontal lobe, and elsewhere, in order of diminishing frequency. Abscesses from ear disease are situated more frequently in the temporal lobe, less frequently in the lateral lobe of the cerebellum, and sometimes in both places, but

always upon the same side. In either position they may be multiple.

Abscesses due to remote causes and to general diseases may be situated either in the cerebrum or cerebellum, and when multiple they may be placed bilaterally, but are usually all in the cerebrum or all in the cerebellum. The medulla oblongata and the inferior vermiform process of the cerebellum are very rare situations.

The size of the cavities varies greatly. Usually it is between that of a walnut and that of a hen's egg. A recently formed abscess is irregular in shape, with ragged ill-defined walls. An older abscess is rounded and may have a definite capsule. It is difficult to say what determines the appearance of the capsule. The writer has seen a thick capsule of almost cartilaginous consistency present at the end of the tenth week in a traumatic case; but, on the other hand, it is certain that after an abscess has existed for many months it may show no trace of a capsule.

The interior of the abscess cavity is usually of a greyish-green colour, and the pus is greenish and often foetid. The surrounding brain tissue is always œdematous and sometimes softened. The convolutions of the surface of the brain and the inferior aspect of the cerebellum may show the results of pressure, and some degree of hydrocephalus may be present just as in cases of intra-cranial tumour.

*Course of the abscess.*—It may become encapsuled and remain for a long time stationary, subsequently extending; or the capsule may become thick and tough, and may even calcify, and rarely its contents may undergo this same change. More frequently the abscess extends before a capsule has formed, and, now stationary, now extending, may destroy much of the cerebral substance, and death may ensue from extensive interference with function. Often if not interfered with, it ruptures, most commonly into the ventricle, less commonly upon the surface of the brain; in either case acute meningitis results. Rupture occurs in about one sixth of all cases that are not afforded the advantage of operative treatment.

*Clinical aspect.*—An intra-cranial abscess has its origin in inflammation and constitutes when developed a foreign mass within the skull. Death may result, as in other forms of intra-cranial tumour, from the effects of continually increasing intra-cranial pressure and wide interference with cerebral function or from the spreading of the infection from the abscess.

Following von Bergmann's description, the symptoms may be grouped into four classes: (1) Those of local suppuration; (2) those due to increased intra-cranial pressure; (3) localising signs dependent upon the position of the abscess; (4) those of terminal extension of the infectious process.

The symptoms which present in a given case are dependent upon the nature of the abscess, whether acute or chronic, extra-dural, subdural, or encephalic. In extra-dural and subdural cases the symptoms are generally acute and the course is rapid; the signs of suppuration are well evidenced; signs of pressure are severe, whilst localising signs are rare, and a state of latency terminating with signs of extension of the suppuration is not common.

In the majority of encephalic abscesses, on the contrary, the signs of initial suppuration may be slight or may be swamped by the symptoms of the preceding disease, otitis media, empyæma, pyæmia, malignant endocarditis, etc., and for this reason may be easily overlooked. A latent period in which symptoms are insignificant or completely absent may follow and may last for weeks or months, sometimes even for years. The latent period is followed either by an acute outburst of symptoms the result of extending infection or the signs of progressive intra-cranial tumour arise.

While the symptomatology of typical cases is very distinct every gradation is met with between the cases of acute abscess with severe cerebral symptoms running a rapid course and ending fatally in two or three weeks and those chronic forms in which the initial disturbance is so slight as to be unnoticed.

## ACUTE (TRAUMATIC) ABSCESS.

Symptoms of the preceding causal factors are often present to which those of the intra-cranial suppuration are added. The latter resemble those of acute meningitis, which sometimes co-exists. The first symptom to arise is usually headache of a severe throbbing nature, often localised to the region of suppuration. It is greatly increased by a dependent position of the head and by the administration of alcohol, but these signs do not distinguish this form of headache from that due to meningitis and intra-cranial growth. Vomiting is invariable and is accompanied by considerable prostration. Pyrexia is always present and is usually high and in the majority of severe cases repeated rigors occur. While there may be considerable restlessness, delirium is rarely an early symptom, neither does paralysis occur except in the final stages. Convulsions are not common, but in cases in which the abscess presses upon the cerebral convolutions local epileptiform attacks may occur. On account of the rapid course of the disease if unrelieved, optic neuritis is not usually met with until the last stage is reached, but it occurs in most cases of abscess, both chronic and acute. These symptoms pass on in the course of three or four days into those which also attend the terminal stage of chronic abscess. Delirium gives place to stupor, which deepens into final coma. Severe convulsions are the rule, and inequality of the pupils and irregular movements of the eye are common, but paralysis of the cranial nerves is rare. Paralysis of hemiplegic distribution and of varying intensity upon the opposite side to the abscess is the rule, but it is rarely more than slight, and when a subdural abscess involves the brain tissue extensively there may be localising symptoms. The pyrexia becomes more severe, and repeated rigors with profuse sweating are common. The pulse is generally slow. In a few cases life has been prolonged until thirty days, but as a rule death occurs in from seven to ten days.

## CHRONIC ABSCESS.

In many cases the initial symptoms accompanying the formation of an encephalic abscess are so slight as not to attract attention, and especially is this the case when symptoms of chronic otitis are troubling the patient. For this reason it is often impossible to fix the date of commencement of an encephalic abscess. The initial symptoms are similar to those of acute abscess, but are, as a rule, much less severe—headache, usually not localised, vomiting, and slight pyrexia. The difficulty of clinically determining the advent of an encephalic abscess with any degree of certainty is rendered obvious by the fact that the most grave cerebral symptoms and many of the signs of meningitis, including even optic neuritis, may suddenly arise in a case of chronic disease of the tympanic bones in the absence of any visible intracranial complications, such as meningitis, sinus thrombosis, or abscess. Several of such cases have rapidly recovered after surgical interference, while in some fatal cases the examination has revealed old tympanic disease as the sole lesion. After its formation chronic encephalic abscess, for a longer or shorter period, does not excite symptoms, or it produces symptoms which are slight. This latent period may last any length of time, from a few weeks to as long as twenty years. Where abscesses have remained latent for a very long period the latency has been complete, and the abscess has always been found thickly encapsulated. But a latency of three to six months does not necessarily mean that a capsule has formed.

During the latent period there may be no symptoms whatever. More often the latency is incomplete, and trifling symptoms are present, of which headache is the most usual. It is not severe, and in cases of ear disease it may alternate with otorrhœa. Slight irregular pyrexia, mental alteration, and recurring convulsions are of occasional occurrence during the latent period.

In rare cases acute symptoms may come on and subside, the latency continuing as before. Such phenomena are probably due to a simultaneous occurrence of local meningitis. The onset of terminal acute symptoms may be sudden or gradual. In the latter case headache, mental change, restlessness, irritability, or depression may be present, and not infrequently optic neuritis precedes the onset of the more acute symptoms. The terminal stage is marked by severe headache, vomiting, and pyrexia—sometimes associated with a rigor. The pyrexia does not as a rule remain high except when suppurative meningitis has been set up by rupture of the abscess. The temperature may be even subnormal a day or two after the onset of symptoms. General convulsions are the rule and are often frequently repeated. Some degree of hemiplegia is usually present on the opposite side if the abscess be cerebral and of considerable size, and some degree of hemi-ataxy will be present upon the same side if the abscess exist in the lateral lobe of the cerebellum, and in this situation an abscess may cause rigidity of the neck muscles and head retraction. When situated in the pons an abscess may cause no symptoms when it is small. But when it is large the usual pontine signs—ocular, facial, and fifth nerve paralysis, with alternate hemiplegia or double hemiplegia—result.

In the temporo-sphenoidal lobe abscess rarely causes localising symptoms, aphasia and alterations of taste and smell being very uncommon and in young children almost unknown. Similarly in the pre-frontal lobe localising signs are almost invariably absent. The pulse is slow, and this is a most important diagnostic indication; shortly before death it may become irregular. Mental symptoms are conspicuous in this stage. Delirium occurs early and may be violent, but it is soon followed by mental depression evidenced by slow cerebration, afterwards by stupor, and finally by coma.

The subjects of intra-cranial abscess may die suddenly and unexpectedly, as do some patients with cerebral tumour.



We have as yet no adequate explanation of the mechanism of this occurrence.

The majority of the cases die in the comatose stage from respiratory failure. Sometimes the abscess ruptures and death rapidly follows the onset of severe signs of meningitis. Rupture into the meninges is not common in cerebral abscess, since cohesion of the membranes prevents the escape of the pus into the membranes; such abscess usually bursts into the lateral ventricle. On the other hand, abscess of the cerebellum not infrequently ruptures into the arachnoid space.

It may be remarked that whatever the unknown determinant of the advent of the terminal stage may be, the *post-mortem* condition found is always one of inflammatory œdema and softening round the abscess, sometimes associated with extension of the suppurative process into the brain tissue. The terminal stage is as a rule shorter than is the course of an acute abscess and death occurs usually in from four to eight days.

Very rarely a case of encephalic abscess presents from first to last the symptoms characteristic of progressive cerebral tumour. Such abscesses are apparently primary, and not the result of secondary infection, local or remote.

*Diagnosis.*—The diagnosis is sometimes impossible, often difficult, and only in rare cases can more than a probable determination be arrived at. The disease has no unequivocal symptoms, and the same clinical aspect may be present in cases of ear disease, sinus thrombosis, acute meningitis, and cerebral tumour. No specific rules can, therefore, be laid down with regard to the diagnosis. The majority of cases of intra-cranial abscess occur, however, as secondary results of conditions the presence of which can be discovered: nasal and tympanic disease, cranial injury, empyæma, etc., and all-important in the diagnosis is the history and the discovery that one of these conditions has existed or does exist. Where local cranial disease exists, tympanic disease, for example, and acute symptoms are present, the diagnosis has to be made between the grave cerebral symptoms sometimes

present in ear disease, meningitis, sinus thrombosis, and abscess. The signs that are useful are given here in the form of a table.

Symptoms . . .	Abscess	Sinus thrombosis	Meningitis.
Mental state . . .	Slow cerebration; hebitude; som- nolence	Acute delirium. Coma	Delirium.
Convulsions . . .	Less common	Usually present	Present.
Focal symptoms	Slight hemiple- gia; a slight h e m i a t a x y sometimes pre- sent	Absent	Cranial nerve palsies.
Optic neuritis . . .	Apt to be present	Unilateral throm- bosis of central vein of retina	Often absent.
External cra- nial signs . . .	Absent	Edema of orbit, of post mastoid re- gion. Cervical tenderness and rigidity; throm- bosis of jugular vein. Cervical glands swollen	Absent.
Pyrexia . . . .	Slight, sometimes subnormal tem- perature	Severe, with wide oscillations and rigors	Marked py- rexia.
Pulse . . . .	Slow	Rapid and small	Rapid.

This differential diagnosis is not of vital importance, and its consideration should cause no delay in summoning the aid of the surgeon. The presence of any symptoms of intracranial disturbance where tympanic disease exists calls for immediate surgical interference, and the surgeon, after cleaning out the diseased tympanum, completes the diagnosis by examining the lateral sinus and both the temporal lobe and the lateral cerebellar hemisphere and proceeds to those measures which the results of his examination indicate.

*Prognosis.*—Cases are on record in which cerebral abscesses with very thick walls or even calcified walls and inspissated contents have been found *post-mortem* in subjects dead of other diseases many years after the presumed time of formation of the abscess. Further, spontaneous evacuation of an abscess through the diseased ear or through a sinus in the area of the local cranial disease causing the

abscess has been followed by recovery. It is obvious that the probability of such results happening in any given case is so small as to be negligible. Further, it is highly probable that no abscess becomes permanently quiescent after it has given rise to severe symptoms. The prognosis is, therefore, that an invariably fatal result will occur unless successful surgical interference is possible. If the abscess is reached and drained, recovery is far from certain, since extensive perifocal softening, acute meningitis, and sinus thrombosis may co-exist with the abscess. Further, when an abscess extends some distance into the brain the resulting sinus heals with difficulty, and especially is this the case in subdural abscess the result of osteitis of the vertex. In consequence a residual abscess is apt to form long after the external wound is healed.

A question of great difficulty and responsibility occurs when after ear-disease slight symptoms of cerebral disturbance such as might indicate a latent cerebral abscess persist in spite of the cure of the ear-disease. Such a case should be kept under careful observation and within easy reach of a surgeon, and the first sign of an acute nature should be the signal for immediate operative procedure.

*Treatment.*—This embodies in the first place the most rigorous prophylaxis, and careful attention to bodily nutrition and sanitation in all children and especially in those convalescent from specific diseases, particularly scarlatina, measles, and whooping-cough, and the greatest care that every child coming under observation suffering with nasal discharge, otitis media, or infective disease of the scalp shall not pass out of observation until such disease be beyond all doubt cured.

The treatment of the pronounced disease belongs to the surgeon.

## HYDROCEPHALUS

THE term "hydrocephalus" denotes a regular distension of the ventricular system of the brain by the accumulation of cerebro-spinal fluid in it; and this distension is associated, sooner or later, with an expansion of the cranial bones and enlargement of the skull.

An old division of hydrocephalus was into acute and chronic, acute being applied to the condition of tuberculous meningitis. But since any marked degree of ventricular distension is unusual in that affection, and enlargement of the head very rarely occurs, this term has fallen out of use. In the majority of cases in which general atrophy of the cerebral tissues occurs, fluid accumulates both in the ventricles and in the subarachnoid space, but such compensatory enlargement is not to be regarded as, in any sense, of the same nature as true hydrocephalus. Such accumulation of fluid is found in cases of cerebral diplegia and general paralysis of the insane in children, and it also occurs in the brains of old people. It is merely the result of wasting and shrinkage of the brain-tissue, and the accumulation of fluid takes place in order to fill up the space which is vacated within the rigid skull.

The enlargement of the head which is not uncommonly found in rickets, has no connection with hydrocephalus. It is probably the result of malnutrition of cranial bones, which grow irregularly, and, being unduly soft, yield somewhat to the intra-cranial pressure. In rare cases, of moderate degree, ventricular distension has been met with, but the enlargement of the head is never progressive, and the symptoms of hydrocephalus are absent.

According to their clinical aspect, cases of hydrocephalus may be placed in three groups: (1) congenital hydrocephalus, in which the enlargement of the head is present

at the time of birth, or begins to be apparent during the first two years of life ; (2) acquired primary hydrocephalus, which may appear at any period of life ; (3) secondary hydrocephalus, the result of intra-cranial growths, or of sclerosing meningitis.

#### PRIMARY HYDROCEPHALUS, CONGENITAL AND ACQUIRED

The clinical manifestations of hydrocephalus, whether congenital or acquired, fall into two groups, which result, respectively, from the effects of the abnormal intra-cranial pressure, first upon the brain-case, and secondly upon the nervous structures. In the congenital form the enlargement of the head is the first noticeable feature ; and this is true also of some cases of acquired hydrocephalus in young children. In most cases of acquired hydrocephalus, on the other hand, the nervous symptoms are first in evidence—namely, persistent headache, vomiting, mental impairment, convulsions, and sometimes optic neuritis. The evidence of cranial enlargement may succeed these symptoms, and the older the subject, and consequently the more resistant the cranial walls, the more severe are the nervous symptoms, and the later is the cranial enlargement in appearing. In some cases of congenital hydrocephalus enlargement of the head takes place during intra-uterine life, and it may be so great as to make delivery impossible without destruction of the head. More frequently the cranial enlargement, not noted at the time of birth, becomes evident during the first few weeks of life. In rare instances the head is unusually small, and hydrocephalus may actually be associated with microcephaly. It is doubtful, however, whether the ventricular distension in such cases is more than compensatory for the atrophy of the cerebral structures.

Enlargement of the head is the most striking feature of hydrocephalus in children. The increase usually affects all the diameters of the cranial cavity, and is most marked on the vertex and least at the base. Trousseau compared the opening out of the cranial bones which occurs as the head enlarges to the falling back of the petals of an opening

flower. The forehead is large, rounded, and projects forwards; the temporal fossæ are obliterated, and the parietal eminences carried backwards. The vertex is often somewhat flattened, as may also be the occipital region. The direction of the external auditory meatus alters with the increasing size of the head; normally directed obliquely forwards, it comes to look directly inwards, or even obliquely backwards in severe cases. The head is frequently asymmetrical. In rare cases, premature synostosis of the sagittal suture occurs, and marked scaphocephaly results.



FIG. 17.—Extreme hydrocephalus. Dr. G. A. Sutherland's case.

In young children the sutures may be widely open, and then there is marked bulging along those lines and at the fontanelles. In advanced cases fluctuation may be present, and dimpling of the calvarium may be so produced. The skull may attain enormous dimensions, and many examples are recorded in which the circumference has been from 60 to 90 cm. The face is characteristically triangular, contrasting markedly with the forehead. Wasting of the facial subcutaneous tissues and retarded development of the maxilla and mandible often render this contrast still

more striking. Bulging of the orbital plates of the frontal bone presses down the eyeballs, so that the pupils become more or less covered by the lower lids, and a band of the sclerotic may be visible between the iris and the upper lid. The hydrocephalic child often uses its hands to depress the cheeks, and so draw down the lower lids out of the position in which they impair the line of vision. The hair of the head becomes scanty in marked cases, the subcutaneous veins of the scalp are often greatly developed and distended, and sometimes a vortex of distended veins radiates from the region of the anterior fontanelle, producing an appearance comparable to the *caput medusæ* of chronic abdominal distension. The general nutrition is poor, and bodily development retarded in proportion to the severity of the effect of the intra-cranial pressure upon the nervous system. In severe cases there may be a considerable reduction in the temperature of the body. Auscultation may reveal a cephalic bruit, but this is neither a characteristic nor a constant sign in hydrocephalus, for it is frequently met with in rickety children, and may be present in a normal subject.

The nervous disorders which appear during the course of hydrocephalus are both variable and inconstant, and acute symptoms are of rare occurrence if the disease appears at an age at which the skull is still yielding. On the other hand, if the ventricular distension commences when the growth and ossification of the skull are complete, the nervous symptoms which arise are very severe, and resemble closely the general effects of intra-cranial growths. In secondary hydrocephalus the symptoms due to this condition emerge more or less definitely from those of the preceding meningitis or sinus thrombosis, or are blended with those of the co-existing intra-cranial growth.

In children the nervous symptoms of hydrocephalus, whether it be congenital or acquired, may be summed up in the following list, the symptoms being frequent in the order in which they are enumerated: convulsion, mental failure, spastic paralysis of the limbs, headache, optic

atrophy, nystagmus, vomiting, optic neuritis. There is no constancy in the occurrence of these symptoms. Convulsion may not occur at all, and mental acuity may be unimpaired. Spastic weakness occurs in less than one half of the cases, whilst optic atrophy is met with still more rarely, and optic neuritis is distinctly unusual.

*Convulsion.*—While it is to be borne in mind that the whole course of hydrocephalus in children may run without the occurrence of convulsion, yet in the majority of cases this symptom is conspicuous. In some congenital cases the convulsions are in part attributable to a condition of lobar atrophic sclerosis or of porencephaly, affecting the hemispheres, which is sometimes associated (see Cerebral Diplegia). In some of the post-natal cases the symptoms of cerebral disorder are ushered in by convulsion, and it is probable that such convulsions are the immediate expressions of the as yet unknown morbid process of which the primary hydrocephalus is the final result.

The convulsions which recur at intervals throughout the course of the majority of cases of hydrocephalus, whatever its nature, result from a condition of functional instability of the cerebral cortex which long-continued increased intracranial pressure brings about. The cerebral cortex is pressed upon and is gradually wasted. The circulation of the blood and lymph is abnormal, and in the later stages of the disease widely-spread anatomical degeneration of the nervous elements occurs. We know that pressure, malnutrition, interference with the normal blood supply, and degeneration of nerve elements when occurring in the cerebral cortex are most potent causes of convulsion. The absence of convulsion in some cases of hydrocephalus, therefore, is a most striking example of the powers of resistance and endurance of the nervous elements to slowly induced abnormal conditions.

The convulsions are usually general, with loss of consciousness. A preceding aura is rare. Local commencement of the spasm which becomes general, associated with loss of consciousness, is not infrequently met with. Local



convulsion without loss of consciousness (Jacksonian epilepsy) may occur repeatedly in some cases and may lead to the erroneous diagnosis of a gross local lesion in the cerebral cortex, and especially is this error likely to occur when hydrocephalus is secondary to intra-cranial tumour. It is a safe rule to hold that Jacksonian convulsion is an all-important localising sign if it occur early in the course of intra-cranial tumour, but if it occur after general symptoms have existed several months it is of little localising value, for it is then not unlikely to be the result of secondary hydrocephalus.

The explanation of the occurrence of such local convulsion is that the effect of the pressure is not equally distributed upon the cortex: the central region tends to suffer earlier and in greater degree than do other regions, and that some *locus minoris resistentiæ* loses its functional stability earlier and in greater degree than does the rest of the cortex.

Simple loss of consciousness is not uncommon, but *petit mal* and mental lapses are rare.

All degrees of *mental reduction* occur, from the least noticeable to complete idiocy. The more severe forms of mental impairment are met with in congenital cases and especially when cerebral agenesis, porencephaly, and teratological defects are associated.

The psychical reduction is less prominent the greater the age at which the symptoms commence, and, as a rule, the intelligence is far greater than the severity of symptoms (cranial enlargement, paresis, etc.) might lead one to expect. Cerebration is usually slow and the disposition placid, and periods of somnolence are of common occurrence.

*Paresis and contractures* may be symptomatic of associated cerebral agenesis, and when this is so they are present from birth. The effect of long-continued ventricular distension in many cases is to cause degeneration of the pyramidal system, and according to its degree the latter entails bilateral spastic paralysis with contracture. The first signs of the onset of this event are exaggeration of the

deep reflexes and the change in type of the plantar reflexes from the flexor response to the extensor response. The lower extremities are affected earlier and to a greater extent than are the upper extremities, and at one period of the disease a case may present the picture of cerebral paraplegic rigidity comparable with that of Little's disease.

The upper extremities are affected later. The paresis of the limbs is almost always symmetrical and equal upon the two sides. Sensibility is generally normal.

*Vision* is interfered with in a considerable proportion of the cases. The frequency of myopia has been already mentioned. The enlargement of the infundibular portion of the third ventricle, by pressure upon the inner borders of the converging optic tracts, may cause bitemporal hemianopia with atrophy of the nasal portions of both optic discs, this condition subsequently progressing to complete blindness and complete optic atrophy.

More often the increased intra-cranial pressure causes atrophy of the optic tracts and secondary atrophy of the optic discs.

In other cases *optic atrophy* is the result of optic neuritis. In late childhood and in adult life optic neuritis is the rule, and optic atrophy seems always to be consecutive to this. In young children, however, while optic atrophy is common, optic neuritis is very rarely seen, and if such optic atrophy is secondary to neuritis it cannot be distinguished from primary atrophy by the ophthalmoscope. The writer inclines to the opinion that optic atrophy occurring in the hydrocephalus of young children need not be consecutive neuritis. Strabismus is commonly present in congenital cases, and it is most frequently convergent. Nystagmus is met with in the subjects of hydrocephalus, who are blind from optic atrophy, and it is of frequent occurrence in long-standing cases in which spastic paresis is well marked. In the latter case it is probably a spastic ocular manifestation indicative of the degeneration of the cortical motor neurons subserving eye movements, and it is a feature of the same order as is spastic paresis in the limbs and trunk.

Headache is often complained of and especially during the early days of illness in acquired cases, but this symptom never dominates the clinical picture in children, and is never so severe and persistent as that arising from the presence of an intra-cranial growth. Cerebral vomiting is of comparatively rare occurrence.

When one considers the profound anatomical alterations which take place in the advanced stages of the disease, the occurrence in some cases of unusual symptoms indicative of interference with the functions of the cerebellum, brain-stem and cranial nerves is easily explicable. Unilateral or bilateral ataxy, vertigo, deafness, anosmia and paralysis of cranial nerves, are the most important of such unusual symptoms.

The signs of failure of the nervous system as a whole usher in the fatal result in severe cases. For some days or perhaps weeks before death, hebetude may become profound; spastic paresis gives place to flaccid paralysis with muscular wasting, the deep reflexes disappear and the sphincter mechanism loses its control and subsequently its tone.

Hydrocephalus which commences in late childhood or in adult life presents an aspect widely different from that just described. At these periods of life the bones of the skull are firm and resistant, and the sutures resist for a long time before yielding to the increased intra-cranial pressure. The general symptoms are acute, and the course of the disease is often rapid to a fatal termination. There is usually no enlargement of the head to aid the diagnosis, and the symptoms resemble those of a non-localisable intra-cranial growth and are headache, vomiting, and optic neuritis.

The headache is severe and usually paroxysmal, and it may be so intense as to cause sudden death, while not infrequently the sufferer loses all control during the paroxysms. Speaking generally, the headache is of much greater severity in adult hydrocephalus than in intra-cranial growth. Similarly vomiting is apt to be more severe and persistent

than that associated with cerebral growth. In many of the cases a fatal result occurs before enlargement of the head occurs, and before cerebral degeneration has produced further signs of spastic paresis than an increase of the deep reflexes, foot clonus, and the change of the plantar reflexes to the extensor type. General convulsions and attacks of coma are not rare.

*Etiology.*—Hereditary influences are undoubtedly of very great importance in the causation of congenital hydrocephalus. This disease frequently affects several children of the same parents, and it may even appear as a striking family disease affecting members of several generations of the same stock. The writer has on two occasions seen hydrocephalic twins. Direct transmission is extremely rare, since the subjects of congenital hydrocephalus scarcely ever attain to procreative capacity. The teratological associations of the malady are so frequent that hereditary morbid influence leading to defective development must play an important part in the causation, both of family and sporadic cases. Partial anencephaly and porencephaly may be met with, and Messing has shown that hydrocephalus co-exists in a large proportion of cases of congenital porencephaly. Spina bifida, meningocele, and hydromyelia are of frequent occurrence in association with this disease, and arrested and irregular development of the brain-stem and cerebellum are the rule. Among other bodily deformities not infrequently associated with congenital hydrocephalus may be mentioned harelip, cleft palate, talipes, rectal and testicular ectopia, and imperforate anus. Of these abnormalities some may be regarded as the possible results of increased intra-ventricular tension at any early stage of intra-uterine life. It is conceivable that spina bifida, meningocele, porencephaly, and defective development of the central nervous system may be produced in this way. The widely-spread deformities of the body, however, must be considered with the hydrocephalus as concomitant effects of some teratological cause. Consanguinity of parents and parental alcoholism have been recorded, and syphilis seems to have a definite connection

with certain cases of congenital hydrocephalus. It is not uncommon for the stigmata of congenital syphilis to make their appearance in the hydrocephalic child after birth. Yet, in the majority of such syphilitic cases, the morbid anatomy of the hydrocephalus presents no special feature. In a few cases definitely syphilitic lesions of the ependyma in the region of the brain-stem have been found. The causation of primary hydrocephalus occurring after the time of birth is often obscure. The majority of the cases occur in childhood, yet no period of life seems to be exempt. History relates that Dean Swift died of this malady at the age of 70, after having suffered from symptoms for three years. In children acute infective diseases, and especially gastro-intestinal infections, may occur as antecedents of hydrocephalus; but the nature of their causal relationship to the disease is unknown. In adults, syphilis stands in important relation in certain cases, some of which have been examined pathologically. In three of such cases there was evidence of old syphilitic disease of the brain, but no obstructive lesion by which ventricular distension could have arisen mechanically. A history of traumatism, such as a severe blow or fall upon the head a short time prior to the onset of the symptoms is obtained so frequently as to have led to the description by certain authors of traumatic hydrocephalus as a special variety. There seems to be little doubt that injury may be the immediate cause of the appearance of symptoms, and that it is not always merely coincidental. Thus, a patient at the age of 18 months was dropped several feet upon the top of his head. The head was severely bruised, and he was unconscious for several days. The head afterwards began to enlarge and assume a peculiar shape. He died at the age of 35 years. The cranial deformity suggested strongly that the weight of the body had driven the base of the skull upwards at the time of the injury. A large patch of ossified dura in the region of the vertex suggested the site of a long antecedent meningeal hæmorrhage. The condition was one of simple hydrocephalus, and hydromyelia was also present.

## SECONDARY HYDROCEPHALUS

Under this name may be grouped together all cases in which there is obstruction in the usual path by which the cerebro-spinal fluid leaves the ventricular cavities, or to the venous outflow from the choroid plexuses. But it is by no means clear that such obstruction is the sole or even the important agent in producing the ventricular distension.

The *causes* of secondary hydrocephalus are, first, by the sclerosing forms of meningitis, especially posterior basic and epidemic meningitis, very rarely tubercular; secondly, intra-cranial neoplasms encroaching upon the ventricular system, especially tumours of the brain-stem and sub-tentorial region; thirdly, adhesive phlebitis of the cerebral blood-sinuses.

*Pathology.*—Hydrocephalus is immediately due to an excess of cerebro-spinal fluid present in the cavity of the central nervous system. Such accumulation may theoretically result from the over-production of cerebro-spinal fluid by the epithelial lining of the nervous canal, or from a partial or complete obliteration of those paths by which the fluid leaves the ventricles to enter the arachnoid space, or of the channels by which the fluid returns to the blood-stream. Our knowledge concerning the origin and fate of the cerebro-spinal fluid, and of the manner in which it circulates, is at the present time far from complete. It appears to be secreted by the ependymal lining of the nervous canal, especially by that portion overlying the choroid plexuses of the ventricles. It seems not to be secreted by the lining membrane of the spinal canal, since the complete obliteration of the latter by a centrally-placed tumour is not followed by distension of the central canal below the seat of obstruction. In some cases of cerebral tumour a part of the lateral ventricles may be completely cut off by the growth from the general ventricular cavities, generally the tip of either the anterior or posterior cornu; and under such circumstances the isolated portion of the

ventricle does not necessarily become distended. This fact would suggest that the cerebro-spinal fluid is chiefly secreted in the region of the choroid plexus, and in small part only by the general lining of the ventricle. There are three pairs of choroid plexuses, situated respectively in the floor of the lateral ventricles, hanging from the roof of the third ventricle, and in the roof of the fourth ventricle. Cerebro-spinal fluid secreted in the region of these is supposed to leave the nervous canal and enter the arachnoid space by certain apertures where these spaces communicate. Such communications exist in the region of the anterior perforated spots,—those opening into the lateral ventricle in the region of the posterior perforated spot, between the diverging crura cerebri, those leading into the third ventricle, and, lastly, in the thin roof of the fourth ventricle. The latter are comparatively large apertures and are three in number, one being situated dorsal to the opening of the central canal of the spinal cord (foramen of Majendie), and the other two are placed one over each lateral recess of the fourth ventricle (foramina of Luschka).

Entering the subdural space, it is supposed that the fluid is absorbed by the vessels of the Pacchionian bodies and by the vessels of the meninges generally.

There are certain strong arguments that much of the cerebro-spinal fluid is absorbed in the lower part of the theca spinalis. If, for example, particulate matter be injected into the subdural space of an animal, it can be found within a few minutes in the lower part of the theca even though the animal is kept in an inverted position. Further, it is the rule in human subjects where blood has escaped into the subdural space of the cranium as the result of injury or operation, to find blood-clots around the nerves of the cauda equina, when opportunity is afforded for examining such cases at an early period after the extravasation has occurred. The blood-clot apparently swept into the lower theca by a constant stream of the cerebro-spinal fluid is found even when the cranial dura has been opened and

free drainage from the brain-case allowed and when the supine position has been constantly maintained.

A further argument in support of the stream of cerebro-spinal fluid from brain-case to theca is that in certain cases of cerebral tumour, as a result of general pressure, the cerebellum and medulla become pushed down and plug the foramen magnum. Distension of the ventricle and urgent symptoms immediately follow, while no cause for such distension other than the filling up of the foramen magnum is discernible.

The wide variability of the Pacchionian bodies as regards number and size tends to discount the assignment to them of an all-important *rôle* in the absorption of the cerebro-spinal fluid. Notwithstanding that many writers have disputed the fact that changes in the lining membrane of the nervous canal can, *per se*, cause hydrocephalus it is undoubtedly true that in many cases such changes are the only abnormalities to be found *post mortem*, and this apart from all obstruction at the point of exit of the cerebro-spinal fluid. It seems, therefore, to be very probable that the general internal surface of the nervous canal has the function of absorbing the fluid.

The quantity of cerebro-spinal fluid present in the normal human subject is quite small, probably less than six ounces. It comes and goes as the volume of the central nervous organs expands and contracts under the influence of the blood pressure and of the relative calibre of the cerebral vessels. In so doing the intra-cranial pressure is kept almost constant. When the subdural space is opened the amount of cerebro-spinal fluid which may escape in the course of twenty-four hours is great, amounting in some cases to several gallons. It is improbable that secretion takes place as rapidly in the intact subject, and it may be that the copious draining of the fluid when the dura mater is opened is simply the expression of the natural mechanism for raising the lowered intra-cranial pressure to the normal.

*Pathogenesis.*—According to the results of the investiga-



tion of the central nervous organs, cases of hydrocephalus fall into two groups. In one, the ventricular distension is the result of a known antecedent morbid condition, either meningitis or some form of intra-cranial growth, and the mechanism of its production, in part at least, is the obliteration of the usual paths of flow of the cerebro-spinal fluid. In addition, the strangling of the veins collecting blood from the choroid plexuses may cause a congestion and consequent transudation to occur. The second group comprises those cases in which the hydrocephalus is apparently idiopathic, the veins and the outlets of the cerebro-spinal fluid being free. There is always some departure from the normal to be found in the structure of the ependymal lining of the ventricle, and sometimes the change is very marked. Meningitis is apt to be followed by hydrocephalus in proportion as recovery from the initial disease occurs, in proportion as cicatrisation follows the inflammatory process, and in proportion as the affection of the covering of the fourth ventricle tends to cause obliteration of the foramina of Majendie and Luschka; and, further, in proportion to the amount of ependymal cicatrisation that may occur. When blocking of the posterior foramina occurs, the only path of exit for the cerebro-spinal fluid is by the fine canals in the perforated spots. These, when patent, appear to be inadequate to prevent a relative increase of intra-ventricular pressure, for they are frequently found so enlarged in cases of hydrocephalus as to admit a fine probe. More frequently, however, they too are easily sealed by the inflammatory process, and a gradually increasing general ventricular distension thus results.

Not infrequently the vascular supply of the large choroid plexuses of the lateral and third ventricles is interfered with by cicatrisation following inflammatory processes which, spreading between the cerebrum and cerebellum, have extended to that double fold of pia arachnoid which extends into the great transverse fissure of the brain and separates the lateral and third ventricles, the *velum interpositum*. This membrane carries the great

choroid plexuses, those of the lateral ventricle upon its upper surface, and those for the third ventricle upon its under surface, and lying between its folds are the veins of Galen which convey the blood from these plexuses to the straight sinus. Cicatrization of the *velum interpositum* of necessity hinders the blood flow in the veins of Galen, and there being no anastomosis with the superficial cerebral veins, the result is an increased production of fluid by the congested choroid plexuses.

In the hydrocephalus following meningitis the phenomena above described often co-exist.

Attention may be justly directed, however, to the other possible factors. It has been suggested above that the general surface of the ependyma is probably an absorbing surface rather than a secreting surface for the cerebro-spinal fluid, and both from direct involvement of the general ependyma by the inflammatory process and from the interference with its functional capacity which the toxins of meningitis may very possibly produce, the ependyma may cease to act as an absorbing surface. Further, if, as is highly probable, some of the cerebro-spinal fluid find its way out of the arachnoid space in the lower part of the theca, it is possible that processes similar to the above may close this path also and give rise to the general thecal distension which is sometimes met with in these cases.

It will follow from the above statements that hydrocephalus is not a sequel of the more acute and rapidly fatal forms of meningitis, but of the more chronic and indurated forms when these affect the posterior fossa of the skull, namely, epidemic meningitis, posterior basic meningitis, and syphilitic meningitis.

Tumours of the brain cause hydrocephalus in the majority of cases by simple mechanical obstruction of the channel of exit of the cerebro-spinal fluid. The growth is usually situated in the mid-brain or pons and either by pressure or actual involvement occludes the Sylvian aqueduct. Less commonly pressure upon the brain-stem from a growth in the immediate neighbourhood may cause

the same result—for example, a tumour of the fifth nerve pressing upon the pons.

In these cases distension above the obstruction alone occurs. Tumours of the cerebellum pressing downwards in the middle line and villous tumours of the choroid plexuses of the fourth ventricle may produce the same effect. A tumour involving and obliterating both foramina of Munro does not seem to cause distension of the lateral ventricles, the reason being, perhaps, that apertures exist or are found in the *column interpositum*.

Tumours which are firm and those which grow rapidly are more liable to obstruct the aqueduct than those which are soft or which grow slowly.

It may be remarked in this place that in the majority of cases of cerebral tumour of long standing some dilatation of the ventricles is found *post mortem*, yet the growth neither involves nor presses upon the aqueduct, but may be situated in the frontal or temporal lobe or may even be extra-medullary and growing from the skull in the anterior fossa. The degree of distension is usually not great and it involves all the ventricles. Sometimes, however, the distension is extreme and the outward manifestations of hydrocephalus are present. The cause of this distension is a subject worthy of consideration, since it may throw some light upon the obscure causal factors of idiopathic hydrocephalus. Obstructive distension of the ventricles of severe degree occurs only in those cases of cerebral tumour which have survived the onset of symptoms a long time (a year or more), and in which pressure symptoms have been constantly present, and the longer the duration of symptoms the greater is the distension. The effect of prolonged intra-cranial pressure in pressing down the cerebellum and medulla into the foramen magnum and so closing that aperture has been already mentioned. This is of invariable occurrence in the cases under consideration, and it will be found *post mortem* that the interior surface of the cerebellum is deeply indented by the edge of the foramen magnum, the portion below the indentation being

elongated and forming with the medulla a cone which with the spinal cord completely fills the upper part of the spinal canal. In one case the cerebellar elongation reached the level of the third cervical roots. Such obstruction alone is doubtless sufficient to produce distension of the ventricles, which in its turn increases the obstruction by further forcing down the plug into the foramen magnum, and it may be said that from this cause hydrocephalus is self-perpetuating. The efficiency of the obstruction at the foramen magnum is rendered very apparent by the fact that in these cases the theca spinalis is not generally distended.

It may not at first sight appear rational that obstruction to the general outflow of cerebro-spinal fluid from the cranial cavity should cause ventricular distension. It must be borne in mind, however, that the intra-cranial pressure is not a simple hydrostatic pressure, but a varying elastic pressure enclosed between the rigid cranial vertebral wall on the one hand and the elastic blood-vessels of the brain substance on the other. It follows, therefore, that the mean intra-cranial pressure can never during life exceed the sum of the maximal arterial blood pressure plus the resistance of the vessel wall to compression, since any such excess would entail immediately cessation of the cerebral circulation.

If the cerebro-spinal fluid is to flow constantly from ventricle to subdural space the pressure at the point of its origin (ventricle) must be constantly higher than at the point of its exit. When obstruction is placed upon the outflow the same condition holds good, and the fluid will tend to accumulate at its point of production within the ventricle, with the result that hydrocephalus occurs.

*Morbid anatomy.*—The quantity of fluid which is found in the ventricles after death varies greatly, a usual quantity being from 15 to 20 ounces. In long-standing cases with great cranial enlargement very large quantities have been met with, and in one instance recorded by Esquirol as much as 720 ounces were found.

The characters of the fluid do not differ greatly from those of normal cerebro-spinal fluid. Its density varies from 1008 to 1010. It is clear, colourless, or slightly yellowish and the reaction is alkaline. It contains a very small quantity of albumen and a comparatively large quantity of alkaline chlorides.

The dilatation of the lateral ventricles is always more extensive than that of the third ventricle, and is usually symmetrical upon the two sides, and it affects the body of



FIG. 18.—Ventricular distension (hydrocephalus) in Sir Thomas Barlow's case of basic meningitis, from a photograph by the writer. A preparation from the same photograph illustrates the article on posterior basic meningitis in Professor Clifford Allbutt's 'System of Medicine,' and this plate is published here with Sir Thomas Barlow's permission.

the ventricle more than the cornua, so that the central cortex is the most thinned. Sometimes one lateral ventricle is much more dilated than the other, and one of the horns may present extensive enlargement while the others are less affected, no cause being apparent for this irregularity, such as occlusion of any of the interventricular passages. The foramina of Munro are greatly enlarged and the anterior pillars wasted. The septum lucidum is often reduced to a thin membrane; sometimes it is perforated. The anatomical limitations and markings which are

normally seen upon the walls of the ventricles become less obvious and often unrecognisable.

The ependyma is sometimes thickened, generally shining, and the network of subependymal vessels stands out obviously. The choroid plexuses are hypertrophied and thickened in less acute cases, but they may be found small, flattened, and bloodless in acute cases. The dilatation of the ventricles occurs chiefly at the expense of the white substance of the hemisphere, the cortex and central ganglia being less affected. The convolutions are flattened, and the sulci indistinct or unrecognisable, according to the degree of distension. The thickness of the cerebral substance is much reduced, and especially in the region of the central convolutions, where in severe cases it may measure a few centimetres only. The corpus callosum is greatly wasted. In advanced cases the cerebral hemispheres have the appearance of a thin-walled sac, which collapses entirely when the contained fluid is allowed to escape. The third ventricle rarely shows a relative distension equivalent to that of the lateral ventricles. The soft commissure invariably disappears. The aqueduct of Sylvius may be much dilated, and when this is the case the fourth ventricle shares in the general ventricular distension, the cavum cerebelli being especially enlarged. In many cases, however, the aqueduct and the fourth ventricle are not markedly distended. In a few cases the aqueduct has been found closed as if by antecedent adhesive ependymitis. The posterior part of the cerebellum and the medulla are pressed down as a plug into the foramen magnum, and there is constantly a well-marked constriction in the posterior inferior part of the cerebellum caused by the pressure of the bony ring of the foramen magnum. The theca spinalis and its processes into the intervertebral foramina may be much distended, but in the majority of cases the interventricular space is not in fluid communication with the thecal space, and no distension is found.

The condition of the pial roof of the fourth ventricle is of great importance considering the widely-held opinion that hydrocephalus is of obstructive origin. In congenital

and in primary hydrocephalus there may be some thickening of the ependyma, both of the roof and of the floor of the fourth ventricle, but there is no blocking of the posterior foramina. (The variability of the foramen of Majendie and its frequent non-existence in childhood must not be taken as evidence of closure of this opening from meningitis.)

Where hydrocephalus is secondary to the presence of intra-cranial tumour the growth may be situated in any position, either above or below the tentorium, and is not necessarily in such a position as to cause direct obstruction to the flow of the cerebro-spinal fluid. Sometimes the lateral ventricle may be completely divided by a growth, and then the isolated portion of the ventricle is sometimes found distended, and sometimes it is found empty. Tumours, which directly or indirectly block the foramen of Munro, and the aqueduct of Sylvius, are almost invariably associated with hydrocephalus, and of these glioma of the pons is the most frequent. The plugging of the foramen magnum by the lower part of the cerebellum and medulla, which has been already referred to, is invariably present wherever the tumour may be situated, and the obstacle which this plugging places upon the free outflow of the cerebro-spinal fluid into the theca is, doubtless, of great importance in the production of hydrocephalus.

When hydrocephalus is secondary to meningitis there is usually much meningeal cicatrisation about the roof of the fourth ventricle at the anterior and posterior perforated spots, and about the edge of the tentorium, where the veins of Galen enter the straight sinus, and extending forwards along these veins in the velum interpositum.

The ependyma is usually thickened and shows evidence of past ependymitis.

*Diagnosis.*—When enlargement of the head is manifest the diagnosis of the disease presents no difficulties. The large head of rickets is easily distinguishable from hydrocephalus by its different conformation and by the association of the other signs of rickets, by the absence of nervous symptoms (the convulsions commonly associated with rickets

being borne in mind), by its non-progressive nature and by the results of anti-rachitic treatment. In rare conditions known as macrocephaly the head is of normal shape and does not undergo progressive enlargement, and there are no symptoms of disease.

The distinction between the primary and the secondary forms of hydrocephalus in children should present no difficulty if a correct history of the early symptoms can be obtained. The initial manifestations in the primary form are slight and cannot be confused with those of meningitis or of sinus thrombosis. Intra-cranial growths which cause early and marked hydrocephalus are situated in some part of the brain-stem from the third ventricle to the medulla, and growth in such a position must of necessity produce such early pathognomonic localising signs as to leave no excuse for erroneous diagnosis save imperfect observation.

The advent of ventricular distension in the course of intra-cranial growth is generally to be detected by the occurrence of widely-spread bilateral signs such as bilateral spasticity, bilateral ataxy, general convulsion, and enlargement of the head.

In the primary hydrocephalus of childhood the disease is not distinguishable until the head begins to enlarge.

In adults the absence of cranial enlargement in most of the cases makes it impossible to separate the malady certainly from intra-cranial growth. It must be borne in mind, however, that headache, vomiting, and optic neuritis of rapid progress and existing alone as signs of disease, are not necessarily signs of intra-cranial growth, but may be the symptoms of primary hydrocephalus. The employment of lumbar puncture is valuable in some of these cases for purposes of diagnosis. The thecal space is more likely to be in communication with the ventricular cavities in hydrocephalus than in intra-cranial tumour. The amount of cerebro-spinal fluid that may be allowed to escape slowly without causing grave symptoms is much greater in hydrocephalus. In both conditions the operation may cause considerable temporary relief of symptoms.



The *prognosis* depends upon the cause of the hydrocephalus and upon the degree of severity of the symptoms and upon whether it is progressive or not. In all severe and progressive cases the prognosis is hopeless, and the same is true of hydrocephalus secondary to intra-cranial neoplasm. In some of the slighter cases, both of the congenital and of the acquired form, the process becomes arrested, and the patient may attain to adult life with the possession of all his faculties. Spontaneous discharge of the fluid through the nose has resulted in cure, but this event is so rare as to be negligible in prognosis.

In cases in which the disease becomes stationary the prognosis as regards mental capacity and as regards the continuance of recurring convulsion has to be considered. If the mental capacity at the time of arrest is fair it is not likely to deteriorate further unless epilepsy is established. When mental reduction is marked at the time of arrest a great degree of improvement cannot be reasonably expected.

*Treatment.*—While some cases of hydrocephalus cease to progress, and the symptoms disappear permanently under medical treatment, a like result has occurred in cases in which no treatment has been applied. It is, therefore, difficult to determine with certainty whether or not treatment has any effect in arresting the disease. Compression of the skull, by means of a tight capelline bandage, was introduced more than a century ago by Goelis, of Vienna, and recently, in the hands of Bourneville, good results have been obtained. Trousseau used strips of diachylon plaster, tightly applied. More recently, Dr. Samuel West has advocated the use of a broad elastic band as more easily applied and more manageable than are plasters and capelline bandages. When such treatment is adopted, the head should be shaved and the pressure applied for periods of a week at a time, a week's interval being given between each application, during which time regular mercurial inunctions are administered to the head. D'Astros considers that this treatment should be persevered in for a year.

The importance of syphilis in the etiology of hydrocephalus suggests the employment of antisyphilitic treatment, and it is certain that the application of mercurial ointment to the head seems to do more good than any other measure, while the administration of iodide of potassium in full doses seems to benefit a few cases.

The results of surgical interference for the relief of pressure and to attempt the re-establishment of a way out for the cerebro-spinal fluid have been, up to the present, so unfavourable that many writers and authorities consider such measures unjustifiable. It must be borne in mind, however, that in severe and progressive cases one is dealing with a necessarily fatal malady, and a few encouraging results have been published which appear to justify farther investigation. Paracentesis of the ventricle is both useless and dangerous, for when relief follows the operation, it is only temporary, and where cerebral tension is very high an immediately fatal result may supervene. Repeated lumbar puncture is advisable in the earlier days, but this is only possible in cases in which the theca is in free communication with the ventricular space.

Various operations have been performed for the purpose of re-establishing communication between the ventricular and the subarachnoid space. The fourth ventricle has been drained and occlusion in its roof rendered patent, the trephine opening being made through the occipital bone; this operation has not been attended with successful results. An aperture has been made in the corpus callosum *via* the anterior fontanelle with the object of establishing permanent communication between the lateral ventricle and the subarachnoid space.

The only operation based upon scientific principles which has yet been suggested for the treatment of hydrocephalus is that which was brought forward a few years ago by Dr. G. A. Sutherland and Mr. Watson Cheyne. The treatment was based upon the physiological work of Dr. Leonard Hill on the relation between the cerebro-spinal fluid and the cerebral venous circulation. Amongst Dr. Hill's con-

clusions were the following: Firstly, that fluid escapes directly into the veins from the subdural and subarachnoid space at any pressure above the venous pressure. The tension of the cerebro-spinal fluid and the cerebral venous pressure are normally the same. Secondly, that the rate of the transudation of the cerebro-spinal fluid depends directly upon the difference between the cerebral venous pressure and the subarachnoid pressure. Diminished subarachnoid pressure produces increased transudation until the cerebral venous pressure and the cerebro-spinal pressure are again equalised. Thirdly, that no pathological increase of cerebral tension can be transmitted by the cerebro-spinal fluid, because this fluid can never be retained in the meningeal spaces at a tension higher than that of the cerebral veins. These conclusions suggested the lines of treatment adopted by Dr. Sutherland and Mr. Watson Cheyne. Their aim was to establish a permanent communication between the ventricles and the subarachnoid space; for then, they concluded, no matter how much fluid might be poured out into the ventricles, it would be at once carried off by the veins; and thus all injurious pressure, whether intra- or extra-cerebral, would be removed, and the brain would be free to develop. Further, by such means matters would regulate themselves in a manner that no method of external drainage could effect: for when the communication was once established, so long as it was kept up, the rest might be left to Nature. The result in their first case was extremely interesting. All abnormal intra-cranial pressure disappeared, the shrinkage in the size of the skull was steadily progressive, until a point was reached when the bones were over-riding to the fullest possible extent, and further diminution in the capacity of the cranium was checked. Unfortunately, the factor necessary for the complete removal of the intra-cranial fluid in that case was absent, namely, the expansion of the brain substance. Consequently, although the ventricles were completely drained, a considerable quantity of fluid remained in the subdural space, as shown at the

necropsy. As there was no increased tension, however, it was not the presence of this fluid which hindered the expansion of the brain. The first case in which this treatment was carried out was one of apparently syphilitic meningitis occurring during intra-uterine life. But it is doubtless the case, as the authors remark, that many cases occur which are more suitable for treatment, and in which this plan of intra-cranial drainage might be worth trying. It seems that the difficulty is to maintain the communication between the ventricles and the subarachnoid space. Catgut is apt to be absorbed; horsehair apparently wanders; so that for such an operation, it is evident that some form of more or less permanent non-irritating drain, which is not likely to be moved, is a very great desideratum. Several operations on the same lines have been carried out by other surgeons. Occasionally the results have been fairly good, but in many cases they have been almost as bad as those of former less carefully thought-out measures. But it is obvious that if anything is to be done in the way of operative procedures for such otherwise hopeless cases, such operative procedures as that just referred to, based, as they are, upon careful scientific observations, are much more to be relied upon than any other measures.

## INTRA=CRANIAL TUMOURS

THE pathology and symptomatology of morbid growths arising within the brain are subjects upon which our present knowledge, though making rapid yearly advancement, is still scanty; and so complex is the anatomical structure and so intricate the physiological mechanism of the central nervous system, that the investigation of this subject is fraught with extreme difficulty. The difficulties which occur in clinical examination are chiefly due to the following causes: *Firstly*, the very incomplete understanding of the nature and localisation of the higher cerebral functions which we possess. *Secondly*, the lack both of constancy and of permanence in the symptoms and signs resulting from local destruction of the brain. This is due to two well-known physiological phenomena, the one being that some other part of the brain may compensate for local destruction of one part by taking over its function; the other is that there is generally not one path, but several paths, for the impulses subserving a particular function, and therefore destruction of the chief path does not entail permanent and complete loss of function.

The first difficulty is the greatest, for it concerns the higher cerebral functions. It will become apparent if we consider for a moment the possible effect of a small local destructive lesion in one of the regions of the brain supposed to be the seat of higher psychological processes. A loss results of psychological elements, but unless the loss is very great, the patient's deficiency is devoid of external expression. Even when very great, this psychological loss may appear to the most careful investigator as slight mental

reduction only. We are able to see the result of a local destruction of the motor cortex since the several parts of the body are locally represented in that region in a preponderant degree, and local motor-paralysis results; but, on the other hand, the body is not locally represented in the psychical areas; *i.e.* local paralysis in consciousness, if the expression may be allowed, has rarely any objective concomitant. The question naturally occurs, Why is an obvious mental derangement so rare in the symptom complex of local lesions of the frontal, temporal, and occipital lobes? The answer is that a negative (destructive) lesion cannot produce positive symptoms. Positive symptoms (delusion, convulsion, etc.) arise secondarily from pressure, partial evasascularisation, and from intoxications, with the products of the degeneration in the region of the negative lesion, and are referable to activity of living elements whose functional capacity is deranged.

*Pathology.*—The nature of the neoplasms arising in the central nervous system is also a matter of difficulty, the more so on account of the loose nomenclature that has been applied to them. Some of the growths are such as occur in other parts of the body, and these presumably arise from the meninges and blood-vessels (endotheliomata, sarcomata, etc.). The majority, however, are "brain tumours," *sui generis*, and are quite unlike growths occurring elsewhere.

Attention may be drawn here to certain growths characterised by the presence of many giant-cells, and usually described as tubercular growths or as gummata. They do not caseate or soften, and tubercle bacilli are never found in them. These tumours are probably of a special nature, distinct from the granulomata above mentioned.

Tuberculosis and syphilitic tumours of the brain and parasitic cysts are the results of the advent and growth of specific organisms in the brain, but little is known of the influences which determine the occurrence of other forms of intra-cranial tumour. Age influences the incidence slightly in that intra-cranial tumour is very rare during

the first two years of life. Sex seems to have no influence. Nor does heredity play any part in the causation except in cases of tuberculous and syphilitic tumours. It is rare for any indication to be obtained of an inherited tendency to morbid growths elsewhere.

A history of a fall or blow upon the head shortly prior to the onset of symptoms is so often obtained in cases of intra-cranial growth in children as to preclude the possibility that the association is a mere coincidence, and there is little doubt that such traumatic influences do act as immediate exciting causes of the appearance of symptoms in all forms of cerebral tumour. It is not difficult, moreover, to conceive the possibility of trauma being the direct cause of tubercular and syphilitic growth, since the focus of the organisms of these diseases may be determined by local lowering of tissue resistance caused by the injury.

The rarely occurring intra-cranial dermoid cyst has some explanation in a perverted process of development.

The essential cause of neoplasms, however, is as yet quite mysterious.

The varieties of tumours met with are as follows:

Simple cysts: Parasitic cysts; echinococcus and cysticercus cellulosæ; tubercle; syphilis; actinomycosis; granulomata.

Neoplasms not peculiar to the nervous system: Lipoma, fibroma, osteoma, angioma, endothelioma, myxoma, sarcoma.

Neoplasms peculiar to the nervous system: Glioma, psammoma, villous ependymal tumour.

Secondary tumours of the brain are not considered here. Always of rare occurrence, they are almost unknown in children. Secondary deposits do not often occur from tumours of the brain itself, and when so occurring they are almost invariably confined to the nervous system. The following case is a striking example of this curious fact. A child aged 12 years died of sarcoma of the middle peduncle of the cerebellum, and at the necropsy there were found no less than forty different tumours, the majority as large as a

hazel-nut, distributed in the posterior root ganglia and along the nerve roots of the cauda equina. A row of these tumours, comparable to a string of beads, were situated along the filum terminale. There was no involvement of any other organ of the body.

Tuberculous tumours occur almost as frequently as all other forms taken collectively. They may be of any size and may grow from any part of the brain-tissue. Often they are deeply seated and have no traceable connection with any process of the pia mater. They are hard in consistency and sharply limited from the brain substance, and it is a curious fact that they rarely soften and break down. They are often multiple and are rather more commonly seated in the cerebellum than in the cerebrum.

Gummata of the brain are very rare in children, and actinomycosis, which has been recorded in a few cases, pertains properly to the subject of cerebral abscess.

Of the neoplasms common to other organs, lipoma is the rarest of all brain tumours; fibroma, osteoma, and angioma are rare tumours growing from the skull and dura mater.

Endothelioma is a common tumour of the brain most often growing from the skull and dura, sometimes arising within the brain. It is a somewhat slowly growing, non-infiltrating, firm tumour. Its flattened cells are many of them arranged in the form of alveoli. It may be described as a neoplasm having the same relation to the endothelial lining of the blood-vessels as carcinoma has to gland epithelium. It is the most vascular of all cerebral growths, a point worthy of remembrance when operative procedures are in question.

Myxoma and fibromyxoma are encapsulated tumours growing always from the membranes. The myxomatous degeneration seen in gliomata and sarcomata is not to be confounded with true myxoma.

The sarcomata are of frequent occurrence in children. Those arising in the brain substance are of peculiar nature, and since every gradation is met with between these and pure gliomata, they may be perhaps more correctly described as tumours peculiar to the brain.



The glioma is by far the most frequently occurring neoplasm (15 per cent. of all cases). Its seat of predilection is the white matter, and it is the typical example of an infiltrating tumour, since it is impossible to define the limit between invaded and non-invaded tissue. The glioma is not very vascular, but its substance is very prone to undergo early degeneration, softening, and even liquefaction—a cyst resulting. Haemorrhage into its substance frequently occurs, and it is often very difficult to distinguish both macroscopically and microscopically between a primary haemorrhage and a haemorrhage into a glioma.

The psammoma is not an uncommon tumour. It arises from the leptomeninges, is firm, well encapsuled, and grows slowly. The characteristic concentric bodies are the ultimate result of a peculiar process of vascular obliteration, each concentric mass representing what was originally a small arteriole in transverse section.

The tumours of the ependyma resemble the papilliferous and proliferous adenomata.

*Comparison between the central nervous neoplasms of children and adults.*—The symptomatology may be said to differ so little in the child from the adult, that one description may be applied to both, if it be borne in mind that compensation for a local lesion occurs more rapidly and to a greater extent in the child, that general and local convulsions are more frequent, as the result of general increased intra-cranial pressure, and that the disease is as a rule not so rapidly fatal. More especially does the last remark apply to growths occurring at a period before the skull has arrived at such stability as to offer great resistance to an increasing intra-cranial pressure.

The frequency with which neoplasms affect the different parts of the brain is widely different, for while subtentorial tumours are much less common than supratentorial tumours in the adult, supra-tentorial tumours are very rare indeed before the age of 16 years, and still more rare before the age of 8 years. Symptomless tubercular masses are excepted.

At the National Hospital, 1895-1901, necropsy was performed upon 155 cases. The relation of situation of growth to age of subject is strikingly shown.

Adult.		Children under 16.	
Supratentorial . . .	112	Supratentorial . . .	5
Infratentorial . . .	13	Infratentorial . . .	25

Of the five cases in which a supratentorial growth was met with, three of the patients were 15 years old and the youngest was 11 years old. Such statistics warrant a more careful reconsideration of premises before the diagnosis of supra-tentorial tumour is determined upon in a child aged 10 years or under.

Intra-cranial tumour would appear to be very rare during the first three years of life—tubercular nodules excepted.

As regards the relation of the nature of growth to age, all that can be said is that tuberculous tumours and villous ependymal tumours are much more common in children than in adults, and that endothelioma seems not to occur before puberty.

*Clinical aspect.*—The symptoms that are associated with the presence of a tumour within the skull are conveniently divided into two groups.

The first group occurs irrespective of the situation of the tumour within the skull. They are called general symptoms, among which headache, vomiting, and optic neuritis are the most important. The second group consists of phenomena dependent upon the disturbance of particular areas of the brain by the tumour, and from the nature of the disturbance the situation of the tumour can be determined. These are termed the "localising signs." To these may be added a third group of signs which may be called the "false localising signs." They occur in the late stages of intra-cranial tumour, and resemble the true localising signs. They are the result of local lesions consequent upon increased intra-cranial pressure and disturbance of the cerebral circulation.

As a rule the general signs precede the localising signs—the advent of headache, vomiting, and optic neuritis is followed by the appearance of localising signs as the tumour increases in size, but it is most important to bear in mind that it is not uncommon for localising signs to appear weeks, months, or even years before the general signs. This is especially the case in tumours situated near the cortex cerebri, where local epilepsy, sensory or Jacksonian, often precedes the general signs.

*General symptoms.*—These are, headache, optic neuritis, vomiting, general convulsion, nystagmus, giddiness, mental alteration, hebétude, coma, and hydrocephalus. Some of these symptoms may, under certain circumstances, also be localising signs. For an example, mental alteration may suggest extensive involvement of the prefrontal lobes; giddiness, if intense, may suggest implication of the vermiform process of the cerebellum, while hydrocephalus in certain cases strongly points to a tumour in the neighbourhood of the Sylvian aqueduct or fourth ventricle.

The three chief general symptoms—headache, vomiting, and optic neuritis—are present in the large majority of cases; headache is least often absent. Vomiting is absent in a fair number of cases, while optic neuritis seems frequently to be absent under certain circumstances. Vomiting and optic neuritis are prone to be absent together, and it is curious to note that vomiting seldom, if ever, occurs in a case not presenting optic neuritis.

Headache is the most constant general symptom. Usually it is described as a dull, general pain, and is referred to the skull. Rarely severe in the early days of the disease, it may be intense when a tumour becomes large or cystic, or when hydrocephalus occurs. When extreme it is paroxysmal, and death has occurred in such a paroxysm, presumably from pain alone. Sometimes the pain is referred to a particular region of the skull, and when associated with local tenderness or deep pressure over the same region it may have some localising value. Otherwise, little reliance can be placed upon local headache as a regional symptom. Impli-

eration of the fifth cranial nerve may give rise to intense pain, darting over one side of the head, and tumours of the base of the skull give rise to pain which is more severe and constant than is usual in encephalic tumours, and which is not relieved to any great extent by analgesics of the coal-tar series. Headache is never such a severe symptom in children as it is in adults, presumably because of the lesser rigidity of the skull. When hydrocephalus begins to be apparent the headache becomes less severe, and not infrequently disappears.

Vomiting occurs suddenly and without effort and has no essential relation to the taking of food. As a rule, a single emesis occurs on each occasion—the child does not empty the stomach. It is generally spontaneous, but a movement of the head—such, for instance, as occurs when the child is turned in bed—may occasion it. It is very prone to occur during an exacerbation of the headache. It is generally stated that vomiting is more severe when the tumour is situated below the tentorium, yet it is rare in cases of glioma of the pons.

Optic neuritis is rarely absent, throughout the whole course of the disease, except when a growth is situated in the medulla or dorsal part of the pons and when a growth early destroys the oculo-motor nuclei. In the former case it may never be present; in the latter if present it subsides with the extinction of the oculo-motor nuclei. The absence of optic neuritis in adult cases of intra-cranial tumour where the growth is not situated in the pons is generally in causal association with advanced arterial degeneration, a condition which never exists in childhood, and this fact probably accounts for the almost unvarying presence of optic neuritis in intra-cranial tumour in children.

The neuritis is almost always bilateral, though it may appear at an earlier period, and may be more marked for a time upon one side. It occurs later where a tumour growing from the meninges or bone compresses the brain, earlier when the growth actually involves the nervous

tissue. Its occurrence is little influenced by the size of the tumour, for intense neuritis may occur when this is no larger than a walnut; but the intensity of the neuritic process as judged by the ophthalmoscopic appearance is a sure indication of the intensity of the disturbances occurring in the neighbourhood of the tumour, whether from rapid growth, acute oedema, or softening in the enveloping brain tissue or from the development of hydrocephalus.

The earliest change to be detected by the ophthalmoscope is hyperemia of the disc, with general fulness of the retinal vessels and tortuosity of the veins. Soon after the disc becomes a little raised and the vessels can be seen to dip over its edges, which become indistinct, first upon the nasal side of the retina, last upon the temporal side, and finally become lost. The centre of the disc is filled up by inflammatory exudation which hides the emerging vessels for some distance and the whole disc becomes raised above the general surface of the retina, like a mole-hill, and it may reach a height of as much as three millimetres. Flame-shaped hemorrhages of various sizes are frequently seen and white exudation appears which may be recognised as white lines along the vessels. There may also be a number of glistening white spots arranged either in a circular manner (asterisk) round the macula lutea or in the form of a fan with its centre towards the disc. In some cases of intense neuritis large irregular woolly-looking patches of white exudation occur in the neighbourhood of the disc. Optic neuritis when once developed may resolve under the influence of medicinal or operative measures, and very rarely it may disappear spontaneously. A neuritis with seven dioptries of swelling may clear up and leave perfect vision provided there be no appreciable amount of white exudation about the optic disc and along the vessels; but if this exudation be present in any quantity, optic atrophy and impairment of vision will almost certainly result, in proportion to the abundance of the exudation. When optic neuritis that has attained a considerable degree (4 dioptries of swelling) resolves, although perfect

vision is retained, the fundus still shows permanent traces of the neuritis—a woolly appearance at the edge of the disc, white lines along the vessels, and filling up of the physiological cup.

When resolution does not take place, the inflammatory exudation becomes organised, contracts, and cicatrises, strangling both nerve fibres and vessels as these pass into the lamina cribrosa, the result being post-neuritic optic atrophy. The period at which such atrophy begins to be apparent varies with each case, and it may be said that the more acute the neuritis and the more severe its degree the sooner does atrophy occur. It may be quite definite six weeks after the first appearance of the neuritis. In chronic optic neuritis of slow onset, on the other hand, it may be months or even years before atrophy is apparent.

The condition of vision at the most intense stage of the neuritis is often unimpaired, and this may give rise to an erroneous prognosis as to ultimate preservation of vision, for in such a case vision may be reduced to light perception only, within two months.

In the present state of our knowledge little can be said as to the way in which optic neuritis is produced. It has been held as due (1) to distension of the optic nerve sheath, with consequent pressure upon the retinal vessels; (2) to extension of "tissue irritation and inflammation" from the brain-tissue along the optic nerve; (3) to meningitis sometimes arising in the late stages of cerebral tumour. It is probable that some other as yet unknown factor is all-important in the production of optic neuritis.

It is interesting to note that optic neuritis and also vomiting are of common occurrence in tumours of the cervical spinal cord.

*General convulsion.*—A certain number of cases of cerebral tumour first came under observation as cases of idiopathic epilepsy, the only symptom present being general convulsion occurring at irregular intervals, and these may occur for months or even years before other signs indicative of intra-

cranial tumour arise. This happens only, so far as the writer's experience goes, when the tumour is situated above the tentorium, but the situation may be in any part of the brain above the tentorium. The explanation of the production of such general convulsions by tumours otherwise without symptoms is difficult.

Whatever the situation of an intra-cranial tumour may be if it is productive of marked general symptoms, general convulsions are likely to occur. In the *first* place the growth must of necessity interfere to some extent with the intra-cranial circulation, and temporary cerebral anaemia is a potent cause of convulsion. *Secondly*, disintegration of cerebral tissue frequently accompanies the growth of a cerebral tumour, and it is associated with the liberation of certain toxic substances (amongst which are neurin and cholin), and these are apt to produce convulsions if they come in contact with the motor elements of the cortex. *Lastly*, some degree of hydrocephalus occurs in almost every case of long standing, whatever be the situation of the growth. The dilatation of the ventricles occurs at the expense of the cerebral tissue, and there is always pressure upon cerebral substance. General convulsion depends upon a condition of general instability and irritability of the central nervous system as a whole, and the presence of an intra-cranial growth entails in every way circumstances favourable to the production of instability, for the brain is badly nourished and subjected to continuous increased pressure, and it is wasted and exposed to the action of self-generated poisons. Further, the variations of pressure that occur may be presumed to act as immediate excitant causes of the convulsion. As a rule such general convulsion is preceded by no aura, and consciousness is rapidly lost. A patient may die in such a convulsion. Closely allied to the general epileptiform attacks is sudden loss of consciousness, sometimes associated with slight general convulsion, and attacks of every gradation may be seen, from the severe general convulsion to sudden loss of consciousness unassociated

with convulsion. It may be here suggested that the not infrequent occurrence of sudden unexpected death in cases of long-standing intra-cranial tumour may be sometimes due to attacks of this nature.

*Giddiness* is of common occurrence and may be constant, or, as is more usual, it may be paroxysmal. It may be described as a sensation of bodily instability referred to the interior of the head, associated with a sinking feeling at the epigastrium, and accompanied always with some impairment of consciousness (mental confusion). It is very apt to occur on suddenly altering the position of the head, and especially upon assuming the upright position after the prone position has been maintained for some time. It must be distinguished from vertigo, which is a sensation of rotation, subjective or objective, and is often accompanied by forced movements of the eyes and of the body, and is a valuable localising symptom.

The principal factor in the production of giddiness is interference with the intra-cranial vascular supply. Small degrees of diplopia from involvement of the oculo-motor nerves, and of the auditory nerve, may also contribute to its production.

*Nystagmus* may result from a tumour in any situation. It is usually marked when the cerebellar system is involved, but its occurrence or absence observes no rule. It is almost invariably present when hydrocephalus has developed, and it is common in the late stages of all long-standing cases.

*Mental alteration.*—When the disadvantageous conditions under which the brain works in cases of tumour are considered, the comparatively trifling nature of the mental alteration which is obvious to the observer in cases of intra-cranial growth is surprising. The nature of the change is usually simple reduction, with slowness of cerebration, which may progress to hebetude and finally to coma. The patients are for the most good-tempered, placid, and content throughout. Mental perversity is rare, and mania when present indicates the occurrence of degeneration or softening in the neighbourhood of



the growth. The latter is probably toxic in origin and it is usually accompanied by pyrexia. It sometimes follows surgical procedure for the removal of extensive growth from the cerebral hemispheres.

*Hydrocephalus* may occur in association with an intracranial tumour in any position, but when occurring as an early sign and when severe in degree it is a localising sign and indicates that the lesion is probably along the course of the brain-stem and is obstructing the Sylvian aqueduct or the fourth ventricle.

#### LOCALISING SIGNS.

*The regional diagnosis of cerebral disease.*—In order to avoid repetition the regional diagnosis of brain disease in general is dealt with, and certain anatomical details chiefly relating to the blood supply of the various regions of the brain are stated. The latter, while more strictly pertaining to the subject of cerebral vascular lesions, are of considerable importance in connection with intra-cranial growth; for vascular obstruction and consequent softening of the area supplied not infrequently occurs from the local pressure which the growth exerts, and focal symptoms of vascular distribution may arise.

The more important symptoms arising from disease of each region of the brain are first described. Subsequently each symptom and the more commonly occurring association of symptoms are discussed with reference to their intrinsic meaning and value in localisation.

*The cerebral hemispheres.*—From a greater part of the area of the cerebral cortex of each hemisphere long axis cylinder processes belonging to large cells stream through the corona radiata and internal capsule or corpus callosum and connect the cerebral hemisphere with more or less distant parts of the nervous system. These proceed to the opposite cerebral hemisphere, to the retina and olfactory mucosa, to the basal ganglia, mesencephalon, pons Varolii, medulla, and to each segment of the spinal cord. They

constitute what is known as the cortical projection system and are called "extrinsic" fibres.

From the whole of the cerebral cortex, however, the shorter axis cylinder processes of smaller cells proceed through the white matter to other regions of the same hemisphere. These lie comparatively closely to the grey matter and are known as the sub-sulcine projection or association system and are called "intrinsic" fibres. The cortical projection system includes fibres conducting towards the cortex (corticopetal fibres) and also fibres conducting away from the cortex (corticofugal fibres) which are motor in function. As examples: the mesial aspect of the occipital lobe receives corticopetal fibres only, which conduct impressions from the retina and subserve vision. Similarly the superior temporal convolution and uncinate gyms receive corticopetal fibres subserving hearing and taste and smell respectively.

The marginal gyms is in like manner connected with impressions of general sensibility, while the central convolutions (motor area) both receive and send fibres subserving muscle sensation and motion respectively.

While those convolutions contributing to the cortical projection system of extrinsic fibres are known definitely to be connected with certain functions (special senses, common sensation and motion), our knowledge as regards the function of those areas of the cortex whose cells furnish fibres of the association system only, is much more limited.

There are four such areas: (1) The prefrontal lobe; (2) most of the temporal lobe (the superior temporal convolution and the uncinate gyms excepted); (3) the inferior parietal lobule; and (4) the whole of the external surface of the occipital lobe. The areas were described by Flechsig, who named them the "sensory annexes," indicating that they were higher sensory centres adjoined to the primary sensory centres. As an example, the angular gyms is a sensory annex of the vision centre of the cuneus. Destruction of the angular gyms upon the left side produces

no loss of vision, but loss of a most important psychical element depending upon vision, for loss of recognition of words seen—word-blindness—occurs.

It is probable that these four areas are concerned with the higher psychical processes, and that they are of similar value, so that where peculiar mental symptoms arising early in a case of cerebral tumour, suggest the implication of an area concerned with psychical function, the indication is not that the lesion is situated necessarily in the prefrontal lobe, for the most marked mental symptoms may exist when it is situated in the temporal, parietal, or occipital lobes.

#### FRONTAL LOBE.

The frontal lobe constitutes the anterior pole of the hemisphere, and presents a mesial, an orbital, and an external surface. It is limited behind by the anterior perforated space, the commencement of the fissure of Sylvius, and the ascending frontal sulcus. Posteriorly, it abuts upon the central convolutions. Beneath its orbital surface lies the olfactory tract, and postero-mesially it is in contact with the optic nerve.

*Vascular supply.*—The whole mesial surface and most of the orbital surface and the upper two convolutions of the external surface are supplied by branches of the anterior cerebral artery. The remainder of the lobe, comprising the third frontal convolution, the posterior inferior extremity of the second frontal convolution, and the external orbital convolution, is supplied by a branch of the middle cerebral artery.

*Connections.*—A large projection system of fibres descends through the corona radiata and anterior limb of the internal capsule to end in the nuclei of the mesencephalon, pons Varolii, and medulla (motor fibres for movements of head and eyes and for speech). A large system of fibres ascends from the optic thalamus to the frontal lobe.

*Functions.*—The anterior two thirds of the lobe are believed to be concerned with psychical functions. In

the posterior third are centres for the movement of the head and eyes. The posterior ends of the second and third frontal convolution upon the left side have well-known and most important functions. The former is the centre for the execution of written speech, the latter the centre for the execution of articulate speech. (In left-handed persons these centres are situated upon the right side of the brain.)

*Symptoms.*—Lesions of the frontal lobe not infrequently exist without obvious local symptoms, the possible explanations being (1) the impossibility of detecting minor mental alterations; (2) the relative rapidity with which compensation seems to occur. Mental alteration is commonly present and may exist in any degree, from the slightest noticeable alteration in manner or habit to pronounced megalomania, and to complete loss of memory and imbecility. Severe degrees of mental alteration are met with in connection with bilateral lesions of the frontal lobe.

Signs indicative of implication of the centre for movement of the head and eyes are rarely present, even as temporary symptoms.

A lesion of the left second frontal convolution may produce inability to write, and a lesion of the left third frontal gyrus will produce motor aphasia in children nearing the age of puberty, who are right-handed.

Epileptiform attacks are common. Of these a sudden impairment of consciousness associated with automatism is characteristic, but similar attacks are met with in lesions of the temporal and occipital lobe. Dr. Hughlings Jackson lays stress upon the occurrence in some cases of frontal lesion of local convulsions, commencing bilaterally, which, as a rule, do not become general and are not associated with complete loss of consciousness. General convulsions are of common occurrence.

The other local signs of importance produced by lesions of the frontal lobe are those produced by pressure upon neighbouring structures. A tumour, for example, commencing in the frontal lobe as it enlarges will press backwards upon or extend into the anterior central

convolutions, and brachial, facial, or crural monoplegia, or a hemiplegia of gradual and late onset, results.

Tumours extending to the orbital surface often involve the olfactory tracts, unilateral anosmia resulting. If the tumours be situated mesially and involve both olfactory tracts, bilateral anosmia occurs. The optic nerve may be pressed upon just before it enters the optic foramen, and unioocular hemianopsia rapidly extending to complete blindness may occur. Similarly unioocular paralysis may result from pressure upon the nerves entering the sphenoidal fissure, and sometimes proptosis appears.

### THE CENTRAL CONVOLUTIONS

The Rolandic or motor region comprises those convolutions which bound the fissure of Rolando, namely the ascending frontal, and the ascending parietal convolutions, the base of the superior frontal and the superior parietal lobule, together with the corresponding portion of the marginal convolution which forms the mesial surface of the hemispheres.

*Vascular supply.*—The motor region receives its blood supply upon the convexity of the hemisphere from branches of the middle cerebral, but upon the mesial aspect of the hemisphere from the anterior cerebral vessels.

*Connections.*—The corticifugal tract is the great pyramidal system which descends throughout the nervous system, forming connections with each segment. The majority of the pyramidal fibres supply the lower centres, and through these the muscles upon the opposite side of the body, but many fibres passing down upon the same side convey incitations to muscles which are habitually used bilaterally or contralaterally. As examples of bilaterally associated muscles those of the tongue, lips, and larynx, and the muscles of the trunk may be instanced, and as contralateral muscles the sterno-mastoid of one side and the splenius capitis of the other act together in the movement of turning the head to one side; further,

the erector spinæ of one side habitually acts with the muscles of the inferior extremity of the other side in walking, and as the result of destructive lesions upon one side of the cortical areas subserving these acts, descending degenerations are found upon both sides in the lateral pyramidal tracts. It follows, then, that such muscles receive a supply from both cerebral hemispheres, and clinically they are never found to be permanently paralysed as the result of a unilateral lesion, however extensive. A large corticopetal system ascends to the central convolutions from the optic thalamus, and conveys sensations coming from muscles, and there is great probability that impressions of common sensation also reach the cortex of the central region by the same path.

In the central convolutions the centres for bodily movement are located and the muscle sense impressions are received together with other impressions pertaining to common sensation. The motor centres are situated in the so-called motor area, but the localisation is not strictly limited, the area of one class of movement merging into the area of another class. Stimulation of the cortex where two areas meet, produces, as a rule, movements pertaining to both areas.

*Symptoms.*—Destructive lesions of the central convolutions produce paralysis of voluntary movement in the corresponding part of the body. Inasmuch as a movement is carried out by the muscles of one side of the body only—the movements of the limbs, for example—the paralysis is complete or not in proportion to the degree to which the centre for such movement is put out of action, and monoplegia or hemiplegia results according to the extent of the lesion. On the other hand, if a movement involve muscles situated upon both sides of the body, there results general weakening of the whole movement, but no paralysis. As examples, the movements of the lips, tongue, and larynx, and of the trunk are not paralysed by a lesion limited to one cerebral hemisphere. Symmetrically situated lesions of both Rolandic areas, however, of necessity entail para-

lysis of the bilaterally associated muscles. The bilateral weakness which a lesion of the central convolutions upon one side entails is not a valuable clinical sign on account of the difficulty with which signs of such weakness can be elicited; for there is no normal with which the strength of a bilateral movement can be compared. Accompanying such paralysis as the earliest indications, or in some cases existing alone as the only marked indication of an incipient or slight lesion, are alterations of the deep reflexes—increase of myotatic irritability and clonus and alteration of the superficial reflexes, the most important being the change in the plantar reflex from the flexor to the extensor type. The plantar extensor response is almost invariably present when a lesion of the central convolutions exists, whatever be its seat. Sensory paralysis of common order often follows sudden lesions of the Rolandic area, but marked degrees do not persist more than a few days unless the lesion be extensive. If anaesthesia persists it amounts to not more than slight tactual impairment. Its distribution is that of a monaesthesia or hemianaesthesia, the latter being sharply limited by the middle line of the body.

Another defect of sensibility which occurs is inability to localise touch. This phenomenon is most marked upon the limbs and is more striking upon the distal part of the affected limb than upon its proximal part.

Irritative phenomena are of common occurrence. Conjugate deviation of the head and eyes to the opposite side is a constant manifestation of a rapidly developed lesion. It is shortly followed in most cases by paralytic conjugate deviation, towards the same side, which soon disappears. Where a lesion develops gradually, conjugate deviation is a very rare phenomenon.

Localised epilepsy, either sensory or motor or a sequence of the two, is often the earliest sign of a small lesion of the motor region. In a majority of cases the motor manifestation exists alone, but in some cases it is always preceded by a sensation. A sensory epilepsy from a central lesion,

however, though it may exist for some time without spasm, is always ultimately accompanied by convulsion.

The Jacksonian convulsion may be entirely local. More often it begins locally and spreads, involving more or less of the same side of the body, sometimes extending to the opposite side of the body and sometimes ending in general convulsion. The march of the spasm is determined by the position of the centres for the various regions in the Rolandic cortex. Bearing in mind that the centre for movements of the face is completely separated from the centre for movements of the leg by the area representing movements of the upper extremity, it will be obvious that the march of a Jacksonian attack commencing in the face will be first to the upper extremity and finally to the lower extremity. Similarly when the convulsion commences in the foot the hand is affected before the face. Where the point of commencement is in the upper extremity, usually the face is affected before the leg; less commonly the reverse occurs. Where the convulsion spreads to the arm the march may be either from hand to shoulder or from shoulder to hand.

When Jacksonian convulsions involve the whole side of the body the trunk muscles are usually affected to some extent on both sides, but in some cases the spasm is strictly limited to the trunk muscles of the same side.

The sensory fit consists in most cases of a tingling sensation referred to the skin, and it generally produces considerable emotional disturbance—the child cries with fright. The march is similar to that of the motor disturbance, but less extensive, rarely involving the half body, and never spreading to the opposite side.

Local epilepsy is often followed by temporary exhaustion of the discharging areas, which is most marked and persists longest in that region in which the discharge began.

When the lesion is progressive, as in cases of tumour, the post-convulsive paralysis remains for a progressively longer period after repeated attacks and finally becomes persistent



and increasing. It is the expression of the transition from the irritative to the destructive lesion.

Of great interest both as regards irritative and destructive phenomena are tumours in the neighbourhood of the paracentral lobe and especially tumours of the falx cerebri. In such cases, since the paracentral lobes are contiguous, double crural monoplegia may result and closely simulate a paraplegia of spinal origin. Further, local epilepsy may occur, now in one foot, now in the other, and in a case of endothelioma of the falx a simultaneous mono-spasm of both feet occurred on several occasions.

### GYRUS FORNICATUS

The limbic or falciform lobe is situated upon the mesial aspect of the hemisphere, and lies between the corpus callosum and the marginal convolution, joining posteriorly with the hippocampal lobe. It receives its blood supply from branches of the anterior cerebral and posterior cerebral arteries.

Its chief connections are with the optic thalamus by a large tract of corticopetal fibres, which are probably the physiological continuants of the system of the fillet or chief sensory conducting path. It is also connected with the Rolandic area by fibres of the cingulum. There is important evidence indicating that the gyrus fornicatus serves as the chief centre for the registration of common sensibility. There is no evidence as yet forthcoming as regards the regional representation of the parts of the body in this lobe.

The gyrus fornicatus seems to be particularly free from the influence of morbid conditions. Its vascular supply from two sources renders it an unfavourable situation for the occurrence of softening. In the few recorded cases of tumour of this region marked sensory impairment upon the opposite side of the body with slight hemiparesis has been present.

## THE INFERIOR PARIETAL LOBULE

This region comprises the supra-marginal and angular gyri. Its vascular supply is from the longest posterior branches of the middle cerebral artery, which inosculate to some extent in this region with branches of the posterior cerebral artery. It receives a large system of corticopetal fibres from the optic thalamus, and is connected with the corresponding region of the opposite hemisphere by fibres of the corpus callosum.

The angular gyrus of the left side contains the usual word-centre or centre for the registration of the symbols of written language. It is probable, also, that the region is the seat also of the higher visual centre. The only important localising sign is the occurrence of word-blindness—inability to appreciate written language in lesions of the left angular gyrus. Epileptiform convulsions preceded by a visual aura may also occur. Experimental evidence and a few recorded cases make it highly probable that the visual defect known as crossed amblyopia may result from a lesion in this region. Crossed amblyopia consists in a general impairment of visual acuity and a general contraction of the visual fields of both eyes, both phenomena being much more marked in the eye of the same side as the lesion. It is probable, as Hughlings Jackson and Gowers believe, that the angular gyrus is a visual centre of the highest level.

## THE OCCIPITAL LOBE

This occupies the posterior pole of the hemisphere and includes the cuneus and the lingual lobe upon the mesial and ventro-mesial aspects. The vascular supply is from the posterior cerebral artery. Corticifugal tracts from this region pass to the pulvinar of the thalamus and to the superior colliculi. The corticopetal system consists of the optic radiation prolonged from the optic tracts and terminating in the cuneus lingualis and external occipital gyri. The lobes of the two hemispheres are connected by many fibres of the corpus callosum. The half-vision centre is

situated in the cuneus and posterior end of the lingual lobe, the lingualis corresponding with the upper quadrant of the visual field and the cuneus with the lower quadrant.

*Symptoms.*—Homonymous hemianopia is the most important symptom of lesions of the occipital lobe, but it occurs only when the mesial aspect or the sub-lying white matter is involved. Quadrantic hemianopia results when the lesion is confined to the cuneus (lower field quadrants lost) and when the lingualis is alone affected (upper field quadrants lost). Hemiachromatopsia has resulted from less extensive lesions of this region.

Sensory epilepsy, consisting of visual hallucinations or visual aura preceding general convulsions, is not rarely met with.

Simple lapses of consciousness associated with automatism are sometimes met with when the convexity is affected.

#### TEMPORAL LOBE

This region lies between the Sylvian fissure on the convexity of the hemisphere and the crus cerebri.

The vascular supply is from two sources; branches of the middle cerebral artery supply the first and second temporal convolutions upon the convexity of the hemisphere, while the posterior cerebral artery supplies the whole of the inferior and mesial surfaces.

*Connections.*—The corticifugal tract passes from the first and second temporal convolutions to the upper part of the pons Varolii. One set of corticopetal fibres passes from the thalamus to this lobe, while another set, arising in the inferior colliculus, ends in the first temporal gyrus. The fibres of the anterior commissure end in the temporal lobe on either side, and each olfactory tract is continued into the uncinate gyrus of either side.

*Symptoms.*—Lesions of the temporal lobe are sometimes accompanied by mental alterations without other localising signs. This is especially the case if the lesion is situated in the anterior part of the lobe. Attacks consisting of sudden impairment of consciousness and the performance of strange

acts, such as have been described in lesions of the prefrontal and occipital lobes, also occur.

Bilateral lesions of the superior temporal gyrus produce complete deafness to sound, but unilateral lesions are not accompanied by any marked impairment of the acuity of hearing; the explanation being that there is in all probability a semi-decussation of auditory afferent fibres in the corpus trapezoides, each superior temporal gyrus being thus in connection with both ears. Extensive lesion of the first and second temporal gyrus of the left side in their posterior two thirds produces "word-deafness," or inability to comprehend words spoken. On account of the action of the auditory word centre as the all-important centre for the origination of speech, word-deafness usually entails also inability to speak and inability to comprehend written language. Bilateral lesions of the upper two temporal gyri produce total loss of language, both receptive and expressive, and the patient's understanding is reduced to the comprehension of gesture (mimetic language) and he can express his thought only by gesture (mimetic expression). Such a condition is necessarily accompanied by great mental impairment.

Partial lesion of these convolutions upon the left side produces varying degrees of word-deafness, paraphasia and paragrammia. Irritative lesions in this region may produce auditory hallucination, and then such phenomena may occur as the warnings of a general convulsion.

Damage in the region of the uncinate convolution may be associated with olfactory and gustatory hallucination or by epileptiform attacks preceded by warnings of a similar nature. In a case of tumour of the uncinate gyrus, the patient was haunted by a persistent disagreeable odour. Loss of smell and taste do not occur, and in explanation stands the anatomical fact that the olfactory tract is connected with both uncinate gyri. The paths, however, of gustatory impressions are not known.

In a considerable number of cases of large tumour of the temporal lobe, loss of the knee-jerk has been observed upon the opposite side of the body, but in many of these cases

the lenticular nucleus was also involved. Tumours of the temporal lobe in the process of growth generally extend towards the optic thalamus, and by pressure on or invasion of the internal capsule produce hemiplegia as a late result.

### INTERNAL CAPSULE

This column of white fibres lying between the lenticular nucleus externally and the caudate nucleus and optic thalamus internally, is the principal path of fibres entering and leaving the cerebrum. The anterior limb consists of the corticopetal and corticifugal fibres of the prefrontal lobe behind which lie the fibres subserving the conjugate movements of the head and eyes. At the knee of the capsule lie the fibres for the movements of the face and tongue—geniculate fascicle. In the posterior limb are situated the pyramidal fibres for the movements of face, upper extremity, trunk, and lower extremity, behind which are placed the tract of common sensation, the auditory radiation and the optic radiation in that order from before backwards.

*Vascular supply.*—The capsule is supplied by three arteries noted for the frequency with which cerebral hæmorrhage is due to their rupture. They are often known as the arteries of cerebral hæmorrhage. The anterior of these is the lenticulo-striate artery, supplying the anterior limb of the capsule and the caudate nucleus. The lenticulo-optic artery supplies the posterior limb, while the external optic artery supplies the optic radiation. These arteries are branches of the middle cerebral artery; from their origin they pass outwards guided by the crus cerebri to the lenticular nucleus, then bending inwards, they take a direct course across the internal capsule to their destinations in the caudate nucleus, optic thalamus and pulvinar thalami respectively. The usual position in which rupture of these vessels takes place is before they enter the capsule, while they are lying in the lenticular nucleus.

*Symptoms.*—The characteristic symptoms produced by lesion of the internal capsule are hemiplegia, hemianæsthesia

(never absolute) and hemianopia, in such combination as the extent and position of the lesion and the anatomical arrangement of the fibres in the capsule determine. Aphasia never results from damage in this region, though some dysarthria may be present when the geniculate fascicle is involved. Irritative symptoms (convulsions, etc.) do not occur as primary symptoms.

#### BASAL GANGLIA

*(Caudate Nucleus, Lenticular Nucleus, Optic Thalamus)*

When localising symptoms of lesions in these regions occur they are the result of secondary involvement of the internal capsule. It has been stated that choreiform movement and intention tremor result, in some cases, from damage to these structures, and the statement may be correct, although such phenomena are in most cases probably the result of multiple small lesions of the cortex in the motor region.

#### MESENCEPHALON

This region includes the quadrigeminal bodies, the nuclei of the third and fourth nerves, the superior cerebellar peduncles, the upward prolongation of the fillet or chief sensory tract and the crura cerebri, with the origin of the third pair of nerves. Close to the crura cerebri are the optic tracts.

Lesions in this situation are often productive of characteristic symptoms. The association of unilateral, or more often bilateral, ataxy with irregular ocular palsy from simultaneous involvement of the superior peduncles and the oculo-motor nuclei is the rule in lesions of the dorsal part of the mesencephalon, and this syndrome is often called "Nothnagel's sign," after the physician who first drew attention to it. Pathognomonic as a local sign of lesion in this region is paralysis of the upward movement of the eyes or of the downward movement, or of both upward or downward movements, the lateral movements being intact. It is only in disease of the anterior part of the corpora quadri-

gemina that the Argyll Robertson pupil phenomenon occurs as the result of a gross lesion. Optic neuritis tends to be absent, or, having been present, it disappears as the third nerve nuclei are, early or later, involved.

Extension of the lesion or pressure may cause hemianopia or hemiplegia, since the optic radiations and the pyramidal tracts are in close proximity. A lesion in the dorsal part of the mid-brain beneath the posterior colliculus and limited to one side is the only lesion capable of producing complete hemianæsthesia to all forms of stimulation, since it is only in this region that the sensory tracts are collected within a comparatively small area. Localisation is often easy when the basal portion of the mesencephalon is involved, for here the crus and the third nerve are likely to be involved together, and the classic syndrome is hemiplegia of the opposite side with third-nerve paralysis upon the same side. Extension of growth across the interpedal space may involve the other third nerve, and the other crus, giving rise to hemiplegia, or double hemiplegia, with crossed unilateral or bilateral third-nerve palsy. From pressure upon the neighbouring optic tract hemianopia with Wernicke's pupil phenomenon may occur in dorsally situated tumours. Hydrocephalus results early in most cases from obliteration of the Sylvian aqueduct.

#### POSS VAROLII

The chief anatomical structures upon the involvement of which localisation depends are the nuclei of the sixth nerve, of the facial nerve, and the motor nucleus and sensory trunk of the fifth nerve, the middle peduncle of the cerebellum and the pyramidal tract.

The most characteristic symptom of pontine disease presents when the nucleus of the sixth nerve is involved. Each sixth nucleus subserves the conjugate movement of both eyes to its own side, and injury of one sixth nucleus produces loss of lateral conjugate movement to the same side, a form of paralysis that does not result except from a lesion

in this situation. The facial nucleus and the fifth nucleus and sensory trunk are often involved and hemiplegia often appears from involvement of the pyramidal tracts, so that the very striking association of hemiplegia with crossed facial anaesthesia, atrophic palsy of the face or of the muscles of mastication may occur; frequently both the latter conditions are present in pontine crossed hemiplegia. Ataxy is not rare, and sometimes intention tremor may occur.



FIG. 19.—Case of pontine tumour, showing paralysis of left sixth nerve and protrusion of tongue to right.

When the lesion involves the pyramidal path above the pontine nuclei, inclusive of the fibres going to the nuclei, a spastic paralysis of the face, masticatory muscles, tongue, and palate may co-exist with hemiplegia either upon the same or upon the opposite side as the lesion involves the pyramidal fibres for these nuclei before or after they have crossed the middle line.

Optic neuritis is frequently absent if a tumour is situated on the dorsal region of the pons and close to the middle line. Tumour of the ventral part of the pons, on the other hand,



usually presents intense neuritis. When optic neuritis is absent headache and vomiting are never obtrusive symptoms.

Hemianæsthesia may result, but is never absolute. Paræsthesia also occurs. Pressure and the traction which the enlargement of the pons may put upon the auditory nerve-roots may give rise to deafness.

#### MEDULLA OBLONGATA

The symptoms of lesions in this situation are unilateral



FIG. 20.—Case of pontine disease, showing paralysis of left sixth nerve and protrusion of tongue to right; there was hemiplegia affecting the right arm and leg.

or bilateral atrophic paralysis of the lips, tongue, palate, pharynx, and larynx, usually associated with unilateral or bilateral hemiplegia (the face being uninvolved). The lips are paralysed and wasted when the nuclei of the hypoglossal nerve is destroyed, because the nerve-fibres supplying the lips, though they rise in the facial trunk, yet have their origin in the hypoglossal nuclei, and join the loop of the facial trunk before it emerges from the medulla.

Involvement of the ascending root of the fifth nerve may cause anæsthesia of the face over the area corresponding

with the distribution of the third division, or third and second divisions, of the fifth nerve, according as the lesion is situated near the cephalic or near the caudal end of the medulla.

Hemiataxy upon the same side results if the inferior peduncle of the cerebellum be affected.

Lesions of the medulla come under observation somewhat rarely, one reason being that they tend to be rapidly fatal from involvement of the respiratory and vaso-motor centres, another that, so far as the incidence of neoplasms is concerned, the medulla resembles the spinal cord in being rarely the seat of tumour, rather than the rest of the central nervous system in which tumours occur commonly. Growth not infrequently occurs, however, alongside the medulla, having its origin in the meninges or bone. In this case the nerve-roots issuing from the bulb are often implicated, together with the spinal accessory, and unilateral bulbar paralysis is associated with paralysis of the muscles supplied by the spinal accessory, the sterno-mastoid, and the trapezius. The facial and auditory nerves are also often involved.

Other signs of note are the presence of head retraction and the occurrence of hydrocephalus.

#### CEREBELLUM

This organ, consisting of two lateral lobes united by a middle lobe, is connected with the brain-stem by means of three peduncles. The superior peduncle passes upwards and, decussating with its fellow beneath the aqueduct of Sylvius, ends in the red nucleus of the opposite side, and serves thus to connect each lateral lobe of the cerebellum with the opposite cerebral hemisphere. The fibres of the middle peduncle cross to the opposite side of the pons, ending among the *nuclei pontis*. The inferior peduncle or restiform body contains the cerebellar prolongation of the dorsal columns of the spinal cord which conduct impressions from the spinal cord to the same side of the cerebellum (chiefly the middle lobe). There is also a large efferent

tract which, interrupted in Decker's nucleus, is continued down the antero-lateral region of the cord, placing the cerebellum in uncrossed connection with the latter.

*Symptoms.*—The experimental researches of Ferrier, Risien Russell, Luciani, and others, have afforded us such knowledge as to make cerebellar localisation comparatively simple if careful attention be paid to certain details.

The symptoms of chronic cerebellar lesions are, for the most, dynamic symptoms—that is, they appear when the muscles are put into their natural actions. Therefore, the physical and mental state of the patient must be moderately good if they are to be clearly demonstrated. Further, the uncertainty of movement which accompanies blindness may confuse the observer.

There are three fundamental points that must be borne in mind.

(1) Compensation takes place readily and quickly in the cerebellum, and localising symptoms arising from a tumour may be now present, now absent, at one time marked, at another time trifling, but tending, as time goes on, to become constantly present and more marked. Repeated observations are therefore essential before it is determined that there are no cerebellar signs present.

(2) Cerebellar attitudes must not be mistaken for compensatory attitudes (*vide infra*). Where bilateral ataxy exists, as is not unfrequent, this point is of especial importance.

(3) The scalp over the cerebellar region must be shaved and careful examination made, for in the majority of instances of cerebellar tumour occurring in children the cerebellar region is bulged upon the side of the tumour. This bulging is often more palpable to the flat of the hand passed over the surface than it is visible. It goes without saying that importance must not be attached to this phenomenon as a negative sign, nor alone as a positive sign when the skull is otherwise very asymmetrical.

*Symptoms—lateral lobe of the cerebellum.*—Hemiataxy upon the side of the lesion is a frequent symptom, and is

more marked in the upper than in the lower extremity. There is an irregular uncertainty and unsteadiness in the performance of fine movements. To elicit these defects of movement, the child is asked (1) to touch with his index finger the tip of the observer's finger held in such a position that he has to raise the arm from the shoulder; (2) to pick up a pin from a hard, flat surface; (3) to put his finger through a ring held at such a distance that he has to stretch to accomplish the act; (4) to put the peg into the holes of a cribbage-board, one after the other. By these means the

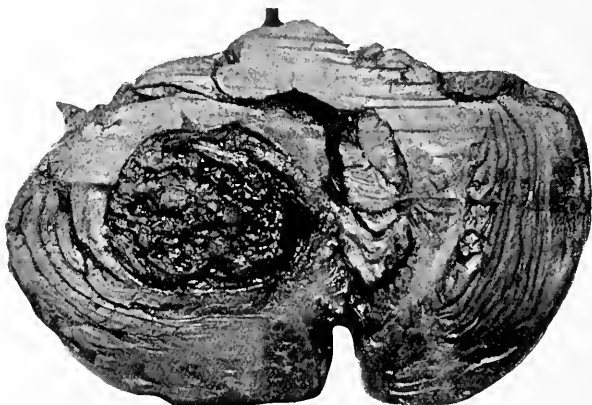


FIG. 21.—Tumour of lateral lobe of cerebellum; at first there was slight ataxy (which disappeared), headache, vomiting, optic neuritis, rapidly causing blindness, and loss of knee-jerks.

characteristic ataxy is easily elicited in the upper extremity; the lower extremity is best tested by asking the child (1) to touch the observer's finger with his toe; (2) to put his heel upon his other knee.

Slight relative weakness is observable upon the hemiataxic side in proportion to the degree of unsteadiness present. The knee-jerk is often peculiar. It is usually increased upon the side of the lesion, but in many cases the knee-jerks may at one time be elicited with the greatest ease, while at another time they may be so difficult to obtain, even with reinforcement, that skilled observers have pronounced them absent. Such diminution and exaggeration may occur

several times upon the same day; it is usually most pronounced upon the side of the lesion.

Foot clonus never occurs.

The plantar reflexes are invariably flexor in uncomplicated cerebellar disease.

Most reliable as signs of lesion of the lateral lobe of the cerebellum are (1) the position in which the head is held, (2) the attitude assumed when the patient is standing, and (3) the manner in which he walks.

*The cerebellar position of the head.*—Though far from a constantly occurring or constantly present symptom, it may be noticed at one time or another in two thirds of all cases of tumour of the lateral lobe of the cerebellum in children. As pointed out by Batten, the head is inclined to the shoulder of the same side as the lesion, and the face is turned slightly to the opposite side. When the patient is sitting up this attitude is always assumed as the rest position. It is most striking when observed from some distance.

*Attitude when standing.*—The body is bent slightly towards the side of the lesion. The patient often lunges or even falls to the side of the lesion, and this is especially liable to happen if, when the patient is standing, his attention and vision are suddenly directed away from his condition of equilibrium.

It cannot be too emphatically stated that if the physical condition is such that the child can only make a feeble attempt to stand and walk with support, or if the examination is made when severe headache or vertigo is distressing him, no reliance whatever can be placed upon the position in which he stands or upon the direction in which he falls.

*Gait.*—The body is bent laterally to the side of the lesion; the lower extremity of that side is used farther outside the vertical median plane of the body, and it is used more slowly than is the lower limb of the sound side. In progression the patient may lunge to the side of the lesion, and when attempting to walk straight and quickly he tends to walk in a curve, the concavity of which is towards the side

of the lesion. Under certain conditions phenomena almost the exact reverse of the above occur; they have been referred to above as compensatory attitudes. They are seen generally when the patient is better, while the true cerebellar gait is seen when he is not so well. The explanation is simple. The patient is desirous of demonstrating how well he can walk, and he is conscious of his hemiataxy and of the half of the body that is apt to play him false. Consequently, he throws the weight of the body over toward the sound side, bending the trunk to that side in so doing. The ataxy present is unfavorable to his balance, and he usually overdoes the correction and tends to lunge to the sound side. From the fact that the body is laterally curved in the attempt to walk straight he tends to walk in a curve towards the sound side. The distinction between the true and the compensatory attitude in walking soon becomes apparent upon repeated examination, for the compensatory attitude is one of effort.

Lesions of the anterior part of the cerebellum usually involve the forward region of both lateral lobes. Bilateral ataxy is present and the patient walks with the body bent forwards, the arms being held forward to save himself should he fall—an event which usually occurs after he has chased his centre of gravity a few steps.

Lesions of the posterior part of the middle lobe of the cerebellum are generally associated with a constant tendency to fall backward, and sometimes head retraction and opisthotonos are present; the ataxy here resulting is bilateral.

Nystagmus is usually marked, and in lesions of the lateral lobe of the cerebellum the slow movements occur towards the side of the lesion.

Pressure upon the cranial nerves in the posterior fossa of the skull may produce cranial nerve palsies, the seventh and eighth nerves being those most frequently involved.

Irritative phenomena are usually very prominent symptoms in vascular lesions of the cerebellum, but in tumour and abscess they are usually absent. Forced movements of

rotation are common in acute lesions of the lateral lobes. The patient rotates round his lesion. The most common variety is seen when the position of the patient in bed is changed. A patient presenting such forced movements usually lies prone in bed with his face turned away from the side of the lesion. If he is placed supine, he at once complains of intense vertigo and often vomits, and then, by a series of somewhat shock-like jerky movements, he rolls over the lesion until the original position is reached. If such a patient be placed in the sitting or standing position, he dives towards the side of the lesion directly support is taken away; he does not fall passively.

In connection with lesions of the middle lobe, Dr. Hughlings Jackson has described attacks of tetanus-like spasm associated with opisthotonos (cerebellar fits).

#### THE RELATIVE IMPORTANCE OF THE INDIVIDUAL LOCALISING SIGNS.

*Mental alteration.*—In children this is often of value as a local indication. Any lesion of the cerebral hemisphere occurring before the age of eight years renders a child "backward" in speech and education, in proportion to the extent of the damage to the brain. Such backwardness is extremely marked in bilateral lesions of the frontal lobe of an atrophic nature (see Cerebral Diplegia).

Marked perversity sometimes occurs in local lesions of the frontal temporal and occipital lobes, and mental symptoms, similar to those seen in the juvenile forms of general paralysis, have been present in some cases of tumour of the frontal lobe.

Slowness of cerebration and lethargy are indications of the effects of prolonged increase of the intra-cranial pressure.

Delirium and mania occurring in intra-cranial tumour are usually the results of some secondary process, such as softening round the tumour, meningitis, or hydrocephalus, with extreme intra-cranial tension.

*Speech defects.*—Aphasia, word-blindness and word-deafness are positive indications of lesions involving the third frontal convolution, angular gyrus, and the two upper temporal convolutions of the left hemisphere respectively.

The absence of these signs, however, is no indication that the corresponding regions are free from involvement.

Articulatory defects are practically always due to a lesion of the brain-stem which either interrupts the pyramidal fibres for speech (lesions of mesencephalon and pons), or damages the hypoglossal nucleus and nucleus ambiguus in the medulla. In rare cases such defects may result from involvement of the hypoglossal vagus and bulbar accessory roots.

*Fits.*—The occurrence of minor attacks, especially lapses of consciousness followed by odd movements, when associated with general signs of intra-cranial tumour are of great importance and indicate that the tumour is situated either in the prefrontal anterior part of the temporal or external part of the occipital lobes, the probability being in favour of a situation in one of the two former rather than in the latter.

Sensory hallucinations are often pathognomonic of the situation of the lesion. Those of vision, such as bright flashes of light, stars, balls of fire, etc., point to the occipital lobe and they are often followed by temporary hemianopia, partial or complete. Hallucination of hearing indicates the temporal lobe (posterior part), provided there is no sign of implication of the auditory nerves or of the labyrinth. Hallucinations of taste and smell are evidences that the region of the uncinate gyrus is involved. Such hallucinations may be associated with slight impairment of consciousness, or with loss of consciousness, with or without general convulsion.

Localised sensory fits may result from a lesion of the gyrus fornicatus or of the central region of the opposite side. If they are followed at a later date by Jacksonian attacks commencing in the same region, or if they are associated with monoplegia, the gyrus fornicatus may be excluded as the seat of disease.

Localised convulsion (Jacksonian or organic epilepsy) indicates a lesion in that part of the central convolutions



corresponding with the part of the body in which the spasm commences. The indication is of great value when it occurs early in the course of intra-cranial tumour, general signs of the latter being present, and especially when the monospasm is followed by temporary monoplegia.

Jacksonian attacks, however, when unaccompanied by general signs of intra-cranial growth, may be simple signs of idiopathic epilepsy. When they appear for the first time late in the course of an intra-cranial growth, they are of no localising value, but are local fulminations resulting from the malnutrition and lessened stability of the cortical grey matter which chronic increased intra-cranial pressure and especially hydrocephalus produce.

Bilateral local convulsion commencing simultaneously in symmetrical parts of the body is indicative of a lesion of the frontal lobe.

Rarely, bilateral Jacksonian attacks, commencing in the lower extremities, have been the result of a tumour of the falx cerebri involving the paracentral lobules of both sides, but in such cases the attacks are usually unilateral, occurring now on one side and now on the other.

*General convulsions.*—Almost every variety of the above-mentioned local fits may end in general convulsion. As early manifestations of intra-cranial tumour they are most common in disease of the frontal and anterior part of the temporal lobe. As late manifestations they may occur wherever be the disease, except when the lesion is confined to the cerebellum.

*Smell and taste.*—Unilateral loss of smell indicates damage to the corresponding olfactory tract as the only possible cause, provided the nasal cavity of that side be healthy.

Unilateral loss of taste may indicate either:

(1) A lesion of the fifth nerve under the skull if it is associated with facial anaesthesia.

(2) A lesion of the facial nerve in the temporal bone, if facial paralysis of peripheral type be present.

(3) A functional condition, if accompanied by hemianaesthesia.

(4) Involvement of the chorda tympani.

*Vision.*—Hemianopia indicates invariably a lesion of the cuneus, optic radiation, or optic tract. If the pupil does not contract to a spot of light thrown on to the blind half of either retina, the pupil reflex fibres which leave the optic path at the external geniculate body are also involved, and, therefore, the point of damage is in some part of the optic tract and associated hemiplegia is a confirmatory sign due to involvement of the contiguous crus cerebri. This sign is called Wernicke's hemianopic pupillary phenomenon.

If the pupil react on stimulation of the blind half of the retina, the optic tract must be intact and the disease situated in the optic path from cuneus to thalamus. If hemianesthesia be present, the disease is certainly involving the optic radiation; if not, the cuneus must be the seat of damage.

Quadrantic hemianopia is nearly always the result of a lesion in the cuneo-lingualis, the cuneus corresponding with the upper retinal quadrants, the lingualis with the lower halves of the retinae. Bitemporal hemianopia is of rare occurrence. It is met with in tumour of the pituitary body which involves the anterior part of the chiasma in the middle line.

Total blindness resulting from optic neuritis renders it impossible to investigate the condition of the visual fields, and several cases have come under the writer's notice absolutely blind from optic neuritis, in which the visual fields had not been investigated before vision was lost, neither was the patient aware of any hemianopic defect so long as his vision lasted; yet the *post-mortem* investigations showed that hemianopia must have been present at an early stage. Such total blindness may, therefore, deprive the physician of one of the most valuable local indications.

*Optic neuritis*, as has been already stated, is of no localising value. Its absence where other signs render the presence of intra-cranial tumour probable suggests a lesion in the dorsal part of the pons or of the medulla. If in a case of intra-cranial tumour optic neuritis disappears with the onset of ophthalmoplegia in one or both eyes, the lesion is

situated in the mesencephalon and involves the oculo-motor nuclei.

Unilateral optic atrophy, especially when the atrophy commences at one edge of the disc and gradually invades the whole disc, with corresponding loss of the visual field, must be due to tumour compressing the optic nerve. Bitemporal atrophy with bitemporal hemianopia is always the result of growth in the pituitary region involving the anterior part of the chiasma.

Crossed amblyopia is in nearly all cases a sign of hysteria, and is associated with functional hemianæsthesia. It is not often met with in children until they approach the age of puberty. Occurring with other signs of intra-cranial tumour, it may indicate a lesion of the angular gyrus of the same side as the most restricted field, or it may be simply an hysterical manifestation associated with intra-cranial tumour.

#### OCULAR PHENOMENA.

Inequality of the pupil is of no localising value. Loss of the reflex contraction to light, with preservation of the contraction upon convergence, is an important indication of a lesion in the region around the posterior commissure, whether the loss be unilateral or bilateral. For the Argyll Robertson pupil so frequently caused by tabes dorsalis and antecedent syphilis in adults is not so caused in children, the only comparable infantile condition in which it is found being the juvenile form of general paralysis.

Paralysis, partial or complete, of the movements of conjugate deviation is by far the most certain localising ocular sign. Loss of the lateral conjugate movements can only occur from a lesion of the sixth nucleus of that side towards which the movement is lost.

Loss of the vertical conjugate movements places the lesion of necessity in the mesencephalon, close to the aqueduct of Sylvius.

Ocular paralysis of irregular type, *i. e.* not corresponding with nuclear peripheral nerve supply, or paralysis of the

vertical conjugate movements when associated with ataxy, constitute strong presumptive evidence of a lesion in the region of the corpora quadrigemina (Nothnagel's syndrome).

Ocular paralysis of peripheral nerve distribution (third, fourth, and sixth nerve palsy) is of importance when occurring early in the course of intra-cranial tumour, *i.e.* within the first six months after general symptoms have appeared. Such paralysis can only be produced by involvement of the nerve either between its nucleus of origin and the surface of the brain, or between its superficial origin from the brain and its exit from the skull. Consequently, the third nerve, having the shortest course, can be involved in a lesion limited to the following situations—the crus cerebri, the interpedal space, the internal lappet of the temporal lobe, the cavernous sinus, and the sella turcica. On the other hand, the fourth and sixth nerves, having a much longer course, are exposed to possible direct pressure from growth in any spot contiguous to the course of the nerves.

The situation of the lesion is certainly the region of the interpedal space when both third nerves are paralysed, and most probably the lower part of the pons, or the dorsum ephippii of the occipital bone when both sixth nerves are paralysed.

*Fifth nerve.*—Anæsthesia of the face over the distribution of the three divisions of the trigeminus associated with paralysis of the masseter temporal and myohyoid muscles (these being mentioned since they are the most easily examined of the muscles supplied by the fifth nerve), indicate either a lesion of the pons close to the exit of the nerve, or a lesion between the superficial origin of the nerve and the Gasserian ganglion, or a complete involvement at the site of the ganglion. Involvement of the branches separately is usually the result of growth involving the dura mater or bone of the middle fossa of the skull. It must be borne in mind, however, that the sensory part of the trigeminus takes its origin from a long column of cells extending from the third cervical segment of the spinal cord

as high as the pons, and that the lower group of these cells supplies the central part of the face, while an upper group (medullo-pontine group) supplies the upper face and anterior portion of the scalp; therefore, lesions of the upper spinal cord, medulla, and pons may cause anesthesia over the lower part of the face, lower and middle portions, or over the whole area of the fifth distribution respectively, according to their position, without any motor paralysis, for the motor nucleus of the fifth nerve is placed in the pons about on a level with the exit of the nerves.

The division of the cells of origin of the sensory fifth into groups is an arbitrary division. Anatomically, the cell column is continuous.

*The facial nerve.*—Involvement of the facial nerve in a lesion causes facial paralysis of an atrophic type, the affected muscles presenting the reaction of degeneration. It is especially important that there should be no confusion between such peripheral facial palsy and facial paralysis resulting from lesion of the pyramidal fibres in any part of their course from the central cortex to the facial nucleus. From certain anatomical peculiarities, it is easy to determine the point at which the facial nerve is involved. The effect of a lesion in the various parts of the nerve here follows:

*Facial nucleus.*—The orbicularis palpebrarum and oris are not affected, since these muscles are probably supplied from the oculo-motor nuclei and the hypoglossal nuclei respectively. (No case has been reported, so far as the writer is aware, of a lesion confined to this nucleus alone.)

*Superficial origin to geniculate ganglion.*—The whole face is involved, and there is no loss of taste upon the same side of the tongue. Affection of the contiguous auditory nerve is common.

*Geniculate ganglion to origin of chorda tympani.*—The whole face is involved, the taste-fibres entering the facial nerve at the geniculate ganglion are involved, producing unilateral loss of taste. Hyperacusis may result if the lesion be above the nerve to the stapedius muscle.

*Below origin of chorda tympani.*—There is no unilateral loss of taste, and as in this situation the facial nerve is dividing into its branches, one of these—temporal, facial, or cervical facial, is likely to be involved to a greater degree than the other.

*Auditory nerve.*—This nerve is often involved with the preceding, and such involvement indicates a lesion either close to its exit from the medulla or in the temporal bone. It is most necessary to ascertain that deafness, when present, is not of old standing, that it is true nerve-deafness, and that there is no tympanic disease present, before a lesion involving the auditory nerve is judged to exist.

*Ninth, tenth, eleventh, and twelfth nerves.*—Paralysis of these nerves from gross central disease is hardly ever met with clinically, since the existence of a gross lesion in the region of the vaso-motor and respiratory centres is incompatible with life.

These nerves are most frequently damaged by growths involving the meninges at the side of the medulla or involving the base of the skull in the posterior fossa. Paralysis of the tongue, palate, pharynx, larynx, sterno-mastoid, and trapezius are the results.

*Hemiplegia* is a most important localising sign, for its presence implies that the lesion is situated either in or close to the course of the pyramidal system. When uncomplicated, it further localises the disease to some situation above the tentorium; for disease of the brain-stem either involves both pyramidal tracts or produces a hemiplegia associated with nuclear palsies or with alternate cranial nerve palsies. In the pontine region hemiataxy is also associated. The following are the chief symptom combinations:

Above the tentorium.	Pure hemiplegia.
Crus cerebri.	Hemiplegia and alternate third nerve paralysis.
Mesencephalon.	Hemiplegia; nuclear ocular palsy; hemiataxy upon the hemiplegia side.
Pons.	Hemiplegia with alternate fifth,

seventh, and eighth nerve palsy; hemiataxy upon either side.

Medulla.

Hemiplegia not involving the face; hemiataxy upon the opposite side to the hemiplegia; alternate ninth, tenth, eleventh, and twelfth nerve palsies.

When it is determined that the lesion producing hemiplegia is situated above the tentorium the following points are of use in ascertaining its probable situation:—

If the hemiplegia commence as a monoplegia and slowly extend, and especially if local convulsion occurs, the lesion is presumably situated either in the cortex or underneath the cortex in the central region; but if, while commencing as a local paralysis, it rapidly progresses, and is associated with later appearing hemianesthesia or hemianopia, the internal capsule is the probable seat of the lesion.

It must be borne in mind that the pyramidal tract in its intra-cranial course seems very resistant to pressure, and it is very rare for a growth to produce hemiplegia unless the pyramidal tract is actually involved, and tumours, especially those growing from the meninges at the base of the brain, may cause great distortion of the brain-stem without any physiological abrogation of the function of the white tracts of the region pressed upon.

Cerebral paraplegia may be due either to a lesion involving both the centres for the lower extremities in the paracentral lobules, a condition which results if a tumour arise in that part of the falx cerebri separating these two lobes, or it may be caused by a pontine lesion situated ventrally and immediately between the pyramidal tracts involving the mesial fibres of each tract, these being the fibres for the lower extremities.

Monoplegia is in a large majority of cases indicative of a lesion of the cortex of the central convolution of the opposite side or of the underlying white matter. It must be remembered, however, that small lesions of the internal capsule,

pons, and brain-stem, if commencing in the course of the pyramidal fibres may cause paralysis strictly limited to one limb, but in such cases local epilepsy never arises, and unless the lesion remain stationary, other symptoms soon arise which obviate any confusion.

Hemianæsthesia is not a common sign of value in localisation, for it would appear that sensory functions are not firmly localised in the brains of young children. Moreover defects of sensibility not amounting to complete loss to touch and pain are very difficult to investigate in young patients, since the outcry resulting from the application of a painful stimulus is almost the only means of judging as to the condition of sensibility. Hemianæsthesia may result either from a lesion of the internal capsule or from a lesion in the tegmentum in some part of its course from the mesencephalon to the medulla. If the sensory loss be severe or absolute, the lesion is certainly in the tegmentum.

Hemiataxy is one of the most important localising signs, and its presence places the lesion below the tentorium. It may result from disease involving either the restiform body, pons, lateral lobe of the cerebellum, middle or superior peduncle of the cerebellum of the same side, or the red nucleus of the opposite side. Bilateral ataxy is usually the result of extension of disease across the middle line of the brain-stem or cerebellum and the involvement of one of the above-mentioned structures upon both sides. Lesion of the inferior vermiciform process of the cerebellum causes ataxy which is bilateral from its first appearance. Intention tremor similar to that occurring in disseminated sclerosis is sometimes seen in connection with lesions of the restiform body; choreiform movements, myoclonic movements, and athetoid movements are most valuable indications, though they are of somewhat rare occurrence. They are due, as a rule, to widely-spread partial lesions of the central cortex.

*Course of the disease.*—With the exception of some cases of tuberculosis and syphilitic tumours and those cases in which the situation and early diagnosis of the growth allow of successful surgical interference, it may be said that the



course is invariably progressive towards a fatal result. There is no doubt whatever that in some cases of tuberculous tumour under appropriate treatment the disease may become obsolete even after pronounced symptoms have appeared; but this happy result occurs so seldom that it cannot be considered in prognosis.

Syphilitic tumour may disappear under treatment, but not infrequently some disability persists, such as recurring convulsion, or monoplegia. These persistent symptoms may be caused either by cicatrization, which follows the healing of the gumma, or by arterial disease so frequently present in the immediate neighbourhood of such a growth.

The progress of a tumour is generally slow, but sometimes it may be rapid. Rarely is it uniform, and periods in which there is exacerbation of the symptoms alternate with periods of diminution. Sometimes the symptoms disappear almost completely and the false hope may be entertained that the tumour has resolved, especially if the disappearance of symptoms has coincided with the exhibition of mercury. How rarely such tumours are really syphilitic can be gathered from the number of cases reported cured or improved in which, subsequently, the necropsy reveals the presence of neoplasm, such as sarcoma, etc. The following case illustrates the disappearance of symptoms for a long period. A boy, aged 11 years, had an attack of "brain fever" associated with optic neuritis, and during the second fortnight of his illness he lay apparently in a dying condition. He had completely recovered at the end of seven weeks. During the next four and a half years he progressed well at his business, but at intervals of about six months he would play truant and be afraid to return home lest he should be scolded. His parents said that this conduct was entirely against his usual nature. At the age of 16 years he came to hospital for defect of sight and was found to have unioocular hemianopia, which was rapidly followed by atrophy of half the disc. No other sign of disease was present. Fourteen days later he fell in a fit, was convulsed, and died. A fibrosarcoma the size of a tangerine orange

was found occupying the mesial aspect of the frontal lobes, and pressing upon the inner part of one optic nerve. A large vein upon the surface of the tumour had burst into the lateral ventricle.

The duration of the disease and its rapidity of progression depend upon the following factors :

(1) *The situation of the growth.*—It is obvious that a tumour which is enlarging will become incompatible with life much sooner if it is situated in the brain-stem than if it is situated in the cerebral or cerebellar hemisphere. Growths of the meninges, especially those situated above the tentorium, are usually the slowest of all in their course.

(2) *The nature of the growth.*—The more quickly growing the neoplasm, the softer its consistency and the more the tendency to degeneration with rupture of vessels, the shorter will be the course of the disease.

The course of glioma and “round-celled” tumour is rapid; that of psammoma, fibrosarcoma, and endothelioma is always slow.

(3) *The occurrence of perifocal œdema.*—Every encephalic growth is liable to the occurrence of a peculiar spreading œdema in its vicinity. We are entirely ignorant of the nature of the process, but it is of great importance as a frequent cause of a fatal exacerbation of symptoms. Two examples may here be useful. A girl aged 15 years, under treatment for two years with epilepsy, was suddenly seized with headache and vomiting. Optic neuritis appeared upon the fourth day, and she died on the seventh day. A hard encapsulated nodule (probably a cicatrised gumma) was found in the left frontal lobe. Extending centrifugally from the focus as far as the parietal lobule, and separated by a sharp demarcation line from the normal brain tissue, was a condition of œdema rendering the brain substance almost translucent.

Agam, a child aged 5 years had had ataxy, optic neuritis, and occasional attacks of headache and vomiting for two years. He was able to walk and play with the

other children. He was suddenly seized with severe symptoms, and died in four days. A hard central sarcoma of the cerebellum strictly encapsulated, was found surrounded by translucent oedematous tissue involving most of the cerebellum and part of the pons.

(4) *The occurrence of ventricular distension.*—In younger children this is to a certain degree compensated for by enlargement of the skull, and when hydrocephalus occurs life is usually prolonged. In older children, on the other hand, in whom the skull is more firmly ossified, the same degree of expansion of the skull does not follow, and life is not long compatible with increasing pressure.

In further explanation of the exacerbation and remission of symptoms which occur in cases of intra-cranial tumour, it should be mentioned that it is probable that the rate of growth of the tumours varies from time to time, and that the administration of mercury and iodide of potassium temporarily interferes with the growth of all neoplasms. Again, variations of the blood-pressure and also of the state of distension of the ventricles, when this exists, occur, and the rule is that symptoms are more prominent when the intra-cranial pressure is high.

Lastly, it is probable that some of the general symptoms of intra-cranial growth are the result of the liberation of toxic substances from the neighbourhood of the tumour, and such liberation may be intermittent.

Death may result in cases of intra-cranial tumour from several causes. The most common of these is exhaustion, induced by the violence of the pain and by the interference with nutrition which frequent vomiting and the difficulty in feeding from mental dulness and dysphagia, entail. Another frequent cause is the lowering of vitality of all the cerebral centres, including especially the respiratory centre, which prolonged increased intra-cranial pressure produces. The clinical evidence of this is coma, with increasing accumulation of mucus in the lungs and resulting interference with respiration. And in any case of intra-cranial neoplasm death may occur quite suddenly from respiratory failure.

Again, the patient may die suddenly without apparent cause, or he may die in convulsion.

Certain complications of cerebral tumour are the immediate causes of death in not a few cases. Hæmorrhage is the most important of these, and it may occur in the substance of a growth or may result from the degeneration of the walls of a vessel which is locally pressed upon by a hard tumour. Meningitis is a common cause of death in tuberculous tumours and not uncommonly in the other varieties of tumour.

Lastly, in very chronic tumour death may occur from intercurrent unrelated diseases.

*Prognosis.*—Except in those very rare cases in which there is high probability that the tumour is of syphilitic nature, *the prognosis may be deemed hopeless unless surgical interference is possible.* Even when the growth is syphilitic the prognosis is not necessarily good as regards life, and by no means to be lightly thought of as regards capacity. For syphiloma does not always resolve on treatment, and when it does, cicatrization and arterial disease may cause permanent disability. Prognosis as regards length of life is very uncertain; for while, in a case in which rapidly increasing symptoms suggesting a situation of the tumour in a vital part (brain-stem) a certain opinion may be formed and expressed that life will not be long preserved, yet in a case with symptoms relatively quiescent, there is always a liability to sudden death as the result of an acute exacerbation of symptoms.

The prognosis as regards operative measures is perhaps best expressed in those statements with which it is customary to approach the friends of a patient. The patient is suffering from a necessarily fatal malady. If he is operated upon he will probably never have any more headache or sickness, and these symptoms must of necessity continue if he is not operated upon. The operation may save his sight. He may die under the operation,—it will very probably hasten death—and he is likely to be mentally reduced and more disabled than at present, but the operation alone holds out a

possibility, however small, of recovery, with partial or complete capacity.

*Diagnosis.*—When optic neuritis, either alone or associated with headache and vomiting, is the sign present, the distinction has to be made between organic disease of the brain and certain other conditions in which these symptoms may occur. These are anaemia, kidney disease, lead-poisoning, and certain blood states of the nature of which we know little. When anaemia is the cause, poverty of the blood is usually so great as to make diagnosis easy; the optic neuritis develops more rapidly than in tumour, while rest in bed, a proper diet, and the administration of iron cause rapid disappearance of symptoms. In renal disease and in lead-poisoning with renal disease, the neuritis may be severe and the fundi usually show other signs characteristic of these conditions. The history of lead-colic, the presence of the lead line and of albumen in the urine should not permit of confusion. In these conditions the headache is occasionally, not often, as severe as in tumour. It is not usually so severe as to keep the patient awake at night. Moreover, there is an entire absence of focal symptoms.

It must always be borne in mind that intra-cranial tumour may co-exist with one of these general conditions, and it may be said that where these symptoms are severe and persistent enough to cause difficulty in the diagnosis the probability is that tumour is also present.

When symptoms of a local lesion exist while optic neuritis and, perhaps, also vomiting and headache are absent, a tumour or some other focal lesion may exist.

The problem includes these questions:

(1) Do the local signs point to a lesion in such a situation as to explain the absence of optic neuritis? (Mesencephalon, pons, and medulla.)

(2) Is the nature of the signs in onset and course such as a slowly progressive lesion would cause?

In certain cases local signs of slow onset and progressive course, associated with headache and vomiting and closely resembling those of intra-cranial tumour, have been due to

an enlarging focus of disseminated sclerosis involving one of the peduncles of the cerebellum. In such cases the optic discs are unduly pale, and other general signs of the disease mentioned eventually appear, though they be delayed for many weeks.

It may be said that progressive paralytic phenomena in children are in nearly all cases due either to the presence of tumour or progressive cell degeneration (the so-called atrophic sclerosis). In the former case they are more strictly focal, whereas in the latter they are always of wide distribution.

Jacksonian convulsion, in the absence of all other signs, not infrequently suggests tumour, but it cannot be too emphatically stated that this phenomenon may be simply a manifestation of so-called idiopathic epilepsy and, in the absence of monoplegia or general signs of tumour it should always be treated as such. It should never be considered as a signal for the immediate opening of the skull. So frequently do cases of cerebral tumour come under observation, in the first instance, as cases of epilepsy, that it behoves the physician to make repeated examination of the optic discs in all cases in which fits have recently commenced. When a tumour is present, optic neuritis usually makes its appearance within six months of the occurrence of the first fit.

When both general and localising symptoms are present, distinction has to be made from meningitis and intra-cranial abscess. The acute forms of meningitis are not likely to be confounded with tumour on account of their acute onset, rapid course, and the associated pyrexia.

The onset of tuberculous meningitis and of posterior basic meningitis is more rapid than the onset of tumour. The prodromata, wasting, and abdominal retraction of the former, and the head retraction and rarity of optic neuritis, although curiously enough the frequency of blindness in the latter, are striking points of distinction. From the chronic forms of intra-cranial abscess tumour can only be distinguished by the presence of an obvious cause for the occurrence of abscess.

When the determination has been made that an intra-cranial tumour is present, it remains to decide where it is situated and of what nature it is. The former question has been fully discussed under the heading of Local Diagnosis; the latter can be rarely determined with certainty. A family history of tuberculous disease, the presence of tuberculous disease in other parts of the body, the recognition of choroidal tubercle with the ophthalmoscope in a case the symptoms of which indicate a tumour of slow growth, make it highly probable that a tuberculous mass is present. So rarely does gumma of the brain occur in children that a tumour, even in the presence of marked signs of syphilis, is almost certainly not syphilitic. If there are indications that the growth commenced in the meninges, fibrosarcoma, psammoma, myxoma, and tubercle have all to be considered. A rapidly-growing tumour is generally either glioma or "round-celled" tumour.

Slowly growing tumours are either myxomata, fibromata, spindle-celled sarcomata, or psammomata. Tumours of the brain-stem are almost always gliomata, sarcomata, or tuberculous tumours.

*Treatment.*—Tuberculous growths are, perhaps, the only intra-cranial tumours in children that medicinal treatment may radically influence. In these conditions cod-liver oil and iron, and an adequate supply of good food, fresh air and sunshine are all-important. It is a remarkable fact that similar treatment appears to improve the condition and lessen the symptoms in almost every case of intra-cranial tumour, wherever it is applicable. Notwithstanding the statement made above that syphilitic growths are almost unknown in the brains of children, yet in every case of intra-cranial tumour that may be deemed of other than tuberculous nature mercury and iodide of potassium should be exhibited; for a temporary amelioration or even cessation of symptoms and a diminution of optic neuritis frequently follow the exhibition of these drugs, even if the event shows the growth to have been non-syphilitic in very many cases.

In children exhausted by vomiting and pain it is very necessary to restore the physical condition by careful feeding and palliative measures before administering these drugs. Mercury should always be given by inunctions of from 10 to 20 grs. night and morning, watch being kept for any appearances of intolerance. Iodide of potassium should never be given when the condition of gastritis and large pale tongue, so commonly associated with persistent vomiting, is present. Indeed, this condition is not infrequently brought about by unsparing administration of iodide of potassium and the vomiting thereby much increased.

*The palliative treatment of symptoms.*—*Headache* is only in rare cases constantly present. It comes and goes. In some cases it appears at such short intervals as to call for continual treatment, and in a few cases it may persist in intense degree for comparatively long periods—24 to 48 hours. Sometimes an attack is so severe as to produce profound collapse, and then life is immediately threatened. As a rule, the lower the blood-pressure the less the headache, and certain remedies which have the effect of keeping the blood-pressure low are of great value in those cases in which headache is persistently troublesome. These are the saline purgatives, mercury given by the mouth, either in the form of grey powder or blue pill, and administered upon alternate nights, and iodide of potassium in small doses. Alcohol almost invariably increases the tendency to headache, and should be avoided.

The application of ice or of Leiter's tubes to the head is frequently advised for the relief of headache, but it is of questionable value. It does not comfort the patient, it rarely relieves the headache, and it is worse than useless when headache is intense and collapse threatened.

By far the most useful measure for cutting short an attack of headache is the administration of one of the coal-tar products. These may be mentioned in the order of potency: Antifebrin (2-5 grs.), antipyrin (7-10 grs.), phenalgin, citrophene, ammonal, and phenacetin. To secure a certain effect a full dose should be given. It should be



remembered that while antifebrin is the most powerful of these drugs antipyrin is by far the most poisonous. The mistake should not be made of ordering alcohol with these remedies in order to avoid collapse, for the action of the drug is interfered with, and by so doing the blue colour of the lips which may result from administration of full doses of some of these drugs is not cyanosis, nor is it a sign of heart failure, but it is due to the presence in the blood of a compound between hæmoglobin and the common base of all the above-mentioned remedies, namely, anilin.

Caffein citrate is often advantageously combined with one of the coal-tar compounds. The combination seems to act more rapidly, and produce an exhilarating rather than a depressing effect. The much used and, indeed, very useful patent remedies, migrainin (antipyrin and caffein) and antikamnia (antifebrin and caffein) are of this nature. A combination of great potency is that of antipyrine and eldoral, but it is very depressing, and should only be used when other remedies fail, and only in sthenic subjects.

When the intensity of the pain produces frenzy, and when collapse from pain occurs, the above-mentioned remedies are useless. It is necessary to put an end to the pain at once, lest death occur. A full dose of morphia, and no less, should be administered hypodermically. Such administration may require some courage on the part of the physician. A full dose of morphia does not seem applicable to a patient who a few moments ago was frenzied with pain and now is pale, pulseless and seemingly unconscious; yet it must be recognised that the cause of the collapse is pain alone, and in such a condition morphia is the best stimulant, the only anodyne that will act, and the only hope for the patient's life is its administration. Should the patient have ceased breathing, artificial respiration should at once be commenced.

*Vomiting.*—All-important in the treatment of this symptom is careful regulation of the diet and attention to any state of disorder of the alimentary canal that may exist—

especially gastritis. It has already been mentioned that administration of iodide of potassium is likely to cause gastritis, which greatly increases the tendency to vomit. During an attack of vomiting the patient should be kept at rest, and should be allowed ice to suck. The coal-tar products employed for the relief of headache are generally efficacious remedies, especially when combined with caffeine. Bromide of potassium is sometimes useful, and so also in certain obstinate cases are atropine, in small doses, and ipecacuanha. Morphia, administered hypodermically, will always check cerebral vomiting, but it is seldom required.

*Epileptiform attacks*, whether local or general, should be treated with bromides either by mouth or rectum.

*Bedsore and cystitis* should not occur if sufficient care is taken in the management of the case. When present they should be treated in the ordinary way. It is important that every bedridden patient should be upon a water-bed.

*Operative treatment.*—Surgical procedures are undertaken in some cases simply as palliative measures for the relief of headache and to prevent loss of sight from optic neuritis. In other cases there is a probability that the growth is in such a situation that it may be successfully removed, while in most cases there is a possibility that such a happy result may follow, though it may be only a remote one. There are certain cases in which operative measures are absolutely contra-indicated: (1) when the tumour is situated in the mesencephalon or pons; (2) cases of long standing, in which it is probable that ventricular distension exists. When a growth is located in the brain-stem there is no hope of successful removal. In cases of long standing, when hydrocephalus exists, the result of opening the skull and dura is to cause such dislocation of anatomical relation and so much alteration of the circulatory conditions that death may ensue rapidly. As an example, a case of tumour of the middle lobe of the cerebellum may be taken when there is marked ventricular distension. If, in such a case, the skull and dura are opened below the tentorium, the intraventricular tension is not necessarily relieved, and the

supratentorial pressure forces down the tentorium and causes the cerebellum at once to protrude from the wound, and produces such disturbance of anatomical relation, if not actual tearing of the tissues, as to be incompatible with the continuance of life.

The earlier the aid of the surgeon is sought in a given case the greater are the chances that his interference will result beneficially to the patient. It is useless to operate for the relief of optic neuritis if the neuritis has proceeded to that intensity which entails inevitable blindness. Again, the process of healing occurs more readily the more nearly the condition of tissues involved by operation approximates to the normal, and the longer a cerebral growth has produced symptoms, the more will the cerebral tissues be found compressed, wasted, and oedematous. The determination as to the advisability of operative procedure in a given case involves the following questions:

Is the presumable situation of the growth such as to allow reasonable hope that it can be removed?

Does the intensity and progress of the optic neuritis suggest that an irremediable stage of neuritis will soon be reached?

It has been already stated that the longer the duration of general symptoms the worse the prognosis as regards the results of operation.

The physical state of the patient at the time of operation does not greatly influence prognosis, for recovery has occurred in the case of several patients who have been brought into the operating theatre apparently moribund.

#### DIFFICULTIES AND FALLACIES WHICH MAY ARISE IN THE LOCAL DIAGNOSIS OF CEREBRAL TUMOUR

The exact determination of the position of a cerebral tumour is beset with many difficulties. A correct knowledge of the paths along which one may be led to erroneous diagnosis is an essential preliminary to the study of clinical cerebral localisation.

The chief peculiarities of the symptomatology of cerebral growth leading to fallacies in local diagnosis may be formulated as follows, and will be dealt with *seriatim*.

(1) The invasion of a tissue by growth without abrogation of the function of such tissue.

(2) The obliteration of the functional defect caused by the destruction of one area by compensation occurring in another area.

(3) The occurrence of false "pathognomonic" local signs.

(4) The concealment of pathognomonic local signs by certain secondary processes arising in the course of the disease.

(5) The impossibility of eliciting signs necessary for diagnosis on account of physical feebleness, mental reduction, and subconscious states.

(1) *Invasion without abrogation of function*.—This condition is of frequent occurrence in the cerebral cortex and centrum ovale, and is especially prone to occur when the growth is a glioma. Less common in the cases in which growths occupy the cerebellum, it is rare when tumours are situated in the course of the main conducting paths, such as the internal capsule and brain-stem.

Growths situated in the cerebral hemispheres may exist for some time either without symptoms or with symptoms so slight as not at once to suggest the nature and severity of the disease. Marked symptoms in many cases do not arise until the growth has reached such a magnitude as to preclude successful surgical interference. Sometimes hæmorrhage occurs in the substance of a growth which until this happens has produced no symptoms, and sometimes if death follow rapidly upon the occurrence of such a hæmorrhage the case cannot be clinically distinguished from one of ordinary cerebral hæmorrhage.

In other cases in which the extravasation of blood remains confined to the region of the growth, severe general symptoms of cerebral tumour may arise simultaneously with the occurrence of the hæmorrhage and the case may run a fatal course of unusual rapidity. As an example of the latter

occurrence the following case may be cited. A girl, aged 15 years, was quite healthy until she was seized suddenly with intense headache and vomiting. She developed optic neuritis, and died six days later. A large tumour of the right centrum ovale with a central hæmorrhage was found *post mortem*.

It may be here remarked that tumours of the cerebral cortex are extremely rare, while subcortical tumours are very common. It is rare for a growth of the latter variety to produce localising symptoms until it reaches the size of a tangerine orange. It follows, therefore, that in a case in which Jacksonian epilepsy is associated with optic neuritis and there is no sign of meningeal involvement, the probability is great that a comparatively large subcortical growth is present even in the absence of paralysis, and the opinion formed as to the possible result of operative procedure therefore must not be over-sanguine.

The explanation of the invasion of a tissue by growth without abrogation of function is twofold. First, the growth as it enlarges may separate and push aside the elements without impairing their function either directly or by interfering with the blood supply. It is obvious that such separation will occur with least damage in the midst of a mass of long nerve-fibres which run more or less parallel to one another, as in the corona radiata. Secondly, an infiltrating growth may grow and extend between the nerve elements without displacing them to a marked extent, and in such a case no symptoms may arise until degeneration of the growth occurs. The most remarkable example of the latter is the so-called "hypertrophy of the pons" (infiltrating glioma of the pons).

(2) *Obliteration of functional defect due to destruction of one area by compensation occurring in another.*—In the cerebrum and cerebellum of the child it is only an extensive lesion that will produce lasting local signs. If the lesion is one of the cortex, compensation may occur by the transference of the function of the lost elements either to the cortex in the vicinity of the lesion or the corresponding part of the oppo-

site cerebral hemisphere or to a distant part of the same cerebral hemisphere which has a function of like quality.

As an example of the first method of compensation: A child, as the result of a small focus of thrombosis, developed a brachial monoplegia which completely disappeared. Some months later he developed a complete brachio-facial paralysis. An old focus of thrombosis surrounded by a recent area of thrombosis was found. The compensation for the primary lesion must in this case have been by that region involved in the second lesion. As an example of

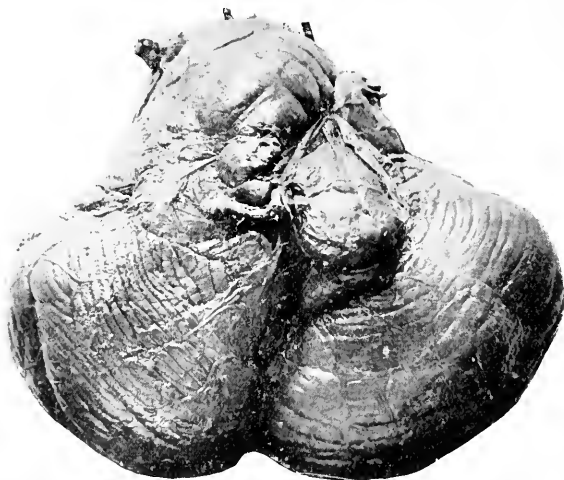


FIG. 22.—Infiltrating glioma of pons, so-called "hypertrophy of pons."

compensation by the opposite hemisphere the well-known case recorded by Sir Thomas Barlow is most illustrative. A boy, who was right-handed, developed complete motor aphasia with right brachio-facial monoplegia. He recovered his speech completely. Some months later he became again completely aphasic and remained so till his death, which occurred many months afterwards. At the necropsy a patch of softening, the result of embolism, of the size of a shilling was found occupying the position of the third frontal convolution of each hemisphere, that in the left hemisphere being obviously the older lesion.

Compensation appears to occur readily for lesions situated in the centrum ovale in inverse proportion to the number of fibres of the corona radiata which the lesion interrupts, and, consequently, the nearer the lesion is to the internal capsule the more likely are permanent symptoms to be produced on account of the wedge-shaped coronal projection converging to the capsule.

The reader may here be reminded that permanent aphasia in any of its varieties does not occur except from actual involvement of the cortical grey matter of the areas concerned with speech. In the cerebellum a non-progressive limited lesion does not produce lasting symptoms, for compensation occurs, as in the cerebrum, by the cerebellar cortex contiguous to the lesion and by the opposite hemisphere of the cerebellum. It would appear that for localising signs of cerebellar damage to be constantly evidenced in the same degree a constantly increasing lesion must be present, and since such a lesion is hardly conceivable it follows that cerebellar localising signs "come and go."

In the conducting tracts of the internal capsule, brain-stem, and spinal-cord compensation is only possible where there is a choice of paths. Where a double path exists when the functions of the nervous system become increasingly developed as age advances from infancy to adult life, one of these paths becomes the usual path, and, increasingly, the only path. Therefore tract compensation takes place easily in the young infant, and the less readily as age advances.

As an example: The cerebral cortex of the central region is connected with the limbs by a path which, from the mid-brain downwards, is double, perhaps multiple. The most ancient of these paths, speaking morphogenetically, is a mediate path in two relays: (1) Cortex-mesencephalon, (2) Mesencephalon-cord. In certain lower animals, the bird for example, this is the only path from cortex to limbs.

The path which has more recently appeared, and which exists only in the higher mammals, is a short circuit placing

the cortex directly in connection with the limb centres in the cord—the pyramidal tract. In the series from lower to higher mammals, and as age advances from infancy in the human subjects, the pyramidal tract becomes increasingly the more important and usual path for volitional incitation of the limbs. Consequently in the higher mammals and in the human infant, complete destruction confined to one pyramidal tract below the mesencephalon does not cause complete or lasting hemiplegia, for the alternative mediate path is available. Compensation for lesions of the essential nuclei upon the outgoing path does not occur, for in these nuclei there is almost complete and exclusive monopoly of function.

(3) *The occurrence of false localising signs.*—When from the presence of growth within the skull the intra-cranial pressure has been persistently above the normal for a long time the following phenomena are apt to appear, irrespective of the situation of the tumour :

(a) Paralysis of any of the cranial nerves. There is no evidence as to what determines the incidence of this paralysis upon any particular cranial nerve, nor is there any order in the manner in which these nerves are affected. The sensory portion of the fifth nerve, for example, may be paralysed, the motor division being intact and *vice versú*. The sixth nerve probably suffers more frequently than any other.

(b) Local epilepsy is prone to occur when there is general ventricular distension, presumably as the result of the wasting and interference with the blood supply of the cortex which results from such distension.

(c) Local vascular lesion may occur in any part of the brain and produce local signs, for the result of long-standing high intra-cranial pressure is to favour the occurrence both of thrombosis and hæmorrhage.

(d) The pressing down of part of the cerebellum into the foramen magnum may cause cerebellar local signs.

(e) Double hemiplegia results in a large proportion of cases in which the ventricles are persistently distended.



(f) As a result of the degeneration in the posterior roots and posterior columns of the spinal cord which occurs in many cases of intra-cranial growth, lightning pains, ataxy, and loss of knee-jerk may occur.

(4) *The concealment of pathognomonic local signs.*—A patient may come under observation for the first time completely blind from optic neuritis, and the presence of hemianopsia and word-blindness—the only sure signs of involvement of the occipital lobes and angular gyrus—cannot be tested. It is remarkable that even an intelligent, observant patient may be completely unaware of the presence of hemianopsia.

(5) The impossibility of eliciting signs necessary for diagnosis on account of physical feebleness, mental reduction, and sub-conscious state is a frequent obstacle to a correct diagnosis. Occasionally, the history in such a case is helpful; but even with a good history the correct diagnosis and localisation of a growth, when a patient is seen in such a state for the first time, is, as a rule, impossible.

#### PECULIARITIES OF THE LOCALISING SIGNS IN CHILDHOOD

It may be said that while the symptoms resulting from local lesions in the cerebellum, brain-stem and basal ganglia are exactly similar in children and adults, those produced by the same lesions of the cerebral hemispheres in children are less constant and less marked than in adults, and the difference is greater the younger the child. The reasons for such differences are not far to seek. The brain of the child at birth has by no means reached its complete anatomical development, and its physiological development is only complete at the age of puberty. Compensation for the results of local destruction of brain tissue occurs readily before the various regions of the cortex are indelibly stamped with a special function, provided that there is a sufficient area of normal cortex remaining. Such compensation may occur from the taking on of the function of the damaged area by: (1) the normal cortical substance in the

immediate neighbourhood of the lesion; (2) by the corresponding locality in the opposite hemisphere; (3) by a region of like quality situated in the same or the opposite hemisphere. As an example of the third method of compensation it has been mentioned upon a preceding page that the frontal, temporal, and occipital lobes are probably concerned with the higher cerebral functions, and it is here suggested as probable that these regions mutually compensate for local lesions incident upon them.

The physiological nature of compensation by centres of equivalent value (of the same level) necessitates the impossibility of any compensation for very widely spread lesions, especially when these involve both cerebral hemispheres; for the lower centres (basal ganglia, etc.) cannot compensate so far as we know for any of the higher cerebral functions.

A local lesion, therefore, of the cortex or subcortical matter in a child, causes less definite localising signs, and such signs occur later than in the adult. In other words, a relatively much larger lesion must be present in the child than in the adult to produce the same degree of paralysis. These remarks do not apply to epilepsy, sensory or motor, for these are irritative symptoms and are never in relation to the size of the lesion as are paralytic phenomena.

The most striking difference between the localising signs in children and adults is the rarity of any variety of aphasia from local lesions. It is almost unknown in connection with cerebral tumour in children below the age of ten years. It does, however, occur in vascular lesions, probably on account of the more complete local destruction which occurs, but even then it is only temporary. Permanent aphasia may be said never to occur as the result of a unilateral cerebral lesion in a child under seven years of age, provided a moderate mental capacity persist, and though exceptions to this rule have been recorded, they occur very rarely. A lesion, however, of a cortical area concerned with speech occurring before the fifth year may greatly retard the development of speech.

The absence, therefore, of motor aphasia, word-blindness,

and word-deafness must not be allowed much weight in a diagnosis.

Mental peculiarities in children are extremely difficult of observation, and it is only rarely and in older children that they are of value as localising signs of frontal, temporal, or occipital involvement. Marked mental defect is, however, always a sign of a cerebral lesion and does not occur in lesions of the brain-stem and cerebellum except as the result of the hydrocephalus which such lesions may produce.

*Local alteration of the conformation of the skull.*—While this event occurs occasionally in association with intra-cranial tumour in adults it is commonly met with in children in association with intra-cranial tumour, vascular lesions, and local atrophy. The frequency of such alterations in the shape of the cranium in children is explained partly by the fact that the cranium adapts itself in the process of growth to the shape of its contents, and partly since the skull appears to yield to local pressure to a greater extent in early years.

In every case of cerebral tumour occurring in childhood a careful external examination of the skull should never be omitted, for in 80 per cent. of a large series of cases of tumour of the cerebellum occurring at the National Hospital, such examination revealed a most palpable bulging of the skull over the diseased cerebellum. Bulging over the situation of a tumour above the tentorium occurs occasionally, but it is not frequent. For the production of such pathognomonic local signs upon the surface of the skull it is not necessary that a growth should implicate the meninges, or that it should reach the surface of the brain, for they sometimes occur with deeply-seated tumours.

Deformity of the skull resulting from encephalic vascular lesions and local atrophic sclerosis occurs when these lesions occur before the eighth year. As it is caused by adaptation in the process of growth, it necessarily does not make its appearance till some time has elapsed after the occurrence of the lesion. In the majority of cases of infantile hemiplegia the skull presents marked flattening in the central region of the side opposite to the paralysis.

Alterations in the configuration of the skull are also very common in case of cerebral diplegia, but since the atrophy of the cerebral substance is, as a rule, symmetrical, the deformity is not so striking, and it is often difficult to determine whether a skull of given shape is within the limits of normal development or not. When, however, the bilateral atrophy affects the frontal lobes alone, or the occipital lobes, most striking deformity results. The peculiarity of the skull associated with agenesis of the brain is the condition known as microcephaly.

## CHOREA AND ALLIED DISEASES

THERE are several distinct diseases of the nervous system in which the occurrence of involuntary arrhythmical movements forms a striking feature. Although some of these diseases are widely separated both etiologically and pathologically, yet the name "chorea" has been in past times applied to all of them, and is still sometimes used, as in the terms "habit chorea," "senile chorea," "congenital chorea." These diseases may be classified as follows :

(1) Chorea proper.

Rheumatic chorea (Sydenham's chorea).

Huntington's chorea.

Hysterical chorea.

Myoclonus.

(2) Diseases in which choreic or myoclonic movements, or movements closely simulating these, sometimes occur as symptoms but which are widely separated from the true choreas by their etiology and pathology :

Cerebral diplegia.

Friedreich's ataxy.

Gross organic disease of the cerebrum (vascular lesions, tumour, abscess).

(3) Diseases to which the term "chorea" has been applied, but in which the nature of the occurring movements is essentially distinct from those of chorea :

Habit spasm.

The several varieties of "tic."

Dancing mania, etc.

When choreic movements occur as symptoms of gross cerebral disease, they are frequently associated with athetotic movements and with myoclonic movements. In cerebral

diplegia, for example, of three cases presenting the same pathological lesion of the brain (atrophic sclerosis) one may present athetotic movements, in a second case choreic movements may occur, while in a third case myoclonic movements may render the clinical aspect peculiar.

It is interesting to consider these three types of spontaneous involuntary movements in relation to one another as symptoms of cerebral disease. So far as we know they occur only when the function of the cerebral cortex of the motor area is directly or indirectly disturbed. If comparison be made between a typical athetotic movement, a typical choreic movement and a typical myoclonic movement, each will be found to present distinct features; yet every transitional form of movement is met with between athetosis and chorea, and between chorea and myoclonus.

In the maladies which are described in this chapter as choreas proper those types of spontaneous movement which are indistinguishable from those of myoclonus, and movements approximating to athetosis, are met with. Myoclonic movements in the face and upper extremities are a marked feature of some cases of Huntington's chorea, and such cases differ but little in their clinical aspect from cases of familial adult myoclonus. In many cases of myoclonus movements of a transitional type occur. This group of maladies is allied, not only by the clinical manifestations which they are so apt to possess in common, but also by the occurrence of cases of an intermediate type which cannot be definitely placed either with chorea or with myoclonus.

The little that is known of the pathology of these diseases further justifies their inclusion in one group.

## CHOREA

*Synonymy.*—St. Vitus' dance; Sydenham's chorea; rheumatic chorea.

Chorea is a spasmodic affection of the nervous system which occurs chiefly in young persons and is generally of limited duration. It is characterised by the occurrence of irregular involuntary movements, by weakness and inco-ordination of voluntary movements, and often by mental weakness. The subject of chorea frequently suffers at some period of childhood from the manifestations of rheumatism, and a history of acute rheumatism in the family is so often obtained that the association with rheumatism is one of the most important characteristics of chorea.

The term "chorea" was originally used in connection with an epidemic of dancing mania which was prevalent in the early part of the fifteenth century in Germany. An epidemic having occurred at Strasburg in the year 1418, the prefect of that city ordered the sufferers to the chapel of St. Vitus, at Zabern, that they might be healed by the saint. This disease was from this time named "chorea Sancti Viti" (*χορεία* = dancing). Subsequently many conditions in which spasmodic movements occurred were known under this name, until Sydenham, in 1686, first described the disease which we are now considering, and applied to it the term "chorea."

*Symptoms.*—While the onset of symptoms is usually gradual and progressive, it is sometimes abrupt, and this is especially the case when emotion has been the determining cause. A few hours after the occurrence of psychical disturbance, perhaps upon the following day, in rare cases within a few minutes, small arhythmic movements of the

face and of the fingers appear and, rapidly becoming general, increase in range and in force.

When the onset is gradual, the appearance of the abnormal movements is preceded by alteration of the mental and physical condition of the child. She becomes more nervous and more impressionable than before; she is irritable, and often laughs and cries without apparent cause. Her facial expression is mournful, she is increasingly unable to apply her attention, and she cannot do her lessons. At this time careful scrutiny will detect slight involuntary movements of the face and of the fingers, often unilateral in distribution. She becomes clumsy in her movements—overturns her glass at table and lets fall objects which she is holding. The inevitable reprimands which she suffers for those faults have an immediate effect in augmenting her clumsiness.

Anæmia, general languor, irregularity of appetite, and oftentimes constipation, are present. It not unfrequently happens that, in children who have years previously suffered from incontinence of urine, this symptom returns. The child frequently complains of pains running down the limbs.

From day to day the movements become more marked and spread to the limbs and trunk. The face is constantly grimacing, and the hands and arms scarcely cease from turning about, and affection of the legs makes the walking irregular and clumsy. The child can no longer keep still, the respiration movements become spasmodic, and the chorea is fully developed.

The symptoms which characterise a well-marked case of chorea are (1) involuntary spontaneous movements, (2) weakness of voluntary movements, (3) loss of precision in voluntary movement, (4) emotional instability, and other psychical disturbances.

The first of these is usually the most conspicuous symptom, but sometimes loss of power dominates the clinical aspect, and when this is so, involuntary movements may be inconspicuous or absent (paralytic chorea, chorea mollis).

(1) *Involuntary movements*.—These are always irregular as regards time and as regards the nature of the movement.



Similar movements are never repeated successively in the same part, and each choreic movement may be said to differ from the one which preceded it and from its successor. Such movement begins suddenly and ends rapidly. Frequently one sees the involuntary movement complicated by the addition of a voluntary movement to cover the fault, as it were. The majority of the movements are complicated movements, involving several muscles and often more than one joint. They resemble volitional movements more closely than do the other involuntary movements met with in nervous diseases, and have been called "*quasi-purposive*" movements. So close is this resemblance that they appear to the parent of the child as voluntary movements, and she is often punished for "fidgeting." The similarity also suggests to the pathologist, that the seat of the disease responsible for such movements must surely be in the neighbourhood of the cortical centres of the motor area from whence the incitations for volitional movements finally emerge.

The complicated movements of chorea are much more rapid than those of athetosis, but they are not so rapid as are the involuntary movements of habit spasm and of the convulsive tics, and they are much less rapid than the shock-like movements of myoclonus.

If, however, a case of chorea be closely watched, with the skin uncovered, much less complicated movements are frequently seen, the simplest of which may involve one muscle only and may be indistinguishable from a myoclonic movement. The nature of the movements varies according to the region in which they occur.

In the face the movements are never confined to one side, even in hemichorea. The slightest movements, such as are seen in the early days of the disease, take the form of asymmetrical twitches about the angles of the mouth and orbits and in the lips. In more severe cases the strangest grimaces may occur. At one moment the angles of the mouth are drawn downwards, then outwards, then the lips are pursed. The forehead is thrown into wrinkles, the eyebrows are brought together, then are released, and the eyelids blink.

Suddenly the face becomes still in the most forlorn expression, to break into a smile or an ugly grimace a moment later. The tongue is thrust into one cheek, then projected, to be rapidly withdrawn just in time to escape the sudden snap of the open mouth. Often when asked to show the tongue the child puts it out rapidly and holds it there by closing the teeth on it. Smacking of the tongue and palate may often be heard at a distance. Lateral movements of the jaws are common. In more severe cases speech is difficult, the words being articulated slowly and in monosyllables.

Sometimes, after standing speechless for a moment, the child takes advantage of a moment of calm to get out a sentence as rapidly as possible. Swallowing is difficult or impossible in severe cases, as no effective sequence of voluntary movements can be produced in the jaw and tongue, and both palate and pharynx are involved in the movements; for this reason nasal feeding is sometimes essential.

The ocular muscles participate in the involuntary movements in that they remain parallel and turn concomitantly when involuntary movements of the head occur.

In rare cases momentary strabismus and diplopia occur. Reading is often difficult. The pupils are generally large.

In the upper extremities the movements always appear first in the hand. The thumb is more restless than the fingers, which are spread and pressed together, flexed and extended alternately. The wrists twist about irregularly, the forearms are constantly agitated with movements of pronation and supination, flexion and extension; while all possible movements of the shoulder occur. Any combination of these movements in any order of succession may be met with. The clumsiness of voluntary movements which the occurrence of such involuntary movements must entail is easily conceived, and is admirably described by Sydenham, who pictures a choreic patient thus: "If a cup of drink be put into his hand, he represents a thousand gestures, like jugglers, before he brings it to his mouth; for whereas he cannot carry it to his mouth in a right line, his hand being drawn hither and thither by the convulsion, he turns it

often about for some time, till at length, happily reaching his lips, he flings it suddenly into his mouth and drinks it greedily as if the poor wretch designed only to make sport."

The lower extremities are less severely affected with involuntary movements than the rest of the body. Such movements are best seen when the child is lying down. The gait is generally affected and tends to be clumsy and insecure, the legs being thrown now too far out, now across one another, while in severe cases walking becomes impossible.

Irregularity of respiratory movements are conspicuous, and spasmodic movements occur. The breath is often taken in rapidly and held for some time, then let go with a loud sigh.

The trunk is always involved to some extent, and movements of a writhing nature are characteristic. In severe cases violent movements of flexion, extension, and lateral flexion occur.

The face is usually the earliest region to present movements, and it is always affected bilaterally. The hand is the next to follow, and the left hand is affected earlier and more often than is the right. The movement may spread to the leg of the same side first, or to the opposite arm. So far as the limbs are concerned, the movements may be confined to one side—hemichorea; but the involvement of the trunk is always bilateral and generally equal upon the two sides.

Choreic movements, after being confined for some time to one side, may involve the opposite side, and the movements of the originally affected side may cease (double hemichorea) or they may persist, general chorea resulting. Hemichorea is more often met with upon the left side than upon the right, and when thus confined to one half of the body the degree of affection is never severe. In severe cases the whole of the voluntary musculature is involved and the movements are incessant. The violence of the movements of the limbs may cause the skin over the prominences to ulcerate from friction against the clothing, and the head and limbs may be badly bruised and cut from the violence of their contact

with adjacent objects, and unless the patient be properly protected wounds may occur which are liable to infection, with such grave consequences as abscess, erysipelas, pyæmia, etc.

The violence of the movements of the trunk is sometimes sufficient to throw the patient out of bed, and the teeth may be broken by the sudden closure of the jaws. The movements of chorea cease during sleep—whether this be natural or artificially induced—except in the most severe cases. In the majority of such cases, however, sleep is impossible; if it occur, the movements are much diminished for the time.

Frequently the patient is much quieter for some time after sound sleep. Mental excitement and the attempt to perform movements increase the spasms, while psychical and physical repose diminish them. There is no doubt, however, that in many of the less severe cases the spasms may be controlled by voluntary attention and also by acts requiring a concentration of the attention, such as writing.

(2) *Loss of power.*—The muscular power, as measured by the dynamometer, may be normal in slight cases; usually a decrease of power upon both sides is noticeable, and this is most obvious in those limbs in which the choreic movements are most marked. More severe degrees of paresis not infrequently accompany or succeed the appearance of spasm, and such paresis is usually localised in order of frequency as follows; brachial paresis, hemiparesis, and paraparesis. The paresis begins to be noticeable a short time after the commencement of the choreic movement.

It may be observed in one limb, or upon one side of the body, that the choreic movements are becoming progressively less marked, and that the limb or limbs are becoming progressively weaker. Soon the arm hangs loosely by the side, and the limb is dragged in walking. The paralysis does not become absolute, and in almost all cases slight choreic movement persists in the paretic region.

Often the mono-paresis, or hemiparesis, corresponds with a hemichorea; but they are met with equally in general chorea, and they may be severe in degree in cases in which the choreic movements are very slight.

Choreic paresis is apt to return with successive attacks of chorea, but not always in the same region; for in one attack monoparesis may exist, in another, paraparesis may be present, while in a third hemiparesis may develop.

It sometimes happens that paresis appears late and after the choreic movements have ceased, and it may persist for a considerable time. While paresis and choreic movements usually disappear together, the former may persist even in considerable degree long after the spasms have ceased. In rare cases loss of power constitutes the first noticeable symptom, and the paresis lessens as the spasmodic movements develop.

Under the name of chorea mollis (limp chorea), a more severe degree of paralysis affecting the whole body is described. It may be preceded by the usual symptoms of chorea. More often the paralysis is the first noticeable phenomenon, and it develops rapidly in from twenty-four to forty-eight hours. This form is characterised by complete flaccidity of the limbs; the child lies upon its back and does not move, and, if one of the limbs be raised from the bed and released, it falls limp and lifeless. The head is no longer held in a natural position, but rolls round on to the ear. Careful investigation rarely fails to reveal, however, some slight choreic movement, either in the face or the fingers. Many French authors state that the reflexes are invariably abolished in this condition and that the paralysis may be almost absolute, but English writers hold that the deep reflexes are not consistently absent, and that severe paresis is the limit of the loss of power. The writer, among a considerable number of cases, has found the deep reflexes present in the majority, and though the appearance of the limbs in some cases has been lifeless indeed, yet some voluntary movement has been possible in every muscle, though no effective act could be performed.

Paretic chorea and chorea mollis run a benign course, and recovery is said to be almost invariable.

(3) *Inco-ordination of voluntary movements* may be the first symptom of chorea to draw attention and it may pre-

cede the appearance of spasmodic movements. It is very marked in some cases when the spasm is slight, and it is most noticeable in the movements of the hand and forearm, which lack precision. The motor centres do not obey the dictates of the will, so that when the patient tries to relax certain muscles for the completion of some act there is marked delay, which interferes greatly with the efficacy of the act. This interruption in the sequential flow of movements which make up an act, is responsible for the sudden dropping of objects which are being carried, and it is demonstrated also by overshooting the mark in the attempt to lay hold of or pick up some object.

The muscles may undergo some diminution in size as a part of the general wasting of muscular tissue, and a considerable degree of hypotonus is always present in parietic chorea and especially in chorea mollis. The electrical reactions sometimes show that irritability is increased and the anodal closure may produce a contraction with the same strength of current as the kathodal closure.

*Sensibility.*—Pain in the joints and down the limbs is frequently complained of. In some cases this may be a rheumatic manifestation, but as its presence seems sometimes to depend upon the severity of the spasmodic movements it is probably a result of the continued movements. In severe cases pains are experienced similar to those occurring in neurasthenia. Headache is common and it may be paroxysmal or continuous; it is apt to be a troublesome symptom when a considerable degree of anaemia is present.

Tenderness of the nerve-trunk has been described and may possibly result mechanically from the muscular spasm.

Blunting of all forms of sensibility exists in a considerable proportion of the more severe cases and it is found chiefly upon the hand and forearm. The defect consists in a slight diminution of perception and of localising faculty, and it is usually most marked in the limb showing the most severe spasm. Considering the difficulty which the impairment of attention, always present in a marked case of chorea, places upon the investigation of slight change of sensibility, it is

obvious that this sign is valueless for diagnostic purposes since it is often absent, and in many cases the psychological condition makes reliable examination impossible.

Hysterical disturbances of sensibility are, however, not rare in patients who have reached puberty and occur usually towards the end of the illness, sometimes after the movements have ceased. Hemianæsthesia is not uncommon and is usually accompanied by contraction of the visual fields, sometimes also by hemianosmia and diminution of hearing on the affected side. Analgesia and hyperæsthesia are occasionally present.

*Reflexes.*—The skin reflexes are generally normal, sometimes diminished, and where hemianæsthesia is present they may not be obtained on the anæsthetic side. The deep reflexes are usually normal in slight cases. The knee-jerks often show an alteration in character which is peculiar to chorea, the contraction of the quadriceps is unduly sustained, and the leg remains at the top of its excursion for several tenths of a second. In severe cases the deep reflexes are often diminished, and in cases of parietic chorea and in chorea mollis they may be greatly diminished or absent in the parietic region. Loss of knee-jerk in cases of moderate severity, where paresis is not conspicuous, has been often referred to the administration of arsenic, but it is doubtful whether arsenic can cause abolition of the deep reflexes without other signs of arsenical neuritis appearing, such as pain, paralysis, etc. It seems certain that the knee-jerk may disappear in cases of non-parietic chorea of moderate severity. In one patient under the writer's care the knee-jerk was found absent in each of several recurrences and returned as the patient became convalescent; in this case arsenic was not used. The sphincters are not affected, but in severe cases, in chorea mollis, and where there is great mental hebetude, the calls of nature are neglected.

(4) *Emotional instability and other psychological disturbances.*—Some degree of emotional instability, failure of attention, and depression, is present in many cases, and for the most part in proportion to the severity of the affection. The

child's behaviour may change. She laughs or weeps without reason. She may become capricious, irritable, and obstinate, and in many ways troublesome. The memory may be impaired. She takes less interest in her surroundings, and this may progress until marked hebetude exists.

Such mental disorders appear and disappear with the chorea, leaving no subsequent trace of their existence.

Severe mental disorders, however, may complicate chorea, and it is argued that they are not dependent directly upon the chorea, but are the result either of intoxication (rheumatic or other infection), or that they are the outcome of the neuropathic heredity which is so often present. The severity of these symptoms in some slight cases of chorea and their entire absence in many of the severe cases support the latter view.

(a) Delirium may occur in acute and grave cases. It is usually violent and loquacious, and resembles other forms of toxic delirium. After its appearance the chorea becomes rapidly worse, and convulsions, followed by coma and death, may ensue.

(b) Hallucinations of vision of a terrifying character are met with only in children from the age of twelve years onwards. They disappear with the chorea.

(c) *Mania*.—This condition is quite exceptional in children, and is practically confined to the chorea of adolescents and adults. The subjects of choreic mania frequently have a direct psychopathic heredity. The form of the aberration may be acute mania, melancholia, moral perversion, or delusions of persecution. Such mental disorders usually appear and disappear with the chorea, and the prognosis as regards mental recovery is good.

*Cardiac symptoms*.—The pulse is usually increased in frequency and especially when the spasmodic movements are severe, and it may be irregular both in force and frequency, and intermittency may be present. Such irregularity has been attributed to choreic spasm of the heart muscle, but there can be no doubt that it is simply the physiological result of irregularity of the respiratory movements upon the



heart. Posture has less effect upon the pulse-rate than in health, the heart beating at nearly the same rate in the upright and in the recumbent position of the body.

Murmurs having all the characters of true hæmic murmurs may be met with in cases in which anæmia is present. They are heard at the base and tend to be conducted along the great vessels of the neck. A venous hum is also present in some cases.

Frequently murmurs are heard which are not referable to anæmia, and which we have no reason to attribute to organic disease of the heart. They are soft in quality and mid-systolic in time, and are heard at the base. They are apt to vary from time to time, sometimes without apparent cause, sometimes as the result of respiratory movement or change of position. Similar murmurs are met with frequently in rheumatism, and sometimes in other toxic conditions. It is possible that they may be the result of the action of some toxic agent upon the cardiac muscle. Such murmurs are very rare in young children, but are frequently met with in children from the age of 8 years onwards.

Severe cases of chorea are very often complicated with endocarditis and myocarditis, and sometimes with pericarditis. Among 73 cases of fatal chorea, Osler found organic disease in 64, endocarditis alone in 43 cases, endocarditis with pericarditis in 19 cases, and pericarditis alone in 2 cases. Sturges found organic heart disease in 75 of 80 fatal cases, and Sir William Gowers states that valvular disease is found in 90 per cent. of all fatal cases.

The signs of an organic cardiac lesion may be present before the attack of chorea, and they are then referable to an antecedent attack of chorea or of rheumatism. More often a murmur indicative of organic disease appears during the attack, and persists after the patient is convalescent. In other cases no murmur is detected throughout the attack, but signs of organic cardiac disease develop soon afterwards. The most common murmur is heard over the mitral area and results from disease of the mitral valve. It is systolic in time. A presystolic murmur may develop when signs of

endocarditis have been present for some time, and when sufficient time has elapsed for cicatrising processes to narrow the mitral orifice, but when a presystolic murmur is heard early in an attack of chorea, endocarditis has certainly occurred antecedent to the attack. Murmurs indicative of disease of the aortic valves are much less frequent, and were found twice only among 250 cases recorded by Sir William Gowers.

Some degree of myocarditis and dilatation are almost invariably present when endocarditis or pericarditis exists. It must be remembered, however, that endocarditis may give practically no sign of its existence, and many cases of chorea have been recorded in which no murmur was present during life, yet the autopsy revealed endocarditis.

*Cutaneous affections.*—The morbid conditions of the skin met with in chorea are the cutaneous manifestations of rheumatism, erythema nodosum, purpura, and rheumatic nodules. In some cases olive-coloured pigmentation of the skin, especially of the trunk, has resulted from the prolonged administration of arsenic, and herpes zoster has also occurred, presumably from the same cause.

*Urine.*—In proportion to the severity of the spasmodic movements, excess of urea may be found and uric acid may be deposited in considerable quantity. Dr. Handfield Jones has described a similar increase of phosphates. Urohæmatoporphyrin, a pigment which occurs in the urine of rheumatic subjects, was found by Dr. A. E. Garrod in fourteen out of twenty cases. Albumen is rarely present except when nephritis exists. In very rare cases glycosuria has been noted.

*Temperature.*—In uncomplicated chorea there is usually no pyrexia, but in some severe cases, and in the acute mania of chorea, the temperature may be raised as high as 104° F. Rheumatic complications (arthritis and endocarditis) are common causes of elevation of temperature. Hyperpyrexia is a very rare event. The effect of the advent of fever upon the spasmodic movement varies in different cases. Most often the movements are diminished

or cease during the fever, while in other cases there is temporary increase of the movements, followed by rapid diminution and cessation.

*Course and duration.*—The disease tends to a spontaneous termination after a variable time. It usually lasts from six weeks to six months. The duration rarely falls short of the earlier period and very frequently exceeds the longer period. The average duration of recent cases treated in hospital has been found by several observers to be ten weeks. Cases which last for more than twelve months are not rare, and slight cases with remissions, in the intervals between which there is not complete absence of symptoms, may last several years. Rarely chorea may become chronic and persist indefinitely. Some of the cases of persistent chorea, after many years, develop signs of pyramidal degeneration—a fact of great interest and significance in the pathology of this disease.

The *course* of the malady is that, after a gradual development of symptoms, there is a stationary period, during which symptoms are well marked, followed by a period of gradual diminution. Almost unnoticeably the disorderly movements become less intense, walking becomes possible, and the legs regain their normal motility, then the upper limbs, and lastly the face. The last of the spasmodic movements are seen as very slight abnormal movements of the fingers or of the face. Inco-ordination and paresis usually disappear before the spasm.

The evolution of chorea may, however, be very variable, exacerbations and remissions of the symptoms alternating.

Death may occur from exhaustion following upon the severity of the movements and the difficulty of nourishing the patient. In chorea mollis, asthenia may bring about a fatal result. The complications of chorea responsible for fatal results are endocarditis and myocarditis, hyperpyrexia, pyæmia, and intercurrent diseases.

The proportion of fatal cases occurring in chorea is probably under 2 per cent. Death occurs most often in first attacks, when these occur about the age of puberty, and

is a very uncommon event in young children and in recurrent chorea.

*Sequela.*—In very rare cases muscular weakness may succeed chorea. It may be general or local, and the loss of power is sometimes very great. There is no change in the muscular irritability but there may be blunting of sensibility. Sir William Gowers suggests that post-choreic paralysis is analogous to the mental weakness which may succeed chorea. It is always transient.

Sudden starts in the limbs similar to those which many people experience on going to sleep may occur and persist for some time after the chorea has ceased. As a rule they follow in cases in which mental disturbance has accompanied the chorea.

Persistent chorea and the development of general spasticity have already been referred to.

*Recurrence.*—About one third of all persons who have suffered with chorea have more than one attack. Osler found among 410 cases that two attacks had occurred in 110, three in 35, four in 10, and six in 3 cases. Females are more prone to a recurrence than males in about the same proportion as they are more liable to original attacks. In many cases no cause for the recurrence can be detected, but sometimes fright and other emotional disturbances and depressing conditions are responsible.

The interval between the termination of the first attack and the recurrence is rarely less than two months or more than two years. Recurrence within a few weeks of an attack is to be regarded as a relapse of the original attack rather than as a recurrence.

The average interval is about one year. If, therefore, a patient has remained well for eighteen months it is improbable that a recurrence will take place.

The heart is more frequently found to be affected in recurrences than in primary attacks, for the reason that, as endocarditis tends to occur in chorea, the greater the number of choreic attacks that have occurred the more likely is the heart to be found affected. The clinical aspect

of recurrent chorea is similar to that of the primary attack, but the symptoms are usually less severe and their duration is shorter. Occasionally, however, a recurrence is much more severe than the original attack.

*Etiology: race.*—The disease is much more prevalent among the white races. It is very rare among negroes and is said to be unknown among the American Indians, but half-breeds of both races are not infrequently the subjects of chorea. It is of common occurrence in the Hebrew race.

*Heredity.*—A neuropathic heredity in the form of hysteria, epilepsy, and insanity is not uncommon. Of much greater importance is a hereditary tendency to rheumatic affections. The incidence in families where both rheumatic and neuropathic heredity occur is often noteworthy. The subjects of chorea do not appear to be subsequently prone to the occurrence of other nervous maladies.

*Age.*—Chorea is perhaps never met with in children under the age of 3 years, and it occurs very rarely before the fifth year. The first attack occurs in the great majority of cases between the ages of 5 and 10 years, after which, although second attacks are relatively common, primary attacks become increasingly rare as age advances. After the twentieth year the disease is almost confined to females. If relapses be included, chorea is most often met with in the thirteenth year. After the twentieth year only 5 per cent. of all cases have their commencement. Instances of the occurrence of true chorea are, however, met with at any period of life from the third decade to old age. A man aged 58 years was recently a patient at the National Hospital with a mild attack of chorea. He had had two attacks of acute rheumatism and one other attack of chorea after the age of 50. The chorea, which was in every respect typical, was rapidly cured.

It appears probable that some of the cases reported as chorea occurring in children under the age of 5 years have been in reality cases of choreic diplegia resulting from structural disease of the brain, and that some of the cases reported as occurring after the age of 40 have been

either cases of Huntington's disease, or cases of organic cerebral disease of which the choreic movements have occurred as symptoms.

*Sex.*—Girls are affected three times as frequently as are boys. The disease is incident upon the largest number of boys in the years from 5 to 10, and upon the largest number of girls in the years from 10 to 15. As the third decade is approached this proportion becomes greater, and in that decade the incidence is almost exclusively upon women.

About three quarters of all cases are from the lower classes; the upper classes are not so often affected. Weakly, nervous, and excitable children tend to be picked out by the disease.

*Climate, season, locality.*—Climate does not appear to influence the occurrence of the disease, but there seems to be a distinct relation to the time of the year. The researches of Morris Lewis in Philadelphia have shown a maximum incidence in March. In London, however, July and August are the months for the greatest occurrence, both of primary and second attacks.

Chorea is more prevalent in towns than in the country. The presence of a large Hebrew element in the population in some towns, such as London, must, however, be taken into account, lest racial incidence be attributed to town dwelling.

*Rheumatism.*—The association of rheumatic manifestations with chorea as antecedent, concomitant, or subsequent events, is of very frequent occurrence. Conclusive evidence of this connection was first published by G. Sée in 1850. The opinion of H. Roger, put forward in 1867, that acute rheumatism, cardiopathy, and chorea, were several manifestations of one and the same disease has much evidence to support it, and in the light of recent pathological investigations, which have shown that a micrococcus can be isolated from the joints of acute rheumatism and can be cultivated, is rendered almost certain. As this when injected into certain animals produces acute arthritis, endocarditis, and chorea, it may be stated that chorea is frequently a nervous manifestation of rheumatism.

The history of a choreic patient frequently brings to light the occurrence of acute articular rheumatism, of cardiac disease, and of other rheumatic signs among the other members of the family. Often the patient has suffered with rheumatic erythema, purpura, rheumatic nodules, and growing pains before the appearance of the chorea; less often an attack of acute rheumatism or cardiac disease has occurred. Signs of rheumatism not infrequently are present during the choreic attack, and of these endocarditis and pericarditis are the most often seen; nodules, erythema, and purpura are not uncommon, but the occurrence of acute articular rheumatism during the attack is unusual. A large percentage of those patients who have never shown rheumatic manifestations before or during the attack of chorea subsequently develop rheumatic symptoms. Among 115 children suffering with chorea who were treated at the Hospital for Sick Children, Dr. Batten found that rheumatism had occurred in 32 per cent. Three years later he followed up those cases that had not suffered with rheumatism previously to the chorea, and among those, notwithstanding that he was unable to trace some of the cases, 113 per cent. of the total number had developed rheumatism in the three years following the chorea. Continuing his investigation three years later, he found that a further 97 per cent. of the total had developed rheumatism within six years of the chorea.

Since the association of rheumatism is so often subsequent to the occurrence of the chorea, it follows that the statistics, which have all been compiled from cases at the time of occurrence of the chorea, must considerably underestimate the proportion of choreic patients who are the subjects of rheumatism.

The Collective Investigation Committee of the British Medical Association found that rheumatism had preceded the chorea in 26 per cent., while in 32 per cent. the rheumatism accompanied the chorea, or occurred subsequently, and to those numbers 14 per cent. must be added if patients who had been the subjects of vague rheumatic pains were

included. If to this total of 72 per cent. the patients be added who have shown no rheumatic manifestation, but whose family history reveals the occurrence of definite rheumatism among near relations, the percentage of cases of chorea which are associated with rheumatism becomes too high to allow of any explanation but that chorea, in a great majority of cases, is a manifestation of rheumatic infection.

*Infectious diseases.*—Almost all of these diseases may have the relation to chorea of indirect exciting causes of the attack. The debilitating effects of whooping-cough upon children make it a frequent antecedent in the history of chorea. Ross found that chorea frequently occurs after scarlet fever; but, as the result of detailed investigation, it appears that the relation of chorea is with rheumatism, itself a frequent concomitant of scarlet fever, and that chorea is not especially incident upon those who have suffered with scarlet fever and have at no time presented symptoms of rheumatism. Measles, variola, diphtheria, pyæmia, and gonorrhœa have all been recorded as direct antecedents of chorea. Féré has pointed out the occurrence of the disease in the course of secondary syphilis. Doubtless, both the bodily and psychological effects of this disease may act as exciting causes of chorea in a predisposed subject.

*Psychical disturbances.*—While these are absent in many of the cases, yet there is no doubt that emotional disturbances are often responsible for the appearance of symptoms as immediate determining factors. The association of St. Vitus' dance with fright is so impressed upon the popular mind that any possible event of this nature shortly prior to the chorea is at once placed in a perhaps unduly important relation to the disease. Sometimes careful investigation will discover that slight choreic movements were present before the shock happened, and that they became more intense afterwards. It is not common for the movements to manifest themselves directly after the emotional disturbance. They usually appear after an interval of from



one day to a week—rarely later. In this connection it is interesting to bear in mind the important relation of sudden emotion to the occurrence of the first fit in epileptics.

*School life.*—It seems only in rare instances that over-pressure in schools is responsible for the occurrence of the attack. No doubt it frequently aggravates it. The anxiety of pending examinations and the emotions produced by punishment at school may, and no doubt often do, aggravate chorea, as would similar psychical disturbances in other circumstances.

*Pregnancy.*—Chorea is apt to occur during pregnancy in women who have not had a previous attack and when there is no other cause attributable. It is met with in first pregnancies, rarely for the first time in second pregnancies, or after the age of twenty-four years. The chorea may appear at any period of the pregnancy, but most often in the third month. Some of the cases prove fatal, and there is a tendency to the development of acute mania in others.

The frequency with which unmarried girls are found among the subjects of the chorea of pregnancy suggests strongly that the attendant mental anxiety and depression play an important part as exciting causes.

Among other conditions which are of interest as causal factors of chorea may be mentioned *iodoform poisoning*, an example of which is recorded by Denme; the application of this drug to a tuberculous wound was followed by the appearance of chorea, which ceased when the drug was suspended and reappeared upon its resumption.

*Errors of refraction* and reflex irritation from intestinal parasites, gastric disturbance, etc., may perhaps act as adjuvant factors. Hysteria is probably not in an important causal relation with the chorea of children. As regards the development of the disease by imitation it is doubtful if true chorea is ever produced in this way, but a hysterical condition closely simulating the malady may certainly arise. The development of chorea in several members of a family successively is often cited as an instance of imitation. The probable explanation is the development of a disease among

subjects hereditarily liable, under the influence of the same exciting cause, acting upon all.

There are other morbid conditions which are met with in connection with chorea so frequently as to deserve attention. *Anæmic states* often precede and accompany chorea. It is highly probable that they are expressions of the rheumatic state and sometimes also of the nutritional failure which is in some cases the determining cause of the choreic attack. Tuberculosis and rickets have been put forward as important predisposing causes, but it is probable that they have in reality no causal relation with chorea. Hysterical manifestations sometimes precede but more often succeed the occurrence of chorea, and in young women neurasthenic symptoms preceding chorea are not rare. The occurrence at any time of epilepsy in choreic patients is very rare.

To sum up the etiological factors of chorea, neuropathic and rheumatic predispositions, hereditary or acquired, constitute the soil in which the malady appears, while rheumatism, infectious diseases, any condition of nutritional depression, and nervous shocks are the immediate determining causes.

*Pathology and morbid anatomy.*—The primary seat of disease in chorea is probably in the motor structures of the hemispheres, and especially the cortex of the Rolandic area, and it is probable that the morbid process is confined to this part of the nervous system. The *quasi-volitional* nature of choreic movements and their association with paresis, the occasional hemiplegic distribution of the spasmodic movements of the paresis, the greater involvement of the arm than the leg, and the cessation of the movements during sleep, suggest a cerebral location. The frequency with which attacks are excited by emotional disturbances, the mental state during the attack, and the occasional association of acute mania, also suggest that the highest parts of the nervous system are involved.

The facts that the manifestations of chorea may be limited to one limb or to one side of the body and that when commencing locally the movements spread to other parts of the

body in the same order as do convulsions which commence locally, and as does also paralysis resulting from an increasing gross lesion of the cortex, suggest strongly that in these cases the *materies morbi* is at first situated in a limited area of the cerebral cortex and that it spreads directly to neighbouring regions of the cortex, such extension being expressed by the spreading of the spasmodic movements from their place of commencement. If chorea were always the result of a general blood state, or of a toxic agent acting generally upon the whole cerebral cortex, it would seem scarcely possible that involvement of the cerebral cortex in a regular order could occur, the order being that of the anatomical localisation of function. It must be remembered, however, that chorea does not in the majority of cases commence locally and spread regularly, but it may be general from its commencement, and it is quite likely that subsequent investigation may determine an initial locally acting cause in the one class of cases and a generally acting cause in the other.

Much light is thrown upon the question of the seat of the disease by the investigation of those cases of gross organic disease of the brain in which choreic movements occur. It has been pointed out by many authors that choreic movements have been associated with lesions in almost every part of the brain, and that such movements are not constantly associated with a lesion in any one position. It is an interesting fact that epilepsy is prone to occur in cerebral lesions situated no matter where and that it is not constantly associated with lesions in any particular position.

It is probable that the appearance of both epilepsy and choreic movements as symptoms of gross cerebral disease is dependent, not only upon the nature and situation of the lesion, but upon the functional condition and stability of those elements which are not involved by the lesion, and upon the indirect effect of the lesion upon the latter. When choreic movements occur from a destructive lesion of the cerebrum—such, for instance, as a softening in the region of

the basal ganglia or of the optic thalamus—it is obvious that the movements must result from the physiological activity of those elements which have escaped destruction. Such movements, however, never occur if the cortex of the central region be destroyed, though, as has been already stated, they may occur wherever the lesion may be, provided the motor cortex is relatively intact.

By far the most common cerebral disease in which choreic movements may appear is atrophic sclerosis (cerebral diplegia). In this condition there is a widely spread degeneration of the cells of the cerebral cortex, but except in the most severe cases the cell atrophy involves some of the cells only. There is a thinning out of the cortical cells, and the various forms of perverse and spontaneous movements which occur in this disease are the expression of the irregular activity of the cells which remain.

Choreic diplegia is symptomatic of a condition of atrophic sclerosis in which the loss of cells is not very great.

Some explanation is necessary to account for the occurrence of choreic movements in cases in which lesions of the crus cerebri, of the thalamus, of the corpora striata, and of the anterior and posterior cortex have existed. The nervous system functions as a whole and the individual elements of which it is composed act together as the units of an army arranged in a set pattern, all being mutually interdependent. The removal of some of the elements or the severance of their interconnections lessens both the stability and the functional capacity of the whole army, and of one division perhaps much more than another. When the loss of elements becomes so great that compensation is impossible, signs of disordered action appear. Again, if as the result of heredity or some other cause, there is a predisposition in the nervous elements to spontaneous and irregular action, the decrease of general stability which results from the loss of some of the elements may cause spontaneous and irregular action to be manifest.

Many different morbid conditions of the brain have been recorded in fatal cases of chorea, such as local meningitis,

encephalitis, vascular lesions, etc. These may be dismissed as the results of complications, and, in the majority of cases, no microscopic abnormalities are to be found. Jakoiwenko and others have described special small bodies in the neighbourhood of the small vessels, which have been called "chorea corpuscles." They are small oval bodies and present a central dark area, which stains deeply with fuchsin, surrounded by a translucent peripheral part, and, according to these authors, they are found all over the nervous system and especially in the globus pallidus of the lenticular nucleus. They are, probably, the results of hyaline degeneration in wandering cells and are not peculiar to chorea.

Alteration in the appearance of the cells of the cortex and of the thalamus has been described, especially swelling, distortion and tigrolysis; but the majority of recent observers have found no abnormality in these structures. It may be also noted that, in functional disorders of any kind, abnormalities of the nerve-cells are conspicuous by their absence, and, further, that most marked changes in the tigroid substance of the nerve-cells are known to result from over-action of the nerve-cells, and from the effects of toxic agents. Moreover, conclusions drawn from such abnormal appearances of nerve-cells as to their functional capacity are of doubtful value.

Perivascular infiltration and small vascular lesions, such as capillary embolism and hæmorrhage, have often been found present, but they are not constant. Similar lesions are the rule in subjects who have died of any infective disease, and this fact is of some importance in connection with rheumatic infection as a cause of chorea.

Most important researches have recently been carried out upon the bacteriology of chorea. Pianese has isolated both a bacillus and a coccus from the central nervous organs, the former of which produced convulsions when inoculated into animals. Many other observers have isolated staphylococci from the central nervous system. Richter and Tribulet found a coccus in the blood, Dana a diplo-

cocci in the meninges. Apert isolated a diplococcus from the blood, but inoculation of this organism gave negative results. Drs. Poynton and Paine have isolated a diplococcus from the arthritic effusion of acute rheumatism, which, when injected into rabbits, produced polyarthritis, endocarditis, and choreic movements. Masses of the rheumatic diplococci were found in the neighbourhood of the small vessels in the cerebral cortex.

In searching for the essential causes of chorea it must be remembered that the symptoms are the expressions of the disordered action of the nerve-cells and that such disordered action may result from an inherent condition of functional instability of the nerve elements, or from the deleterious effect upon the nerve elements of locally acting micro-organisms or generally acting blood states, and further, that both of the causes may act concomitantly.

The importance of functional instability of the nervous elements as a cause of chorea is shown by the frequent neuropathic heredity, and by the age and sex incidence of the disease, for it occurs after the functional development of the nerve centres has been effected, but before functional stability is complete, and while the motor processes are still prone to become active independently of volition. The incidence of the disease is greatest at that age and in that sex in which emotion is most active, and in which the stability of nervous elements is most easily influenced by emotional disturbances, which as exciting causes of chorea are to be traced with far greater frequency than are all other exciting causes. It is interesting in this connection to call to mind Sturges' observation that the physiological expression of emotion is often by movements which bear no distant resemblance to chorea, and the shy and nervous child in the presence of a stranger frequently shows movements almost choreic.

It has been already stated that in hysteria a clinical aspect may occur which is almost indistinguishable, in some cases perhaps quite indistinguishable, from chorea, and it is quite probable that some cases of true chorea, where neither

rheumatic manifestations nor other causes are traceable, are examples of purely functional disturbance.

The state of disordered action, whatever be the essential cause, sometimes becomes ineradicably impressed upon the nerve elements, and chronic chorea results.

There seems to be little doubt that in a large proportion of cases chorea is the result of an infective process, and perhaps the nature of the infection is not always the same. That rheumatism is dependent upon a specific organism is proved, and the specific organism when inoculated into animals has produced arthritis, endocarditis, and movements like those of chorea. The frequent association of chorea and rheumatism in the same subject in the light of these recent researches upon the bacteriology of rheumatism suggest strongly that when associated, these diseases are due to the presence of the same micro-organism, the appearance of chorea being determined by the condition of functional instability of the nervous elements.

The question arises, however, as to whether infections other than rheumatic may give rise to chorea, for many infectious diseases have been reported as the immediate precursors of chorea. It seems possible that future investigation will show that other infections may bear the same relation to chorea as does rheumatic infection. It must be remembered, however, that the association of chorea with rheumatic infection is far more frequent than is its association with all other known infections, taken collectively.

When chorea is the result of an infective process, do the nerve elements suffer as the result of a general toxic blood state, or do the organisms act locally upon the nerve elements? Many authorities, and among them Sir William Gowers, incline to the first of these alternatives, but it may be pointed out that the micro-organisms of rheumatism have been demonstrated in close proximity to the cells of the cerebral cortex, and the restriction of the spasmodic movements to one limb on one side of the body and their method of spreading make it extremely probable that in some cases

the virus acts locally. Possibly the action may be both local and general. What we know of the pathology of chorea may be briefly summed up thus: There is a functional derangement of the nerve-cells of the cerebral cortex, chiefly of the central region, and this is not accompanied by any perceptible primary morbid changes in the nerve elements nor in the other elements of the cerebral cortex. Functional instability of the nerve elements, both inherited and dependent upon age and sex, is a most important cause, and emotional disturbances are common excitants, and in some cases no other causes are present, the disease being purely functional. Infective processes, especially rheumatic infection, produce the disease as the result of a general toxic blood state, and by the local action of micro-organisms in the cerebral cortex—a view which approximates to the vascular theory of Dr. Hughlings Jackson, promulgated many years ago.

*Diagnosis.*—The recognition of the malady in its usual form presents no difficulty, for the character of the spontaneous movements at once suggests the diagnosis. Where such movements are inconspicuous and the clinical aspect is dominated by some other symptom, such as paralysis or acute mania, the nature of the disease is not so readily determined.

In the majority of cases of paretic chorea, the history of the existence of spontaneous movements before the development of the paralysis is forthcoming, and careful examination always reveals slight movements in the face or in the fingers, especially when the patient attempts a sustained act, such as keeping the mouth widely open or holding the hand out with the fingers spread.

In other cases the paresis is not preceded by spasmodic movements, and difficulty in diagnosis may arise, the more so since in such cases the paralysis is usually more severe.

The paresis of chorea, however, is in itself highly characteristic. It is of gradual onset; it is a flaccid paralysis which is never absolute; the arm is usually affected more severely than the leg, and the hand more than the shoulder.



The face generally escapes. There is no local muscular wasting, and no pain. While signs indicative of spasticity are absent, the deep reflexes are, in most cases, present. If the other causes of hemiplegia or monoplegia of gradual onset in children and adolescents be considered, gross cerebral lesions, such as tumour, abscess, and sclerosis, and gross spinal lesions will include all except hysteria.

Where cerebral lesions exist the resulting paralysis is a spastic one and other signs of intra-cranial disease (head-ache, vomiting, optic neuritis, convulsion, etc.) are present. Gradual paralysis of spinal origin is associated either with pain and spasticity or with muscular atrophy and absence of the deep reflexes. The difference between paralysis so arising and paresis from chorea is very striking, and confusion of the two should not arise. All causes of sudden hemiplegia or monoplegia are at once distinguishable, since the onset of choreic paralysis is always gradual. Moreover, in the paralytic form of chorea careful observation will in most cases discover some slight and typical involuntary movements. Once aware of the possible nature of the case, the diagnosis is not difficult, and the observation of Sir William Gowers should be borne in mind, that when a child between the age of 7 and 12 years is said to have gradually lost the use of one arm or of one side the disease is most often chorea. In the severe condition of chorea mollis, the clinical picture of complete helplessness with retention of slight voluntary movements in all muscles is highly characteristic and can hardly be mistaken.

Maniacal chorea may be mistaken for acute mania. Characteristic movements are, however, usually present, and the subjects are younger than most of the subjects of acute mania. There is less continuous garrulity than in acute mania, and not infrequently there is speechlessness. The history of antecedent involuntary movement must serve to identify those difficult cases in which all spasmodic movements cease when the mental disturbance becomes considerable.

Hysterical chorea may so closely resemble true chorea

that the greatest difficulty may arise in diagnosis. In some cases the departure of the spasmodic movements from the choreic type, and the occurrence of rhythmic movements may betray the presence of the great simulator of other diseases—hysteria. Again, paretic chorea may be mistaken for hysterical paralysis and especially since the development of the latter condition may be quite gradual. Hysterical paralysis, however, if flaccid is generally complete and both this condition and hysterical chorea are apt to be associated with characteristic stigmata of hysteria, such as hemianæsthesia, “stocking” and “glove” anaesthesia, amblyopia, functional attacks, etc.

Huntington's chorea, in which the involuntary movements are identical with those of chorea, is at once separated from the disease by the history of direct inheritance and by the age at which it occurs, and also by the psychical condition, which is one of smiling and complacent mental reduction.

Electrical chorea is known in Northern Italy only. The movements are sudden and shock like and there is associated rapid muscular wasting and pyrexia, and the aspect of the disease has nothing in common with chorea. Myoclonus is easily distinguished from chorea by the short shock-like and simple character of the movements. There is little difficulty in distinguishing conditions of organic disease in which choreic movements appear as symptoms. In cerebral diplegia to certain forms of which the name “congenital chorea” has been applied, the malady dates from birth or from the first three years of life—a period during which chorea is infinitely rare, if truly it ever occurs. Bilateral athetosis, diplegia with choreic movements, diplegia with myoclonic movements, and diplegia with intention tremors, are the clinical varieties which have been included under the term of congenital chorea. They are chronic maladies and are often progressive; moreover, with them are associated signs of grave organic disease, bilateral spasticity, contractures, paralysis, and recurring convulsions.

Similar grave signs of organic disease are present in

cases in which choreic movements occur as symptoms of cerebral tumour and vascular lesions of the brain.

*Prognosis.*—The prognosis in uncomplicated chorea in children is almost invariably good, and choreic paresis even in its severe form does not alter this outlook. The presence of signs of rheumatism, and especially of organic heart lesions, involves the prognosis of those conditions as apart from that of the chorea. Inclusive of these complications, the British Medical Association Investigation Committee found that of 437 cases only 2 per cent. were fatal. In the most severe cases of chorea, in which swallowing is impossible and the violence of the movements has produced many wounds, the prognosis is good if skilled treatment be obtainable and if there be no secondary infection. According to Anstey the malady is more serious after puberty, but in-patients treated at the National Hospital between the years 1888 and 1903 presented no peculiarity in this respect either as regards severity or duration of the illness. The chorea of pregnancy is more often fatal, and usually from the result of abortion, whether spontaneous or artificially induced.

Where pronounced psychological disturbance exists the prognosis is serious in proportion to the severity of the mental change, but many severe cases recover completely. The danger is that the condition may be followed by progressive and permanent mental deterioration.

Sir William Gowers, from an analysis of a large number of cases, finds that the more severe the attack the longer will be its probable duration; that a recurrence will probably terminate sooner than a first attack, and that etiological conditions of age, sex, preceding rheumatism or heart disease, and present endocarditis, afford no indication of the probable duration of the disease.

*Treatment.*—It is all-important in the treatment of chorea from the mildest to the most severe cases, that physical and mental tranquillity, with freedom from emotional disturbances, should be secured. The child should be at once removed from school and all lessons interrupted. She

must not be scolded for her faults, or subjected to the laughter and imitations of other children, and her surroundings should be made as light and happy as possible. It is well in every case to commence treatment with several days' absolute rest in bed, provided that such treatment can be carried out without entailing the loneliness and fretting which the enforced imprisonment is apt to produce. While a few days' rest in bed is advantageous, it is not advisable to prolong this for more than a week, unless some complication or the severity of the case necessitates enforced rest.

When absolute rest is considered inadvisable, or after it has been carried out, the ordinary period of rest should be prolonged. The child should rise late and go to bed early, and should rest for two hours in the early afternoon. A bright room, an interesting companion, and varied amusements during the hours of rest are very desirable, and isolation from other children during this treatment is advantageous. It is better to abandon enforced rest than to allow it to become irksome to the patient and result in mental depression—a condition above all things to be avoided in chorea. The child should be well clad in woollen garments, especially at night, when the spasmodic movements are liable to leave her uncovered while she sleeps. Tepid sponging or light massage are often usefully applied just before the child goes to bed, as they tend to promote a feeling of contentment and to induce sleep.

The engagement of a trained nurse is always to be recommended when circumstances permit. The introduction of a stranger need never be so abrupt as to produce emotional disturbance in the patient, and a tactful nurse can be on such terms with the child in a few hours that she may be given absolute charge without danger. In this connection it may be stated that it is very unusual to see a choreic child become definitely worse as the result of admission into hospital.

Improvement in the condition of bodily nutrition is to be aimed at in all cases of chorea. Among the London poor

the choreic children are mostly ill nourished, and the effect of a liberal supply of good and nutritious food upon the course of the disease is very striking. Often, however, the appetite is capricious, and the child being allowed to eat only what she likes, is consequently ill fed. It should be impressed upon those who have care of the child that the ordinary diet must be supplemented with as much milk and farinaceous food as she will take.

Any possible causes of ill health should be sought for and remedied, such as errors of refraction, dental irritation, intestinal parasites, constipation, phimosis, etc.

In the hygienic and dietetic treatment of chorea are embodied the most important weapons at our disposal for combating the disease, and in the slighter cases these remedies are sufficient alone.

Severe cases, in which the movements are violent, call for skilled attention, and a trained nurse is required both night and day. Provision has to be made that injury shall not occur from the violence of the movements and that the child shall be adequately fed. The patient should lie upon a water mattress if possible, placed upon a large guarded bed, the sides of which are everywhere protected by pillows, which must be fixed. When a cot is a necessity it is easy to pad all the ironwork with cotton-wool over which bandages are wound. An excellent bed in cases of urgency may be made by placing the mattress upon the floor in a corner of a room, the walls being protected by the two halves of a straw mattress placed upright and the two other sides surrounded by pillows. If the limbs are bruised and cut they should be covered with cotton-wool applied with a light bandage. When swallowing is difficult it is best at once to resort to nasal feeding, which rarely causes as much discomfort as the ineffectual and exhausting endeavours to take food with the spoon. A china feeding-cup must never be used, since from sudden closure of the teeth the spout may be broken off and swallowed, with a fatal result. An enamelled metal cup is safe. Since vomiting may follow nasal feeding if a full meal be given, it is advisable

to administer a smaller quantity at shorter intervals, and not more than three quarters of a pint should be given at one time to a child of 8 years.

The nostrils should be used alternately, and signs of irritation of the nasal passages should be treated with the local application of boracic lotion. Glycerine should never be used as a lubricant for the nasal tube, for it causes great pain and subsequent swelling of the nasal mucous membrane. The best lubricant is olive oil.

Sleeplessness is the rule in the class of cases under consideration, and, according to Dr. Hughlings Jackson, is the expression of the exhaustion which the constant movements and lack of nourishment bring about. Inquiry will generally show that if a child has not slept for three nights it has had no adequate nourishment for four days. In this sleeplessness of starvation sedatives such as morphia, chloral, and bromides are useless and may be harmful; but the administration of a proper meal by the nose combined with a full dose of alcohol is in most cases soon followed by sleep.

The use of drugs for sleeplessness in chorea is to be avoided, and is for the most part unnecessary. Indeed, these may do positive harm, and should be rigidly avoided in severe conditions. Chloral, however, is sometimes valuable in less severe cases where sleep is broken.

Warm and tepid baths and douches, applied regularly and in such a way as to be grateful to the patient and to cause no fright, are very useful adjuncts in the treatment of chorea.

Pyrexia when present requires the usual treatment in the form of tepid or cold sponging and tepid baths. Hyperpyrexia requires the cold pack and the cold bath.

The use of arsenic is followed by a marked beneficial effect in many cases of chorea. It is supposed to act by improving the general condition of nutrition, and especially the nutrition of the nervous tissues. There are, however, almost as many cases in which this drug seems to have no beneficial effect. It should be given in doses of from two to five minims of Fowler's solution, three times daily,

after meals. An increase of the dose to fifteen minims or more does not appear to be followed by such increased benefit as to render it justifiable. Large doses of arsenic (five to ten minims) should never be given; the danger of setting up neuritis is very great, and the writer has never seen any benefit from their administration, and has frequently seen neuritis as a result of such dosage.

The subcutaneous injection of arsenic has been recommended because much larger doses can be given without intolerance than can be given by the mouth. The advantage of large doses of arsenic has not been proved, and irrecoverable arsenical paralysis has resulted in several cases.

The value of antipyrin in chorea was first pointed out by Hübner, and it has been largely used on the Continent in doses gradually increased until two drachms in the twenty-four hours are taken. It is chiefly of service in uncomplicated cases of moderate severity. It should be at once discontinued if signs of intolerance, and especially if albuminuria, appear, but these events seem to be rare, and were not observed in the above-mentioned series of cases. Marked anemia, malnutrition, and active cardiac and rheumatic complications are contra-indications to the use of antipyrin.

Exalgin, emalgin, antifebrin, and many drugs of the same class, have been similarly used, but they have no advantages over antipyrin.

Salicylate of soda in large doses has been recently advocated by Dr. Lees, who has given us as much as two hundred grains daily with, it is said, beneficial results. The writer has little experience of this treatment. What little he has is distinctly against its efficacy, and in the hands of Leroux and others it was not followed by encouraging results.

Chloral is useful in many cases. In chronic and intractable forms of the disease Sir William Gairdner and Dr. Charlton Bastian have obtained good results. The patient is isolated, and with doses of from ten to fifteen grains, taken four times a day, remains continually asleep, the object being to obtain arrest of the spasmodic movements by prolonged sleep. This treatment is continued from four to six weeks. The tempo-

rary mental derangement which may follow this treatment is a strong contra-indication to its adoption.

Hyoscin, administered hypodermically, has proved a useful sedative. The writer has used it with excellent results in less acute cases which had proved intractable under other treatment. One hundredth of a grain of hyoscin hydrobromate was injected three times a day. As a rule the injection is followed immediately by wide dilatation of the pupils and slight flushing and sleep lasting from one to two hours. In acute cases, in cases in which endocarditis exists, and when the patient is badly nourished, hyoscin and other depressing drugs are contra-indicated. Indeed, in all such cases food and alcohol are the only drugs admissible.

The bromides are of little value.

Iron is useful when anemia exists. Other tonics, such as cod-liver oil, phosphate of lime, malt, etc., are of great value in weakly children. Strychnine and quinine are not of value in the acute stage, but may subsequently be given with advantage.

Electrical treatment has not been followed by good results, and it is apt to frighten children and so aggravate the disease. Counter-irritation to the spine is not beneficial and no benefits seem to have resulted from hypnotism. During convalescence massage is distinctly useful as an aid to general nutrition.



## ELECTRICAL CHOREA (DUBINI'S DISEASE)

Two diseases entirely distinct from one another are described by different authors under the name "electrical chorea." In England the term is restricted to a fatal and presumably infectious malady which is endemic in Northern Italy and which was described in 1846 by Dubini as "Chorea electrica." In France and Germany the term has been applied to a benign form of chorea or of hysterical chorea in which shock-like movements, approaching the type of the movements of habit spasm, are a marked feature.

Dubini's chorea seems never to have been met with except in Lombardy and adjacent parts of Piedmont, and even in these localities it is a rare disease. It affects both sexes equally and may occur at any age. Heredity seems to play no part in the causation. The onset of symptoms may be acute, subacute, or slow.

In acute cases the malady is ushered in by severe pains in the neck and back and the temperature is considerably raised. Irregular pyrexia of severity proportional to the acuteness of the symptoms is the rule throughout the course of the illness. Spasmodic movements are early symptoms and are first noticeable in the upper extremity. The lower extremity of the same side is soon affected and subsequently the spasm becomes general. The movements are sudden and shock-like and are often of wide range and powerful, resembling very closely the movement which results from the stimulation of a motor nerve by a single induction shock. Gradual loss of power occurs in the affected muscles and in the late stages of the disease paresis becomes severe and general.

The muscles waste progressively and lose their Faradic

excitability and the deep reflexes disappear. Epileptiform convulsions not infrequently occur, and are sometimes confined to one side of the body; they may be followed by transient paralysis. Progressive mental hebetude occurs.

The disease is probably the result of an acute infection, the incidence of the micro-organisms or their toxins being presumably upon the cerebral cortex. On account of a prejudice prevailing in the popular mind in Northern Italy but few autopsies upon the subjects of this disease have been performed. No constant morbid changes have been found in the central nervous system and the pathology of the condition is quite unknown.

Treatment seems to have no effect upon the course of the disease.

## HUNTINGTON'S CHOREA

ALTHOUGH this malady is unknown in children and is very rare in young adult life, yet certain of its features have such an important bearing upon the pathology of choreic movements in general that some mention of the disease is essential.

The symptoms make their appearance between the ages of 25 and 50 years, and in the great majority of cases the malady commences in the fourth decade of life. The sexes are equally affected. The affection is in most cases familial, rather less than half of the children of affected parents who survive the fourth decade of life escaping. Direct transmission from either parent to the child is invariable, for the malady does not skip one generation to appear in the next, and it may be transmitted through many generations. In two affected families the ancestors were affected as far back as history is obtainable—five generations in one family and four in the other.

Rheumatism and other diseases seem to play no part in the causation, nor do emotional disturbances seem provocative of its appearance.

The onset is gradual, and the first noticeable symptoms are movements similar to those seen in ordinary chorea. Commencing in the face and upper limbs, they slowly extend to the rest of the body and become more severe as they extend. While in the course of some years their intensity becomes considerable, yet it never reaches a severe degree as in Sydenham's chorea, and in some cases the movements remain slight throughout. Voluntary movement usually arrests the spasmodic movements

while excitement increases them, and they cease during sleep. Articulation is often much impaired by the spasm. Sensibility is normal, as is also reflex action.

Mental alteration is a marked feature of this disease. Progressive psychological reduction with happy complacency is the rule, but after the disease has existed some years many patients become insane. In several cases an attack of acute mania has ushered in the symptoms. In the late stages of the disease signs of general spasticity with exaggerated tendon jerks may be present. The symptoms continue to the end of life, which is only shortened by the malady through the indirect influence of the mental state conjoined with the exhausting effects of the spasm.

Recent investigations into the pathology of this disease have shown that a sclerosis of the cerebral cortex, and especially of the central region in small widely-scattered patches, is constantly present. Often, some thickening of the pia arachnoid is associated. It has been suggested that the sclerosis is due primarily to an overgrowth of the neuroglial elements, while other investigators have referred it to pathological vascular conditions. These views appear, however, improbable, and, considering the hereditary character of the disease, it is justifiable to consider hereditary chorea as an abiotrophic condition, and that the essential pathology is a slow degeneration of the cortical nerve elements.

The morbid anatomy of Huntington's chorea adds much to the probability that all choreic movements are the expressions of interference with the function of the cells in the grey matter of the motor cortex. We have seen that choreic movements may result from gross organic lesions of the brain—for example, tumour and atrophic sclerosis, which directly or indirectly interfere with the central cortex—without complete destruction of its elements in any region, or complete abrogation of their collective functions. Complete destruction of any part of the motor cortex is never associated with the presence of choreic movements in the corresponding part of the body.

In gross cerebral lesions, however, when choreic movements occur, other symptoms of damage to the motor cortex, such as paralysis and rigidity, also exist. In Huntington's chorea, where lesions of the motor cortex are constantly found,—viz. widespread degenerative changes in the cortical cells—the only expression of interference with the elements of the motor cortex is the choreic movement. Further, in rheumatic chorea, collections of the diplococcus of rheumatism have been demonstrated in the immediate neighbourhood of the small vessels of the motor region. These facts point, in unison with so many of the clinical characteristics of chorea, to the cerebral cortex as the seat of disease in those maladies in which choreic movements occur.

## MYOCLONUS

SYNONYMS : Paramyoclonus multiplex ; myokimia.

The characteristic symptom of this condition is the occurrence of sudden shock-like simple contractions of the muscles, which may vary in intensity from simple fibrillary twitching to contraction causing a violent movement of a limb. They are often symmetrical and may also be isochronous in corresponding muscles on the two sides of the body.

Attention was first drawn to this malady by Sir Russell Reynolds, who described a single case. Friedreich in 1881 described a second case and applied the term "paramyoclonus" to the affection. Ten years later Unverricht published a monograph upon the subject, using the term "myoclonus." Recently Sir William Gowers has written on the subject, and Biancone, under the name of "myokimia," has described several cases in which the clonic contractions were very small, hardly exceeding in range those movements of the muscles known as fibrillary twitchings.

*Symptoms.*—The spasmodic movements of myoclonus are simple sudden movements, and may exactly resemble the movement resulting from the application of a single induction shock to the nerve supplying a muscle. Each movement usually involves a single muscle only, and it may involve no more than a few fibres of the muscle, resembling then the fibrillary twitchings so characteristic of progressive muscular atrophy. In other cases many muscles may be implicated in some of the shock-like contractions, which may then be of so violent a character as to throw the patient to the ground.

When several muscles contract together in a myoclonic

movement, the distribution of the contraction is never determined by the nerve supply of the muscles, nor do the muscles contract according to their synergic association, but the single spasm involves a group of muscles which cannot possibly be thrown singly into contraction by volition.

Myoclonic movements show no regularity as regards rhythm or as regards range of the successive movements. Sometimes an isolated movement recurs at comparatively long intervals; in other cases periods of almost incessant myoclonic movements are separated by intervals of comparative quiet, or the spasms may occur constantly, now in greater and now in less degree, as many as forty clonic contractions of the same muscle appearing in one minute. When many contractions rapidly succeed one another in the same muscle a tendency to tonic contraction of the muscle is occasionally seen. The movements are very often strictly symmetrical upon the two sides of the body, and may then be isochronous. The upper limbs are more often affected than the lower limbs; the trunk is less often the seat of the spasm; and the face escapes in about half the cases, while the ocular muscles are the only voluntary muscles which are never affected.

The proximate segments of the limbs are more severely affected than the distal parts, while the most peripheral parts, the hand and the foot, often show no movements; but there are exceptions to this rule. In the upper limbs the pectorals and deltoids are especially prone to be affected. In the lower limbs the quadriceps extensor, and in the trunk the abdominal recti and obliqui muscles and the erectores spine are most frequently involved. In slight cases and in those cases which have been described by Biancone under the name "myokimia," the contractions produce no movement of the limb and are not obvious to the observer until the patient's clothes are removed. From such mild cases to the severe cases in which spasms occur of such violence as to cause the patient to fall suddenly, every degree of interference with movement may be met with. The movements cease during sleep in the great

majority of cases, but in rare instances they persist in much diminished intensity.

The effect of volitional movements upon the clonic spasm varies widely in different cases. In some instances, mostly mild cases, voluntary exertion inhibits the occurrence of the spasm. On the other hand, voluntary movements may excite and augment the spasm, and especially is this the case in severe types of the disease; and in the two extremes of the disease, as regards severity, one sees in the mild cases that the movements are most marked when the patient is quiescent, and in the severe cases least marked or absent during rest.

The myoclonic movements interfere with volitional movement in proportion as they have a loco-motor effect. In severe cases the patients may be unable to dress themselves on account of the clumsiness which the involuntary movements entail.

The electrical excitability of the muscle is unaltered, and there is no muscular wasting, but the mechanical excitability is increased, and percussion of a muscle may evoke the movements. The sphincters are not affected. The superficial reflexes are usually increased. The deep reflexes may be normal, diminished, or increased, and in severe cases of long standing a condition of general slight spasticity may develop.

Idiopathic myoclonus is thus brought in line with those cases of cerebral disease in which myoclonus occurs as a symptom, for these are invariably spastic.

Sensory phenomena are, as a rule, totally absent, but in the class described by Biancone as "myokimia," subjective sensory disturbances form an important part of the symptomatology.

Speech may be seriously interfered with when the muscles of the jaw, tongue, palate, and larynx are involved, and spontaneous laryngeal and pharyngeal noises may occur.

Epileptic attacks are often associated with myoclonus, and the term *epileptic myoclonus* has been used to distinguish these cases from those of myoclonus in which



epileptic attacks do not occur. Several cases have been recorded as epileptic myoclonus in which general and progressive spasticity existed from the commencement of the symptoms, and it seems highly probable that these cases were, in reality, cases of widely-spread cerebral disease of which general spasticity, epilepsy, and myoclonus were symptoms.

Mental changes are exceptional in cases of pure myoclonus, but the disease is apt to appear in neuropathic subjects, and the intellectual faculties are, as Raymond has pointed out, rarely quite normal.

*Course and duration.*—Myoclonus, as a rule, is a slowly progressive affection up to a certain stage, and when this is reached it may remain stationary for years, having little tendency to shorten life, and the patient may ultimately die of some other affection without any period of freedom from the spasms.

A few cases have been recorded in which the disease has ended fatally within a few months of the onset.

Recovery may take place either spontaneously or as a result of treatment, but the affection is very prone to recur, especially if a patient be exposed to the influence of those agents which were presumably the exciting cause of the first attack.

*Etiology.*—The causal factors of the malady are obscure. In a few cases direct neuropathic heredity has been traced, but this is exceptional. Alcoholism in the parents is not rarely present. Many instances of the affection of several or all of the children of the same parents have been recorded, and the fact that in such cases the symptoms have commenced at the same age in children who had been previously healthy argues that the disease is congenital.

The malady appears in children usually between the ages of 5 and 15 years, while in adults it generally commences between the ages of 25 and 40 years. Both sexes are liable to the affection.

*Pathology.*—No morbid changes have been found in the nervous system in the few cases which have been examined

*post mortem*. It is, however, probable that the seat of the morbid process is in the cerebral cortex of the central region, for when this region is affected with atrophic sclerosis and other pathological conditions myoclonic movements may be conspicuous symptoms. The frequent association of myoclonus with epilepsy also suggests this seat for the disease, as do also the clonic nature of the movements and the abnormal mental state so often present in the subjects of myoclonus. The cessation of the movements during sleep favours a cerebral origin as against a spinal origin for the malady. Many authorities, and especially Unverricht, have brought arguments to bear against a cerebral seat for the disease. They point out that the movements are simple, symmetrical, and often isochronous, that they are never of unilateral distribution, and that muscles are often involved in one movement which never act synergically in any volitional movement; whereas in spasms known to be of cerebral origin the muscles are involved in the order of their natural synergic association and an isolated contraction of a single muscle is never met with. Again, electrical stimulation of the cortex cerebri invariably produces contraction in a group of muscles concerned in one particular act. Against these arguments it may be said that the necessity of synergic combination of muscles in spasm of cerebral origin is very doubtful if the spasm which occurs in an epileptic attack is taken as the type of spasm of cerebral origin. For surely the spasms which occur towards the end of an epileptic attack are not the contractions of muscles in strict synergic association. Although the result of electrical stimulation of the normal cortex is the production of complex co-ordinated movement, it does not follow that stimulation of a pathologically deranged cortex would produce the same result.

It is important to recognise the mutual interdependence of the elements of the nervous system and to realise that as the result of a pathological condition of the motor cortex the functional condition of the whole motor tract corresponding becomes widely altered; and while the cortical seat of the disease in myoclonus is probable, a profound

change in the functional state of the motor cells of the spinal cord, as a secondary result of the cortical derangement, is also likely.

The nature and mode of action of the morbid agent which is responsible for this disease are entirely unknown to us.

The *diagnosis* of the disease is not difficult in a typical case, since the simple shock-like movements in symmetrical muscles on the two sides of the body, without any resemblance to volitional movements and altogether destitute of rhythm, occur in this disease alone. In some cases, however, the movements may be less characteristic, and may depart somewhat widely from those above described as typical of myoclonus. It has happened, for example, that among several children of the same parents who have been affected with the disease the movements present in one case have been typical, while in another the movements have departed so far from the type that but for the familial association one would hesitate to place them in the category of myoclonic movements.

Myoclonus is distinguished from chorea by a more gradual onset, by the nature of the movements, by the frequent exemption of the face which is always involved in chorea, and by its association with epilepsy. On the other hand, the frequent rheumatic manifestations of chorea are not present in myoclonus. Electrical chorea, in which the movements may closely resemble those of myoclonus, is readily distinguished by the rapid course, high pyrexia, paresis, and great muscular wasting which occur.

In certain forms of hysteria movements of a shock-like nature may cause great difficulty in diagnosis. In hysteria such movements tend to be less persistent than in myoclonus, and other hysterical stigmata are usually present which serve to distinguish these affections, for myoclonus is not often associated with other functional derangements. While in a few cases of hysteria movements are present which cannot be distinguished from those of myoclonus, yet in the majority of cases in which the latter disease is

simulated by hysteria a careful investigation of the movements will reveal some peculiarity which is foreign to myoclonus.

The convulsive "tics" can hardly be confused with myoclonus, for in the former disease the movements are large movements, and are like voluntary movements. Moreover, the spasmodic utterances and fixed ideas which are commonly present in the more severe form of tic do not occur in myoclonus.

Myoclonic movements, when occurring as symptoms of gross cerebral disease, are almost always associated with spasticity and paresis, usually bilateral in distribution, less commonly unilateral, and other signs of intra-cranial disease are often present. In some cases, however, the movements may be the earliest manifestations of cerebral disease, and the subsequent clinical development of the case can alone distinguish them from idiopathic myoclonus.

*Treatment.*—The malady is an intractable one in proportion to the length of time the symptoms have persisted. Some cases recover spontaneously, and this may occur after treatment has been abandoned as useless. The removal of any discoverable exciting cause and the improvement of the general condition of nutrition is of great importance. The application of electricity has been credited with the recovery of many cases, but it is of doubtful value except in hysterical myoclonus. Hydropathic measures have been useful in some cases. Arsenic has also been of decided benefit occasionally, but the effect of chloral in lessening the movements is only temporary, and bromides are of no value.

## THE TICS

UNDER this name are comprised several distinct yet nearly allied diseases characterised by the occurrence of sudden, rapid, involuntary, coördinated movements, the peculiar nature of which has suggested the application of the most descriptive term "tic" (twitch) to these maladies. They have been clearly separated from other affections characterised by muscular spasm, and their clinical features ably described by Gilles de la Tourette, Guinon, Noir, and others of the Salpêtrière School. They are classified as follows :

(1) *Simple tic or habit spasm.*—In this condition sudden involuntary movements, occurring usually in the face, neck, and upper extremities, make up the clinical picture. The disease often runs a favourable course.

(2) *Convulsive tic.*—The muscular twitches are more severe than in simple tic, and imperative ideas and explosive utterances are most important symptoms. The affection is nearly always progressive and intractable.

(3) *Psychical tic.*—The affection resembles the foregoing except that the motor manifestations are absent, and the spasmodic movement is replaced by psychical changes.

(4) *Coördinated tic.*—Here there is habitual performance of a coördinated act, which generally originates from some habit. These movements are often unconsciously performed and are much more under the control of the will than are the imperative acts of psychic tic, and the disorder frequently admits of complete cure.

*Simple tic ; habit spasm.*—This malady is signalled by the frequent repetition of some simple muscular action in the face, neck, or upper extremities—rarely in the lower

extremities. The movements, which are of lightning-like rapidity, are always of the same nature, and are not varied, though two separate movements may co-exist.

*Etiology.*—Simple tic is a common disorder of late childhood, and occurs most often between the ages of 5 and 10 years, while 80 per cent. of the cases occur between the ages of 5 and 15 years, after which a few cases are met with, but in decreasing frequency, till the end of the fourth decade. Sex appears to have no influence upon the incidence.

Heredity is traceable as direct transmission in a few cases only, but a history of the occurrence of other neuroses in parents and relatives is common. Sometimes several children of the same parents are affected.

The onset of the affection is often preceded by deterioration of general health, which may result from acute illness, unhealthy hygienic surroundings, overwork, or bad feeding. In some instances mental emotion or fright appears to be the exciting causes.

Not infrequently the movements have originated from some local cause of irritation and have become involuntary. Among the common local causes of irritation may be mentioned adenoid vegetations in the naso-pharynx, nasal catarrh or obstruction, conjunctivitis, and errors of refraction. Sometimes the source of irritation is far removed from the region of the spasm—for example, the presence of phimosis or of oxyurias. In many cases the affection originates in some trick or habit, repeated so often as to get beyond conscious control.

Some cases owe their origin to that half-conscious imitation which is apt to occur in a neurotic child who is impressed by constantly witnessing the occurrence of peculiar movements in another. A neuropathic tendency in the child is of great importance in causation, and not infrequently the affection develops when he or she is first sent to school.

*Symptoms.*—The movements are of the nature of a simple act; they show themselves suddenly and are executed very rapidly. One kind of movement only occurs in most cases,

but sometimes two or more movements may recur, and in some instances the nature and seat of the recurring movement may change, a second movement taking the place of the first.

The seat of the spasm is most frequently the face, head, and neck. Blinking, winking, alternate elevation and depression of the eyebrows, side to side movement of the mouth, sniffling, tossing the chin in the air and sudden movement of the tongue, palate, or larynx, accompanied by an unpleasant fidgeting sound, are of frequent occurrence. Any movements of the head upon the trunk may occur, while shrugging of the shoulders and movements of the arm are often met with.

Respiratory movements are often associated with those of the tongue and larynx. Spasmodic movements of the legs are not common, but sudden flexion of the leg and thigh and stamping of the feet may occur. In rare cases the tic takes the form of a sudden start, passing with electric-like rapidity through the limbs and trunk.

In the less severe cases a few minutes elapse between the succeeding movements; in the more severe cases the movements may follow one another almost unceasingly. The movements are increased by excitement and under observation, and can be usually controlled by an effort of will, but only for a limited time. They invariably cease during sleep.

*Diagnosis.*—It is usually easy to distinguish the tics from chorea if it be remembered that the movements of chorea are not of the sharp, shock-like character of those of tic, and that in chorea there is not infrequently the association of pains and even of actual rheumatism.

*Prognosis.*—The majority of cases of simple tic occurring in childhood recover if appropriate treatment be adopted early.

The later the age at which the symptoms appear the more difficult it becomes to effect a cure, and when the malady commences after middle life it is usually permanent.

The duration of the malady greatly influences the prog-

nosis, since the longer the functional disturbance has existed, the more firmly the vicious habit becomes impressed upon the nervous element and the more difficult is its eradication. A favourable prognosis can only be given with certainty after the ameliorating effects of treatment have been observed, for in any given case the condition may continue indefinitely, and, when persistent, the spasm may spread to other groups of muscles than those originally involved.

*Treatment.*—Any cause of local irritation should be carefully sought for and remedied. Adenoids should be removed if present, and any existing errors of refraction corrected. The patient should be placed in the best possible hygienic surroundings, and the system should be strengthened by good food, fresh air, and tonics. The increase of the spasm which attention causes and its aggravation by fright and depression, demand that the movements shall be as far as possible disregarded by those who have to do with the child, and that threats or punishment when he is unable to control the spasm, should never be made or inflicted. It should be kindly impressed upon him by someone who has his confidence that if he continue in his attempts to control the spasms they will get better, and he should be induced by a judicious combination of rewards and deprivations to make a strong effort to conquer the habit.

In severe cases, mental exertion and all kinds of excitement should be interdicted, and rest in bed is indicated. If the limbs are affected gymnastics are useful, and it is advisable that these should be carried out in front of a full-length mirror so that the patient may see and correct his faults. Arsenic is of value in many cases, and strychnine is often useful.

Hammond advocates atropine in doses of  $\frac{1}{100}$  grain, gradually increased to  $\frac{1}{40}$  grain, and conium in doses of five minims of the fluid extract gradually increased. These drugs may be advantageously combined with bromides.

*Convulsive tic* (Gilles de la Tourette's disease).—The characteristics of this malady are the repeated occurrence



of spasmodic movements of a similar nature to those occurring in simple tic, but, as a rule, of greater severity, and either limited in distribution or generalised, and the tendency to explosive utterances and irresistible impulses to repeat words or sounds or to imitate gestures. Further, imperative ideas are often present.

*Etiology.*—The subjects of convulsive tic nearly always show the stigmata of physical degeneracy. A neuropathic heredity is often traceable, and sometimes direct heredity is present. The disease is rather more common in males than females, and the first symptoms are usually noticeable in childhood between the ages of 10 and 15 years.

The immediate exciting causes are most often mental and physical shocks. In some cases the affection follows some acute illness, while instances in which the affection resulted from imitation have been recorded.

*Symptoms.*—The clinical picture is made up of the following symptoms: involuntary movement, spasmodic utterances, impulsive imitation of gestures, words, and sounds, and psychical derangement with imperative ideas.

The spasmodic movements occur suddenly, and are rapidly executed, and irregularly repeated. They are of the same nature as the movements of simple tic and favour the same situations, but the spasm is not restricted to the repetition of one movement, but successive movements may vary widely in nature, and there may scarcely be a muscle which is not sometimes involved in the spasm. General spasm sometimes occurs, but is exceptional. It is almost impossible to convey in brief a clear idea of the great variety of facial grimaces, head-jerking, grotesque attitudes and movements of the limbs, and ridiculous pantomime which occur in this affection, for almost every kind of movement may occur. A patient may perform such extraordinary antics in walking that at first sight one cannot but think that he is "shamming."

The spasmodic movements are not continually occurring. They are present as attacks, and in the attack each movement may involve a different group of muscles, and there is

a tendency for the same order of involvement in successive attacks, so that the same pantomime occurs in each attack. Notwithstanding that the patient is fully conscious of the movements, and that he is ridiculous to the spectator, he has no lasting voluntary control over the spasm. A powerful effort of will, or more often the performance of some voluntary movement, may cut short the attack, but the effort is so fatiguing and induces so much *malaise* that the patient does not exert himself in this way.

The movements cease during sleep and are less troublesome during repose and when the sufferer is alone. They are increased by excitement and observation. In the interval between the spasms voluntary movement is not impaired, and acts requiring the greatest delicacy of co-ordination may be accomplished without the least evidence of any defect.

(3) *Psychical tic*.—The nervous derangement which is usually expressed in sudden contractions of the muscles may give rise to phenomena of a different order, but very similar in their physiological nature. Of these the most important are :

- (1) Explosive utterance.
- (2) Fixed ideas.
- (3) Impulsive acts.

These conditions, which may be termed “exclamatory” ties, and “psychic” ties, may each be the sole or chief symptom of the malady. More often they occur in association with convulsive tic in the same case, and it is then usual for the convulsive movements to appear first, and to be followed some time later by exclamatory tic, and finally by fixed ideas and impulses.

In more than half of the cases of convulsive tic, however, the muscular spasms alone occur ; it is only in the minority of cases that exclamatory utterances and psychical disturbances are met with.

The exclamatory tic takes the form of some sound, word, or group of words, habitually and explosively uttered with complete irrelevancy of time, place, and sense. The sound

may be an unpleasant inarticulate noise, or it may resemble the cry of some animal. When words and sentences form the recurring exclamation, they are frequently of an obscene nature (coprolalia).

The utterances may occur singly, or may be repeated several times in rapid succession, and they may be synchronous with the occurrence of spasm, in which case they are apt to be spoken loudly and explosively.

Echolalia is sometimes met with. It consists in an uncontrollable impulse to repeat certain words which are heard spoken, or the cries of animals. The word or words which are echoed are usually those which especially call the attention of the patient by their unfamiliarity or for some other reason. The patient may echo the end words of his own sentences. In all cases the echoes are repeated several times.

In some patients suffering with convulsive tic movements and gestures are imitated. This echokinesis is a phenomenon which occurs in the epidemic disease "latah" of the Malay peninsula, and also in the condition which occurs in Siberia known as "myriachit," and it is probable that both these maladies and also the jumping malady of Maine are closely allied to convulsive tic.

It is characteristic of all impulsive movements occurring in convulsive tic, whether coprolalia, echolalia, or echokinesis, that though the patient may strenuously desire to prevent their occurrence, he is utterly unable to do so by any effort of will, and although he may learn to circumvent the occurrences by the performance of some other movement, or by speaking quickly, he only succeeds in putting off the evil moment for a short time.

The psychological peculiarities which are met with are mainly imperative ideas and impulsive acts. Insanity of doubt, arithmomania, agoraphobia, acrophobia, and mysophobia may occur.

In cases in which such symptoms are very conspicuous, grave signs of mental deterioration slowly supervene, judgment and memory fail, will power and attention diminish, and the patient becomes incoherent.

*Diagnosis.*—The nature of the spasmodic movements of convulsive tic is so peculiar that there is little liability to confusion. The several forms of chorea and myoclonus are at once distinguished by the widely different character of the spontaneous movements. Moreover, explosive utterances and imperative ideas and impulsive acts do not occur in these maladies.

The separation of convulsive tic from hysteria is rarely a matter of difficulty; for, while both diseases may co-exist, they are totally different affections. The spasms of convulsive tic may be to a certain extent simulated by hysterical manifestations, but this is of rare occurrence, and impulsive utterances and ideas never occur as symptoms of hysteria. Further, the stigmata of hysteria serve to distinguish this disease.

*Prognosis.*—The course of the malady is that the symptoms progressively increase, in some cases continuously, while in others stationary intervals occur. In very rare instances recovery has been recorded, but most of these cases have subsequently relapsed. Psychological disturbances are of great importance in the prognosis, since in cases in which these symptoms are absent life may not be shortened by the disease. In every case of convulsive tic, however, it must be remembered that psychological symptoms are likely to make their appearance, and, when they occur, insanity commonly results.

*Treatment.*—There are no measures from which one can confidently expect improvement in this disease. Apart from general tonics, drugs are useless. Change of circumstance and isolation, with strict moral and physical discipline, with the encouragement of healthy pursuits and congenial intellectual and mechanical occupation, are indispensable.

When fixed ideas are present, all precautions must be taken which are desirable for a patient of unsound mind.

*Coördinated tic.*—This name has been applied to a group of cases in which complicated coördinated movements are habitually repeated without cause or purpose.

Many of the cases are the results of habits which have become indelibly impressed and recur spontaneously—the condition hardly amounting to a disease. These habit movements and tricks may be of so varied a nature as to defy a single description. They can, in most cases, be controlled by an effort of the will, and do not occur so long as the patient keeps the matter in mind. Those measures are applicable for the treatment of this condition which have been described for the treatment of simple tic.

## STAMMERING

STAMMERING or stuttering is a spasmodic affection leading to a sudden check in the utterance of words or to a rapid repetition of the consonantal sound, less frequently of the vowel sound in connection with which the difficulty arises.

*Etiology.*—The disorder is rarely, if ever, congenital. It may appear at any period between early childhood and puberty, but its earliest manifestations do not appear until after the child has learned to talk fluently. Sometimes defects of articulation almost exactly resembling the stammering of children come on comparatively late in life as the result of organic brain disease, especially of general paralysis of the insane.

The sexes are very unequally affected, boys being the subjects of the disorder five times as frequently as are girls. There is no explanation for this incidence, but the fact is of interest that boys acquire speech more slowly and at a later age than do girls. Heredity is a common etiological factor, usually in the form of an inherited highly strung nervous temperament.

In cases in which the disorder appears among the children of parents who stammer or when several children of the same family suffer, the causal factor in most cases is imitation, and not heredity. Stammering is not in most cases associated with a low standard of mental development, but the reverse is the rule. Among imbeciles and idiots, while other varieties of speech defects are common, stammering is rare.

The most common cause is undoubtedly imitation, which, though in most cases unconscious, is sometimes commenced

for the purpose of teasing and annoying a companion, and is persisted in until the "biter is bit." In this connection Dr. George Cathcart records a most interesting example. The eldest child of a family contracted the habit from a stammering nurse and the five younger members in turn became stammerers. The youngest child, having had the disadvantage of five bad examples, was much the most afflicted. In two of the five children the stammer disappeared completely on removal for a short period from the family circle and reappeared upon their rejoining it.

Severe emotions such as shock or fright have been the immediate antecedents in some cases, while in others the affection has come on during convalescence from severe illness, such as typhoid fever or diphtheria. Sometimes the affection, after being cured, has returned during convalescence from such illnesses.

As possible predisposing causes to stammering may be mentioned certain causes of peripheral irritation, such as the presence of naso-pharyngeal adenoids, the eruption of the molar teeth, intestinal parasites, and phimosis.

The condition is not associated with any anatomical defect in the organs of articulation or with any structural changes in the nervous system except in certain rarely occurring cases in adults which have been already referred to.

*Physiology.*—In the production of articulate speech three separate muscular mechanisms are concerned (1) the muscles of the lips, tongue, and palate for articulation; (2) the intrinsic and extrinsic muscles of the larynx for the production of the voice; and (3) the respiratory mechanism for supplying the blast of air which sets the vocal cords into vibration.

For distinct speech there must be absolute co-ordination, both as regards time of action and as regards power between these mechanisms. Professor Wyllie uses the happy comparison between the production of articulate speech and the production of notes upon a violin. He compares the bow hand of the violin with the larynx and the hand manipulating the strings with the articulatory mechanism.

Each hand has its own work to perform. The right hand wielding the bow is the producer of sound; in proper time and with correct emphasis of touch, it calls forth the vibration of the strings in rapid succession. The left hand moulds the sound into music by the manipulation of the strings. Its fingers must not only keep pace upon the strings with the movements of the bow, but they must also touch each string to a nicety in the right place, otherwise the note will be out of tune. The two hands must act in perfect coördination with one another; each hand, in assisting the other, must perform its part of the work exactly at the right moment, neither too late nor too soon; since the slightest inexactitude in this respect would be utterly destructive of the music.

In the movements which produce articulate speech those of the larynx are comparable with the movements of the bow hand of the violinist, while those of the tongue, lips, and palate are comparable with the movements of the fingers upon the frets, and as it is common for one who is commencing to learn the violin to pay too much attention now to the bow hand, now to the fingers of the left hand; so, in the common variety of stammering, the speaker neglects the laryngeal mechanism, and, when no speech is emitted by his attempt, he unwittingly throws more force into the wrong quarter—the buccal mechanism, which appears to him to be at fault. The nerve centres for the buccal movement are thus put into a condition of physiological embarrassment, and become surcharged with energy which may overflow into other centres, and produced those spasmodic complications which are so commonly seen in those who stammer.

In the great majority of cases it is the laryngeal mechanism which is at fault. Its action is delayed, and the necessary synchronism of action of laryngeal and buccal movements so necessary for distinct utterance is lost. Thus stammering occurs over the consonants and more especially over the voiceless consonants, and not upon the attempt to produce the vowel sounds.



In rarer cases the action of the buccal mechanism may be delayed, and then the subject stammers over an initial vowel sound. Moreover, in other cases, both mechanisms may be equally at fault so far as can be judged.

An exact knowledge of the manner in which the various sounds of articulate speech, vowels and consonants, are produced is all-important to the observer in investigating articulatory defects, and to the stammerer in order that he may be put into the right way to overcome his defects by a knowledge of the manner in which they are produced.

A vowel has been defined by Ellis as a modification due to resonance in the cavities above the larynx, of an original quality of tone produced by the vibration of the vocal cords in the larynx. Each vowel is a continuous voice-sound, and calls both the respiratory laryngeal and buccal mechanism into play.

In the natural vowel series, "i," "e," "a," "o," "u," the first is produced with a small buccal cavity and with a small anterior opening, and in the production of the succeeding vowels the size of the buccal cavity is progressively increased; the anterior opening of the cavity being progressively increased in size in "i," "e," and "a," and progressively diminished in "o" and "u."

Consonants are produced by the sudden and more or less complete interruption of the breath by the contact of the walls of the buccal cavity in certain positions known as the primary stop positions, and they are divided into two main groups: (1) The voiceless consonants, which are purely buccal sounds, and (2) the voiced consonants, which have, in addition, a preceding or concomitant voice-sound.

Voiceless.		Voiced.		Voiced and nasal.
p	...	b	...	m
f	...	v	...	
wh	...	w	...	
t	...	d	...	n
th	...	dh	...	
-	...	l	...	
-	...	r	...	

Voiceless.	Voiced.	Voiced and nasal.
s	z	...
sh	zh	...
-	y	...
k	g	ng

The stop positions are as follows :

(1) The anterior or labial stop position, where the lips come in contact. Here are produced the voiceless explosive "p," its voiced equivalent "b," and nasal resonant "m," the voiced fricative "w," the voiceless fricative "wh."

(2) The middle stop position is the anterior linguo-palatal. Here are produced the voiceless explosive "t" and its voiced equivalent "d" and nasal resonant "n," with the voiceless fricative "sh" and the voiced fricatives "zh," "r," and "z."

(3) The posterior stop position is the posterior linguo-palatal.

Here are produced the voiceless "k" and its voiced and nasal equivalents "g" and "ng," with the voiceless fricative "y."

There are two other positions in which fricatives are produced.

(4) The labio-dental position, between the upper teeth and lower lip. Here the voiceless "f" and voiced "v" are produced.

(5) The linguo-dental position. Here the voiceless and voiced "th" are produced, and the sibilants "s" and "z."

The difficulty in articulation that stammerers experience arises first in connection with the utterance of initial consonants, especially the voiceless explosives, and it is always greatest with these. The stammerer makes the attempt to speak often without first filling the chest with air, thus showing a lack of co-ordination of the respiratory with the other mechanism. He closes the oral canal at one or other of the closing points, according to the nature of the consonant to be articulated, and this he does as well as one possessed of a normal faculty of speech. Instead, however,

of allowing the vowel to follow the initial consonant without delay, he presses his lips or his tongue and teeth or his tongue and palate more firmly together than is necessary; the explosive escape of breath does not take place; the other muscles of the face and those of the glottis, and even the muscles of the neck, become spasmodically affected, as are those of articulation; gesticulatory movements are made, the abdomen is retracted, the head may be drawn backwards, and the larynx drawn forcibly upwards until finally the sufferer may work himself up into a terrible condition of agitation; his heart beats forcibly, his face becomes congested, and his body is bedewed with perspiration, and he may present the appearance of a maniac.

The above graphic word-picture of Kussmaul illustrates admirably a severe degree of the ordinary variety of stammering where the laryngeal mechanism for the production of the voice does not come into play synchronously with the buccal mechanism, and the stammerer, not knowing what is at fault, misdirects his energy into the oral mechanism. The surcharge of this mechanism produces physiologically embarrassment in consciousness, the excess of energy overflows into the centres supplying neighbouring muscles and causes spasm in these. When the initial fault is with the laryngeal mechanism the patient may stammer over initial vowels, and when attempting to speak he may stand with mouth wide open and tongue protruded, unable for some time to utter a sound. When at last he manages to get over the stile the words pour out rapidly until he has no breath left. Knowing the difficulty that will arise with the next breath, he gets as many words out with the one breath as he can. A most excellent word-picture of this habit of festination in speech so commonly associated with stammering occurs in Shakespeare:

“I prithee tell me who it is quickly and speak apace. I would thou couldst stammer that thou mightst pour this concealed man out of thy mouth as wine comes out of a narrow-necked bottle either too much at once or none at

all. I prithee take the cork out of thy mouth that I may drink thy tidings" ("As You Like It." Act III, Sc. 2).

The lagging behind of the laryngeal mechanism in most cases is well exemplified by the fact that even the worst stammerers have no difficulty in singing or when intoning, for here the laryngeal mechanism is first set a-going and moreover the chest is well filled.

Sometimes the patient uses tricks to prevent the stutter, and these may become engrafted upon him as habits difficult to eradicate even when the stammer is cured. These may take the form of (1) associated sounds; whooping, grunting, crowing; or (2) habit spasms; contortions of face, limbs, or body.

*Prognosis.*—The majority of cases of stammering are cured if appropriate and diligent treatment be adopted. The presence of mental deficiency renders the prognosis much worse.

An important factor is the age of the patient. The most favourable period for treatment is from 12 to 16 years, when the stammerer is old enough to realise the importance of getting rid of his defect, and can intelligently comprehend the principles of his treatment, and before the defect is very deeply rooted by force of habit upon his organs of speech.

There are, however, some patients who have mastered the elements of their teaching and who do not stammer when reading or reciting to themselves or when speaking to their teacher, yet the moment they are called upon to take their part in daily life or to talk to strangers the stammer reappears.

It is stated that hypnotism has been of signal service in some of the most intractable of cases.

*Treatment.*—In all cases the general condition of health should be attended to, and any cause of local irritation, such as defective teeth, enlarged tonsils, nasal obstruction, pharyngeal adenoids, and phimosis should be removed. Nervous and shy children should certainly be removed from school, and perhaps all cases are more profitably treated

away from school. The treatment should be individual, and the child should not be brought into contact with other stammerers.

In the case of very young children a cure may often be obtained by gently and firmly insisting upon the repetition of the letter or letters over which the child has stumbled until they can be pronounced quite smoothly.

In older children two fundamental points are to be observed: the one is that the teacher must obtain the full confidence of his patient; the second is that the patient must grasp the fact that his energies are to be directed towards the vocal element and that his voice must be made as far as possible a continuous one.

The patient should be removed as far as possible from causes increasing self-consciousness of his defect.

The respiratory capacity and control of air within the chest should be increased by breathing exercises, the chest being deeply filled and the air being then allowed to escape over the false vocal cords with a slightly hissing sound as slowly as is comfortable to the patient.

He should speak slowly, with a full resonant voice. When he comes to a word upon which he tends to stutter he should raise his voice and direct his energies to vocalisation and not to articulation. If the difficulty is with a voiced consonant he must be directed to voice it; if a voiceless consonant, his attention should be turned to the vocalisation of the subsequent vowel sound. He should also intone his speech. An admirable series of test phrases for repetition according to each consonant with which the difficulty arises will be found in 'Disorders of Speech' by Professor Wyllie.

Gymnastic and singing exercises are valuable additions to treatment, and should associated movements be present, the reading exercises should be carried on in front of a mirror, so that the patient may be aware of these himself, and endeavour to suppress them.

*Idioglossia* is a condition in which on account of difficulty in learning the pronunciation of certain consonants a child substitutes other consonants, and so seems to speak a

language entirely its own. The language is sometimes rendered still less intelligible by the retention in speech of words of a child's own invention, which are sometimes very numerous. The following admirable illustration is given by Dr. Colman of one of his patients who thus repeated the Lord's Prayer: "Onë Tahde na ah in edde, anno de Di na, I tidde tah, I du de na on eet a te edde, te ut te da onë dade ded e didde onë tetedde a ne ahdin to te tetedde adase nt, ne no te tetate, ninne ntle enu, to I a te ninne pouë e dordy to edde e edde, Amé."

This patient was unable to pronounce many of the consonants, and substituted for them "t," "d," or "n."

*Lalling* is a defect due to the want of precision in the oral articulatory mechanism. It is a persistence of the characteristic speech of children before articulation is completely learnt, and it occurs chiefly in connection with defective intelligence.

*Lisping* is a defect resulting from the indistinct enunciation of certain consonants or from the substitution of wrong consonants. It is commonly met with in infants learning to speak, and may persist from a clumsiness in articulation. More often it is engendered by loss of the first milk teeth, and becomes a habit. Sometimes it is due to defective conformation of the mouth and especially to backward displacement of the upper incisor teeth, which prevents the tip of the tongue from meeting the palate in the middle stop position.

The *prognosis* in idioglossia and in lisping is invariably good. The treatment consists in patient and careful teaching of pronunciation, with in the latter case attention to any oral or dental defect that may exist.

*Aphthongia* is a rare disorder in which the attempt to speak sets up severe spasm in the muscles of articulation, chiefly in those of the tongue. Nothing is known as to the pathology of this rare disease, which seems to be of the same class as writers' cramp, violinists' cramp, etc.

## CONVULSIONS IN CHILDHOOD

A CONVULSION has been defined by Dr. Hughlings Jackson as a "sudden and excessive discharge of many nervous arrangements representing movements at once or nearly together, because the cells subserving such movements have become highly unstable."

Such a definition being accepted, the investigation of convulsions as they occur in early life resolves itself into an inquiry into the causes of, or conditions attending the presence of, instability in nerve-cells leading to convulsion, and a further inquiry into the localities in which such cells are situated, and whether the character of the convulsion gives any indication of this.

It was for long supposed that the medulla oblongata was the seat of the discharge in convulsion. Later the belief gained ground that the only seat of discharge was in the cortex cerebri, but now both extreme views have been modified, and it is believed by the highest authorities that while most fits have their origin in cortical discharge, some depend upon discharges from cells situated in lower structures, such as the nuclei at the base of the brain. Such a view accords with the evolutionary levels of the nervous system as described by Dr. Hughlings Jackson, to whose teaching on this and other neurological subjects medical science is under deep obligations.

These levels are three in number:

(1) The lowest level or series of lowest centres situated in the spinal cord, medulla oblongata, and pons varolii, and this series of sensori-motor centres represents all parts of the body in the simplest combinations. In this lowest level are also comprised regulating centres, micturition centre,

vaso-motor centre, respiratory centre, etc., which are really recombinations of elements of the centres already mentioned, viz. the anterior and posterior spinal horns, and their homologues at a higher topographical level—the cells of medullary and pontine nuclei.

(2) The middle level, consisting of the motor-centres in the Rolandic area, and the corresponding sensory area of doubtful localisation. In this there is a re-representation of all parts of the body, sensory and motor.

(3) The highest level, probably situated in the frontal and occipital regions of the cortex, in which there is a re-representation of all parts of the body, sensory and motor.

In the new-born infant, practically the only developed part of its nervous system is the lowest level—the level subserving reflex processes. The higher levels are still very imperfect. As the child grows, however, these levels become more and more active, being gradually evolved so as to possess among other functions controlling and regulating influences on the lower centres.

Thus reflex processes become much less automatic and more and more under the control of the impulses which originate in and are conducted from the higher centres.

Such a view as this gives us an indication for a classification of fits. There seems every reason to suppose that there are fits which have their starting-point in the cells of each of these three different levels. Thus laryngismus stridulus is a lowest level fit, a fit due primarily to a discharge commencing in the respiratory centre situated in the lowest level. An attack of this kind may remain simply this or may spread from the respiratory centre so as to become universal by leading to the discharge of the other cells of this level. In many cases the discharge does not spread beyond this centre just as in fits of the middle level (Jacksonian fits) the discharge may remain limited to certain cells in which it starts. Besides laryngismus stridulus, it is likely that a rigor is really a lowest level fit. So is an attack of spasmodic asthma.

The characteristic fit of the middle level is the Jack-



sonian fit—the fit which commences with a discharge in the cells of the middle level—the motor and sensory areas of the cortex, and in its simplest and most common form consists of a spasm in the small muscles of the hand on one side. Such a fit may, as already stated, remain limited; but it may spread and become universal, even leading to loss of consciousness by the involvement of other centres and cells.

In fits the result of discharge in the highest level, the effect of discharge in the much more complexly constituted elements of this highest level is the production of much more rapidly evolved and widespread movement, leading to generalised convulsion. Such a convulsion may be preceded by an "aura," motor or sensory, which is of great importance because it indicates the point at which the discharge commences—may, indeed, indicate the only part at which a condition of disease exists. And as in the lowest level fits, the convulsion may be only a slight attack of laryngismus stridulus *without* any spreading of the discharge so as to induce a general fit, and as in middle level fits the discharge may be limited to the cells subserving the small hand muscles, and also not spread so as to give rise to general convulsion, so in the highest level fits the fits may consist of merely an aura (sensory) or a slight, perhaps complex motor, effect, and the discharge may not spread so as to cause a general convulsion. It must always be remembered in reference to these fits that only those cells which first discharge may be the morbid ones—that they are the focus, so to speak, and may act as the fulminate, capable by their influence of leading to discharge in cells and centres which are quite healthy. Such a consideration as this is of great importance in reference to the probable effect of operative measures undertaken with the view of removing an area in which diseased cells exist in a state calculated to give rise to discharges, not only in the diseased cells themselves, but in contiguous healthy structures.

Convulsions are sometimes present in the newly-born, occurring either immediately after birth, or in some cases a few days later. There seems no room for reasonable doubt

that first-born children are more liable to suffer from such convulsions than are the later born children. Cyanosis is frequently present in such cases at birth, and is the result of the difficulty or abnormality which has attended the birth of the child. Such difficulty or abnormality, leading to long duration of the labour, is naturally more likely to occur in first labours, and in conditions of abnormal presentation. Occasionally, however, a child who has been born very rapidly suffers from convulsions either because of some local injury or because a condition of instability is induced in cells by the rapidity of the birth and the consequent loss of equilibrium between the different parts of the nervous system. In many such cases the convulsions are severe; they may persist with great frequency for several days, they may be confined to one side, or, starting on one side, may become general; and even if they are unilateral in commencement, they do not always begin on the same side. Where the convulsions are always limited to the one side such a condition is strongly suggestive of some local cortical injury; but even in such cases and still more in those in which the convulsions are general, and not of constant commencement on the same side, the convulsive tendency may be merely the result of some irritative blood state, such as one of supervenosity, and the convulsion may pass off, leaving absolutely no trace of local weakness or paralysis or other evidence of structural changes in the cortex. In some cases—those chiefly in which the commencement is local and always on the same side—when convulsions have ceased, a varying degree of weakness remains on the side which has been first and chiefly affected in the convulsion.

Such a condition no doubt depends upon some local lesion in the cortex, the result possibly of the difficulty in the labour inducing meningeal hæmorrhage, perhaps the result of the turgidity, and even rupture or thrombosis, occurring in a cortical vein. When such a local lesion is present, the tendency to convulsion is apt to persist, the attacks being nearly always one-sided, and such a condition is usually associated with a varying degree of hemiplegia, which does

not entirely pass off after the convulsion. So that, in these convulsions which occur in the newly-born, we have two conditions which may induce the instability which gives rise to convulsions: (1) The supervenosity or other morbid condition of the blood which is so apt to be present in children whose birth has been difficult—frequently the first-born, and (2) actual structural changes inside the skull—vascular lesions, especially venous, causing irritation and consequent discharge in cortical cells, or hæmorrhage from the meninges setting up similar irritative phenomena.

In a certain number of cases, also, convulsions occur quite early in life, not necessarily at the very commencement, unassociated with any evidence of difficulty or abnormality in the labour. In these the convulsions seem to be the result of some dietetic abnormality leading, possibly, to some toxic irritation of cells and consequent convulsion. They are not uncommon during attacks of diarrhoea. In these cases all the signs of irritation pass away when the child has been accommodated with suitable food, and it is rare to have any recurrence in such cases, or even if the fits have been of local commencement and one-sided, to have any unilateral weakness remaining.

There can be no doubt that a large proportion of the cases of convulsion which occur in early life, later than its very commencement, are associated with the condition of perverted nutrition which we know as rickets. Such convulsions frequently occur during the first dentition, and are ascribed, probably with good reason, to the irritation of that process as an exciting cause. To the question, Why should a rachitic condition induce convulsions? different answers have been given. Rickets is undoubtedly a diseased condition, the result of perverted nutrition. The most common causes of it are unsuitable hygienic conditions, unsuitable food, especially starchy food given at too early an age and in a not easily digested and assimilated form. The most obvious phenomena are, of course, the enlargement of the bones at the points at which growth is going on, and this enlargement is the result of excessive growth of car-

tilaginous tissue and insufficient bony modification of this tissue. It is probable that a similar want of co-ordination exists between the development of the nervous tissue proper and the fibrous tissue, which acts as a scaffolding for it, in the cases of rickets in which convulsions come on, and this presumably leads to an instability of cells and a consequent tendency to discharge.

Again, it must be remembered, as has already been set forth, that the lower centres in the hierarchy of the nervous system are the more automatic, and are under the control of the higher centres. Any interference with the development of the latter may have one of two effects: (1) It may induce in these higher (cortical) centres an instability and consequent tendency to discharge resulting in convulsion; or (2) it may prevent the higher centres from developing sufficiently to control the lower; and so we may have produced a condition of convulsion the result, not of irritation of higher centres, but of discharge in uncontrolled lower centres, so that the fits occurring in rickets may be lower level or higher level fits. And in the rachitic condition it is quite possible that there may be a toxic blood condition present which induces in cortical cells a liability to discharge, to which any difficulty with dentition, or any other form of peripheral irritation, may act as the exciting cause and lead to severe convulsions.

These convulsions in rachitic children are not often unilateral. They are much more frequently general, and although not in themselves very dangerous to life, they are of the utmost importance in regard to the future, for there is little doubt that they leave behind them a residual condition of the nervous system which frequently leads to the occurrence of epileptic fits in later life. It must always be remembered that a discharge along one path renders the occurrence of a similar discharge in the future comparatively easy. This may seem almost a truism, for it is a condition which lies at the root of the formation of all habits. Its practical bearing is to emphasise the importance of energetic and efficient treatment of these convulsions in early life, so

as to prevent as far as we can the occurrence of similar attacks, then designated *epilepsy*, in the near or even distant future.

There is another large class of cases of convulsion in early life to which reference must be made here, although this condition is described under "Infantile Hemiplegia"—those, namely, in which in a previously healthy child there supervenes a condition of severe illness, with high temperature, occasionally delirium, and the occurrence of either a severe convulsion or a series of convulsions commencing unilaterally but frequently spreading so as to involve the other side as well. Such convulsions are the result of a local lesion on one side of the brain, a lesion determined by either an acute inflammatory process, by thrombosis in an artery, or by clotting in a surface vein. The onset is similar to that of acute anterior poliomyelitis, and it may be incidentally mentioned that there is some reason for supposing that the morbid process is essentially the same. The condition left after the acute illness has passed off is one of unilateral paralysis affecting the face, arm, and leg, and it is at first, as a rule, much more extensive and severe than it ultimately becomes. In some cases the weakness left is extremely slight, but the question with which we are here concerned is, What tendency is left to the occurrence of convulsions? In these cases, as already mentioned, the onset is usually with a severe convulsion or a series of convulsions. It occasionally happens, however, that only faintness and general *malaise* attend the onset of the hemiplegia; so also it occasionally happens that no tendency to convulsion remains after the acute illness has passed off, even although the hemiplegic weakness may be very distinct. In many cases the occurrence of subsequent convulsions is delayed for months or years, in many they occur almost at once and are repeated at varying intervals. The degree of hemiplegia also varies. In most cases it is distinct and unmistakable, and when convulsions are present their origin in the condition giving rise to the hemiplegia is obvious. In a small number of cases the unilateral weakness is so slight as to be

scarcely noticeable, and the case may be regarded as one of so-called idiopathic epilepsy until the history is carefully inquired into, and a minute physical examination carried out. The true nature of the condition is especially apt to be missed if the attacks which are present are momentary and slight—attacks of *petit mal*—as they sometimes are. For it must be observed that in these cases a condition of *petit mal* may be that assumed by the attacks, and there may be also a condition after such attacks of the exceedingly important state of post-epileptic automatism—important not only from the interest attaching to it, but also from its medico-legal aspect. A careful study of these cases of convulsion or *petit mal* associated with hemiplegic weakness is of the utmost importance in the elucidation of the nature of epilepsy; for it may be stated that in those cases of convulsions the result of some definite structural alteration in the brain we have analogies, and very close analogies, to the various convulsive conditions met with in so-called idiopathic epilepsy, a fact which probably is calculated to throw much light upon the true pathology of epilepsy.

Besides those now enumerated, convulsions may occur in childhood in connection with almost any acute illness. Infantile spinal paralysis is not infrequently attended in its early stages with convulsions. Pneumonia may be ushered in with convulsions, and any feverish condition, in short, may give rise to a convulsion. It has been stated that a condition which in an adult would give rise to a rigor in a child is attended with convulsion. The convulsions in connection with tubercular meningitis are, of course, the result of the cortical irritation which goes with the meningeal inflammation. And convulsions (probably lowest level fits) are apt to occur in any condition of cyanosis, such as whooping cough, for example, may induce.

*Treatment.*—From what has already been said, it will be understood that the convulsions of early life are, for the most part, symptomatic of either gross structural intracranial disease, of an unstable condition of cells due to a loss of equilibrium in the development of different parts of

the nervous system, or to some toxic condition, which in some way induces an unstable condition of cells and leads them to discharge. In all these different conditions the radical treatment of the condition resolves itself into a correction, so far as is possible, of the underlying cause. In cases in which traumatism may have led to the effusion of blood on the surface of the brain, operative measures, if the indications are clear, may be called for. In certain cases of birth-palsy with convulsions such measures, it is said, have been to some extent successful. If the convulsions are associated with supervenosity, the application of leeches or the actual practice of blood-letting may be resorted to. If they are associated with the obvious signs of rickets, a correction of diet and surroundings with the view of remedying this must be tried. But in all these conditions some immediate measures are necessary if convulsions are frequent and severe. If the convulsions are associated with pyrexia, the patient should at once be placed in a warm bath, which is to be gradually cooled down to below the body temperature, if necessary even as low as 10° F. The relief is not infrequently immediate and complete. As has already been stated, convulsion in a child is not infrequently the equivalent of a rigor in the adult. In the pyrexial cases, as well as in others, the administration of from two to five grains of calomel by the mouth is often extremely useful, and may, indeed, be regarded as the necessary routine treatment. Bromide should also be given—freely—in one dose by the rectum. Even quite a young baby may have as much as twenty to thirty grains administered in this way. The strontium or sodium salt is probably preferable for this purpose, being less depressing and less apt to give rise to cutaneous disturbance. And this is the mode of treatment adapted for every case of convulsion, no matter what the cause. For in every case we have to deal with instability of nerve-cells, and this instability is best counteracted by bromides. In some cases, those especially in which the convulsions lead to what is practically a *status epilepticus*, stronger measures will have to be used. Hydrobromide of

hyosine in doses of  $\frac{1}{200}$  to  $\frac{1}{100}$  of a grain should then be given hypodermically. In conditions of frequent and persistent convulsion it is most efficacious. But it is a strong remedy, and its effect must be carefully watched. In all cases in which treatment has been effective in controlling the convulsions the habitual use of bromide must be continued for at least several days and only gradually discontinued. In children, as already said, the strontium salt is probably to be preferred on account of the less tendency it possesses to give rise to cutaneous troubles. Chloral hydrate may also be combined with the bromide, but in minute doses to a child—one to five grains at most, and by the rectum. The inhalation of chloroform is seldom necessary, but may be tried in persistent cases, the sedative—bromide, or bromide with chloral—being simultaneously administered by the rectum.



## NIGHT TERRORS

Various attempts have been made to classify night terrors and to draw a distinction between them and nightmares and dreams. These attempts are not satisfactory. A division, for example, of night terrors into those which are idiopathic and those which are symptomatic, although it seems feasible and reasonable, breaks down at once on an appeal to facts. There seems to be no reason to doubt that all night terrors are symptomatic—symptomatic of probably two conditions: a sensitive and somewhat exhausted nervous system on the one hand, and some exciting condition either in the peripheral mechanism or in an excited, overworked brain, or a condition produced by some terrifying or disturbing appeal to the imagination on the other hand. Night terrors, in short, are apt to occur in so-called nervous, sensitive children, and are particularly prone to occur in them at times when “over-pressure” has been present or some terrifying experience has been passed through which brings the child into a condition of nervous tension; and the acceptance of such a view as this enables us to understand the occasional close relationship (which some writers have unduly magnified) of night terrors to sleep-walking and epilepsy. There is no doubt that in the relatives of patients suffering from epilepsy the history of sleep-walking is much more frequent than amongst the relatives of normal people. Yet it does not follow that they are interchangeable. Similarly, night terrors are occasionally present in children who afterwards suffer from epilepsy. Yet the development of the more serious malady is rare, and the association of these three conditions of sleep-walking, night terrors, and epilepsy merely shows that in all

the conditions a somewhat unstable and sensitive nervous system is the essential condition, and that the direction and nature of the morbid development are probably determined by different factors.

Although it may be true that night terrors in their most obtrusive form, with screaming, etc., only occur in children, there is little doubt that the equivalent nervous disturbance is present also in adults. Thus the medical student who was conscious of much respiratory difficulty, and woke with an effort, to see the hind quarters of a large black dog disappearing through the door of his room, had an experience which, to a child, would have been most terrifying, and would certainly have ended in screaming. So vivid was the vision on the first occasion that he got out of bed and went into the next room to search for the dog. The vision or nightmare recurred frequently, but soon came to lose the really terrifying character which it had at first. It occurred originally during a period of unusually hard work. Of similar significance is the tale told of Spinello Aretino the artist, who was at work on a fresco (part of which is now in the National Gallery), depicting Lucifer changed into a most hideous beast. He took immense pains over the work, and was intensely absorbed in its execution. We are told that the hideous figure he had fabricated appeared to him in his sleep demanding to know where he had seen him looking as hideous as he had made him. The shock was so great that he only survived it a short time.

Such examples illustrate well the tendency to the onset of those disturbances at times of overwork and nervous tension. They also carry the suggestion that the causes of night terrors (frightful dreams, etc.) are present in adults just as in children, but that it is only in the latter, as a rule, that these dreams and visions give rise to recognised signs and outward manifestations of terror, although the adult may experience very real terror on awaking from one of those dreams.

The age at which night terrors are most common is between 3 and 8. A characteristic attack usually occurs

within two or three hours of going to bed. Its presence is made known by the piercing scream of the patient; he is found to be huddled into a remote part of the bed, perhaps crouching on the floor, with widely dilated staring eyes fixed intently on some part of the room. He may also point in the direction in which his eyes are directed, and his exclamations of "Black dogs!" "Black man!" "Hound faces!" clearly indicate the nature of the vision which terrifies him. He may get out of bed, he may attempt to get out at a window, or run screaming along a passage, falling exhausted at the end of it, again huddling in a corner to escape his visionary but terrifying pursuer. He may speak to some one, he may be induced to return to his room by means of soothing utterances, and may drop off to sleep and wake up with no recollection of his fearsome experiences of the night. Usually he does not recognise anyone during the attack, and yet he is amenable to the soothing cajoleries of a mother or nurse. In some attacks there seems to be, soon after the awakening, a recognition of familiar people and places. Such attacks are quickly over, and yet of these, too, there may be little, if any, recollection in the morning.

As has already been stated, the necessary condition for night terrors is an unusually sensitive nervous system. But various forms of peripheral irritation seem to be not only the exciting cause, but may determine the nature of the terrifying sensation. It is said that refraction errors may serve as such a cause, and induce complex visual hallucinations which give rise to terrors. Such a cause is only likely to be effective in cases in which there has been ocular strain from reading, etc. Aural sensations probably determine night terrors in some cases. Sir William Gowers suggests that the sensation of falling from a height, which most of us have experienced in our dreams, may be the result of slight attacks of auditory vertigo. Similar vertigo may conceivably occur in children, and the terrifying dream may induce an attack of screaming and sobbing, and more or less vague terror in a young child. To a child such a

dream is very vivid. If it should be the first of its kind, it may easily induce a condition of intense terror. If it occur repeatedly, the child's experience will soon lead to a suppression of at least the outward expression of alarm or fear.

Bodily pain also serves as an excitant of night terror and may determine its character. Dr. Colman relates the case of a boy who woke up screaming that some one was in the room hurting his finger. Although recognising his parents, he continued to see this phantom and in a second similar attack he said his finger was not only hurt but bleeding, and pointed to imaginary spots of blood on the sheets.

Intestinal disturbance is the most commonly alleged cause of night terror. It is doubtful whether it is really so effective as it is supposed to be. But it is probable that, like other forms of peripheral irritation, it may excite an attack. Similarly any febrile disturbance may be a cause. A high temperature in a child may cause a convulsion. It may give rise also to cerebral disturbance which may be associated with such psychological disturbance as attends night terrors.

Night sweats are said to be an occasional cause of night terror or nightmare. Dr. Leonard Guthrie mentions that as a child he used to have the horrible dream of being captured by Red Indians, devils, and masked burglars, and slowly tickled under the arms, whilst he was unable to move hand or foot or even to cry out. This dream he associates with profuse sweating, and in confirmation of this view he mentions the fact that he still occasionally gets it if the weather is hot or the bed-clothes unusually heavy.

But even if we were to enumerate many more forms of peripheral irritation which may be the exciting causes of night terrors, many cases would still remain for which no such cause could be found. The existence of this large class may justify, at all events as convenient, the use of the term "idiopathic" to describe these night terrors. The exciting cause of them seems to be something psychological acting upon a child with a sensitive nervous system and possessed of a vivid, not to say morbidly active, imagination. In such a

class must be placed the child who is "working sums in his head all night"—sums which will not come out right—who wakes up in a state of terror and alarm similar to that which, if he were younger and more sensitive, would haunt the devotee of billiards, who in his dreams plays a game, "with a twisted cue, on a cloth untrue, and elliptical billiard balls."

Similarly, also, we have the highly imaginative sensitive child, who is afraid in the dark, and dreads being left alone because of the vague terrors of the night which his imagination conjures up, the kind of child who, when assured that God would be with him, naïvely asked that God might be taken away and the candle left. It is little wonder that children falling asleep with vague terror still present should wake up screaming and terrified at their alarming dreams. Such children, also, are often haunted by gruesome or horrible pictures, very often seen in illustrated bibles or other good books. Thus in one child the nights were rendered hideous by a haunting picture of the raising of Lazarus, and parents cannot be too careful in the selection of picture-books, or too charitable in the allowance they make for nervous or timid children.

The *treatment* of night terrors and terrifying dreams is the removal as far as possible of their exciting cause and the correction of the sensitiveness and instability of the nervous system which permit their occurrence. The removal of all kinds of peripheral irritation and of mental anxiety or over-pressure at school is essential. No sensitive child who is afraid in the dark should be compelled to remain alone in the dark. Night-lights are cheap and safe, and better than calomel or bromide. Yet drugs must not be despised, and in persistent cases of night terrors, possibly closely allied to epilepsy, a nightly dose of bromide is very effective. It is naturally essential that such children should be carefully fed, and that they should have plenty of fresh air and moderate exercise.

## EPILEPSY

EPILEPSY may be defined as a disease of the nervous system which manifests itself in various phenomena, depending upon discharges occurring in nervous structures. Such discharges result in sensory or motor effects, and are associated in most cases with impairment or loss of consciousness. As has already been indicated in the article on convulsions—for convulsion is one of the most usual, although not invariable, signs of epilepsy—that disease is best regarded as of three different varieties, depending upon the evolutionary level of the nervous system in which the nervous discharge originates. The first variety depends upon discharge in the lowest level of the nervous system—the spinal cord and ponto-bulbar nuclei. The character of the fits in this variety is not very well known, for it is not easy to study them, and they are probably not very common. The movements are mostly trunk movements, or at least gross movements; consciousness is usually lost, and the fits are of brief duration. Experimentally, a liability to convulsion has been induced in guinea-pigs, and such convulsion can be produced when the brain has been removed. Similarly in rabbits, from whom the brain proper has been removed, convulsions can be produced by rapidly bleeding the animals. It may be asserted that such convulsions are not epileptic. They are, doubtless, analogous to attacks of laryngismus stridulus and spasmodic asthma, which probably depend upon discharges induced in lowest-level structures by supervenous conditions of the blood; and it may be desirable to restrict the name “epileptic” to the phenomena depending upon discharges occurring in the highest-level structures, in the same manner as it is perhaps best to restrict the term “epileptiform”

to the phenomena which depend upon discharges occurring in middle-level structures. But in their essential character, as depending upon pathologically induced discharges from nervous structures, they are really identical in character with both middle-level (epileptiform) and highest-level (epileptic) discharges. The results of such lowest-level discharges are mostly motor in their manifestations. Doubtless there are sensory equivalents, for each evolutionary level of the nervous system consists of a sensori-motor mechanism.

The middle level of the nervous system consists of the motor area of the cortex—roughly speaking, the Rolandic region and the corresponding sensory structures, the position of which is still indefinite. It is no doubt in intimate anatomical as well as physiological connection with the motor structures. In the pathological discharges the result of disease at this level to which the name “epileptiform” or Jacksonian has been applied and which were studied by Bravais and elucidated by Dr. Hughlings Jackson, the characteristic feature is their local commencement, usually in movements of the fingers or toes, of the hands or feet, or of the face or tongue. Sometimes these Jacksonian fits may commence in shoulder movement or in leg or even in trunk movements if the lesion is situated in such a position as to irritate the cells subserving these parts. But given a condition equally affecting the cells of this level those subserving the finest and most highly evolved movements (which are probably, as was suggested by Dr. Hughlings Jackson, subserved by smaller cells) will be the first to discharge, so that most Jacksonian fits certainly commence with movements of the fingers. It must always be remembered that a discharge commencing locally—the result of local instability no matter how induced—may spread from the diseased focus and lead to discharge in structures which are quite healthy and normal, and so a discharge commencing locally may lead to a general convulsion. Such a fact emphasises the extreme importance of studying convulsions in reference to their commencement, for it is probable that, in the great majority of fits, the manner of commencement of the con-

vulsion indicates that point in the brain the instability at which, *i.e.* the disease in which, leads to discharge.

In these Jacksonian or epileptiform fits—fits of the middle level—the commencement is usually in the hand or foot or face or tongue, as has been said. This discharge may remain localised and then the fit is a partial one; or it may, by some influence, lead to the discharge of contiguous cells and so spread as to become a general convulsion involving loss of consciousness. The commencement, however, it must be remembered, indicates the part of the cortex in which the cells are unstable.

After a Jacksonian fit there is usually paralysis, and careful observation of the paralysis is as necessary as careful observation of the commencement of an attack. The two, in fact, should supplement each other, for paralysis is always greatest of the parts first and most convulsed. It varies in degree from abolition of fine movements only, to complete powerlessness, and it varies in range from monoplegia affecting a hand to profound and universal but temporary paralysis. One point is of interest and importance, *viz.* that a right-sided fit in a right-handed person may, indeed usually does, induce temporary aphasia. So that these Jacksonian fits have their analogues in the varying paralyses which occur as the result of brain disease, *viz.* monoplegia, and hemiplegia with aphasia; and the localisation of the lesion determining the fit, and the subsequent temporary paralysis, is to be arrived at by a study of the commencement of the fit and of the distribution of the paralysis, especially in regard to its degree.

Fits of the highest level are much less simple. For the level of the nervous system, on disturbance of which they depend, is much more complex in its structure and functions. That level also, like the lower levels, is sensorimotor in its functions, and disturbance or discharge occurring in it is much more likely to involve impairment or loss of consciousness. Indeed, it may be stated that loss of consciousness is almost invariable in highest-level fits. The fits also are much less deliberate in their march. There seems to be almost a



contention of movements, a struggle, so to speak, on the part of many movements to take place simultaneously, so that it is very difficult, if not impossible, to divide a highest-level fit into definite stages. Fits also are of two kinds, fairly distinct, although in many cases it is not possible to separate them definitely. Yet the *grand mal*, or severe fit, with generalised convulsion and loss of consciousness, is more or less broadly distinguished from the attack of *petit mal*, in which loss of consciousness occurs, but in which convulsion is slight and unimportant.

The severe fit is usually ushered in by an aura, and this is important just as the commencement of a Jacksonian fit is important, for it indicates the point of greatest instability, the part of the brain at which the discharge commences. After the aura there is usually turning of the head or eyes to the right or left, with loss of consciousness, the limbs on one or both sides are drawn up in tonic spasm, the patient becomes deeply cyanosed, froth appears at the mouth, and he seems to be at the point of death. The tonic spasm relaxes, and is replaced by repeated clonic jerks, affecting all the part which had just been tonically contracted, and the tongue may be severely bitten during the clonic contraction of the masticatory muscles, and the froth at the mouth becomes blood-stained if this happens. There succeeds complete flaccidity of all the muscles, the breathing is loud, rapid, and stertorous, and the sphincter of the bladder and even of the rectum may be relaxed. Occasionally there is a brief period of consciousness, and the patient may vomit, or he may sink into a deep stuporous sleep which lasts for hours. At first the knee-jerk is abolished, possibly because of the supervenosity, but it presently returns, and may be excessive, and ankle clonus even may be present. When the patient wakes there may be intense headache.

The auras which usher in fits are various. In many instances no aura exists, or at least the patient denies its existence. The most common is the epigastric, which starts usually as an uncomfortable sensation in the epigastrium or in the left side of the stomach. It travels upward, and

when it reaches the neck a feeling of suffocation is induced, and the patient then, as a rule, loses consciousness. Occasionally an attack is preceded by giddiness,—by a feeling as if objects are passing before the patient, or as if he himself were turning round. In such cases it is not unusual for the patient actually to turn round, fall down, and become unconscious. In other cases the vertigo may be accompanied by a noise, or the noise, usually rushing in character, may be the only aura. In either case it is to be presumed that the aura is related to the auditory and equilibrating centres. As has been indicated, however, the vertigo may be ocular; and a visual aura, simple it may be, or even so complex as actually to be associated with a vision of a ball of fire or a more or less quaint figure, may usher in a fit. A crude sensation of smell also, a “chemical smell,” a “smell as of a druggist’s shop,” may precede the fit, constituting an olfactory aura. And there may be associated with this, besides champing or masticatory movements, a vague dreamy state, a state of reminiscence, in which the patient has the distinctly pleasing sensation that he is passing through a familiar experience. Such an aura has been called an “intellectual aura,” but Dr. Hughlings Jackson’s term of “dreamy state” is much more descriptive and more non-committal.

Such are some of the varied auras of epileptic fits. Innumerable variations in these are met with, and it will be recognised that they may clearly indicate a focus of disease in more or less definite parts of the cortex. And it may be incidentally mentioned that in cases in which fits have commenced, *e.g.* with olfactory auras, gross lesions have been found in the tip of the temporo-sphenoidal lobe, and in cases with auditory auras further back in the same region. In many cases of epilepsy, of course, even those in which special sense auras have been present, no detectable disease has been found, but there is little doubt that the central structures subserving such special senses have been morbid. And, as already stated, it is most important to inquire in each case into the presence of an aura, and if it be present

into its character, for there is little doubt that the primary discharge takes place at the part of the cortex subserving that sensation which the aura indicates. And it must also be remembered that the discharge may not spread beyond this point. We have already seen that in lowest-level fits and in middle-level (Jacksonian) fits there may be no general convulsion following the initial discharge. So also in these fits we may have, and in many cases do actually have, the aura occurring alone without any subsequent general convulsion. This fact would confirm, if further confirmation were needed, the view already expressed, that the seat of the initial discharge may be the only really diseased part, and that the discharge which occurs in other contiguous portions of the nervous system is a discharge in some way induced in healthy structures by undue activity in morbid structures.

Besides these severe attacks of epilepsy there are others, of much slighter character so far as their outward manifestations are concerned, but of no less importance to the patient. These are attacks which are known as *petit mal* and are vaguely described as "sensations," "turns," "lapses," "spells," etc. Sometimes these attacks are the only ones which occur. In other cases there are also attacks of severe epilepsy in the same patient. One of the most common forms which they assume is a momentary loss of consciousness, occurring suddenly, interrupting for a moment the patient in the middle of a sentence. In such an attack the sentence may be resumed where it was broken off and the patient remain quite unconscious that anything has occurred. Sometimes there is mental confusion, lasting only for a few seconds, and of this and this only the patient may be aware when he regains consciousness. Usually there is a slight change of colour, the eyes may become fixed and the pupils dilated. Sometimes there are vague and indefinite movements of the hands, not infrequently if the patient is walking or standing he suddenly falls but automatically picks himself up again, rubbing in an absent-minded way the part he may have bruised in his fall. Sometimes the attacks

are more than momentary. They may occur suddenly and unexpectedly and lead to a sudden and often severe fall. Occasionally they only occur when a patient is seated. Sometimes they are preceded by an aura similar to that occurring before severe fits, and the patient at once sits down, thus saving himself from many awkward accidents.

In short, these attacks of *petit mal* vary in severity and duration, from merely momentary lapses little noticeable by others and frequently unnoticed by the patient, to attacks in which consciousness is lost for minutes, and which may be accompanied by slight twitchings or local rigidity or deviation of the eyes, *i.e.* attacks scarcely to be distinguished from the milder form of severe epileptic fits. It must also be remembered that these attacks of *petit mal* may be preceded by any of the auras already described, and one of the most interesting varieties is that already alluded to in which "reminiscence" occurs, with, it may be, a crude sensation of smell and "smacking" or "champing" movements of the mouth. In some attacks of *petit mal*, even the slight ones, there may be involuntary passage of urine.

There is one condition of great medical interest and of the highest medico-legal importance which may occur as a sequel to attacks of *petit mal*, that, *viz.* which is known as "post-epileptic automatism." After a slight attack—and it is to be noted that such attacks are often so slight as to pass almost, if not quite, unnoticed—the patient may go through some elaborate and apparently purposive act. He may, *e.g.* begin to undress himself, and if he is interfered with, or resisted, may become violent. One patient of the Queen Square Hospital had attacks of severe epilepsy, and also of *petit mal*. On one occasion he was seized with a slight attack in the street. It was apparently momentary, but after it was over he began to undress himself where he was. A rough interference with him rendered him, not actually violent, but resistive. He was taken in charge by the police, brought before a magistrate, who ridiculed his story and fined him five shillings, or twenty-four hours' imprisonment. Another patient, a poor young woman, subject

to attacks of *petit mal*, had one in a shop to which she had gone to buy some trifling article. After the attack she made her way behind the counter and began rearranging the articles on the shelves. She, also, was given in charge, but fortunately encountered, not a superior magistrate, but an intelligent inspector, to whom she showed her hospital card. Another instance which illustrates the great medico-legal importance of this automatism happened at Queen Square a few years ago. A patient was shown into the consulting-room, and while telling his story he was noticed to stop, suddenly become pale, and to look absent-minded. This only lasted for a few seconds, when he immediately made a dart for the fireplace, seized the poker, and was about to assault the physician, whom he had never seen before, when he was fortunately seized and overpowered by the students who were in the room. Such a case illustrates the great medico-legal importance of this condition of post-epileptic automatism, for there is little doubt that crimes are occasionally committed—even homicide—by patients while they are in this irresponsible condition, and every case in which such a plea is urged as an explanation of apparent murder has to be carefully scrutinised.

There is little doubt that most cases of what has been called "proemsive" epilepsy are instances of automatic uncontrolled movement after an attack of *petit mal*. In this variety the patient suddenly starts running with great vigour and at his utmost velocity, occasionally also shouting loudly. He usually runs until he is exhausted, and when he comes to himself he is quite unable to account for his behaviour.

The explanation of these cases, whether of violent physical effort or of apparently purposive and deliberate elaborate actions after an attack, is probably to be found in the paralysis of higher structures which follows discharge in them, and the consequently uncontrolled action of lower and more automatic structures.

*Time and occurrence of fits.*—The time of the twenty-four hours at which fits occur is of considerable importance

in reference especially to treatment, as we shall see later. In some cases—probably in most cases—the time is variable, and the patient may have a fit at any hour of the twenty-four and the onset may be during the sleeping or waking hours. In a large class of cases the fits occur only during the day: in another smaller class the fits are called “nocturnal” because of their common occurrence at night. But a careful inquiry in such cases will usually establish the fact that the determining cause of a fit is not the time, but the condition of sleep, for which, of course, the night is particularly the time. In many of these cases of so-called nocturnal fits it will be found that there is a history that a fit has occasionally occurred during the day, but only during a sleep in which the patient has indulged in the daytime. So that in such cases the determining cause of a fit is apparently some condition which arises during sleep, most probably a modification of the circulation occurring under such conditions, giving rise to instability in certain cells. This, as we shall see, has an important bearing upon the treatment to be adopted in such cases.

In another class of cases the fits occur only in the early morning while the patient is dressing. In such cases also it is probable that some circulatory condition is the determining cause of the onset of the fits at such a time, and the recognition of the definiteness of the time at which fits occur in such cases is also most helpful in determining the treatment.

*Conditions modifying the frequency of fits.*—There are several conditions which modify the frequency with which fits occur. Of these the most important as affecting all ages is acute illness. During an attack of pneumonia, *e.g.*, a patient the subject of epilepsy, even if the fits have previously been of daily occurrence, is not likely to have any. So long as the temperature remains up, so long will the convulsive tendency remain in abeyance. The same is true of most other feverish conditions, although it is equally true that in certain kinds of fever, *e.g.* scarlet fever, probably on account of the toxic condition of the blood, convulsions may

arise *de novo*, and an epileptic weakness already present may be rendered more definite. Still it is broadly true that feverish conditions occurring in an epileptic patient have the tendency to abolish fits during their persistence.

Although we are here chiefly concerned with epilepsy of the young, it may also be mentioned that menstruation and pregnancy have a considerable effect on the occurrence of fits. In many young girls the epileptic fits to which they are subject occur only in relation to menstruation. Sometimes they occur before the period, sometimes immediately after it, sometimes only during it. Even if fits occur at other times in such cases, there is such an obvious tendency for them to be grouped around the period that we are forced to the conclusion that the physical disturbance occurring at that time has a very close association with the onset of the fits. In a similar way, pregnancy in many cases has a curious and variable influence upon the fits in an epileptic. In some cases this influence is of the slightest—fits occur during pregnancy neither more nor less frequently than at other times. In other cases—and these are somewhat unusual—the fits are increased in frequency during pregnancy. This, having regard to the occasional occurrence of eclampsia and to the nervous conditions generally present during pregnancy, is only what was to be expected, and it is surprising in some ways that it is not the rule. In another class of cases no fits at all occur during pregnancy, although they may have been frequent before and may resume their frequency when the pregnancy is past. It would thus seem that in certain cases of epilepsy pregnancy has the effect on the fits which we have seen that acute disease has in most cases. The reason for this is not clear. It may, conceivably, be related to the cardiac hypertrophy which is present in pregnancy, and, at least, it is probably to be associated with that feeling of vigorous health and unusual *bien-être* which so many women experience during pregnancy.

*Etiology and pathology.*—There is very little doubt that many cases of epilepsy owe the origin of the epileptic attacks

to the convulsions which occurred in infancy from one of the various causes alluded to under that heading. In many, no doubt, the convulsive tendency remains hidden and unobserved, as it were, until some crisis, such as puberty, or some mental or physical shock, gives rise to a condition of what we may somewhat indefinitely call general nervous tension, which brings out the latent instability in certain nerve-cells. In all cases of epilepsy there is, we believe, this instability of cells in one or other of the levels already alluded to, and in most this instability is due to the impress made upon such cells in early life. It is not likely that in many cases there is any anatomical change in the cells, such as can be recognised by even our finest methods. It is not improbable, however, that future research may disclose even structural alterations in some cases of epilepsy. Such a view brings all cases of epilepsy into line. We know that in many cases of Jacksonian epilepsy there are gross structural changes present, changes which we are able to recognise as morbid. In some cases of epilepsy, of the so-called idiopathic variety, changes have also been found. Thus in one case of epilepsy, with dreamy state and a crude sensation of smell, recorded by Dr. Hughlings Jackson and Dr. Colman, a small cyst was found in the uncinate gyrus of the temporo-sphenoidal lobe. And it is at least not unlikely that in other cases, those especially in which the fits arise in adult life in a patient without any history of predisposing or determining illness, the actual cause is to be found in some, probably recognisable, morbid change in nervous structures. And there is little doubt but that in the great majority of cases of epilepsy such changes are situated in the cortex cerebri. Ponto-bulbar fits undoubtedly occur: we know that fits occur as a result of gross irritative lesions in the Rolandic area: we know also of fits occurring as a result of lesion in certain other parts, *e.g.* the temporo-sphenoidal lobe, and there can be no reasonable doubt that the majority of cases of what has been called idiopathic epilepsy have their cause in some morbid condition of the cerebral cortex.



*Diagnosis.*—The diagnosis of epilepsy is not always easy. This is especially the case when hysteroid manifestations are associated with true epilepsy. Such manifestations are peculiarly obtrusive, and, unless it be recognised that they are very prone to follow very slight attacks of *petit mal*, their true significance is apt to be missed, and the preceding epileptic attack to be ignored. The convulsions of rickets, and in association with dentition, have been alluded to already. Their close relationship to epilepsy is undoubted, and they are frequently in early life the determining cause of the epilepsy which supervenes later. It is not likely that the convulsions, sometimes general, more frequently of the Jacksonian variety, which occur in tubercular meningitis, will be mistaken for anything but what they are. Uremic convulsions are, as convulsions, not to be distinguished from epileptic fits, and the same is true of the convulsions which are, not infrequently, associated with juvenile general paralysis. But an examination of the patients and the recognition of associated signs and symptoms should prevent any error. Similarly, the localised, usually unilateral, fits which occur in cases of infantile hemiplegia will be easily distinguished on account of the associated paralysis. When, however, the paralysis is very slight, so as almost to escape notice, and when, as sometimes happens in such a case, the fits are rather of the nature of *petit mal* than of severe attacks, the real nature of the attacks may escape notice. In one case of this kind there was only slight weakness of the hand, and the attacks were slight and momentary, followed by automatism. Such a case clearly indicates the essential identity of epileptic and epileptiform convulsions. Reference need only be made to the convulsions which occur in association with lead-poisoning, from alcohol, and from absinthe. These are seldom, if ever, met with in young subjects.

*Prognosis and course.*—In order to arrive at any decision as to the prognosis in any particular case of epilepsy, the course which the disease follows must be recognised. Great variation in this is met with. In some cases fits occur at

long intervals of months or even years, quite apart from any treatment. In other cases the patient may have five or six fits in a day and then go for months without any. Such possibilities must be recognised in order not to attach undue importance to the effect of drugs that may be administered. It seems to be fairly certain that in cases in which fits have started in very early life, in which there has been persistent and frequent repetition of such attacks, there is less likelihood of materially modifying the course of the disease than in cases in which, even if there have been convulsions in early life, a long interval of freedom has been experienced. And in the former class of cases mental deterioration is much more likely to take place than when fits occur only at rare intervals. Undoubtedly, also, in many cases a spontaneous cure occurs; so that, while every case of epilepsy is necessarily of such a nature as to give rise to considerable anxiety in reference to the patient's future, there are many points in most cases, a consideration of which enables one to take a not by any means hopeless view of any particular case.

*Treatment.*—Good hygienic conditions are, of course, essential in the treatment of epilepsy. It is often necessary to give advice as to the future of an epileptic child and, other things being equal, an open-air life is to be preferred. Diet is unimportant, except that it should be simple and wholesome, mixed animal and vegetable, with, on the whole, a small allowance of red meat. A number of hospital patients tried on vegetarian diet did not improve. The best results—these, indeed, extremely good—are obtained when the patients are taking a generous mixed diet.

In regard to drugs, the bromides are our sheet anchor. It is the writer's custom to avoid now the bromide of potassium, giving instead the bromide of sodium, for the reason that it is equally efficacious in controlling the fits, and less depressing. The dose, of course, varies with age, but should be large. Ten grains three times daily is not too much for a child aged 6 years if the fits are severe and frequent. Bromide of zinc, which can only be given in small doses of

two or three grains at a time, because of its emetic properties, is useful, in association with the other bromides, especially in cases of *petit mal*. Bromide of strontium is the best salt to use if acne should be troublesome. It is given instead of the bromide of sodium. In some cases equal quantities of the mixed bromides of ammonium, potassium, and sodium seem to be more efficacious than a single salt. Borax, in doses of half the number of grains of bromide with which it should be given, is very useful. It is not efficacious alone, except in doses which are apt to set up severe gastro-intestinal symptoms. Nitro-glycerine in small doses is also sometimes useful. It should be given with bromide in an acid mixture. Valerianate of zinc seems to have some influence in hysteroid conditions.

It is a point of much importance to prevent the depression which all bromides are apt to induce. The combination with bromide of small doses of *nux vomica* is very useful for this purpose. It does not interfere with the effect of the bromide on the condition of the nervous system underlying epilepsy, and the painful depression with which one was very familiar at one time is not now nearly so frequent if this drug is used.

In cases in which there is reason to suppose or suspect that circulatory modifications are related to the fits, the use of digitalis as well as of bromide and *nux vomica* is extremely useful. This is particularly true of cases of so-called nocturnal epilepsy—cases in which the fits occur during sleep. These are most satisfactory cases to treat, and a single dose of bromide, with *nux vomica* and digitalis, given at night is usually quite effective in controlling the attacks. Belladonna is an ancient and most useful remedy: it is not easy to say in which class of cases it is most useful. Chloral hydrate in small doses has also been found useful in cases of *petit mal*.

Much can be done to render drugs most efficacious by a consideration of the times at which the fits in a given case are most apt to occur. If they occur indiscriminately, recourse must be had to the regular administration of the

remedy decided upon, three or four times in the course of the day. If they occur at a special time, a larger dose should be given an hour, or two hours, before this. Such a plan sometimes has the effect of moving the fits to another time, and then they must be, so to speak, followed up and attacked in their new position. For nocturnal fits, the medicine should be administered in one dose, at bedtime, and no food should be allowed during the last hour the patient is out of bed. If fits occur during the time the patient is dressing, some food, *e.g.* a little milk, with a dose of medicine, should be administered half an hour before the patient gets out of bed. Observation will convince anyone of the extreme importance of following out such details of treatment as these examples indicate, and the results will be such as to gratify all concerned.

One word may be said in reference to the removal of peripheral irritation. If phimosis be present it should be corrected, but the writer has never yet known fits cured by circumcision. The same is true of intestinal and every other form of irritation. These should be removed, for in a patient with an unstable nervous constitution a straw may turn the balance for or against him.

In reference to the larger question of operation on the brain for epilepsy, it need only be said that operations for symptomatic or traumatic fits have not been so strikingly successful as to lead one to urge the extension of surgical measures to cases in which it is impossible to definitely indicate the part of the brain in which the discharge determining the convulsion starts.

## JUVENILE GENERAL PARALYSIS

This is a condition which remained undescribed until in 1877 Clouston published the first case, under the name of "developmental general paralysis." Since then, and especially in the last few years, many cases have been described clinically, and in a considerable number the diagnosis has been confirmed by *post-mortem* examination. So that the disease is now well recognised, and with the increasingly definite description which a wider experience has made possible, the frequency of its occurrence will, no doubt, be found to be greater than has been suspected.

*Symptoms.*—Advice is frequently sought in such cases either on account of mental change, or of bodily weakness, or because of the occurrence of fits. Occasionally an affection of vision, due to commencing optic atrophy, is an early symptom. Sometimes the patient is said to have been always backward; in other cases quite the opposite has been the case, the child has been unusually sharp and intelligent, has won numerous prizes, and indeed is of the same intellectual type as the adult general paralytic. At or about the time of puberty usually—although the age of onset varies between 8 and 23, strikingly similar in this respect to cases of juvenile tabes dorsalis—a change occurs which may be produced by an acute illness or by an accident, or may come on gradually and apparently spontaneously. The affection is found with almost equal frequency in the two sexes, thus differing in a very striking manner from the adult disease, which occurs so much more commonly in males. The mental symptoms are essentially those of a progressive dementia. Sometimes, as has been said, these are gradually engrafted on a mental condition which has never been normal. Occa-

sionally they arise in a boy or girl who has had fair or even unusual intelligence. The onset may be so gradual as to attract little or no attention for a time, unless the patient has duties to perform which call for intelligence and skill, and his inability to perform those duties reveals the mental and perhaps even the physical change. Occasionally mental depression may occur early in the disease. Delusions of persecution have also been present. So have delusions of grandeur, although, compared with the frequency with which these are present in the adult disease, they are rare. Dulness, apathy, loss of memory, sometimes stupor, are the marked mental symptoms, although these may occasionally be varied by an outburst of excitement or even of passion. But such variations only leave the patient mentally weaker and the condition gradually becomes one of profound dementia.

*Physical symptoms.*—Probably the most striking physical condition is the imperfect development of the body in the great majority of cases. The patients as a rule are undersized and the state of development of limbs and trunk is poor. Frequently the reproductive organs are infantile in type. In boys the testicles are usually very small, occasionally one or both are undescended or imperfectly descended, and the condition of the breasts may closely resemble that in females about the same age. Weakness of the limbs is almost constant, and in the late stages the lower limbs may be almost completely paralysed and have undergone contracture. Tremor of the hands, tremor of the tongue and in the facial muscles are almost invariable; occasionally marked ataxy is present and sometimes lightning pains. The knee-jerks as a rule are very active, Babinski's sign may be present, but rarely ankle clonus. In a certain number of cases the knee-jerk is abolished. This was the case in four out of Dr. Mott's twenty-two cases, and it probably represents the usual proportion in which this occurs.

Pupillary symptoms are almost invariable. Sometimes there is only inequality or irregularity: as a rule the light re-

action is very sluggish or completely absent; sometimes the reaction during convergence is also absent. Optic atrophy may also occur, and this may go on to cause total blindness. In cases in which it is an early symptom it may persist alone for a long time. Many of the cases bear on them the definite stigmata of congenital syphilis—Hutchinson's teeth, scarring about the mouth, interstitial keratitis, or disseminated choroidal atrophy. In thirteen out of Dr. Mott's twenty-two cases such stigmata were present; and in others the family history was more than suggestive, as will be pointed out when the etiology is considered. And it should also be remembered that in some cases of congenital syphilis in which paraplegia is present from cord affection, there is also frequently some mental change which may at any time assume the type of that present in juvenile general paralysis.

*Course and termination.*—The disease, of course, always terminates fatally. Its duration has varied from six weeks to seven years. But in the cases which terminate early this result has been due to some intercurrent and accidental complication. The usual duration is from two to three years, as in the adult, and, as a rule, male cases end fatally sooner than female, and the cases which begin early last as a rule longer. The course is one of gradually increasing mental and bodily weakness, and death usually is the result of some intercurrent complication or of exhaustion. Occasionally convulsions precede the end and hasten it, and in females phthisis is a frequent complication and cause of death.

*Etiology and pathology.*—While there seems to be no doubt that some illness, or accident, or some physiological strain, such as that involved in overwork at school, or of pregnancy, or lactation, may be the immediately exciting cause of the onset of the symptoms, the preponderating influence of inherited syphilis as the cause of the condition seems with increasing evidence to become more and more certain. In not one of Dr. Mott's 22 cases could syphilis be excluded. In 13 of these there were definite signs of syphilis on the body. In 4 the history was doubtful, but

syphilis could not be excluded in these. In the others, although there were no signs on the body, there was a definite maternal history of miscarriages, still-births, etc., and in one case there were signs of infection in the mother, and a sister had keratitis and Hutchinson's teeth. Similarly Thiery, in his analysis, finds syphilis in 64 per cent. of the cases—a proportion which, he says, is too great to permit of syphilis occupying merely a secondary place in the production of the disease. Occasionally, syphilis acquired in early life from a wet nurse, or from an accidental infection—*e.g.* a kiss on the lip—seems to be the cause of the disease. Of course it is possible that some neuropathic taint may determine the direction of degeneration, so that in some instances, under similar conditions, general paralysis may develop, in others juvenile tabes dorsalis, and in others merely imbecility, associated, it may be, with symptoms of bodily disease. Yet the influence of neuropathic heredity is small compared with that of hereditary syphilis. In three of Dr. Mott's cases the fathers had suffered from general paralysis. Although there was no sign of syphilitic residua in any one of those three fathers, there were in the children of two of them undoubted signs of hereditary syphilis, and the maternal history of the other, made its existence practically certain. In 12 out of his 22 cases there was no history of direct or collateral insanity at all, and he is probably correct in his view, contrary to that of Thiery, that hereditary neuropathy is not a prominent factor in the production of the disease.

*Morbid anatomy.*—The characteristic changes in the brain in this disease are marked atrophy, especially in the frontal and cerebral convolutions, thickening and adhesion of the pia-arachnoid membranes, dilatation of the ventricles, and a granular condition of the ependyma. Occasionally there is an internal pachymeningitis, with the formation of a subdural false membrane. It is noteworthy that in six cases in which the hemispheres were weighed there was an average difference of 28 grammes, the right being in every instance the heavier. In all these cases there was a marked



affection of the speech function, which, as we know, is chiefly situated in the left hemisphere—the more diseased and atrophied. Microscopical examination of the brain reveals a condition closely similar to, if not identical with, that in the adult form of the disease, viz. atrophy of the tangential fibres, and atrophy and disintegration of the neurons of the cortex, especially of the frontal and central convolutions. The occipital region seems to be but slightly affected. There is also a marked overgrowth of glia cells and a cellular proliferation around the vessels. According to Alzheimer there is atrophy of the cells of the third and fourth layers, while those in the superficial layers escape. Mott could not confirm this change. Degenerative changes, although less marked, are also found in the nervous and connective-tissue elements of the cerebellum and the basal ganglia, and not infrequently changes characteristic of congenital syphilis are found in other organs. In four of Dr. Mott's cases there was pericellular and perivascular fibrosis of the liver, such as is met with in cases of hereditary syphilis in which the nervous system is not affected.

*Diagnosis.*—The diagnosis of this condition is by no means easy in certain cases. As has already been mentioned, in some of the cases a defective or at least somewhat backward mental condition has been present from birth. When dementia becomes established, if its association with other symptoms suggesting general paralysis is not yet clear, it will not be easy to be quite sure that the case is of this nature. But an examination into the condition of the reflexes, especially of the pupillary light reflex, will often be sufficient to suggest at least, the true nature of the disease. When fits are present, the condition most likely to be mistaken for general paralysis is epilepsy, and the association of mental deterioration will not, of course, negative such a diagnosis. But not infrequently the nature of the mental changes, and probably the presence of some of the physical signs and symptoms already referred to will be sufficient to lead to a correct diagnosis. In that class of cases in which there has been a period of active, perhaps occasion-

ally of almost precocious, intelligence, the diagnosis is not so difficult. There are comparatively few diseases which are characterised by the onset of gradual mental deterioration at such an early age. Epilepsy is, of course, one, and has been already alluded to. Another is cerebral or cerebellar tumour, and when localised convulsive seizures are present the diagnosis may offer some difficulties. But the condition of the pupils, the absence of optic neuritis, and usually also of both headache and vomiting, and perhaps the presence of tremor and articulatory difficulty, will indicate the true nature of the condition. And it must also be borne in mind that the presence of definite stigmata of congenital syphilis is, on the whole, confirmatory of a diagnosis of general paralysis. Some cases of disseminated sclerosis resemble closely, in certain particulars, cases of general paralysis. But, although sometimes there is a slight degree of mental change in disseminated sclerosis, and usually in the direction of the "facility," which is the most common alteration in the early stage of general paralysis, the nature of the tremor is different in the two diseases. There is usually no change in the pupil reaction in disseminated sclerosis, and there are no fits.

*Prognosis.*—The prognosis is, of course, bad. Patients with this disease all die, although, as a rule, they live longer after its onset than do the subjects of the adult disease. A patient may live as long as seven years. As a rule the disease is fatal in three or four, even if no intercurrent complication or acute exacerbation, such as a severe convulsive seizure, bring about a fatal result at some earlier time—occasionally, indeed, so early as only a few weeks after the recognition of the early symptoms or signs.

*Treatment* so far has proved quite inefficacious. The use of mercury and iodide of potassium is, of course, indicated, but the constant experience of the uselessness of these drugs points clearly to the probability that the disease is the result, not of true syphilitic lesions, but of post-syphilitic degeneration.

## CRETINISM

THE condition to which this name—obscure in its derivation—is applied has received much attention of late years in consequence of its relations to myxedema and other diseased conditions of the thyroid gland, and its amenability, in some degree at least, to treatment by the administration of thyroid substance. It occurs as an *endemic* disease in various parts of the world, especially in such mountainous regions as Switzerland and the Austrian Tyrol. In this endemic variety a strong family tendency is present, and the stunted growth and other appearances peculiar to the condition are frequently associated with the presence of an enlarged but diseased thyroid gland. The relation of cretinism in the child to goitrous disease in one or both parents in these countries is also, according to Koehler, very close, and there seems to be no doubt that the distribution of endemic cretinism is similar to that of endemic goitrous disease—a relation which is regarded by the same authority as the result of infection through the water of the districts.

Besides this endemic variety of cretinism there is also a *sporadic* variety which occurs fairly frequently in this and in other countries. In this condition it is, as a rule, impossible to verify the presence of any thyroid gland, and even if it is present it is diseased, or degenerated, or undeveloped. So far as appearances go this absence of thyroid in cases of sporadic cretinism seems to constitute the only distinction between the sporadic and endemic varieties. In the latter, as has already been stated, there is frequently present a goitrous swelling, indicating, not an enlarged, but a diseased or degenerated thyroid.

*Symptoms.*—In cretinism the mental and physical de-

velopment is delayed. An untreated cretin at the age of 18 or 20 may have the appearance of a child of 2 or 3. The bridge of the nose is depressed, there is a small palpebral aperture and a protruding tongue. The abdomen is large, there is nearly always a prominent umbilicus, not infrequently an umbilical hernia. The fontanelle is usually late in closing and the eruption of teeth is delayed. The skull may, however, assume a correct form and even an ordinary size. Speech is much delayed, often very imperfect, and the voice has a peculiar harsh croaking character, and there is usually snoring during sleep. There is no definite paralysis, but the muscular power is weak, the vital powers are low, there is much sensitiveness to cold, and the temperature is nearly always subnormal. The skin is usually swollen, dry, and scaly, and the hair is hard and stunted and scanty. The viscera, as a rule, display no abnormality.

It must, however, be remembered that there are considerable differences, both in the physical and mental characteristics of different cretins. Many are physically exceedingly small and infantile in their proportions. Others are of somewhat larger proportions, although none approach anything like the normal size. Similar differences are found in their mental qualities and characteristics. Occasionally a cretin is quiet, placid, and good-natured: much more frequently, however, restlessness, destructiveness, and viciousness are the outstanding qualities, and under the very best conditions the mental state is one of great backwardness.

Before the introduction of thyroid treatment for myxœdema it had been recognised that cretinism was really a form of myxœdema, and this view is borne out by the effect of thyroid treatment in the disease. In the older cretins the treatment is almost completely ineffectual. This, of course, was only to be expected, but even in those the modification which is induced by the administration of thyroid—especially on epidermal structures—is often remarkable. In the young cretin, however, the result is

most striking. The writer had under his care a typical cretin aged  $4\frac{1}{2}$  years. From the illustration (Fig. 23) it will be seen that she exhibited all the characteristics of the disease—dry scanty hair, thick hard skin, a prominent umbilicus, and a fatuous expression. She also had a harsh croaking voice, was dirty in her habits, and had never had any teeth. Two months of treatment with thyroid powder effected a complete transformation. Her hair became soft



FIG. 23.—Cretin, aged  $4\frac{1}{2}$  years, before treatment was commenced.  
From a photograph by Dr. Guy Wood.

and abundant, her skin moist and natural, the umbilicus receded, she had an eruption of her milk teeth, and she became bright in her mind and clean in her habits (Fig. 24). It is now ten years since the treatment was started; and, while she is smaller and less intelligent than girls of her age and than her own younger sisters, she is fairly developed, is able to help in housework, and so far as her physical condition is concerned is quite normal. She still continues taking thyroid daily, and only once relapsed, when it was intermitted for a few weeks. Of course such

progress in a case in which the treatment was started at  $4\frac{1}{2}$  years suggests a very hopeful outlook for cases in which treatment can be begun when the condition is first recognisable.

The *etiology* of the condition, except that it is related to absence or disease of the thyroid gland, is obscure. Why certain localities, as in the endemic variety, should favour its occurrence is possibly due, as Kocher thinks, to infection



FIG. 24.—The same child after two months of treatment with thyroid substance. From photograph by Dr. Guy Wood.

through the water. And in this variety also the relation to goitrous disease in the parents seems to be undoubted. In the sporadic cases various influences have been invoked as explanatory—alcoholism in parents, shocks to the mother during pregnancy, etc. The one constant condition, however, is absence or disease of the thyroid gland, and isolated cases occur in large and otherwise absolutely healthy families. The case referred to above was the sixth child in an otherwise quite healthy family of ten.

The *diagnosis* is not as a rule difficult. Cases of so-called

Mongolian imbecility have sometimes been mistaken for cretins, and the smallness of the child, its backward mental state, and its large tongue and fatuous expression, excuses such a mistake. But in Mongolian imbeciles the condition is present at birth, the thyroid gland is normal, the child is smiling and usually active, and deformity of the fingers, and not infrequently congenital heart disease, are present. In cretinism, on the other hand, the condition is not, as a rule, obvious before the sixth month, there is no deformity of the fingers, and visceral disease is absent. The inefficacy of the treatment by thyroid substance in Mongolian imbecility is also in startling contrast to its marked effect in cretinism. The condition known as achondroplasia (*q. v.*) has also been mistaken for cretinism, but to anyone who has seen a case of this disorder the clinical picture, showing a large head, short humeri and femora, and the characteristic deformities of the hands, is so definite as to make its recognition easy. Certain conditions of infantilism also may occasionally resemble cretinism, but are, as a rule, to be readily distinguished by a consideration of their distinctive features.

The *prognosis* in cases of endemic cretinism seems to be very bad, and thyroid substance, although effective to a certain extent, cannot be considered curative. In the sporadic cases, on the other hand, the prognosis is good if the condition is recognised early and suitable treatment adopted. Whether complete cure, involving the building up of a strong body and a healthy mind, is attainable in any case remains to be seen. It seems to be at least a possibility, if the condition is recognised quite early and a suitable dosage of thyroid given continuously.

The only *treatment* to be adopted, apart from the adoption of suitable hygienic conditions and the administration of good food, is the administration of thyroid gland substance in some concentrated and effective form. There are now numerous and very reliable preparations, both liquid and solid. The quantity to be given varies with the individual, and must be arrived at by patient trial. It is well to begin

with a dose twice a day, small at first, and to go on increasing it until physiological effects—rapid pulse, sickness, etc., are produced. As soon as this stage is reached reduction may take place, but regular administration must be continued, and even after the characteristic features of cretinism have all disappeared a quantity—usually small, but variable in different cases—must be given, and will probably have to be continued during the life of the patient. It must always be remembered that it is not a drug that is being given, but that a certain physiological substance, necessary for the bodily economy, which the body itself cannot, or at least does not, manufacture, is being supplied from an artificial source.



## MONGOLIAN IMBECILITY

This name may be neither a scientific nor a particularly suitable one, but it is convenient, and it is to a certain degree descriptive of a fairly large group of cases, familiar enough to the alienist physician, but seen so often in the ordinary course of medical work amongst children as to make its recognition important. Dr. Langdon Down is responsible for the name, and to anyone who has seen one or more of the cases to which it is applied, with their round, moon-like faces, their oblique and narrow palpebral fissures, so suggestive of Mongolian or Kalmuc features, the name will certainly appear not inapt.

As a class these imbeciles are marked by certain physical and mental peculiarities which necessitate their separate description. These are so striking as to definitely mark out the group, and individual cases of the disease resemble one another so closely as to be readily and easily mistaken one for another. At birth the infants are small, and growth is unusually slow. The skull is small and short at the base, globular and smooth on the vertex. The narrowness of the palpebral apertures and their obliquity have already been referred to. The mouth is small and is usually kept open, the tongue is large, often protruding, and the nasopharynx small, so that respiration is difficult, and during sleep snoring is almost invariable. These features are present and recognisable at the time of birth. The chest is usually normal, the pelvis is small, and the abdomen, consequently, protuberant. Umbilical hernia is common. The long bones of the extremities are shorter than usual, the joints are unusually loose and mobile—the result partly of laxity of ligaments, partly of hypotonia of muscles, and this can be

shown in a striking way by the ease with which the child can be placed almost in the intra-uterine position. The hands are short and square, and one very common deformity has been described by Dr. Telford Smith,—viz. a marked shortening of the second phalanx of the little finger and much lateral displacement of the terminal phalanx, producing a marked bowing of this digit. The thumbs also are often small in proportion to the other digits. Dr. John Thomson and Dr. Archibald Garrod have both directed attention to the frequency of congenital cardiac disease in these children. Dr. George Sutherland, whose writings on



Figs. 25 and 26.—To illustrate the features of Mongolian imbecility—Dr. Sutherland's cases.

this subject are of much interest and value, has also found evidence of congenital cardiac defects in 20 per cent. of his twenty-five cases. Occasionally, other evidences of maldevelopment are present, *e.g.* congenital club foot, imperforate anus, etc.

*Symptoms.*—The expression of the infant during rest or sleep may not appear at all peculiar or immature. When the child smiles the facial muscles cause a marked wrinkling of the skin, and the expression really becomes a grimace. In crying the lips become swollen and protruded, the eyelids also swell, and their margins may be inverted. While the child is awake, facial movements are active, and there is

frequently a habit of protruding the tongue, and drawing it back with a smacking noise. The lower jaw also is often protruded, and the upper lip sucked in by the lower. The ordinary crowing sounds of infancy are replaced by peculiar throaty noises, and the child is content to lie on its back for very long periods contemplating its own toes and uttering those throaty noises, apparently its mode of expressing pleasure or contentment.

The mental condition of these children is not characterised by any definite defect or perversion. The condition is merely one of placid backwardness and non-development of the natural interest and intelligence of the child. The mother is usually ready to give it a high character, as "the best baby she has ever had." It does not often cry, it requires little or no effort to amuse it, and is, in short, unnaturally free from vice. It is only when it reaches the age of 9 or 10 months without developing any attempts at talking, and without showing any of the natural perverseness of the infant, that suspicions are aroused that there is something amiss. Even then the mother is ever ready to repel any aspersions on its mental condition and stoutly maintains its intelligence. Yet in such a child the expression is obviously fatuous, and it has defective power of attention, as shown by its inability to follow objects with its eyes. The absence of the dulness and lethargy often associated with mental defect in children makes the recognition of its backward mental state all the more difficult.

The bodily and mental development proceed slowly; the teeth are cut late; walking is not usually learnt before the third or fourth year; and the physical condition remains weak, and the bodily powers feeble. There is usually a subnormal temperature, and the physical condition ultimately attained, should the child reach adult life, is not one of any degree of vigour. At puberty, it is said, there is frequently a tendency to the deposition of fat. The mental condition is one of backwardness and weakness. Education to a slight degree is possible, but ability to read more than short words of a few letters is rarely, if ever, attained. But the sufferers are un-

objectionable in their habits, although quite unable to earn their own living, and are merely pleasant and affectionate imbeciles.

*Etiology.*—From the nature of the condition and the fact that its features are present at the time of birth it would appear that there is some agent at work interfering with development from a very early period of intra-uterine life. Whether this is the result of some defect inherent in the embryo or a result of parental disease we do not know. Occasionally Mongolism is present as an isolated phenomenon in an otherwise healthy family, just as sporadic cretinism may be. Some observers have noted it in the last member of large families which may have no other diseased member. Others have been inclined to invoke syphilis as an underlying factor, regarding the condition as a parasymphilitic degeneration or non-development. Dr. Sutherland in his twenty-five cases had definite evidence of syphilis in eleven and suspected it in three more. It is certainly a very striking proportion. From his own observations the writer is not inclined to attach such importance to it as Dr. Sutherland does, yet it is difficult not to suspect its influence if more extensive statistics should bear out these figures. It is conceivable that the growth and development of the embryo may be interfered with in definite directions from different causes, and it is quite likely that parental syphilis may be one of those causes. And the fact stated above, that it sometimes occurs in the last member of a perhaps large and otherwise healthy family, would suggest defective vital endowment as a cause of the condition.

*Pathology and morbid anatomy.*—From what has already been said, it will be clear that no grave morbid changes are likely to be discerned underlying the mental and physical peculiarities described. The changes in the joints or ligaments, the deformity of the little fingers, the subnormal temperature and the cardiac defects are all signs of the defective development which underlies the condition. But it cannot be said that any definite change in the central nervous system has yet been found which explains these

defects. The small size of the base of the skull in the antero-posterior direction has been referred to. This, as has been shown by Fraser, who examined the skull of a female patient who died at the age of 40, does not depend upon the early ossification of any of the sutures. On the other hand, the structures at the base of the brain have been found by Wilmarth to be unusually small, the defect showing itself in the pons and medulla especially. Sutherland found a similar condition in a case which he examined. Such a defect might certainly explain the smaller size of the basal bony structures, and it is not improbable that it will be found to be associated with defects in the cellular groups not only at the base, but also in the cortex cerebri. At least such a condition of obvious want of development in certain parts of the nervous system indicates a line of investigation which may throw much light on the true nature of this condition.

The *diagnosis* of the condition is not difficult when one is at all familiar with the type. The resemblances to cretinism have already been mentioned. The differences are distinct. In Mongolism the condition is present at birth, the infants are smiling, grinning, and good-natured, and the skin is natural. The characteristic condition of the palpebral fissures has already been alluded to, and also the condition of the lips and tongue, and of the little fingers. Congenital heart defects are common, whereas in cretinism they are rare. Cretins, too, are dull and impassive, with dry skin and thick immobile lips and a mental condition by no means placid. As Sutherland remarks, "the smiling face of the Mongolian imbecile suggests the possession of some secret source of joy, while the somewhat sad countenance of the cretin suggests the cherishing of a secret sorrow."

The *prognosis* is essentially bad. No cure is to be hoped for, and while the educability varies in different cases it is never great in degree. The physical conditions of defective vitality and strength also are such as to predispose to disease (especially respiratory troubles) and early death.

Many of the children die in infancy from diarrhœa. And, even if infancy and childhood are passed and adult life is reached, the same mental condition persists, and is also associated with physical weakness, rendering survival to anything like advanced age almost impossible.

*Treatment* by antisyphilitic remedies, by thyroid, thymus, and other drugs, is quite inefficacious. Indeed, the only measures to be adopted are measures of general hygienic care, and for the mental state, the educative influences which are found at their best in special institutions. The cardiac condition gives rise, as a rule, to no symptoms, and the signs of its presence usually become less marked as the child grows.

## ACHONDROPLASIA

This is a condition to which the above name is now that most commonly applied, characterised by the large size of the head and the shortness of the limbs, both upper and lower, resulting in a condition of dwarfism of a characteristic type. Whatever justification is needed for its inclusion in a work like this must be sought for in the fact that it is apparently a condition depending upon mal-development, and that it has points of similarity to cretinism, so striking as to have occasionally led even good observers into an erroneous diagnosis. Parrot, in 1878, was the first to describe the disease clinically, and to insist upon the origin of the deformities in a defect in the development of the cartilaginous bones. Previous to this the condition had been described by anatomists and by obstetricians merely from premature or still-born children, and had been regarded as a kind of "fœtal rickets," differing essentially from ordinary rickets in the characters of the deformities produced, and in the fact that these were actually present at birth. Already, in 1856, Virchow had his attention occupied by certain fœtuses with unusually large heads and short limbs. Winkler, in 1871, distinguished the condition from ordinary rickets, and suggested the name of *rachitis micromelia*. Depaul also insisted upon the necessity of distinguishing the two diseases. Other observers, Müller, Porak, Kuebbing, Marchand, Pierre Marie, and Kaufmann, also wrote on the subject, the last-named suggesting the name of *Chondrodystrophia fœtalis*. This, however, is more cumbersome, and has no particular advantage over the name achondroplasia, which is that now most generally used.

The essential and obvious features of the condition are

the enlargement of the head combined with the shortness of the limbs. The trunk is practically normal and of a size suited to an individual of much higher stature. These



FIG. 27.—Case of achondroplasia, showing large head, characteristic facies, and shortening of humeri and femora.

characteristics are very well brought out by Pierre Marie, who contrasts the various measurements in two cases of achondroplasia with those in a normal child of 8. It will be sufficient if one of the subjects of achondroplasia



be used for comparison. This was a man aged 47 years, whose height was 122 centimetres compared with 116 in a child of 8. In the child, the upper limb, from the acromion to the tip of the middle finger, measured 51.5 c.; in the patient of 47 the corresponding measurement was 44.2 c. This shortening is apparently the result of disproportionate shortening of the upper arm as compared with the forearm, for whereas in the child of 8 the upper arm is 21 c. and the forearm 17.5 c., in the adult patient with achondroplasia the upper arm is 13 c. and the forearm 17.7 c. Similarly in regard to the lower limbs. In the child, from the great trochanter to the sole the measurement is 58 c., in the patient 51 c., and here, again, the greater shortening of the thigh as compared with the leg is quite obvious. In the child the thigh is 27 c., the leg 26.5 c.; in the patient the thigh is 23 c. and the leg 24 c. It is interesting to compare with this the trunk measurements from the pubis to the episternal notch. In the child this is 34.5 c., whereas, in the achondroplastic patient it is 47.5 c. It will be at once seen that with limbs in proportion to his trunk this patient would really have been above the ordinary height.

In regard to the head also, the measurements were very instructive. In the case to which reference has already been made the circumference was 67 c. The transverse diameter was 25 c., and the antero-posterior the same, giving a cephalic index of 100, which is a figure practically unique. As to the characters of the head, it resembles very closely a hydrocephalic head, being rounded with very well marked frontal and parietal bosses. The face is not relatively so small as one would expect. The features are large, the nose at its root flat with a big extremity and gaping nostrils. The palatal arch varies, being sometimes small, sometimes apparently normal. The teeth show nothing unusual.

Certain other peculiarities should be referred to. Lordosis in the lumbar region is almost invariable. Possibly this accounts for the tilting of the pelvis which is so often present, and which has probably some relation to the frequent difficulties with labour to which reference will be

made presently. The upper limbs also seem to have their attachment at a level posterior to that which is usual, so that they seem to hang back very much.

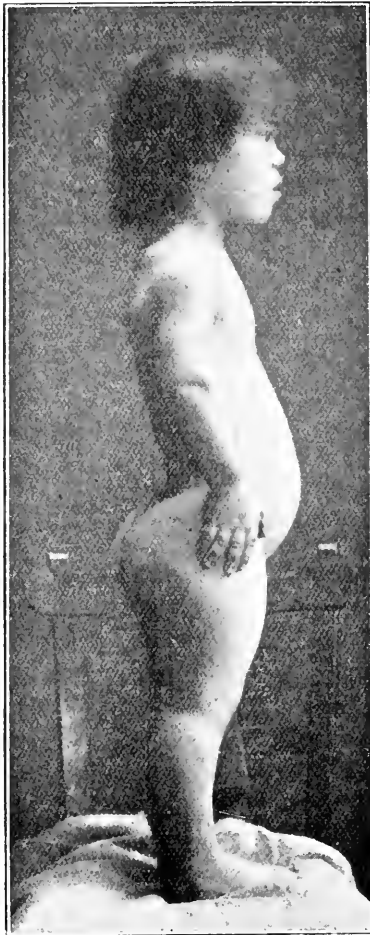


FIG. 28.—The same child, showing lordosis, etc., and the characteristic *main en trident*.

The peculiarities of the hands are very striking and constant. They are reduced in all their dimensions and the fingers are almost equal in size. The ring finger seems to be on a posterior level to the others, so that it is partly

concealed from the front by its neighbours. One very striking peculiarity is the fact that when the fingers are extended they are not approximated except at their metacarpal extremities, and so at their distal extremities there is an interval between them. This deformity has been named the *main en trident*. This condition may be compared with the tendency usually present in patients suffering from this disorder to have the upper limbs more or less separated from the trunk even if hanging down—the result, apparently, of the large size of the head of the humerus. The lower limbs themselves are curved as in rickets, but the curvature seems to be the result, not so much of actual bending of the bones, as if there were softening, but rather of a displacement at or about the level of the knee of the two parts of the lower limb. There is little or no bending of the upper limbs. The tendency to keep them not quite approximated to the trunk has already been alluded to.

The muscular development of these cases is usually extremely good, and the short thick-set body with marked musculature gives an appearance of strength which is not altogether deceptive. The weight of the body is also, as a rule, much greater than one expects, probably because of the greater density of the shortened bones. The sexual power varies. Occasionally the sexual organs are infantile, but some cases have undoubtedly been quite normally developed, and more than one achondroplastic woman has had a child.

In regard to intellectual features a few words will suffice. In all cases there seems to be a lowering of intelligence; in some this is so marked as to justify a description of the patient as weak-minded. But even in those in whom intellectual development has gone so far as to make it possible for the patient to read or write, the level of intelligence attained is always a low one and not usually sufficiently high to enable the patient to earn his own living.

*Etiology.*—The condition seems to arise between the third and sixth month of fetal life. Apparently the ossification which should go on in cartilage in the long bones ceases, while the bone seems to go on developing normally from

the periosteum. Hence the disproportion which undoubtedly exists between the length and the thickness of the long bones. On the other hand, the bones which are developed without passing through a cartilaginous stage—the clavicles, ribs, frontal and parietal bones, etc.—attain to normal form and dimensions, and hence arises the disproportion between the trunk and the limbs. The cause of this dystrophy of the primordial cartilage we do not know.

In reference to heredity it should be mentioned that the cases of achondroplasia are, as a rule, isolated instances of this peculiarity in a family otherwise healthy, the patient's brothers and sisters being usually of normal stature and development. There is no doubt that there have been instances in which an achondroplastic woman has had a child with the same conditions present. In some cases the child has had to be delivered by Cæsarean section, and it is possible that but for the deformity and tilting of the pelvis already alluded to such women might reproduce several children with the same disease or deformity. If this were so, it would justify the analogy to which some authors have drawn attention between achondroplasia in man and the curious anomalies characterising such dogs as bassets or dachshunds, in which the body is long and well-developed, yet in which the limbs are very short and the individual bones much stunted and bent.

*Diagnosis.*—When one is once familiar with the appearance of a patient with achondroplasia, its recognition is not difficult. The mistake has been made of regarding it as simple rickets, but a consideration of the actual condition and of the history will prevent such an error. Cases have also been taken to be a form of cretinism, and such a mistake might easily be made. But a consideration of the peculiar deformities in achondroplasia and of the appearance and general physical condition in cretinism will prevent such an error.

*Prognosis.*—This is, of course, bad so far as any hope of cure is concerned, and it is not likely that any form of *treatment* will be found to modify the condition.

## HEMICRANIA WITH THIRD NERVE PARALYSIS

A CONSIDERABLE number of cases have now been recorded of what has been called "migraine with recurrent paralysis of the third nerve." Charcot designated them with the name *migraine ophthalmoplegique* to distinguish them from the ordinary cases of migraine with visual symptoms—*migraine ophthalmique*, but it is probably better to dissociate altogether from such cases the name "migraine," which has a definite clinical connotation. In no case of true migraine in which any of the ordinary visual symptoms has been present—hemianopia, fortification lines, etc.—has third nerve paralysis ever been described; and conversely in no case of unilateral headache with third nerve paralysis have any of these visual symptoms been present. So it is better to regard the cases of ordinary migraine and of so-called "migraine with third nerve paralysis" as of distinct varieties, and probably preferable to mark such a distinction by applying the classical term "migraine" only to that class of cases with which it has been so long associated.

*Symptoms and clinical course.*—In a patient in whom paralysis of one third nerve supervenes after a severe unilateral headache, there is usually the history that unilateral headaches of a somewhat severe character, occasionally accompanied with vomiting, have been present for some time. After one of these headaches it is found that the eyelid on the affected side droops over the eye, and further examination reveals the fact that the ocular movements depending upon the third nerve are also in abeyance, or at least are interfered with. After a variable lapse of days the paralysis begins to diminish and finally may entirely disappear. In the course of time another headache comes on

and a similar paralysis is re-established. This also may clear up completely, or not quite completely. Each successive attack of paralysis usually leaves a residue of more and more weakness, until finally a time comes when complete paralysis of all the parts supplied by the third nerve is established. After this has happened the unilateral



FIG. 29.—Third nerve paralysis in hemiplegia. In this patient the paralysis passed off completely in three weeks. There has been slight recurrence since. From a photograph by Dr. Howland.

headaches still persist and often become increasingly severe. But frequently no further signs or symptoms are super-added, and the patient remains with ptosis, external strabismus, defective inward, upward and downward movement of the eye, and a liability to unilateral headaches.

In some cases, apparently similar in nature, paralysis of other cranial nerves—fourth, fifth, sixth, or seventh, as well as of the third—has been present. In one case both third

nerves suffered, and a slight degree of optic atrophy has also been described. Yet by far the most common variety—so much the most common as really to constitute a class—is that in which the third nerve is the only one affected, and that only on one side.

*Pathology and morbid anatomy.*—The commonly accepted pathological explanation of ordinary migraine is that it is a vaso-motor neurosis—some vascular condition, probably one of spasm of vessels—affecting temporarily a certain area of the visual cortex. In these cases of hemicrania with third nerve paralysis, however, as we have already said, no visual symptoms are present, and the only sensory effect is the severe pain on one side of the head. The remaining effect is a motor one, the result of interference with the functions of the third nerve. Mr. Holmes Spicer and Dr. Ormerod have contributed a very exhaustive paper on this subject,<sup>1</sup> and have quoted all the cases available at the time their paper was written. It is a significant fact that in all the cases in which there has been an opportunity of examining the condition *post mortem* some morbid condition—tumour, or exudation or granulation—has been found involving the trunk of the third nerve itself. It is probable that in all cases in which third nerve affection is present with hemicrania there is some actual change in the nerve trunk, and this condition explains why the recurrent paralysis so often tends to become a permanent affection. It can easily be supposed that at first the affection of the nerve may be slight, and that temporary congestion or inflammation may so interfere with it as to abolish its function for a time. When the nerve affection becomes more severe and more extensive, the function of the nerve will be more and more interfered with, until it is finally abolished, and so permanent paralysis of the structures supplied by it is established. Such a pathological explanation corresponds with the clinical history of most of the cases recorded.

*Diagnosis.*—A distinction will usually have to be made between this condition and one in which the third nerve is

<sup>1</sup> 'Ophth. Soc. Trans.,' vol. xvi.

the seat of an inflammatory—often a gummatous—process. Unilateral headache is not common in this latter class of case. It may, however, be present with the paralysis, but it will not usually have been present as a recurrent hemi-*crania*. Third nerve paralysis may also occur as an early sign of tubercular meningitis. It is also present in some cases of pontine tumour, but in neither of these conditions is there likely to be a history of recurring one-sided headaches, nor is there likely to be the clearing up of the paralysis which almost invariably occurs at first in these cases. There will also, of course, in both tubercular meningitis and pontine tumour, be present the general symptoms and signs of each condition.

*Prognosis.*—When once the diagnosis is made, the future of the patient, at least as regards the condition of the third nerve, is necessarily regarded with considerable apprehension. Even if the initial paralysis has entirely disappeared, it is to be feared that subsequent and recurrent attacks may leave the structures supplied by the nerve permanently paralysed. The danger to life may not be imminent. That must depend upon the nature of the morbid process which affects the nerve. But in most cases it may be presumed not to be any malignant growth, although, of course, it might easily be of that nature.

*Treatment* must unfortunately be symptomatic—for the sensory symptoms, *e.g.* by phenazone, phenacetin, migranin, etc. It is probably also desirable, in some cases at least, to administer iodide—either iodide of sodium or iodide of iron—on the chance of influencing the assumed inflammatory condition giving rise to the temporary symptoms.



## CONGENITAL DEFECTS IN OCULAR MOVEMENTS

THERE seems to be very little doubt that in certain families there exists a tendency to the occurrence in different members of a defect in ocular movements. Cases of this character have been described by observers in different countries, and in many instances the defects have been very similar. These congenital defects are apparently the result of developmental anomalies affecting the ocular muscles; but, so far as we are aware, no really searching investigation has been made into the condition of the nervous system. The usual characteristics are the existence of ptosis, usually on both sides. This, of course, leads to a constant wrinkling of the brows in the attempt to raise the eyelids. With the ptosis there is usually associated a defect in the upward, and also in the downward, movement of the eyeballs. The lateral movements are usually of moderate extent, and the best movement of all as a rule is convergence. Frequently there is an appearance of absence of any of the movements effected by the oblique muscles. Occasionally, however, rotation is present of such a character as to indicate the probable presence of one or other, or both, of these muscles. As a rule, diplopia is not a symptom. Visual acuity has been good, and no evidence of an affection of the internal ocular muscles has ever been present. The children suffering, also, have in no case been affected with any of the usual congenital developmental defects, such as hare-lip, cleft palate, or colobomata. As has already been said, the condition, as shown by *post-mortem* examination, has been, apparently, the result of complete absence of the muscle or muscles of which the movements are defective; or if the muscles have been present they have been ill-developed, of insufficient

length, or attached to the eyeball in an abnormal position. No exhaustive examination of the nervous system in any case has been made, but it is highly probable that these muscular anomalies are associated with defects in the cells of the ocular nuclei.

## HEREDITARY OPTIC ATROPHY

(LEBER'S ATROPHY)

SEVERAL diseases have already been considered which are characterised by a tendency to occur in several members of the same family, and often, also, to occur in successive generations. It has been known for many years that there are cases possessing those two features, in which impairment of vision is the important morbid condition. In 1871 Leber wrote a most instructive paper referring to such conditions generally, but dealing particularly, as a result of his own observation, with a remarkable series of 15 cases, occurring in four families. He mentioned at the same time 3 cases reported by von Graefe from a single family. In a later paper he added others, bringing up the total to 55 cases, occurring in sixteen families.

In the preliminary part of his paper Leber referred to several instances previously recorded of visual affection of different members of the same family, and even in different generations. In one series a curious tendency had been marked to the occurrence of the visual failure in the dark-eyed members of the family; in another series of cases the sons became affected, and the daughters remained exempt. But as all these cases either occurred in days before the ophthalmoscope was used, or no satisfactory ophthalmoscopic examination had been made, they must be put on one side as of an indeterminate character, and not necessarily of the same class as those which Leber himself described.

As has already been stated, the disease described by Leber affected several members of the same family, and occurred in several generations. He found that it com-

menced, as a rule, a few years after puberty. In one case, however, he found it present as early as 5, and in another it commenced as late as 43. Occasionally no previous history was to be obtained in the family, although several brothers and sisters in one generation might be affected. In no case was the father affected, and the mother only in two cases; and he found that males were almost exclusively affected, although the disease was transmitted through the female line, so that in some instances children born of the same mother, but by different fathers, were found to be affected. No definite history of consanguinity in the parents was obtained, and if it did occur it must have been so unusual as not to be of very much consequence in determining the etiology.

The dimness of vision was usually of sudden onset, the impairment often became intense in a few days or weeks, and there was seldom any headache or other associated symptom. Occasionally there were subjective sensations of light or colour, and in addition to the true visual affection it was found that the colour-vision was either abolished or much diminished. The ophthalmoscopic appearance consisted in a haziness at the margin of the disc, the vessels were normal in size, or appeared even a little larger than usual, there were white lines along their margins, occasionally there were spots or streaks in the fundus. Pallor of the disc succeeded the disturbance of vision, and became distinct when the visual defect was at its height. The impairment of vision might remain permanent and unaltered. It frequently, however, underwent a certain degree of improvement, and in three cases occurring in one family, complete recovery was said to have taken place. In some of those cases certain tendencies to nervous disturbance were present, such as tremor, vertigo, or even fits. In every case organic cerebral disease could, it is said, be excluded. Both eyes were affected in all, unequally as a rule, and the affection of one occurred before that of the other.

Since Leber's time cases of a similar character have been described by Fuchs, Higgins, Norris (of Boston), Habershon,

and others. It may be stated that the only definite addition to our knowledge in reference to the disease is, that it may occur in even more than the three generations mentioned by Fuchs, and that a mother may convey it to a daughter, she herself having been affected. Its relation to toxic conditions, such as those induced by tobacco and glycosuria, has not yet been made perfectly clear. The disease, in short, is characterised by its occurrence in different members of a family, by its occurrence in—as many as five—successive generations; by its predominating, but not exclusive, affection of males, and by its preference for transmission in the female line. The ophthalmoscopic appearances are not usually attended with any severe nervous disturbance, are characterised in the early stage by signs of slight neuritis, followed by pallor and other evidences of atrophy, and accompanied with considerable impairment of vision. Apparently this loss of vision does not go on, in many instances, to complete blindness, and in many cases there is a tendency towards improvement. No definite evidence of organic nervous disease is present in the great majority of cases, although occasionally, in isolated instances, epileptic fits have been described, and in a few intense headache has ushered in the symptoms.

There is very little doubt that in this disease one is dealing with a more or less definite entity; but there is considerable probability that a good many cases have been included which are really of a different character. The question whether in some instances a condition of retinitis pigmentosa was present would, of course, refer especially to the cases described before the era of the ophthalmoscope. The ophthalmoscopic changes in the fundus in that condition are, as a rule, fairly characteristic, so that now cases are not likely to be confused with those of hereditary optic atrophy. In retinitis pigmentosa also, as is well known, night-blindness is one of the most definite features. In hereditary optic atrophy, however, the vision does not seem to be worse in twilight. Cases of optic atrophy occur in different members of the same family undoubtedly, having as their causal background congenital syphilis. No doubt some of the cases that have been

described as cases of hereditary optic atrophy are really cases of congenital syphilis with optic atrophy. These have already been referred to in the article on juvenile general paralysis, and will be referred to again in the article on hereditary tabes dorsalis.

The relation of the disease to epilepsy is by no means definite; and with regard to the cases in which insanity has been alleged as an associated condition, it is not improbable that these have really been cases of juvenile general paralysis. It is exceedingly difficult to say definitely what the relation of tobacco is to this disease. In many of the cases described there has undoubtedly been excessive use of tobacco: in many others the influence of tobacco could be excluded. One other fact has to be borne in mind also; and that is, that in cases of diabetes mellitus, even when the sufferer does not use tobacco, and does not take alcohol, characteristic scotomata for colour may be present. In cases, also, of diabetes insipidus, optic atrophy frequently occurs; and the same is true of cases of disseminated sclerosis. In these also colour scotomata, identical with those of tobacco, may occur in non-smokers. But even after all such possibly complicated cases and conditions have been excluded, there still remains a fairly large class of cases in which optic atrophy, with associated visual defect, remains as a family disease, affecting often more than one member of a family, occurring, it may be, in several generations, and with a preference for affecting males rather than females. Yet there is a tendency to transmission in the female line, just as in muscular dystrophy, and as in that disease also, isolated cases in an otherwise healthy family are met with, and although males suffer more than females, the latter are by no means entirely exempt.

## MICROCEPHALY

THE condition of microcephaly is one concerning which it is desirable to say something, especially in view of the fact that during recent years, attempts have been made by operative measures to remedy what is probably a developmental defect incapable of being relieved by such measures. It must be recognised that there are two distinct varieties of microcephaly: (1) that condition of smallness of the head, associated with mental defect, which is associated with the presence of actual gross structural disease. In some cases of diplegia there is a condition of microcephaly. In a distinct proportion of cases of infantile hemiplegia a microcephalic defect is present. In the former condition the smallness of the head is more or less symmetrical, in the latter there is usually flattening in the frontal or parietal region on the side on which the lesion of the hemisphere causing the hemiplegia exists, and also in the occipital region corresponding to the opposite cerebellar hemisphere. We have seen that in those cases there is shrinking and reduction in size of the affected hemisphere, and a corresponding and probably proportional reduction in size in the opposite lateral lobe of the cerebellum. The reduced size of the head in those regions is secondary to the structural changes in the intra-cranial contents.

(2) The other variety of microcephaly is that in which a small and usually deformed head is associated with idiocy and great bodily weakness which cannot be described as local paralysis. The child's head is narrow, there is a ridge in the middle line, the eyes are unusually prominent, and the face small and expressionless. The limbs are weak, there is usually inco-ordination of movement, the child is

unable to feed itself, is noisy and dirty in its habits, is unable to speak, and its mental condition is one of marked imbecility.

Some years ago it was supposed that this condition of microcephaly was the result of premature synostosis of the skull bones, leading to pressure upon and consequent interference with the development of the brain. Operations of varied complexity were therefore devised by Lannelongue and others, consisting essentially in removal of parts of the skull cap, with the object of giving the brain an opportunity of expanding and developing. In some cases improvement, it was asserted, took place, but it was soon recognised that whatever improvement did occur was only such as might be expected in an imbecile child under the influence of such discipline and training as it might receive in a hospital or other institution. And it is now generally acknowledged that operation in such cases is not a measure which offers any chances of improvement. The premature ossification of the skull is probably to be regarded not as a cause of mal-development of the brain, but rather as a condition of defective development associated with the defective cerebral development, so that any operative procedure on the skull is not likely to result in improvement of the cerebral condition.

Microcephalic idiots, in short, are to be regarded as the subjects of defective bodily development, and of weak-mindedness, and as not likely to be benefited by any treatment except such special education and disciplinary care as are to be found in idiot asylums. Operation is not to be recommended, for, as has already been stated, any improvement that has followed such operation (and in no case has it been very marked) has only been such as was likely to follow the special care and nursing which the child enjoyed in a hospital ward.



# SPINAL CORD AND ALLIED DISEASES

## INFANTILE PARALYSIS

(INFANTILE SPINAL PARALYSIS, ACUTE ANTERIOR POLIOMYELITIS, ACUTE ATROPHIC PARALYSIS, ESSENTIAL PARALYSIS OF CHILDREN)

THE disease which is so well known under these different denominations was first described by Dr. Underwood in England in 1774. But the first really good clinical account of it was given in Germany by Heine in 1840. Eleven years later Ralliet and Barthoz described it in France, and by them the name "essential paralysis of children"—not really a satisfactory designation—was first used. In 1865 Prevost first demonstrated the anatomical changes in the spinal cord underlying the clinical symptoms, and soon after this Lockhart Clarke, Charcot, and Joffroy corroborated and amplified Prevost's description. In 1861 Duchenne had furnished a characteristically clear clinical description of the disease and had invented still another name for it, viz., *paralysie atrophique graissouse de l'enfance*. But up till 1885, when Drummond published an account of the anatomical changes in a recent case, the description of the changes in the spinal cord consisted only of an account of the condition found in old-standing cases. Since that time several cases have been published in which the *post-mortem* conditions were described as they were found in patients dying soon after the onset of symptoms. The disease has received its most common name of infantile paralysis on account of the preponderating frequency with which it attacks young

children. The more anatomically descriptive name of "anterior poliomyelitis" has been used since the discovery by Prevost and Lockhart Clarke that the grey matter in the anterior horns of the spinal cord was the seat of the morbid changes, and such a name has the advantage of being applicable, not merely to the disease as it occurs in children, who are the most frequent victims of it, but also as it occurs in those of maturer years, whom it also occasionally attacks.

*Age and sex.*—In 120 cases under the writer's care 74 were males and 46 females—a proportion which shows the much more frequent incidence of the disease in males than in females. Out of 113 patients regarding whom accurate information was forthcoming, 105 cases occurred in the first decade of life, 4 in the second, 1 in the third, 2 in the fourth, and 1 in the sixth; of the patients under 10 years, the onset in 33 was in the first year, in 37 in the second, in the third in 14, in the fourth in 8, in the fifth in 6, in the sixth in 4, in the seventh in 2, and in the ninth in 1. It is thus seen that the first two years of life are *par excellence* those in which the disease occurs. Of the 33 cases occurring in the first year of life 9 were under six months, while 24 were six months or over.

It has been pointed out by various authorities, both in this country and in America, that the onset of the disease is far more common in the hot months of the year. In sixty-nine cases it was possible to discover the exact month of the onset, and the result strikingly confirms the observations of others. In January there were two, in February four, none in March or April, in May three, in June seven, in July sixteen, in August seventeen, in September six, in October six, in November five, and in December three. So that, although the great majority occur in July and August, there are scattered cases in most of the other months.

The *mode of onset* of the disease varies. In the vast majority it is ushered in with *malaise*, high temperature, vomiting, sometimes severe pain in the limbs, occasionally convulsion, and it is often only during convalescence that the true nature of the disease is discovered, on account of the patient's inability to use a limb. In a certain number

of cases, however, the onset is unaccompanied by any constitutional disturbance. The child goes to bed apparently well in the evening, and is found to have one or more limbs in a state of flaccid paralysis in the morning. In such cases there is usually no pain. In another series of cases—not a large one—the paralysis succeeds some fall or other accident. Sometimes the onset of the paralysis is immediate, as in one case in which there was dislocation of one of the paralysed limbs. Occasionally the paralysis succeeds the accident after an interval of a few hours, as in a case in which the patient fell and twisted a leg, but was able almost at once to resume its play. Next morning the affected limb was paralysed. Occasionally the paralysis succeeds over-exertion, as in the case of a child of 3 years, who, the day after a long and fatiguing walk of eight miles, was found to have paralysis of both lower limbs. Sometimes the paralysis closely follows a severe fright.

The relation of infantile paralysis to other diseases is uncertain. In three of the writer's cases it is said to have been an effect of rheumatic fever, but as this was unaccompanied by swelling of joints or other unequivocal sign of acute rheumatism, it is possible that the pains in the limbs, which are so often present at the onset of uncomplicated infantile paralysis, may have been by mistake regarded as rheumatic pains. In only one case did the onset follow measles, and in one it is said to have followed influenza. This name is now applied to so many somewhat indefinite feverish conditions that it would not be justifiable to attach any importance to a single case. In one case the onset occurred during convalescence from diphtheria, and in another it followed an illness characterised by delirium with head retraction. Convulsions are usually mentioned as an occasional accompaniment of the onset of this illness. They were present in five cases. Such convulsions are to be regarded as either reflex or as a result of the cause of the paralysis affecting temporarily, cerebral structures. Considering how slight may be the exciting cause of reflex convulsion in all children, but especially in rachitic children, such as are most London children from whom hospital

statistics are drawn, it is not unlikely that the convulsions in those cases are most properly to be regarded as reflex.

*Course and complications.*—As has already been stated, the condition of paralysis is often only recognised when the general constitutional symptoms are beginning to pass off and the child attempts to resume its former amusements. It is then found to have impairment of the movement of one or more limbs. As regards the parts which suffer, there are accurate notes in 115 cases. From these it appears that in fifty-two cases one leg was affected—the left in thirty-



FIG. 30.—Infantile paralysis, affecting anterior tibial muscles.  
Mr. Jackson Clarke's case.

four, the right in eighteen,—and in forty-three both legs. Of these forty-three one had, in addition to the affection of both legs, an affection of the left thenar eminence and the left arm as well, one of the left arm, one of both hands, three of the right arm, and one of both hands, the right more than the left. In eighteen the arms only were affected, the left in eight—in one of these the face was temporarily affected—the right in seven and both arms in three. Two patients had one arm and one leg affected, one the right arm and left leg, and one the left arm and left leg. From these various particulars only one definite fact stands out,

viz., that the lower limbs suffer much more frequently than the upper. In only three was there any definite affection of the sphincters, and in none was this permanent. In the cases in which both legs were affected one always suffered more than the other, and in the cases in which one leg was affected the knee-jerk was usually lost. But in three cases the leg below the knee only had suffered, and the knee-jerk was preserved—a fact which completely disproves the statement occasionally made that in a leg affected by infantile paralysis the knee-jerk is always lost. The fact is, that if



FIG. 31.—Infantile paralysis, affecting chiefly calf muscles of left leg, causing talipes calcaneus. Mr. Jackson Clarke's case.

the vastus internus escapes, the knee-jerk will almost certainly be present.

The affected limb, or part of a limb, is then found to be paralysed; there is unusual flaccidity of the muscles, as a rule without pain or discomfort. In some cases there is severe pain in the affected parts at first, and this may occasionally persist for some time. The paralysed part is cold and deeply cyanosed, but there is never persistent sensory impairment. If slight anaesthesia is present at first, as it occasionally is, it is only temporary. Contractures are apt to become established because of the unantagonised

contraction of the healthy muscles, and various forms of talipes may develop according to the muscles affected. If the affected muscles are now tested by the faradic current, there is found to be either complete absence of faradic reaction in them or very great diminution in the response. The galvanic reaction, however, will probably persist,



FIG. 32.—Infantile paralysis, affecting upper limbs. From a photograph by Dr. Collier.

and there may be an altered polar reaction. A typical case of infantile paralysis is thus characterised by wasting of muscles, accompanied by diminution in the size of the bone, and perhaps, although not invariably, shortening of the limb, loss of faradic reaction in the muscles, coldness and cyanosis of the affected limb, loss of the reflex

action, if the muscles subserving it are involved, complete retention of the sensory functions and the presence of a certain degree of contracture in the muscles which are least affected, and consequent deformity of the limb.

As regards the course of the disease it must be recognised as a general rule that the paralysis is at first always more severe and extensive than it subsequently proves to be. The presumption is that at first, probably on account of inflammatory exudation and congestion, the function of certain structures is temporarily placed in abeyance, and as soon as the acute process has subsided these temporarily impaired structures are able to resume their function. The same mechanism may probably also be invoked to account for the occasional occurrence of sensory impairment in an early stage. Such impairment, as has already been stated, is never permanent, and it is likely that its temporary presence is the result of transient interference with sensory structures in the cord. The occasional transient affection of the sphincters is also probably to be similarly accounted for. The occasional presence of severe spontaneous pains in the limbs affected and of tenderness to touch and during movement is a fact of importance and significance in this disease. Sir William Gowers associates such pains with the probable occurrence of changes of an inflammatory character in the peripheral nerves simultaneously with the changes in the cord to be presently alluded to. Cases have been described in children in which there was paralysis with such pains, from which complete recovery has taken place, and the suggestion has naturally been made that in such cases it is not unlikely that a purely neuritic condition has been present. For it is rare to get complete recovery in cases in which there is strong evidence of an affection of the anterior horns.

Of complications in the acute stage the most common is bronchitis, and this may become a serious danger to life. Convulsions have already been referred to as occurring occasionally in connection with a feverish onset. Of complications in the later stage the only one that may be men-

tioned is the rare condition in which a chronic degeneration of anterior horn cells—with consequent atrophy of muscles—progressive muscular atrophy—is superimposed upon a cord injured by an attack of acute anterior poliomyelitis in early life. This is rare, as has been said, but ought to be kept in mind.

*Pathology and etiology.*—The pathology and etiology are necessarily bound up together. When we regard the conditions under which the symptoms of the disease are manifested—in otherwise healthy children, nearly always during hot weather, usually with a feverish onset, and occasionally with severe pains—it is difficult to escape from the conviction that we are dealing with a toxic disease in which there is some local lesion. What adds probability to such a view is the occurrence of this disease occasionally in epidemics—in one of which at least not only human beings but also the lower animals suffered—and the occurrence of the same disease simultaneously in more than one member of the same family. It would seem as if temporary climatic conditions, together with, perhaps, some individual favouring state, the nature of which we do not know, gave rise to a set of conditions in which the *materies morbi* was able to become active. It need scarcely be pointed out that in these respects the disease is very similar to many others of the infantile diseases. It may be objected to such a view that it fails to account for those cases in which a local injury is followed by a diseased condition in the spinal cord. It may also be objected that some cases at least recover completely in spite of severe initial pain and temporary paralysis. No doubt the former class of cases are not easily accounted for without invoking theoretical considerations of some complexity. The latter, however, may be more easily accounted for by supposing either that the poison exerts an influence on the peripheral structures only, or that the sensory symptoms are the result of an unusual restriction of the spinal cord lesion to sensory structures in it. That such structures are affected together with motor structures will be evident presently when we deal with the morbid anatomy. The



occasional restriction of the disease to sensory structures calls for no essential change in our conception of the morbid process. It may also be stated in reference to the cases in which local injury is followed by paralysis of central origin, that such cases of local injury determining changes in central structures are by no means unknown in other diseases, such as alcoholic peripheral neuritis and perhaps even more strikingly in tubercular meningitis, general paralysis of the insane, and tabes dorsalis.

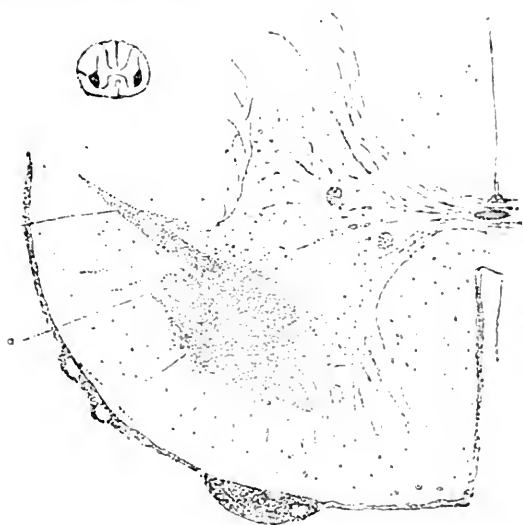


FIG. 33.—Acute anterior poliomyelitis in the adult; inflammation and exudation in anterior horn in cervical region. The opposite horn was similarly affected. Dr. R. T. Williamson's case.

*Morbid anatomy.*—The morbid anatomy of this condition has to be considered in two aspects: (1) The changes which are found in the central structures and the peripheral organs when the paralysis has existed for a period of months or years; (2) the changes present in cases in which an opportunity of examination has been afforded in the first few months of the illness.

Chareot in 1879 formulated the hypothesis that the disease was a primary parenchymatous degeneration of the nerve-

cells, the changes in the interstitial tissue being regarded by him as secondary. This theory was founded upon the observation of cases of long standing. Ten years later Rositer examined three recent cases and described accurately the changes met with in the ganglion cells and interstitial tissue. His observations led him to confirm Charcot's



FIG. 34.—Transverse section of lumbar region in the cord of a child who died thirteen days after the onset of infantile paralysis, showing thrombosed vessels with perivascular exudation and scattered hemorrhages. From a drawing made from one of his own sections and kindly lent by Dr. F. E. Batten.

theory. Rogers and Damaschino from observations on recent cases concluded that the disease was a primary interstitial myelitis. Pierre Marie in 1892 was the first to call attention to the vascular origin of the disease, and his views were definitely confirmed by Goldscheider. It is known that the anterior horns are supplied by branches of the anterior spinal artery, and in cases which have ended fatally in the first few

days and have been examined the regions of supply of those branches have been found necrosed and softened, the vessels blocked, much extravasation of leucocytes around them, and the nerve-cells completely disorganised. Many degenerated fibres were also found in the anterior commissure, ground fibres, and root zone. The antero-lateral ascending tract and the direct cerebellar tract also showed degenerated fibres, and they were also present in the posterior root, although affecting mostly the endogenous system



FIG. 35.—Transverse section of lumbar cord in a child three months after the onset of infantile paralysis, stained by Marchi method. The area of softening is limited to the anterior horns. On the left side the fat, stained black, lying in the perivascular space, marks out the position of a vessel. From preparations and photographs kindly lent by Dr. F. E. Batten.

of fibres. Sections of the peripheral nerves also showed by the Marchi method many degenerated fibres.

Of course these changes have been found only in cases proving fatal at a very early stage. It may be claimed that such changes do not necessarily occur in cases not proving fatal; yet it must be said that the pathological changes found in these old-standing cases are not incompatible with such a commencement, although the very slight nature of the initial illness in many cases may throw a certain degree of doubt upon this severe inflammatory origin of the disease in those.

With reference to the peripheral structures it need only be mentioned that the nerves are degenerated, the muscles affected become practically masses of fibrous tissue, the bones cease to grow always in thickness, and frequently in length, so that there is always marked diminution in the size of the affected limb. It is not unlikely that in the cases in which coldness and blueness of the affected limb or limbs is very marked there is paralysis of the vaso-motor

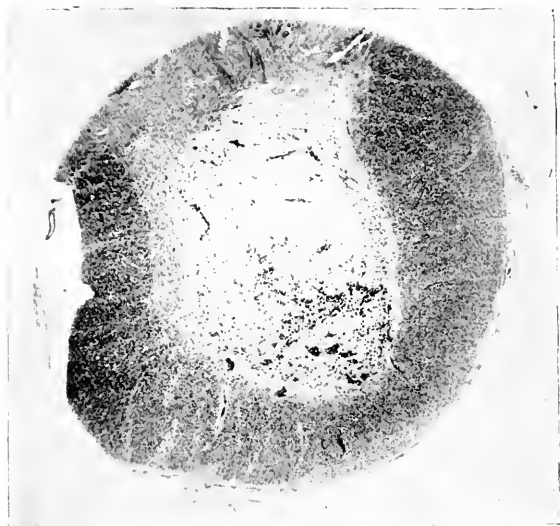


FIG. 36.—The right anterior horn, under a higher power, showing the black stained granules scattered in the softened area of the anterior horn. Also from preparations and photographs by Dr. Batten.

mechanism in the cord, but the situation of this mechanism is so uncertain that its lesion must, in the meantime, be a matter of conjecture.

A further question remains to be asked, and, if possible, answered, What is the cause of this thrombosis in the branches of the anterior spinal artery on which the cord changes in acute anterior poliomyelitis depend? It is almost certain that it is some form of toxæmia which leads to this result, for there is no likelihood that the vessel walls

are thickened or diseased in any other way. The poison is probably a poison of a peculiar character. It is probable that it is the same or a similar poison which causes the so-called encephalitis in children, on which many cases of hemiplegia depend. Dr. F. E. Batten, however, who has done much excellent work on this subject, is of opinion that the thrombosis may be the result of different forms of infection, and that the disease is not the effect of some special poison. He also thinks that such an effect is more likely to be found in the lumbar region because of the more distal position of this point from the heart, and because of the long course of the reinforcing arteries. Yet the existence of the disease in an epidemic form certainly favours the view of a special and peculiar poison.

*Diagnosis.*—The diagnosis of this condition does not in the great majority of cases present many difficulties. When the onset of the disease is marked by pyrexia, *malaise*, and general constitutional disturbance, the difficulty that is likely to arise is in discerning that a paralytic disease is in question and not merely some pyrexial attack of a specific or non-specific character. In cases of this nature, as has already been mentioned, the true nature of the disease is sometimes not recognised until the patient is able to begin to move about again, when paralysis, either of slight or severe character, is recognised. It therefore behoves the practitioner to remember the possibility of the occurrence of this condition in all cases of illness of severe but ill-defined character in children. If there is much pain at the onset acute rheumatism is most likely to be suspected; when there is no pain the condition may be mistaken for pneumonia, bronchitis, or other feverish condition. In the cases in which infantile paralysis is supposed to follow rheumatic fever it is certain that in some, and it may be in all such cases, the initial pains, etc., are really part of the disease. Another condition which is somewhat difficult to distinguish from infantile paralysis is the commencement of some joint disease. This difficulty, of course, is only likely to occur in those cases in which the paralysis is limited to one limb.

and the writer has known several cases in which the definite diagnosis was at first exceedingly difficult and, indeed, occasionally impossible. He has also known a case of commencing double hip-joint disease regarded as a case of infantile paralysis, but in such cases an electrical examination, not always an easy undertaking in children, will usually suffice to clear up the diagnosis. A distinction which is not always easy is that between this spinal cord affection and the analogous cerebral affection—infantile hemiplegia. In the latter, however, both the arm and leg on the same side are affected—an unusual combination, as we have seen, in the spinal disease—the onset is usually with convulsions, and there is an absence of any electrical change in the muscles of the affected limbs. The reflexes, too, are exaggerated, whereas in the spinal disease they are absent or much impaired if the muscles subserving them are attacked, or, to state it more correctly, if any part of the reflex arc is damaged.

There is not likely to be any difficulty in distinguishing cases of myopathy from cases of the disease with which we are at present concerned. Occasionally, however, a doubt or difficulty may arise if the cases are seen long after the onset, and if a reliable history of the sudden occurrence of the symptoms is not obtainable. But in myopathy the disease is usually one affecting all the limbs; there will be, in cases of the pseudo-hypertrophic type, the enlargement of certain muscles, and there is not the coldness and blueness of the affected limbs so common in infantile paralysis. Of course if a reliable history is obtainable, this difficulty is scarcely likely to arise, as the exceedingly gradual onset of the myopathic disease is in marked contrast with the sudden occurrence of the spinal affection.

One somewhat rare condition should be mentioned in regard to diagnosis—that, viz., of infantile scurvy, or, as it is usually called, scurvy rickets. In many cases of this affection the pain in the limbs, the extreme wasting of the muscles, and the indefiniteness of the symptoms generally, not unusually make it difficult to be quite sure that the

condition is not a spinal one. But the association of evidences of scurvy elsewhere, especially in the gums if there are any teeth, and a history of errors in the feeding of the child are usually sufficient to mark the distinction. Yet in some cases the difficulty is a very real one, and the writer has known at least one case in which after recovery in other respects the atrophy in one thigh was so striking as to make it doubtful for a time whether there was not some spinal lesion associated with the constitutional condition. But the thickening of the bone—the result, no doubt, of hæmorrhage under the periosteum—was always very marked, and the ultimate complete recovery of the child proved the condition to have been only the scorbutic one.

The *prognosis* in cases of anterior poliomyelitis must, in the early stages of the disease, be a somewhat guarded one. This is especially true of the cases in which the upper and lower limbs are both affected. In such cases the respiratory muscles may also be involved, and a slight bronchitic attack may be a serious and even a fatal complication. In some cases there is reason for believing that a condition similar to that affecting the anterior horns of the spinal cord affects the analogous nuclei in the medulla. If such a condition is recognised in any case it will necessarily be of the gravest significance, as we know that these nuclei are closely related to structures the intactness of which is necessary to life, and there is little doubt that these cases nearly always end fatally in the early stage. The writer has known one case in which there was simultaneously with an attack affecting the spinal structures related to the shoulder muscles of one side, paralysis of one side of the face, partial in character. This patient recovered completely so far as the facial muscles were concerned. The deltoid, supra- and infra-spinatus, however, remained permanently paralysed. In the ordinary case of anterior poliomyelitis, however, the prognosis as regards life is nearly always favourable. As regards complete recovery, however, this must not be expected, and the recognition of this fact is necessarily a great shock to the parent. Yet in nearly all cases a very

great degree of recovery ensues, and in many instances in which the paralysis at first affects two limbs one of these completely recovers. Similarly in cases in which a whole limb is paralysed, the paralysis frequently becomes confined to a group of muscles—perhaps to those subserving a single movement—so that in the ordinary case of infantile paralysis, although it is extremely rare for complete recovery to take place, one can usually state with assurance that some, and it may even be an unexpectedly considerable, degree of recovery will probably take place. But, as has already been said, complete restoration of the functions of the paralysed muscles is scarcely to be looked for.

*Treatment.*—In reference to the treatment we must first of all consider the measures which it is necessary to adopt at the time of the onset of the illness. At first, as we have seen, the condition may be one of little or no constitutional disturbance; or, on the other hand, there may be much pain, with high fever and general *malaise*. In either case rest in bed is in the first instance desirable—in the one case it is essential, in the other it is prudent, as an extension of the original morbid condition may conceivably take place. When a high temperature is present, the patient must be treated on the general lines observed in any case of acute febrile illness, with light, easily digested food, cooling drinks, and occasional sponging. If we are dealing with one of those cases in which pain in the affected parts is very marked, it will be well to protect the painful limbs by means of a cage to prevent the bed-clothes from increasing the pain. If the pain be very severe, it may be even necessary to give opium. Antipyrin and salicylates have also been recommended, and so long as their use is not attended with too much depression of the circulation, they may be given with advantage and relief to the patient, and they may aid in the elimination of whatever *materies morbi* is at the root of the symptoms. Although there is little or no tendency to trophic disturbance, yet the possibility of its occurrence should be borne in mind and the necessary precautions taken. It is doubtful whether drugs are capable of favourably affecting the arterial changes



which occur in the spinal cord. Ergot and belladonna have both been recommended. The latter, by slightly stimulating the circulation, might conceivably do a little good; the administration of the former is probably not unattended with some degree of danger. The treatment, indeed, necessary in this stage may be summed up in good nursing, keeping the affected limbs warm, the administration of light food, and the use of such drugs as may relieve pain, if this is severe, or ameliorate the symptoms and perhaps tend to elimination. As soon as the pain ceases to be very severe the position of the limbs should be looked to, and any malformation or distortion as far as possible corrected. If this is attended to early, it may prevent the occurrence of deformities which may be troublesome later. Passive movement and massage should, therefore, be commenced as soon as the subsidence of pain permits.

In regard to the later treatment, massage and passive movements, as well as movements against resistance are very important, with the view, not only of preventing deformities, but of increasing nutrition and favouring recovery. The affected muscles are not able to respond to volitional impulses, but their manipulation by a trained rubber tends to keep them in a condition in which they are ready to respond to such impulses if, in the process of recovery, they ultimately do reach them. This also is the object of electrical treatment, and, that being so, it will at once be evident that the use of an induced current is quite inefficacious. Such a current only gives rise to a muscular contraction by stimulating the muscle nerves, but if these are degenerated, as they no doubt are if the cells in the anterior horn are destroyed, it is obvious that no effect is to be obtained by attempting to stimulate them with such a current. In the galvanic or constant current, however, we have, as we know, an agent capable of stimulating muscular fibres to contraction even when these are disconnected from their nerve, and the paralysed muscles in a case of infantile paralysis are essentially in this condition. And there is no doubt that in every case of this disease the constant current should be carefully

and assiduously used. It must be used, however, also with discrimination. Its application, if effective, is not without some discomfort, and even pain, so that at first a very gentle strength must be applied. Indeed, with especially nervous children it is often necessary to use the battery two or three times without any current passing, and then very gradually to increase the strength as the child's confidence is gained and its fears overcome. And in some cases we must recognise that, on account of the fear and even terror produced in a nervous child by the use of the current, its application cannot be urged, and if it is persisted in, is capable of doing real harm. But whenever it is possible it should be used, as its effect undoubtedly is to stimulate the muscular fibres and so to keep them ready to respond to voluntary impulses as soon as the nervous structures have sufficiently recovered to be able to transmit those. One thing is frequently noticed with reference to the responsiveness to the constant current of the affected muscles in cases of anterior poliomyelitis, viz. that on the first or even second or third application of a good current there is no appearance of contraction in the muscle to which the electrodes are applied. But by-and-by, if the application is persisted with, visible contraction does take place on stimulation, and it has been stated that such contraction may ultimately be evoked even when it has been impossible during as long as a year to obtain any response.

It is frequently asked, How long ought one to persist with such treatment? In the writer's experience all the good that is to be got is obtained in the majority of cases within a year of the onset. In exceptional cases it is possible that it may be desirable to continue treatment longer, and if at the end of a year improvement is manifestly taking place under treatment it should, of course, be persevered with. But the statement made above will be found to be generally correct.

Even after the most careful and skilful and prolonged treatment, it is usually found that certain muscles are irretrievably damaged, perhaps even completely destroyed and

represented only by fibrous tissue. The disappearance or weakness of such muscles necessarily leads to contraction in their antagonists, and this may cause deformities. If these have occurred, tenotomy of the unantagonised muscle may be necessary and is often performed with great benefit. Especially is this the case, *e.g.*, when some mechanical support to the limb is necessary in order to make locomotion possible or, at least, easier. And the use of such apparatus is in many cases necessary. It would be impossible to discuss the varied forms of appliances used. These are as numerous as the different varieties of paralysis produced, and the mechanism necessary in any given case must be decided simply by a consideration of the structures paralysed and the best mechanical means of compensating for these. One point also is important to remember, *viz.* that in a certain number of cases of infantile paralysis the growth of the bone lengthwise is interfered with. Its increase in thickness always suffers, but in those cases in which its increase in length is much affected this is probably from interference with the epiphysis. The result, of course, is marked shortening of the limb, which if uncorrected in some way may lead to lateral curvature. So that in considering the question of mechanical supports for limbs this question of the possibility of shortening should always be considered, and taken into account early in the history of the case.

During the last few years tendon-grafting has been tried, with the view of minimising the paralysis from this disease. The tendon of a healthy muscle is split and connected with the tendon of the paralysed muscle in various ways. In some cases the result has been good. Attempts have also been recently made, by splitting a nerve going to a healthy muscle and inserting some fibres into the paralysed muscle, to transmit impulses in this way and so to re-stimulate the paralysed fibres. It is not unlikely that the future may show some results considerably better than the moderately successful ones already obtained.

## OBSTETRICAL PARALYSIS

ALTHOUGH such a term as this should be used to describe all diseased conditions which result to the child during its birth—whether these are due to lesions of brain, spinal cord, nerve-roots, nerves, or muscles—it has come to be applied almost exclusively to lesions which do not affect the central nervous system. There can be no doubt that certain forms of paralysis are the result of brain lesions which occur at the time of birth, and are directly associated with a difficulty, or at least an abnormality, in the labour, due to an abnormal character or presentation of the fœtus or to a maternal deformity. In 130 cases of still-birth examined by Dr. Herbert Spencer hæmorrhage into the substance of the brain was found in only one. Hæmorrhage on the surface of the brain into the arachnoid or pia mater was found, however, in no less than fifty-three cases, or 40·7 per cent. Hæmorrhage occurred over the convexity in forty-nine cases, over the base as well in thirty-five; in six cases only the hæmorrhage was limited to the base. Hæmorrhage into the ventricle occurred seven times. These statistics, it is true, refer to cases of still-birth in which, in many instances no doubt, the hæmorrhage was the direct cause of death. Yet it is scarcely open to doubt that in many cases cerebral or cerebellar hæmorrhage occurs, which is not fatal, but leads to paralysis, varying both in degree and in distribution.

Similarly he found in forty-four cases in which the spinal cord was examined that in no less than thirty hæmorrhage had occurred. It is therefore also probable that occasionally children are born alive in whom some lesion of the spinal cord has been produced during birth leading to some

impairment of power in one or more limbs or in the trunk. It should also be remembered that so-called congenital cavities are occasionally met with in the spinal cord, and it is possible that the starting-point of such cavities may be some injury to the cord produced in the process of birth. The brain conditions which may occur during birth and give rise to paralysis are referred to in the section on Cerebral Palsies, and a further reference to possible spinal cord lesions at birth will be found under Syringomyelia.

Facial paralysis is one of the common forms of obstetrical paralysis. It is of the usual peripheral type, affecting all parts of one side of the face, the upper as well as the lower. When it is complete the child is unable to close the eye and the paralysis imparts to it a most ludicrous expression when it cries. The affected side of the face then remains quite immobile. The paralysis does not often interfere with the child's ability to take the breast or the bottle, and irritation or inflammation of the cornea from non-closure of the eyelids on the affected side is rare.

The cause of the paralysis in the majority of cases is direct pressure of the forceps, leading to bruising of the nerve, but not effecting any break in its continuity. In rare cases the continuity is, however, permanently interrupted, there is not complete recovery from the paralysis, and the growth of the affected side of the face is strikingly interfered with, causing much deformity. In some cases in which the forceps has not been used facial paralysis has occurred. It must be supposed that in such cases some local compression of unusual character, probably from some deformity or peculiarity in the maternal pelvis, has caused the paralysis, or that some œdema and bruising of the nerve has occurred, as might conceivably take place if a *caput succedaneum* were situated so as to cause this.

Ptosis has been recorded as occurring in new-born infants, the result of the pressure of forceps on the nerve to the levator palpebrarum.

The *diagnosis* of such a condition of facial paralysis is not likely to cause much difficulty. At the same time, unless

the practitioner is familiar with it, it may cause him more embarrassment and even alarm than it need do, for in almost all cases a complete recovery can be confidently predicted. The duration of the condition varies, but it is more often a matter of days than of weeks. Of course, as has already been stated, in very rare cases complete recovery does not take place. In all such it is well to examine into the condition of the other cranial nerves, so as to be sure that the condition is not one of congenital mal-development of certain parts of the nervous system. The treatment need not be very energetic. Gentle rubbing of the facial muscles with the finger will usually be sufficient. If unusual delay should seem to take place in the recovery, the use of a constant current of very gentle strength might be tried. It is not advisable to use blisters or any other form of counter-irritation.

The condition of so-called "sterno-mastoid tumour" may be referred to here. It is a hæmatoma occurring in that muscle when it has been exposed to pressure during birth. It results probably from a tearing of the muscular fibres, and is important because it is apparently, at times, the starting-point of a congenital torticollis.

The most important form of obstetrical paralysis is that in which the upper limb suffers. It is nearly always unilateral, and the muscles affected as a rule are the deltoid, biceps, brachialis anticus, and subscapularis. In severe cases the whole of the plexus may be affected, causing complete paralysis of the upper limb. Sensation is never permanently impaired.

This form of paralysis is likely to occur in cases in which the arm is out of its usual position and so placed that there is pressure or unusual traction upon the brachial plexus. Such a condition of affairs is likely to occur in cases in which the child presents abnormally. When it is necessary during birth to change the position of the arm, either with a finger or instrument, if the pressure to do this is exerted in the axilla, paralysis is very likely to follow. This is most likely to happen in cases in which the legs are born first,

especially after turning, and it may also occur in cases in which after the head is born it is necessary to exert strong traction to extract the body.

The *diagnosis* is not likely to offer much difficulty; the flaccid condition of the limb, especially in any case of abnormal labour, will usually indicate the nature of the paralysis. The distribution also of the paralysis is very suggestive, the muscles involved being always those about the shoulder and sometimes only those. It is always, however, advisable to examine into the condition of the bones and joints. Dislocation of the head of the humerus has sometimes occurred, with or without this paralysis. Fracture of the clavicle has also been described, and it is said that separation of the epiphyses of the humerus may simulate this paralysis. In all such cases, however, it is not unlikely that injury of the plexus has been combined with the bony lesion.

The nature of the lesion is no doubt a traumatic neuritis, and the distribution of the paralysis depends upon the nerves or parts of the brachial plexus which are injured.

The *prognosis* in all such cases is bad—in this respect strikingly different from that in the facial affection. Complete recovery is very rare: permanent paralysis, especially of the deltoid, or at least of its posterior part, is the rule, and the biceps, brachialis anticus, and subscapularis are also likely to be permanently affected, and this affection is, of course, combined with marked atrophy of the affected muscles.

The *treatment* in a new-born child cannot be energetic. It must be almost restricted to rubbing, as the use of the battery, while theoretically desirable, is scarcely practicable.

Paralysis of the inferior limbs as a result of obstetric proceedings is rare. When it does occur it is usually the result of violence, possibly necessary violence, exerted on the vertebral column and consequent injury to the cord. According to some authorities the important condition of congenital hip dislocation is the result of malposition of the lower limbs of the fetus *in utero*.

## FRIEDREICH'S ATAXY

THIS disease, known also as "hereditary ataxy," is so named after the Heidelberg professor who first described it in 1861. In some cases which had a few years before that date come under his notice, he recognised a close resemblance to the condition which had been recently described by Duchenne under the name of "locomotor ataxy." These cases, however, although similar to those described by the great French observer, differed in several details, more particularly in the two points that in Friedreich's cases there was evidence of the presence of the same condition in more than one member of a family, and that there was an absence of impairment of sensibility. Various attempts have been made to stamp the disease thus differentiated by Friedreich with a name more or less descriptive of the clinical condition or the anatomical changes underlying it. But the various names suggested have not met with any wide acceptance, and "Friedreich's disease" or "Friedreich's ataxy" still remains its most usual designation. Occasionally, also, the name "hereditary ataxy" is used as a synonym.

*Symptoms.*—Since the original observation of this disease by Friedreich many cases have been described under this name, and it is quite certain that a considerable number have not been true examples of the disease described by the Heidelberg professor. This has, unfortunately, produced considerable confusion in regard to the symptomatology of the disease, and when all doubtful cases are excluded from consideration it is remarkable to note the marvellous completeness of the clinical picture drawn by the original observer.



The usual age at which the first symptoms arise is between 6 and 10 years. Cases have been recorded as cases of Friedreich's ataxy in which the age of onset of symptoms has been a much more advanced one. Some of these cases it is already possible to classify under another heading; others must still be regarded as unclassified, although they can be excluded from the group we are at present concerned with. Some are undoubtedly cases of Friedreich's ataxy. The onset is characterised by some difficulty in walking. In some cases, occurring in the younger members of a family of which older members were already affected, before any obvious difficulty appeared, the kneejerks have been found to have disappeared. The gait, instead of being uniform and regular, is characterised by an occasional lurch, the feet come down heavily, and are not planted where it is desired to place them, and the patient is thrown off his balance. The efforts made to recover this cause still greater irregularity in the mode of progression. In standing the feet are placed widely apart to obtain greater stability, and the eyes are fixed on the ground, the attitude imparting to the patient an aspect of preoccupation. This difficulty in standing and walking commences usually, as already stated, before the age of 10, and as time goes on it gradually becomes greater. At length a period arrives when the patient finds it difficult to walk without support, and he slowly but gradually reaches a stage in which he is unable to walk at all, or even to stand. As the difficulty in walking increases other phenomena are superadded. There is a loss of rhythm and regularity in utterance and phonation, the final syllables in words are "clipped" or imperfectly pronounced, and a certain explosive character is imparted to the articulation. Speech proper does not suffer, and intelligence may remain unimpaired, although some emotional instability, especially a readiness to laugh, is usually superadded, and in some cases mental weakness has been present. In the cases in which grave mental change has been present other signs have existed, rendering the nature of the case doubtful. The

movements of the hands, also, become coarsely tremulous and uncertain, and even the head and trunk share in this unsteadiness—an unsteadiness which is not unlike that in chorea. In some cases there is a distinct *main en griffe*,



FIG. 37.—Friedreich's ataxy—the characteristic deformity of the feet.

not extreme in character, and in most cases there is weakness of the interossei. This is also the case in the feet, and accounts for the deformity to be observed in them. The lightning pains so characteristic of tabes are rarely, if ever, present, but occasionally slight cramp-like pains, or even

actual cramps, are complained of. There is no interference with the action of the sphincters except occasional slight weakness, such as is evidenced by difficulty in starting the act of micturition, and there is no marked sensory impairment or trophic disturbance, except the impairment of the muscular sense already alluded to. In some cases there



FIG. 38.—Lateral curvature in Friedreich's ataxy. From Gowers' 'Manual of Diseases of the Nervous System,' third edition, by permission of Sir William Gowers.

has been present some slight impairment of the sensation of the skin, particularly for heat or pain, but this is not common, far less is it constant.

With the difficulty in walking changes amounting to deformities in the back, or in the feet, or in both, are frequently associated. In the latter there is a peculiar

form of talipes to which the name of "humped foot" or "pes cavus" has been applied. It is characterised by an excessive arching of the dorsum, a hollowing on the inner and inferior aspect, and some inversion. The deformity in the back is a lateral curvature, which is often extreme, but neither of these changes is constant, although the latter is almost invariably present.

There is no interference with the normal pupillary reaction, and optic atrophy, although sometimes present, is extremely rare. Nystagmus is very frequent, although in many cases the slight movements on extreme deviation scarcely justify this name, and would be more correctly described as those of nystagmoid jerking. The state of the sphincters of the bladder and rectum has been already referred to. They are, generally speaking, unaffected, although a slight difficulty in starting or controlling the act of micturition is sometimes complained of. There is no difficulty with swallowing.

The deep reflexes are abolished. Not only the knee-jerk is lost, but also the wrist- and elbow-jerks. Yet in a few cases of Friedreich's ataxy, otherwise typical as regards gait, articulation, and deformities, the knee-jerk was found present, but diminished. The plantar reflex has been noted as very active in many published cases. In cases observed since the significance of alteration in this reflex has been observed an extensor response (Babinski's sign) has been evoked. Besides these more strictly nervous symptoms, a considerable degree of general weakness is frequently present, and a condition of distinct, although not excessive, anaemia may be said to be the rule. Glycosuria was present in one case described as Friedreich's ataxy. The patient died at 10 of diabetic coma. Examination of the heart usually reveals the presence of a very distinct basic systolic murmur.

A few years ago, from a consideration of cases published by several different observers and of some which he had himself seen, Marie concluded that there exists a type of disease in many points resembling Friedreich's ataxy, but differing in several important particulars. This he called

"hereditary cerebellar ataxy," and he regarded the symptoms as the result of some congenital defect in the development of the cerebellum. In the ataxy the articulatory difficulty, the frequent presence of nystagmus, and the fact that more than one member of a family might suffer, it closely resembled Friedreich's disease, but important and striking differences were manifested in the occasional presence of the Argyll Robertson pupil, the not unusual occurrence of optic atrophy, and the invariable exaggeration of the knee-jerk. The onset of symptoms also in this disease was usually at a much later date than in Friedreich's ataxy. The remarkable series of cases described by Dr. Sanger Brown probably belongs to this class. These were characterised by the occurrence of a similar condition in many members of the same family throughout several generations, namely, gradually increasing weakness and inco-ordination in the legs, with marked tremors of the head and body, and impaired articulation. In these points they resembled Friedreich's ataxy. They differed, however, in the fact that the age at which the symptoms commenced was more advanced—in one case it was as late as 45 years—the knee-jerks were exaggerated, and ankle clonus was occasionally present, optic atrophy was of frequent occurrence, and there was neither nystagmus nor deformities. Nonne and Menzel have also described similar conditions occurring in more than one member of the same family, but in Nonne's cases the ocular movements were impaired, the speech was loud, nasal and explosive, and there was some mental weakness. Gee, also, in this country, has described three cases of hereditary infantile spastic "paraplegia," and Tooth has met with instances of a family disease in which the symptoms were those of spastic paraplegia of gradual onset at or soon after puberty. The relation of these groups of cases to Friedreich's ataxy will be discussed in dealing with the pathology of the disease, and the diseases themselves will be considered separately.

*Etiology.*—One point of predominant importance in considering the origin of the disease is the tendency which is

manifest in the great majority of instances for the affection to attack more than one member of the same family. Alcoholic over-indulgence and syphilis in the parents of victims have each been invoked as possible and likely factors in its causation, and the evidence of the presence of one or other of these conditions, sometimes of both, has been in many instances indisputable. On the other hand, there does not seem to be anything like conclusive proof of the constant presence of either or both; and the fact that an isolated case has occurred in a large family, the other members of which, both older and younger than that one the subject of Friedreich's ataxy, were quite healthy, would seem to diminish the importance of such a history. In some instances the first symptoms have been manifested after some debilitating disease or acute illness. Two particular diseases, diphtheria and small-pox, have been supposed to be peculiarly efficacious in evoking the early symptoms.

*Pathology and morbid anatomy.*—The characteristic lesion which has been described in the cases examined *post mortem* has been a sclerosis affecting especially the posterior columns of the spinal cord, but in a less degree also the lateral and anterior—a condition, in short, of wide-spread sclerosis affecting all the different regions of the white matter of the cord, but falling with greatest force upon the posterior columns. Lissauer's tract usually, but not always, escapes—a feature of marked distinction from tabes, in which it is always degenerated. The cells in Clarke's column have also been found atrophied and the posterior roots and peripheral nerves degenerated. But another condition has been observed in several of the cases, and it is interesting that Friedreich himself should have noted this—namely, a peculiar smallness of the spinal cord. Taking into account what has been called the familial character of the disease, it seems highly probable that this condition of the nervous system really depends upon a tendency in certain parts of that system, especially on the afferent side, to premature decay and death—a tendency doubtless depending in turn upon a defect or perversion of the ordinary developmental

process. No doubt the condition of sclerosis and consequent contraction would account in some degree for the small size of the cord, but it is highly probable that this is



FIG. 39.—Friedreich's ataxy, cervical region, showing sclerosis in the posterior columns and also in the lateral. From sections kindly lent by Dr. Newton Pitt, prepared and photographed by Dr. Collier.

not the only cause of this peculiarity. Déjérine and others have also described a condition of peri-ependymal gliosis around the central canal.

Besides the evidence of defective development exhibited in the spinal cord, some observers have been of opinion



FIG. 40.—Friedreich's ataxy—dorsal region of the same cord.

that the cerebellum also is not so well developed—the condition supposed to underlie the manifestations of Marie's hereditary cerebellar ataxy. In one of Menzel's cases, already briefly alluded to, there was degeneration in the posterior column and in the lateral and anterior as in

typical Friedreich's disease. But there was also degeneration of the hypoglossal and facial nuclei and atrophy of the cerebellum and pons and of the olivary bodies. The atrophy of the cerebellum was associated with a deficiency in the cells of Purkinje. In Nonne's case, very similar clinically, the anatomical condition was the same as regards the cerebellum and pons, but there was no cord degeneration. These were apparently not undoubted cases of Friedreich's ataxy. In three of Sanger Brown's cases which have been examined *post mortem* widespread posterior sclerosis was found in the cervical region and slighter lateral changes. Marked changes in the cerebellum, however,

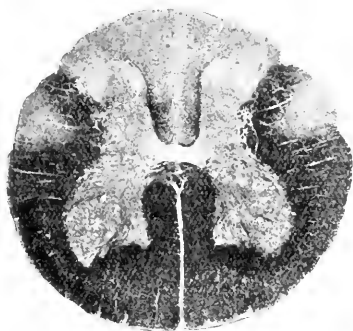


FIG. 41.—Friedreich's ataxy—upper lumbar region of same cord.

were not apparent, although the cells of Purkinje were not over-abundant. A case similar in its clinical aspects to that of Menzel, and almost identical as regards the cerebellar changes, the cord showing no alteration, was described several years ago by Fraser and Coats. This case may be regarded as a connecting-link between these different types. Whether future observation will be able to bridge over whatever small gulf remains between those cases and cases of Friedreich's disease it is impossible to say; but it seems not unlikely that it may be possible to demonstrate at some not very distant time that all belong to a group of cases in which there is defective development of the nervous system, especially on its afferent side, and that the slight differences between them are dependent upon the defect being mani-



fested in different degrees in different parts of the same system. It has been already stated that in a few cases of Friedreich's ataxy otherwise typical the knee-jerk has been found present. It can easily be conceived that if the sclerosis in the lateral column is considerable, while that in the posterior column is slight, the knee-jerk may be present and even exaggerated. Further, it might be that as the disease progresses and the posterior columns become more and more sclerosed the knee-jerk might in time disappear if the reflex arc were disorganised to such an extent as to make the transmission of impulses impossible. In the cases to which allusion has been made, in which the symptoms of lateral sclerosis are distinct, we may assume that the tendency to degeneration has manifested itself in its full force in that region, while the posterior columns have been comparatively spared. The cases of Friedreich's ataxy, otherwise typical, in which the knee-jerk is present must be regarded as forming a connecting-link between the two extremes.

*Diagnosis.*—To anyone unfamiliar with the disease the diagnosis is not easy. The early age at which the symptoms commence, the absence of the characteristic pain and of the pupillary inaction to light, are usually sufficient to distinguish it from the ordinary form of locomotor ataxy. In many points, however, as is only natural considering the changes in the spinal cord, it presents a close resemblance to combined posterior and lateral sclerosis—the ataxic paraplegia of Gowers. But in the latter, also, the age at which the symptoms commence is usually much more advanced than it is in cases of Friedreich's ataxy, and there is an absence of the deformities, one or other of which is usually present in the latter disease. Cases of disseminated sclerosis offer the closest resemblance to cases of hereditary ataxy. In the latter the knee-jerk is nearly always absent, while in the former great exaggeration is the rule, and the tremor is different in being in the former so much exaggerated by voluntary movement. The articulation in the two diseases is, in some instances at least, closely similar, and the

presence of nystagmus in each further increases the resemblance. So that reliance must, as a rule, be placed on the general picture presented, and attention be particularly directed in any doubtful case to the family history, the state of the deep reflexes, and the presence or absence of deformities. It may also be remarked that cases of disseminated sclerosis are, as a rule, characterised by periods of improvement and deterioration, while in Friedreich's ataxy the progress is steadily downwards.

*Prognosis.*—From the nature of the disease the prognosis is necessarily bad. There is no immediate danger to life, and patients often live for many years after they have become completely helpless. But arrest of the process, far less improvement, is, with our present knowledge and resources, not to be expected. Death usually results from some intercurrent complication, which soon proves fatal to a patient already greatly emaciated and enfeebled.

*Treatment* chiefly consists in attention to general health and hygiene and to symptoms as they arise. The anæmia is usually benefited by the administration of iron and arsenic. There is no contra-indication to strychnia, which often does good by improving the patient's appetite and general condition. The cramps are usually not so severe as to call for any treatment except gentle rubbing. It is doubtful if much good is effected by dividing the tendons of the muscles whose traction produces the foot deformity.

## HEREDITARY CEREBELLAR ATAXY

THE conditions described under this name a few years ago by Pierre Marie, although in many important points bearing a close resemblance to Friedreich's ataxy, differ from it in so many of its features that it must be regarded as a different disease. The cases so far described are a somewhat heterogeneous collection, and with further experience it is not unlikely that the clinical picture which has been drawn will be altered in some of its outlines, and it is almost certain to have many additional details added to it.

*Symptoms.*—The first symptom of the disease is unsteadiness in walking or standing. This does not, as a rule, appear until about the age of 20, and it has first shown itself as late as 45. The difficulty slowly but steadily increases, the patient walks with his feet apart and sways from side to side as if drunk. There is also unsteadiness in standing, but this is said not to be increased when the eyes are closed. After the lapse of a varying time (from one to three years) the hands become affected. The affection first shows itself in a certain degree of tremor or unsteadiness on voluntary movement. This increases so as to exhibit in time a close resemblance to the so-called intention tremor of disseminated sclerosis. The writing is much altered and the act becomes more and more difficult. Meantime the weakness in the lower limbs may increase so that the patient becomes unable to walk. Besides the difficulties in the limbs curious choreiform movements may be present, somewhat resembling those in Friedreich's ataxy but more constant and more extensive. These movements frequently affect the head and trunk and face as well as the limbs.

Jerky spasmodic movements may also be present, and a curious difficulty is experienced by the patient in letting go anything he has grasped in his hand. This prolongation, and even exaggeration of muscular contraction, shows itself also in the facial movements so that the naso-labial fold becomes exaggerated and the mouth may expand into a very marked grin. A similar muscular spasm is also shown in certain associated movements, as for example when the patient is speaking the occipito-frontalis becomes violently contracted and the eyes become widely opened and staring. The speech is jerky and explosive, not elisive or syllabic like that in Friedreich's disease or disseminated sclerosis, and the voice as a rule is low, guttural, and monotonous, with an occasional irregular and unexpected break into high notes.

Sensory impairment is not present. Subjective sensations of pains in the limbs or back, headache, and vertigo occasionally occur. Trophic and vaso-motor disturbances are not present, and deformities such as pes cavus or lateral curvature are by no means constant and are probably rare.

The sphincters are said not to be definitely affected. In at least one case, however, incontinence of urine was present. In some of Dr. Sanger Brown's cases there was difficulty with micturition, and when this was present there were frequently shooting pains in the limbs. There is occasionally some difficulty in swallowing, the knee-jerks are active, and ankle clonus may be present. The mental condition need not be affected. Occasionally there seems to be some dulness and lethargy, but this impression may be erroneous on account of the dejected expression, which is not always correlated with mental impairment. Indeed, in one series of cases the mental condition was unusually and unexpectedly good.

Important ocular symptoms are present. Atrophy of the optic disc with impaired vision and contracted fields is the rule in this disease. Nystagmoid jerkings also occur, not definite enough or constant enough to be designated nystagmus. In Dr. Sanger Brown's cases they were absent.

Weakness of certain ocular muscles—external rectus, superior rectus, levator palpebrarum—has also been present. This is not constant, although a tendency has been noticed to the occurrence of the same ocular weakness in different members of an affected family.

*Etiology.*—In considering the etiology of this condition the family tendency to the disease is the most striking phenomenon. What determines this we do not know. Consanguinity, alcoholism, syphilis, and tubercle seem to have no part in its causation. The late age at which the symptoms first show themselves, usually after 20 and as late as 45, explains the fact that the disease may be transmitted through several generations. The earlier age of onset of the disease in Freidreich's ataxy probably precludes marriage and procreation. Accidents and mental and physical shocks do not seem to be even exciting causes.

*Pathology and morbid anatomy.*—The disease seems to depend pathologically upon defective development or premature decay of certain structures especially related to equilibration and connected with the cerebellum. In all the cases examined until now the unusually small size of the cerebellum has been obvious. In some this has been very marked, as in one case in which it weighed 81 grammes instead of the usual 160 or 170. In others the diminution in size has been slight. In most cases the diminution has depended upon a general reduction of all the elements. In some the diminution in the number of the cells of Purkinje has been a striking feature—a condition similar to what has been met with in certain of the lower animals, while in others these cells have been apparently normal in size and in number but the tissue in which they are situated has been found to be smaller in extent and more fibrous than usual. In these cases, also, the grey matter generally of the cerebellum has been reduced in amount, and the sulci have been deeper and wider. The cerebellar white substance has also been reduced in volume, but the dentate nucleus remained unaltered. In other cases in which the changes in the cerebellum have been apparently slight there have been distinct alterations

in the connections between the organ and the spinal cord and the generally small size of the cord has been obvious.

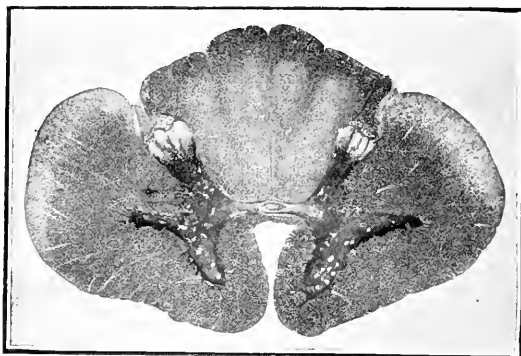


FIG. 42.—Cervical region of cord in one of Dr. Sanger Brown's cases. From Dr. Barker's paper in 'The Decennial Publications.'

This reduction in size of the cord has been associated with atrophy of its grey substance and degeneration in the

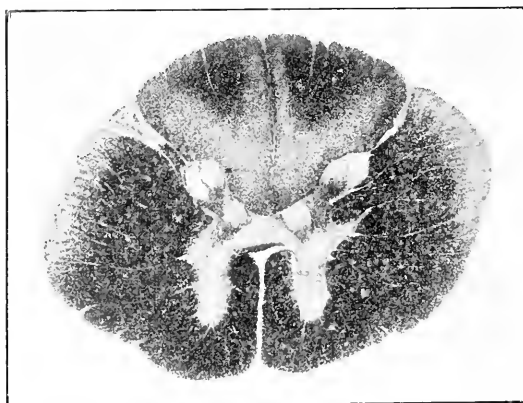


FIG. 43.—Dorsal region in same case.

posterior columns, in the column of Gowers, and in the restiform bodies. The column of Clarke has also been altered both in its cells and fibres. In Sanger Brown's three cases

examined *post mortem*, one by Meyer, and two by Barker, the degeneration was in the dorsal cerebellar tract and not in the ventral-cerebellar tract of Gowers. The condition of the cord, indeed, has been strikingly similar to that found in Friedreich's ataxy, but the changes have been less extensive. So far, then, as the histological examination is concerned, it would seem that in this disease we are dealing with a condition in which the afferent structures on which co-ordination and equilibration depend are the parts affected.



FIG. 44.—Lumbar region, same cord. In all sections the degenerated part (lightly stained) is similar in distribution to that in Friedreich's ataxy.

In some of the cases the cerebellum alone seems to be the part at fault; in other cases the alterations in that organ itself are comparatively slight, but those in the tracts leading to it from the periphery are somewhat extensive. This latter class, in the anatomical changes present, closely resembles cases of Friedreich's ataxy; the morbid anatomy in the former class only resembles that of Friedreich's ataxy because of the diminution in size of the cerebellum and cord. Whether future observation will show that the two diseases are essentially the same remains to be seen. It is not impossible that as in the remarkable cases of Sanger Brown, three of which have been examined *post mortem* with

strikingly similar results, changes in the cerebellar afferent tracts of the cord especially, will be found to constitute the most distinct feature of the disease.

*Diagnosis.*—The closest relations of this disease clinically (and, as we have also found, pathologically) are with Freidreich's ataxy. While the two diseases are strikingly similar in the presence of ataxy of the cerebellar type, of nystagmus occasionally, and of choreiform movements always, of inco-ordination of the hands, and of articulatory defects, the disease under consideration differs very strikingly in the later stage at which it commences, in the activity of the deep reflexes as contrasted with their absence in Friedreich's disease, and in the frequent occurrence of optic atrophy. Deformities in the spine and in the feet are also, at least unusual, in cerebellar ataxy. Yet it would be rash to say that the knee-jerk is never present in Freidreich's ataxy and optic atrophy certainly does occur, although very rarely, in that disease. The writer has under his care at present two cases of Freidreich's ataxy in the same family, each of them showing all the characteristic signs and symptoms of the disease, but one having in addition well-marked optic atrophy which has caused almost complete blindness. These considerations show the close resemblance between cases of the two diseases. They suggest a relationship, and it may be that future observations will show that the relationship is even closer than now appears. Certain forms of cerebellar disease may also closely resemble this condition clinically. These, however, are usually the result of inflammatory or other conditions characterised by an acute onset. The family form of spastic paralysis also offers certain resemblances in the condition of the reflexes and the frequent presence of nystagmus and perhaps of optic atrophy. But the gait is merely spastic, not ataxic or reeling, and although the articulation may be slurring it is not so markedly explosive and there are no choreiform movements present. Certain cases also of disseminated sclerosis resemble closely cases of cerebellar ataxy. The age of onset, the presence of ataxy and weakness of the legs, and tremors in the hands,



of articulatory difficulty and of optic atrophy may all correspond. But the absence of family history, the frequent presence of sphincter trouble in disseminated sclerosis contrasted with its rarity in cerebellar ataxy and the probably more rapid progress in disseminated sclerosis, will usually serve to distinguish the two.

*Prognosis.*—The prognosis, from the nature of the disease, is necessarily bad. The disability gradually increases, but the absence of any condition, such as bladder trouble, likely to endanger life renders the outlook as regards the duration of life quite good. The encroachments of the disease are slow, and a patient may remain able to get about, at least indoors, for many years after the commencement of the illness.

*Treatment.*—Treatment with a view to modifying the diseased condition is obviously in the present state of our knowledge quite hopeless. All that we can do is to treat symptoms such as pain, cramp, etc., as they happen to arise, and to endeavour to keep the patient under the best attainable conditions as regards food, nursing and hygiene.

## JUVENILE OR HEREDITARY TABES DORSALIS

THIS has hitherto been regarded as a somewhat indefinite condition closely resembling in many points the disease which we have just considered—Friedreich's ataxy. A careful consideration of the cases under this name as well as of some which have been looked upon as anomalous cases of Friedreich's ataxy, makes it clear that it is now to be regarded as a definite clinical entity, with, however, variations in the symptomatology of the clinical varieties of the disease comparable with those which occur in the cases of ordinary Tabes dorsalis.

Its first and most important characteristic is its occurrence in early life. It may occur as early as 8 or be discovered as late as 23, although in a case described at the latter age it was present in a fairly developed form. It had probably, therefore, first manifested itself some years earlier. In one variety the early symptoms are lightning pains in the legs. These are usually accompanied by some weakness and ataxy of the limbs, abolition of the knee-jerk, and perhaps some slight affection of the sphincters. The Argyll Robertson phenomenon is usually present. Another clinical variety is characterised by an early affection of vision, the result of optic atrophy. At first there may be no ataxy, although the knee-jerk may be impaired or even lost. The optic atrophy may go on to cause complete blindness, but ataxy and other symptoms may remain in abeyance, temporarily or permanently. In an unusually large proportion of the published cases optic atrophy has been present, sometimes with ataxy, more frequently without it. But even if ataxy be absent in such cases the knee-jerk is usually found to be lost, or at least defective, and the pupil is almost always,

under such conditions, inactive to light. Not uncommonly different members of the same family are found to be affected, usually in different degrees, at least when affected simultaneously. In one very interesting family, of which the details have been published by Dr. Brooksbank James, there were eight surviving children out of fourteen born alive. Of these eight the eldest was a young woman of 20 with paralysis of the left external rectus muscle, absent knee-jerks, and Argyll Robertson pupils. The second was a boy of 17 with Hutchinson's teeth, scars at the angles of the mouth, gross central choroido-retinal changes and sluggish knee-jerks. The third was a girl of 15, with sluggish knee-jerks and pupils fully active to light. The central incisors were separated and narrowed at the extremities. The fourth was a girl of 10 with normal pupils and active knee-jerks, but somewhat suspicious incisors. The fifth was a girl of 8 with central incisors separated and narrowed inferiorly, unequal pupils which were inactive to light, and sluggish knee-jerks. The other two living children were aged respectively 4 years and 18 months, and showed no abnormality—at least at the time of publication.

In another case the patient was a lad of 17 years, the son of a tabetic father; the knee-jerks were lost, the pupils were inactive to light, there was marked ataxy of the arms and legs and optic atrophy with blindness. In another of the recorded cases the father of the patient had been tabetic. One striking feature in many of the published cases is the co-existence with signs and symptoms of tabetic affection in the patient or other members of the family, of definite evidence of congenital syphilis. The importance of this will be alluded to in reference to the etiology and pathology of the condition.

*Etiology and pathology.*—There seems no room for doubt that juvenile tabes is a result of congenital syphilis. The cases which are briefly alluded to above are random but characteristic examples of all the recorded cases, and one cannot but be struck by the close analogy which such cases

bear to those of juvenile general paralysis. In these also the influence of congenital syphilis is strikingly marked by the presence, in such a large proportion of the cases, of definite stigmata of that disease or of a history which more than suggests it. Occasionally the two conditions—as frequently happens with ordinary *tabes dorsalis*, and general paralysis—are associated in the same patients. Strumpell records a case of a girl of 13 who had juvenile *tabes* and also symptoms of general paralysis. The father had had syphilis, and secondary symptoms were present two years before the patient's birth. The writer had under his care, a few years ago, a young man of 17, who had commencing optic atrophy and sluggish knee-jerks. His knee-jerks entirely disappeared and he became blind. Soon after his blindness was complete he developed slurring articulation and mental exaltation, which necessitated his removal to an asylum. He himself had no definite signs of congenital syphilis, but his sister had Hutchinson's teeth, scarring about the mouth, and disseminated choroidal atrophy. So that in this case also, in which there was a condition of combined juvenile *tabes* and juvenile general paralysis, there was evidence of the influence of parental syphilis. Of course, it is not easy to furnish conclusive proof that these diseases are the result of inherited syphilis. Yet the evidence is so strong in favour of such a view as to suggest the question whether it might not be well to recognise the fact explicitly in the descriptive designation of the two diseases. It would, then, appear that there are two degenerative diseases of the nervous system especially prone to occur in children who are the subjects of congenital syphilis: one, the disease known as juvenile *tabes* characterised by one or more of the following symptoms—loss of knee-jerks, ataxy, lightning pains, incontinence of urine, optic atrophy; the other, juvenile general paralysis of the insane, with mental symptoms similar to those that are present in the ordinary general paralysis of the adult, together with Argyll Robertson pupil, occasionally optic atrophy, and either

an active or an absent knee-jerk; and as is the case also in the ordinary form of these diseases as they occur in adults, the two conditions may co-exist in these juvenile patients.

The pathological anatomy is not yet definitely established. Sclerosis of the posterior column is almost certainly present, and there is probably a sclerosis in other parts of the cord as well, just as there is in Friedreich's ataxy.

*Diagnosis.*—A difficulty may arise in the diagnosis of this condition from Friedreich's ataxy. Yet in the latter the absence of lightning pains and of pupillary phenomena and the presence of the nystagmus and of the deformities already described in that disease, will usually be sufficient to distinguish the two. It must, however, be remembered that occasionally in juvenile tabes a lateral curvature of the spine has been described. Sometimes a difficulty will arise in deciding whether a patient, *e.g.* of 20 years, is suffering from the congenital form of weakness or from the acquired form. A consideration of the condition and all the circumstances of the patient and of the family history will usually be sufficient to allow of a correct decision being arrived at. The condition of the reflexes will usually be sufficient to distinguish the disease from the hereditary type of spastic paralysis and of cerebellar ataxy. The distinction between juvenile tabes and juvenile general paralysis may occasionally be somewhat arbitrary, for in the cases in which both diseases co-exist it is not easy to say when one ends and the other begins.

*Prognosis.*—The prognosis is distinctly bad, although there is much variation in the rate at which the disease progresses. It may be very rapid; it may, on the other hand, gradually lead to a condition of almost complete disablement in which the patient may remain for years. When blindness is also present the condition of the patient is very distressing, and may remain practically unaltered during several years.

*Treatment.*—The treatment which naturally suggests itself, having regard to the probable influence of syphilis,

is that by mercury and the iodides. Unfortunately, it seems to have little, if any, effect, but it is certainly desirable to try such treatment with care and completeness. Any other line of treatment must simply attempt to alter or alleviate such conditions as arise in the course of the disease.

## FAMILY SPASTIC PARALYSIS

REFERENCE has already been made in the section on "Cerebral Diplegia," to conditions to which Sir William Gowers has applied the name "abiotrophy," conditions, that is, in which certain structures—and in this instance we are dealing with structures of the nervous system—seem to have been endowed with a short tenure of life, and in which those structures tend to decay prematurely, and, in the course of their decay, to give rise to certain symptoms. In many cases this tendency is present chiefly in the pyramidal system of fibres, so that the condition which arises is one of spastic paralysis, and the anatomical condition in the spinal cord associated with it is one of lateral sclerosis. In some cases, as has already been indicated in the article referred to, such a condition seems to date from birth. The child is born with a pyramidal system undeveloped, which never develops, or, it may be, is born with a pyramidal system fairly normal, which tends, however, to decay very quickly—that is, in the first few months or years of life. But in other cases—and those are the cases with which this section is concerned—the tendency to decay does not assert itself until later in life, frequently about the age of puberty, or even later, and a condition of spastic paralysis then gradually develops. A good many instances are now on record in which such weakness has developed, sometimes in a solitary member of a family, sometimes in several members of the same family, at or about the age of puberty, and has gone on to cause complete disability in the course of a comparatively short time.

The *symptoms* characterising such a condition are great stiffness and rigidity of the lower limbs of gradual onset,

exaggerated reflexes, ankle clonus, with extensor plantar response. There is, as a rule, no sensory impairment, and the upper limbs may not be affected, or if they are the affection is only of very slight degree. Nystagmus is occasionally present, and a difficulty in articulation similar to that which occurs in disseminated sclerosis has been noted, not only in the isolated cases, but also in the cases in which more than one member of the family has suffered.

The *pathology* of the condition has already been indicated in the introductory remarks. Certain parts of the nervous system, although they may be apparently developed normally, are endowed with that short tenure of life which renders their early decay almost inevitable. And the morbid anatomy shows that the decay which takes place is chiefly in the pyramidal system of cells and fibres. Besides such degeneration it is not unusual to find in those cases degeneration in the posterior columns as well, extending from the lumbar to the cervical region, and also occasionally in the cerebellar system. But the degeneration in those parts is not nearly so profound as it is in the pyramidal system, and it is probably secondary to the interference with movement which the pyramidal sclerosis necessarily entails.

*Diagnosis.*—The diagnosis in such cases is necessary from other conditions of spastic paralysis resulting from lateral sclerosis. A tumour of slowly growing character may, of course, give rise to spastic paralysis, and may lead to the symptoms of lateral sclerosis. So may pressure from tubercular or syphilitic disease, but those conditions would be distinguished by the absence of the family history, and by the presence, frequently, of more or less severe pain. It is exceedingly difficult in many cases to distinguish such a condition from commencing disseminated sclerosis. Indeed, at certain stages the distinction is impossible, and it may be necessary to wait and watch the further progress of a case before it is possible to be quite certain.

The *prognosis*, of course, is bad as regards anything like improvement taking place, but it is not necessarily bad as



regards duration of life, for life in such a disease is often prolonged for many years, the degeneration seeming to set in, to reach its maximum in the structures affected in a comparatively short time, and after this the other structures which are left undegenerated apparently remain normal.

*Treatment*, unfortunately, is inefficacious. No drugs and no measures with which we are as yet familiar can be regarded as likely in any degree to alleviate the symptoms of this inevitable disease.

In certain cases there seems to be not only an affection, such as that indicated above, of the pyramidal system, or upper motor neuron, but the lower neuron also seems to be affected, resulting in the production of a condition of wasting, of the extremities especially, similar to that which occurs in progressive muscular atrophy. In those rare and unusual cases, of which several are now on record, occurring in several families, and affecting perhaps more than one member of the family, the disease must be regarded as probably also of the atrophic type, in which not only the upper neuron has been affected, but in which the lower neuron has suffered also. Such cases are, indeed, instances of amyotrophic lateral sclerosis of a family type. They occur at a much earlier age than the usual form of that disease, and, as has already been said, one or more members of the same family may be affected.

The *prognosis* in this is by no means good, and the patients inevitably become completely helpless.

The *diagnosis* has usually to be made from cases of ordinary amyotrophic lateral sclerosis, and the age at which the disease occurs is a great help in making the distinction. They have also to be distinguished from cases of syringomyelia, the distinction from the latter being chiefly in the absence of any sensory phenomena.

*Treatment* in this condition also is quite inefficacious.

## PARAPLEGIA FROM SPINAL CARIES

(POTT'S PARAPLEGIA)

THE occurrence of this condition has acquired fresh interest and importance in recent years on account of the operative measures which have been advocated for its relief. These in some instances have been crowned with almost dramatic success; in others no improvement has been effected, and it is of the utmost importance to try to distinguish if possible between cases which are likely to benefit from operative measures and others in which contra-indications to operation exist. The disease is not uncommon in children; and while in many respects the condition as it occurs in children and as it occurs in adults is practically the same, it must be understood that what is about to be said, in reference especially to treatment, applies in a peculiar way to the condition as it occurs in young people. A characteristic case in a child may be described as follows: The child, previously healthy, possibly not regarded as robust, is found to become much more easily tired than formerly, and not to be willing to use its legs so freely. With this languor, and what is looked upon sometimes as indolence, no pain may be associated. Occasionally, however, it is found that the limbs "start" at night, and it is not uncommon for the child, to its own great distress, to wet the bed. The weakness becomes more and more marked, the disinclination and inability to get about become more striking, and it may sometimes be noticed that a stooping attitude is assumed, and that the back is kept more rigid than usual. An examination of the limbs at this time would probably reveal a complete intactness of sensory functions.

It is usually found, however, that the knee-jerks are exaggerated, ankle clonus may be present, and frequently the plantar response is of the extensor type. On examining the back, a slight angular curvature may be visible. If this is so, the nature of the condition is at once revealed. On the other hand, there may be no such curvature, but it is usually found that there is a degree of rigidity about the spinal column which is of similar significance. Of course, if the condition is recognised, treatment is at once adopted. But in a case in which it is not recognised the child usually tries to get about until it is no longer able to walk. It is then found to be in a very spastic condition; the legs are closely locked together, extremely rigid, there may be even some slight sensory impairment. The sphincters also are frequently affected, there is usually difficulty in controlling the action of the bladder, and the sphincter ani may cease to act, especially on the occasions on which some opening medicine has been taken. Girdle sensation is fairly common, and in a case such as that described would be almost certain to be present. It must be remembered, however, that the deformity of angular curvature need not necessarily show itself. Such a deformity is produced by the falling together of the anterior parts of the bodies of the vertebrae, and the consequent protrusion of the spinous processes. But it is quite conceivable that a good deal of inflammation of the membrane of the cord, secondary to bone disease, may be present—sufficient, indeed, to give rise to complete paraplegia, yet the bone disease may not be so severe as to give rise to any visible deformity of the spinal column.

Such a condition as that described relates, chiefly of course, to caries as it occurs in the dorsal region. When it occurs in the lumbar region we are much less likely to have any obvious deformity of the spine present; but then the rigidity of the spinal column will be very marked. Again, when caries affects the lumbar region it is quite possible that the reflexes, especially the knee-jerks, instead of being exaggerated, may be actually abolished, because the reflex arc, on the integrity of which the knee-jerk

depends, may be interrupted by the inflammatory process. Similarly, if the caries is situated in the cervical region, deformity in the shape of an actual acute angular curvature rarely occurs, and can only occur if the carious process in the spinal column is of extreme degree. What most frequently happens is that there is much thickening about the spine in the cervical region, and great infiltration of the soft tissues of the neck. So that on feeling the patient's neck one is at once struck by the great thickening and hardness of all the tissues about the spinal column. Further, in those cervical cases not only are the lower limbs affected, but also the upper limbs, and from the fact that the actual nerve-roots are frequently involved in the disease, local wasting of muscles, especially the small muscles of the hand, is a very frequent accompaniment of cervical caries.

To sum up, then, the clinical history in a case of spinal caries is characterised by a gradual onset of weakness, in the lower limbs only if the caries is in the dorsal or lumbar region, frequently by girdle sensation, occasionally by shooting pains around the waist and even into the limbs, by an interference with the function of the sphincters of the bladder and the rectum, and by some degree of anæsthesia in the severe cases. It must be remembered, however, that even in cases in which the motor paralysis is complete there may be no sensory change at all. If the caries affects the cervical region, then we may have weakness or paralysis of all four limbs. There is, as has already been stated, not necessarily any actual angular curvature of the cervical spine, but there is much thickening, especially in the soft tissues of the neck. There is also frequently wasting in the hand muscles—it may be also in the arm muscles—from involvement of the nerve-roots which supply them, and there may be anæsthesia of the distribution which characterises an affection of the nerve-roots. The knee-jerks are usually exaggerated—they may be absent in lumbar caries—ankle clonus is frequent, and the plantar reflex is usually of the extensor type.

*Etiology.*—The etiology of such a condition as this is, of course, related to the family history indicating a tendency to tuberculous disease, and to the personal history of the patient in reference to other manifestations of tubercle. It is quite possible that there may be no evidence of any tubercular lesion in any other part of the body. Occasionally, however, some indication of tubercle affecting the lungs may be present, and in such cases the nature of the condition is at once obvious. As regards the immediate cause of the caries, one cannot help feeling that the occurrence of falls, and of jars to the back, is of some importance. Just as a blow on the head is so frequently the precursor of tubercular meningitis, apparently causing such a local lowering of vitality as to give the bacillus its chance of invasion, so also a blow on the back or an injury to it through a fall or strain seems to determine, in many instances, the onset of caries of the spine.

*Morbid anatomy.*—From what has been said it will be clear that the condition which gives rise to paraplegia in association with spinal caries is really one of gradual myelitis. The myelitis, the actual affection of the spinal cord itself, is apparently secondary to pressure exerted from outside, or inflammation spreading from membranes. This pressure is not necessarily primarily by the bone itself; but bone disease, by irritating the membranes and probably pressing upon them, gives rise to an inflammatory hypertrophy, especially of the dura mater, which is so striking in some instances as actually to have been named by Macewen a "tumour"; and this it is which, by its pressure, and possibly by the inflammation which it communicates to the spinal cord, gives rise to myelitis. Sometimes the pressure exerted is extremely severe, so that the cord becomes narrowed to an almost incredible degree, and when one has seen in a patient with complete paraplegia such a cord, which afterwards apparently entirely recovers its functions, it is a matter for great surprise how such a marvellous regeneration in such a delicate structure can be effected. In many instances not only is the cord affected,

but the nerve-roots also, both sensory and motor, may be involved. This will be understood from the nature of the disease, which causes collapse of the bodies of the vertebrae, and consequent pressure upon the nerve-roots at their exit from the canal and actual inflammation of them. But from the fact that the disease is so frequently present in the dorsal region of the cord, it can be understood that such interference with the function of the nerve-roots need not necessarily be very obvious. In cases, however, in which the lesion is in the cervical region there is frequently, as has already been stated, definite wasting of the muscles, especially the small muscles of the hand. An examination of the cord itself in a case of spinal caries reveals the existence of an inflammatory condition, as is shown by increased vascularity, and infiltration of leucocytes at the seat of pressure. There is also, of course, a breaking down of the nerve-tissue proper, and the tendency to its replacement by fibrous tissue. Below the lesion there is the usual descending degeneration, which shows itself most markedly in the pyramidal tracts of the cord. Above the lesion there are areas of degeneration of ascending tracts.

*Diagnosis.*—The diagnosis of paraplegia from spinal caries in the early stage may not be at all easy. It is especially difficult in the cases in which no deformity of the spine is present. Indeed, in such a condition as that it may be impossible to diagnose it, and one can only come to the conclusion that the patient is suffering from a form of spastic paraplegia which may be the result of pressure from caries of the spine or of one of the many other conditions giving rise to that form of weakness. Where, however, there is the co-existence of weakness of the legs, with deformity of the spinal column, the diagnosis is not difficult. And it is always important to recognise the necessity for examining, in every case of spastic paraplegia, into the condition of the back. In no condition is the old dictum truer, that "more things are missed by not looking than by not knowing," than in cases of spastic paralysis of obscure origin. In every such case a careful

examination of the back is of the utmost value and importance. But even with one's eyes open to the possibility of spinal caries the diagnosis is by no means easy in cases in which deformity of the back is not obvious. Thus, in one instance the diagnosis of disseminated sclerosis was made because the patient was suffering from spastic paraplegia without any sign either of deformity or rigidity of the spine. Six weeks later the patient had a very distinct angular curvature, and the true nature of the disease was manifest. Wherever, then, in any child previously healthy there are signs of a gradual weakness of a spastic character affecting the legs, caries of the spine should be suspected. Where the condition is one affecting all four limbs, an examination of the cervical spine, both from outside and also from the pharynx, ought at once to be made.

*Prognosis.*—The prognosis in cases of spinal caries must depend to a great extent upon the general health of the child and upon the conditions which are attainable for its treatment. Most cases in children, fairly healthy otherwise, do quite well, even in hospital. In sanatoria, or under conditions in which fresh air, good food, and careful nursing can be obtained, the results are even better. Of course, any involvement by tubercular disease of other organs necessarily complicates the outlook very much, and it is not uncommon for general tuberculosis to supervene if the lungs have once become involved. It is rather a curious fact, but the onset of tubercular meningitis in cases of caries of the spine is undoubtedly rare.

*Treatment.*—The treatment of the condition is, first of all, by means of rest, made as absolute as possible by the use of extension, and counter-extension if necessary; by fresh air, the best possible hygienic conditions, good food, and careful nursing. A child who has commencing weakness in the legs in whom the condition of caries is recognised should be at once put to bed, extensions applied to the legs, and, if it is impossible to keep the child quiet otherwise, counter-extension also to the head. It is quite remarkable with what ease such treatment is borne by even

quite young children, and how quickly they reconcile themselves to the conditions, and how rapidly they improve. In the great majority of cases of caries in children no other treatment than this is necessary. One may often have to wait a very long time before the result is obtained. Thus, a child lay absolutely immobile, quite unable to move either of its feet, for a whole year. Improvement then began, and within six weeks the child was walking about. So that one must not be discouraged by the length of time which is sometimes necessary before improvement takes place. The same treatment, of course, must be applied to cases of caries of the cervical spine. These are much more difficult to treat, and on account of the proximity of the disease to structures which are essential for life, the child is actually in more imminent danger from the chance that the inflammation may spread to these structures.

The question of operation in caries of the spine in children is one which has not very often to be considered. There is no doubt that in many of the cases of cervical caries operation is urgently demanded on account of the disease involving the phrenic nerve and the condition of respiration becoming thus very much embarrassed. In most dorsal and lumbar cases, however, treatment by rest and immobility is, as a rule, quite effective, and one must always remember that, even in cervical cases, in which the symptom may be extremely urgent, one would not be quite sure that by relieving pressure in the cervical region one is really relieving all the conditions present. In one case of the kind in which operation was undertaken the child, unfortunately, died, and two other foci of the tubercular disease in the spinal column were discovered. In most cases in which the disease affects the dorsal region surgical treatment will be rarely, if ever, necessary if only one is prepared to go on long enough with treatment by rest, good food, and the best possible hygienic conditions. It must also be remembered that when the patient is once more able to get about some rigid support for the spine is desirable—at least for a time.



## SPINA BIFIDA

ALTHOUGH the chief clinical interests of the condition known as *spina bifida* have been surgical, its medical and especially its developmental aspects, are of peculiar importance. Some years ago the Clinical Society of London appointed a Committee to investigate the condition, and although the work of this committee was undertaken in the first instance with the view of investigating the results of treatment by means of Dr. Morton's injection of iodo-glycerine solution, and other methods, the results arrived at, after a careful pathological examination of many specimens, are extremely important, and have been the means of rendering our knowledge of the condition at once extensive and exact. The condition is described as of three varieties:

(1) That in which the membranes only are protruded—spinal meningocele.

(2) That in which there is, besides its membranes, protrusion of the spinal cord itself and its appertaining nerves—meningo-myelocele.

(3) That in which there is protrusion of the membranes and of the spinal cord, the central canal of which is dilated so as to form the sac cavity—the inner lining of this being constituted by the expanded and atrophied substance of the cord—syringo-myelocele.

The second variety is by far the most common. Besides these three forms there is also a condition known as *spina bifida occulta*. In this variety there is no protrusion through the defective canal, although the vertebral column may be felt to be defective. There is usually a spinal cord extending considerably lower than normal, and over the

spot where the defect in the arches exists—usually in the lumbo-sacral region—there is frequently an excessive growth of hair. In this form also there is often a dimple just below the coccyx—the post-anal dimple, and it is not uncommon to have only the functions of the sphincters interfered with, and perhaps the existence of some anaesthesia in the perineal region.

The condition of spina bifida is essentially a developmental defect, and the varieties of the condition already alluded to depend upon the degree to which this defect exists. In normal development the medullary plates unite to form the normal tube—the embryonal spinal cord. There is then an ingrowth of the mesoblastic tissue from each side to enclose the cord and form the neural arches of the vertebrae and the membranes of the spinal cord. If fluid now accumulate in front of the spinal cord—and it seems as if defect in some way led to this—the cord, unsupported by membranes behind, gets pushed back and flattened out, thus forming the posterior wall of a cyst, which protrudes through the defect in the bony arch. The nerve-roots and ligamentum denticulatum, in consequence of this displacement of the cord, become enormously lengthened out. The epidermis is continued over the sac wall, but in the middle line the skin is destitute of hair and other appendages. The absence of corium, muscle, etc. (which grow from the mesoblast) is due to a defect in this, the same as leads to absence of the neural arches; but, curiously enough, in spina bifida occulta there is an excessive growth of hair in this part, as if the part of the mesoblast on which this depends had undergone excessive growth, without leading to the development of the bony arches of the vertebral column. In some cases of spina bifida not only are the arches defective, but even the bodies of the vertebrae have not been developed, and there is a deficiency in the bony canal in front as well as behind.

(1) *Spinal meningocele*.—It has been stated that this is a very uncommon form of spina bifida, but there is a

certain fallacy underlying this statement. There is no doubt that it is not common as a museum specimen, for out of the 125 specimens of spina bifida examined by the Committee of the Clinical Society only ten were examples of meningocele. But it must be remembered that this form of lesion, which sometimes remains *in statu quo*, and sometimes undergoes spontaneous cure, is much less fatal to life than others, and so its occurrence as a museum specimen will be correspondingly rare. The deformity may occur in any region. The deficiency in the neural arches is usually limited to a small area, and, indeed, the protrusion may take place between the arches of adjacent vertebrae. Occasionally, however, the bony deficiency is extensive. The typical form of meningocele may be assumed to consist of a sac composed of dura mater and arachnoid communicating with the general cavities of the spinal membranes, invested with normal skin and tending to be pedunculated. But such a specimen the Committee did not see. Occasionally the sac is double within, composed of distinct portions under a single cutaneous covering. Sometimes the spinal cord becomes prolapsed so as to lie in the neck of the sac and more or less occlude it. Occasionally at the summit of the sac there is a smooth longitudinal depression unpigmented, devoid of hair and glands.

Clinically such a condition may be, and probably as a rule is, quite free from symptoms, and considering that the cord and the nerves are not involved in the protrusion, this is not to be wondered at.

(2) *Meningomyelocele* is a condition which is much more complex, both pathologically and clinically. It is common as a specimen, and was illustrated in no less than 76 out of 125 museum specimens. In a typical specimen of this kind the bodies of the vertebrae are intact. The neural arches are not present for a varying distance, usually in the lumbar or lumbosacral region. The wall of the protrusion is formed of skin and dura mater, except at the summit, where there may be only a thin membranous covering. The dura mater is continuous with the theca

surrounding the spinal cord within the intact neural canal. The arachnoid is continued over the interior of the sac, the cavity of which thus corresponds to the sub-arachnoid space. The spinal cord runs across the upper part of the sac to its posterior wall, with which it becomes incorporated a little above the middle. Some nerves may arise from this part of the cord and pass horizontally forwards to their intervertebral foramina. Of such nerves both roots are present, separated at their origin by a process of pia mater—a continuation of the ligamentum denticulatum. The ganglia of the posterior roots are present, and there may be small additional ones (ganglia aberrantia). In such an arrangement the anterior and posterior nerve-roots of each nerve are separated horizontally by a considerable interval (in which lies the process of pia mater already alluded to), and the same is true of the anterior roots of each pair of nerves. Probably, therefore, the nervous tissue of the cord much thinned out extends commensurately with the area so included. The sac may contain lymph ensheathing the nerves passing through it, and there may be an external depression in the middle of the swelling, due probably to the dragging of the nerve-roots which pass through the middle of the sac to emerge at their intervertebral foramina.

Such a pathological condition necessarily implies a clinical state characterised by paralysis of varying extent and degree. The character of the paralysis will depend upon the part of the cord involved and the extent of its involvement.

(3) *Syringomyelocoele*.—In this condition, which is exceedingly rare, the interior of the sac is the dilated central canal. This internal lining is fibrous, and on dissecting this off it may be possible to display the nerves running round the cavity towards the foramina. The sac may communicate by a small opening with the substance of the cord, and it can easily be seen from such a condition of things that such a protrusion might easily be mistaken for a simple meningocele. But the wall of the sac, which in a

meningocoele consists, as we have seen, of skin and dura mater, in such a specimen would really consist of skin and much attenuated cord substance and nerves. Such a condition necessarily implies severe paralysis, and it is possible that in certain cases there is, in the portion of the cord inside the vertebral canal, a dilated central canal (syringomyelus) with perhaps the clinical condition which we know as syringomyelia (*q.v.*).

From what has just been stated it will be understood that the condition of spina bifida is of medical interest and importance chiefly in reference to the symptomatology. If the condition is a severe one, in which the cord itself and certain nerve-roots have been involved, the symptoms will depend upon the extent and degree of such involvement. The upper limbs do not, as a rule, suffer except in so far as the condition of spina bifida may, as sometimes happens, be associated with some degree of dilatation of the central canal (syringomyelia). Besides the actual dilatation of the central canal there may also exist some tissue of an embryonal type around the canal, some connective tissue in which the development of nervous elements has not taken place. This is probably the condition which, in cases of syringomyelia, has been termed *gliosis*, and the symptoms of interference with the elements of the spinal cord in the cervical and dorsal regions will depend upon the degree in which this exists. The condition of the cord where the spina bifida actually exists will determine the condition of the lower limbs. In some cases there may be wasting of both limbs, sensory and trophic disturbance, and a condition of flaccid paralysis with loss of control over the sphincters. Occasionally the affection may be only of one limb, or at least of one in a much more severe degree than of the other. The sphincters are very apt to suffer, and in some cases may be the only structures involved. In one case in which the condition was one of spina bifida occulta there was paralysis of both sphincters, a patulous anus and anaesthesia in the anus and over the scrotum, penis, and perineum. There was a post-anal dimple in

this case. In another, in which also there was a post-anal dimple, there was paralysis of the sphincter of the bladder with some anæsthesia about the vulva and perinæum. In this case, as in the previous one, there was no affection of the lower limbs except a condition of weakness giving rise to inability to walk far, and to a tendency to occasional falls.

The *treatment* of spina bifida is a matter of surgical therapeutics. It is obvious that in the second and third varieties of the condition alluded to, operation is scarcely likely to be satisfactory, for surgical procedures involve interference with structures which, if defective, may not be quite functionless, and such interference is not likely to do anything more than to render still more defective the function of such parts. In some cases of the first variety it is conceivable that operation might remove the deformity and do good, but most cases of spina bifida are scarcely capable of amelioration by any means at our disposal.

## SYRINGOMYELIA

SYRINGOMYELIA, by which is to be understood broadly that condition of the spinal cord in which cavities exist, was known as a pathological condition long before the symptoms clinically associated with it were recognised and their relation to it verified. It was even supposed by those who described it first that a central canal in the spinal cord was in itself a morbid condition; and it was not until 1859 that Stilling recognised that a canal normally existed in the spinal cord. As a result of this, the condition formerly described as syringomyelia was regarded as one in which the central canal of the spinal cord contained fluid and was possibly dilated, and hence a new name was invented, namely hydromyelia, and that of syringomyelia began to fall into disuse. Some years later, however, Hallopean, Charcot, and Joffroy demonstrated that there were conditions of the spinal cord with cavity formation in which the cavities have no relation—or, at all events, no necessary relation—to the central canal. We may say at once that the condition now recognised as syringomyelia is one in which there are at least two different kinds of cavities present—*first*, cavities in connection with the central canal, either originally related to it, or which, by a process of disease, have been led to communicate with it. These are characterised by the presence, in some part of the cavity, of the peculiar epithelium which normally lines the canal. These cavities may, as has been stated, result from a process of disease leading to absorption of tissue and consequent communication with the central canal, but, on the other hand, such a cavity may really be nothing more or less than the very much dilated central

canal, having, it may be, around it some tissue of an embryonic character forming what has been described as a wall to the cavity. Such cavities are found in connection with the condition of spina bifida. They are also found, it is said, in connection with spinal tumours existing above the position at which the tumour is exerting pressure. Besides this class of cases, in which the central canal is, as has been stated, an essential part of the cavity which is present, a *second* class of cases of great importance exists, also designated by the name of syringomyelia, that, viz., in which a new growth is present, it may be throughout the whole length of the spinal cord, but a new growth in which are present cavities of varying extent, and it may be also variable in number, sometimes in different numbers and degrees at different levels. These cavities in this type of syringomyelia would seem to be the result of some curious tendency in tumours of the spinal cord (a tendency also present in tumours of the brain and the cerebellum) to the formation of cavities, possibly related to the fact that the structures in which these tumours grow are enclosed in resistant bony chambers, and therefore subject to very considerable degrees of pressure. It is also possible that one kind of cavity in the spinal cord, although dating from early life, is not strictly congenital, but is the result of hæmorrhage into, or laceration of, the cord arising during a long and difficult labour. Dr. Herbert Spencer has shown that in a considerable proportion of cases of still-birth, cases in which a long or an otherwise abnormal labour has taken place, hæmorrhage occurs into the spinal cord, and it is conceivable that in cases presenting similar difficulties at birth, but which survive, a similar condition may give rise to a cavity in the spinal cord.

*Symptoms.*—From what has already been said it will be understood that the symptoms in cases of syringomyelia, while presenting on the whole very broad features of resemblance, will vary in individual cases, and especially that the course and progress of the disease will vary according as the case is to be classified pathologically in the first or in



the second group. The following three classes of symptoms, however, are to be found in varying degree in all of them: *first*, evidence of involvement of the structures which subserve the muscles, leading to muscular atrophy; *secondly*, evidence of involvement of sensory structures, causing anaesthesia, analgesia, thermal anaesthesia, or other sensory anomaly; *thirdly*, evidence of interference with the trophic functions of the cord, leading to vaso-motor disturbances, variations in secretion, ulceration and the formation of abscesses, and interference with joint nutrition. While it is true that syringomyelia as regards its symptoms, and also as regards the morbid anatomy, is a condition mainly related to the spinal cord and its functions, yet it must be noted that in some cases the process, whether of a new growth or of dilatation of the canal and its consequences, extends up so as to affect the structures at the base of the brain, thus giving rise to involvement of the nuclei related to phonation, articulation, and deglutition, and also of the nuclei related to ocular movements, and of nuclei of other cranial nerves. Indeed, it may be said that evidence of interference with structures subserving ocular movements, namely, nystagmus, is one of the most constant symptoms or signs in syringomyelia.

*First*, symptoms of interference with the muscular apparatus. There can be very little doubt that many cases which were described before 1886 as cases of progressive muscular atrophy were really instances of syringomyelia. The atrophy in cases of syringomyelia most commonly affects the small muscles of the hand, and the historical claw hand is very frequently present in that disease, just as it is in progressive muscular atrophy. Besides the wasting of the small muscles of the hand, there may also be wasting about the shoulders. And there is frequently combined with this a marked degree of lateral curvature of the spine, probably due to some interference with the innervation of the muscles supporting the vertebral column. Besides the muscular atrophy in those situations we have also occasionally wasting of the tongue, and even paralysis of one side of the face,

the result of the process spreading up to the highest structures of the cranial nuclei, as already indicated. It is somewhat unusual, but by no means unknown, for the muscles of the lower limbs to become affected in this way, yet the lower limbs are frequently the seat of symptoms pointing to an affection of the pyramidal tracts subserving those limbs. This is particularly true of cases of syringomyelia in which the central canal is involved; and when one recognises that the morbid process is usually most marked in the cervical region, it can readily be understood that, by means of pres-



FIG. 45.—Case of syringomyelia with facial paralysis, sixth-nerve paralysis on the right side, and hemiatrophy of the tongue. There was much muscular wasting at the shoulders and anaesthesia (see Figs. 46, 47, and 48).

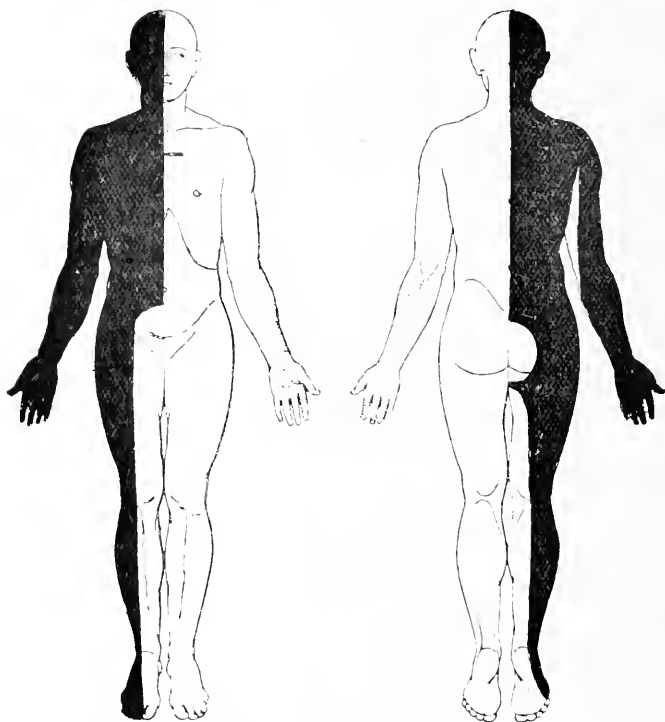
sure on and interference with the function of the cells in the anterior horns of the spinal cord consequent on such dilatation, such wasting as we have described may arise. It will also be understood how in such cases the pyramidal tracts related to the lower limbs come to be pressed upon, and how such pressure may give rise to signs indicating sclerosis of those tracts—namely, exaggeration of the kneejerks, the presence of ankle clonus, and an alteration of the plantar reflex as well.

*Secondly*, as regards sensory symptoms which are present, it may be said at once that there is no degree of constancy. Sometimes the limb which is most affected with the wasting

suffers comparatively little from sensory disturbances, whereas the opposite limb may have the sensory phenomena which are usually associated with syringomyelia. The characteristic phenomenon is not so much impairment of sensibility as that curious dissociation of sensation which may be said to be almost characteristic of this disease. For a limb in which this peculiar condition is present may have perfectly normal sensation as regards light touches, yet it may be that in such a limb the patient is unable to perceive the prick of a pin, except as a light touch, and that heat and cold are quite indistinguishable. Such a state of matters leads to occasional disasters. Thus a patient with syringomyelia who took hold of a kettle by a handle which was very hot perceived no discomfort or pain or excessive heat in his hand, yet the heat was sufficient to burn his hand severely. In another case, a patient in the habit of smoking cigarettes frequently had his fingers burnt without experiencing any sensation, and this led to the production of small but troublesome and persistent ulcers. Not at all uncommonly such a condition of dissociated anaesthesia exists over the face, sometimes at the back of the head also, and occasionally as a more or less complete zone around the chest. It is rare that such anaesthesia extends as far down as the pubes, and it very rarely affects the lower limbs. Occasionally there is no disturbance of sensation in cases apparently of syringomyelia, and this is probably to be explained by the fact that the sensory tracts are not encroached upon by the cavity.

*Thirdly*, trophic disturbances. The trophic and vasomotor disturbances are sometimes very striking. Excessive redness of a limb—not necessarily associated with coldness of it—is one of the commonest signs. The limb most affected usually is that which suffers most in its muscular structures. Not uncommonly this may be associated with excessive sweating over this limb, and it is not at all rare, especially in cases of very extensive new growth in the spinal cord associated with the formation of cavities, to find profuse sweating and redness all over the body, with

perhaps the escape of some particular area, such as one side of the face, or one hand, or one side of the chest. Besides these vaso-motor and secretory disturbances, joint conditions, apparently depending upon interference with their nutrition, may be present; and these are of the nature described by Charcot as occasionally present in tabes.

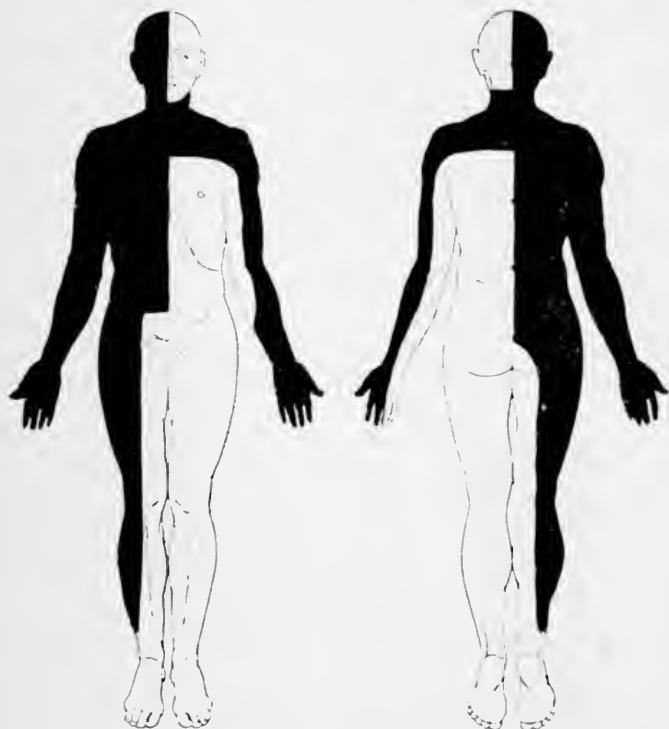


FIGS. 46 and 47.—Distribution of anaesthesia in a case of syringomyelia.

They are rare, but one occasionally sees a disorganised condition of a joint in a case of syringomyelia. Sometimes the elbow is affected, sometimes the shoulder, not so frequently the joints of the lower limbs, and it is possible, of course, that the condition leading to the marked spinal curvature, which is so frequently present may depend in some degree upon interference with the nutrition of the vertebral joints.

*Etiology.*—There are three different views as to the

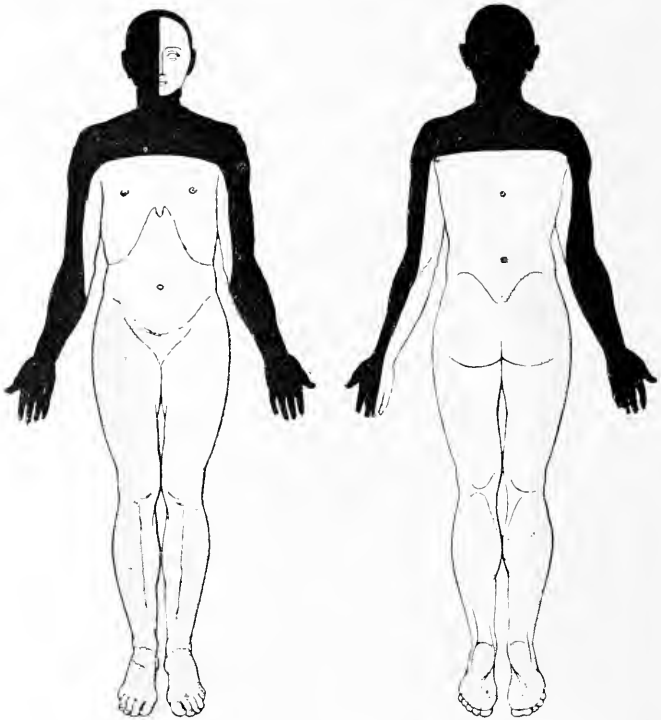
etiology of syringomyelia which have to be considered. First, that view according to which a congenital abnormality is supposed to be the determining cause of a central cavity—that is, that a condition of syringomyelus starts as one of hydromyelus. Secondly, the view that a cavity or cavities in the cord are the result of a central growth. Thirdly,



FIGS. 48 and 49.—Distribution of analgesia in the same case.

that which supposes that there is a chronic inflammatory process around the canal, or that there is inflammation apart from the canal, and that holes are formed in the cord as the result of softening. In the cases in which we find a cavity lined either partially or completely with the epithelium which is characteristic of the central canal, there seems to be no reason for doubting that the cavity is either an extension of the central canal or that some newly-

formed cavity has been brought into communication with that canal. In such cases the tissue around the cavity is somewhat anomalous in structure, partaking more of the character of the embryonic tissue from which the cord is developed. In some instances, also, there is an appearance of sclerosis, or at all events an absence of the



FIGS. 50 and 51.—Distribution of thermoanesthesia in the same case.

ordinary nervous elements around the posterior horns and in the posterior columns. This is occasionally very marked at the central end of the postero-internal columns; and if the explanation which has been offered to account for the tissue around the canal be the correct one, namely, that it depends ultimately on some abnormality of development, it is significant that this condition of excess of connective-tissue elements at the expense of nervous

should be present in the posterior columns also, the last large division of the cord to be supplied with nervous elements, and should actually be most marked at the point at which these columns are last furnished with nervous tissue proper, namely, the end next the central cavity. These facts seem to point strongly in the direction of the view that in such conditions, at all events, as those in which

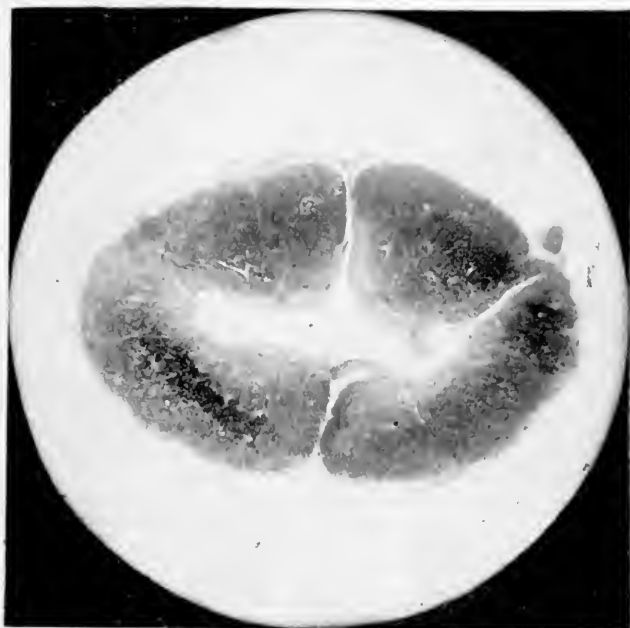


FIG. 52.—Syringomyelia: the cavity had in places a lining of the epithelium characteristic of the central canal.

the central canal, with part of its epithelial lining, is either in whole or in part the cavity in syringomyelia, that condition depends upon some congenital defect. Of course it is quite possible that while this congenital defect may have existed all through the patient's life without giving rise to any symptoms until, at the time the symptoms first arose, some process of new growth, slow it might be, became engrafted, so to speak, upon the congenital abnormality, and the

tissue, by growing instead of developing, gave rise to the symptoms for which the patient had to seek advice.

But while such an explanation may be offered for cases of this nature, an entirely different etiology must be looked for in another class of cases—those, namely, in which there is a condition of new growth extending throughout the



FIG. 53.—Syringomyelia: cavity involving central canal and reaching into posterior root. Sclerosis in posterior columns, cervical region. This condition gave rise to no symptoms, and was discovered accidentally. The patient died of intra-cranial tumour. From preparations and photograph by Dr. James Collier.



FIG. 54.—The same cord as in last fig., but in dorsal region. Here are two distinct cavities.

whole of the spinal cord, leading to what is sometimes an enormous increase in its size, but associated with the presence of cavities in it. This we must regard as almost certainly of the nature of a sarcomatous growth. The secret of the formation of cavities we do not yet know. It is possible that, as has been already suggested, the formation of cavities in a growth in the spinal cord, as in a similar growth in the brain, may be related to the conditions of excessive pressure under which such tumours grow. But the



origin of such growths is just as obscure in the nervous system as their origin is elsewhere.

*Morbid anatomy.*—There is no doubt that cavities of considerable size may exist in the spinal cord without giving rise to any symptom or recognisable signs. In some cases the first evidence of the existence of a cavity has been the occurrence of some hemorrhage into it, setting up a condition not easily distinguished from an ordinary myelitis. This is probably the result either of a weakening of the tissue usually supporting a vessel, or of some condition interfering with the nutrition of the wall of a vessel situated in the periphery of the cavity. Such cavities are probably congenital in origin, and they may have, therefore, some embry-



FIG. 55.—The same cord in lumbar region.

onic tissue in their walls, with possibly thin walled or ill-developed blood-vessels, one of which gives way.

These cavities may have a distinct cellular wall, and they are not infrequently merely a dilation of the ordinary central canal. When this is considerable in size it may so encroach on neighbouring structures, both grey and white, as to set up signs and symptoms which we recognise as those of syringomyelia. Another kind of cavity—it might even be considered another variety of that just described—is connected with the central canal, and has a distinct lining, usually in part only, of the characteristic epithelium lining the central canal. This cavity may branch in different directions; especially may it extend into one or other, or even into both, posterior horns. It also usually encroaches

on the grey matter of the anterior horns, causing distortion of these, destroying some of the cells, injuring others, and

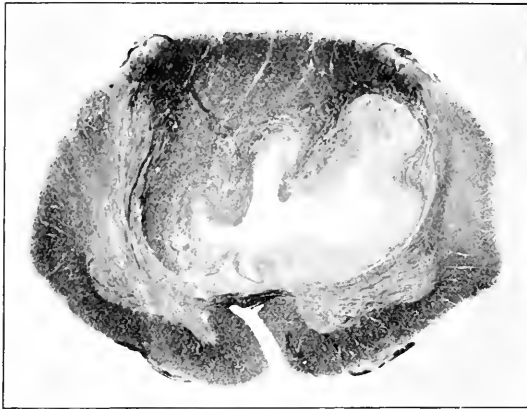


FIG. 56.—Syringomyelia, cervical region, with branching cavity causing much distortion of the grey matter and sclerosis of the lateral column. From preparations of Dr. Collier.

so giving rise to the motor and sensory symptoms associated in most cases of the disease. Besides the epithelial lining at some part of this canal, the remains, as already mentioned,

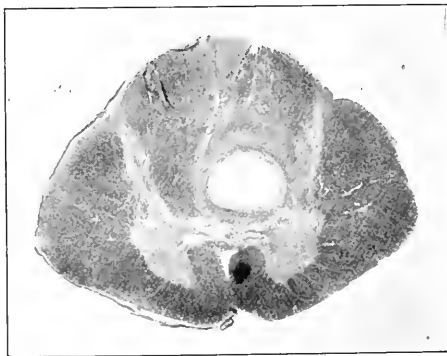


FIG. 57.—Syringomyelia with defined cavity. Same cord as last figure.

of the characteristic epithelium which lines the central canal, the cavity has a cellular wall, and has often also a wavy

fibrous envelope, possibly merely a development of the cellular elements. The tissue is not well-developed neuroglia, but seems to be an embryonic form of this tissue which has never actually developed into neuroglia proper, but rather seems to tend to overgrowth and breaking down. In such a condition we have not only the posterior horns and anterior horns affected and their functions interfered with, we also have a sclerosis of the pyramidal tracts, possibly the result of pressure, and some sclerotic condition also in the posterior columns, apparently due to defective develop-



FIG. 58.—Syringomyelia with large one-sided cavity with cellular wall. Same cord as last two figures.

ment in those. Such a condition as that described has been called by some a condition of gliosis, to distinguish it from that which is about to be described, known similarly as a condition of gliomatosis.

This last form of syringomyelia is definitely a form of new growth, gliomatous or sarcomatous, infiltrating the cord throughout its entire length, and interfering greatly with all its functions. The formation of cavities in it is probably the result of a breaking down of tissue, and may be determined by the severe pressure to which it is subjected by the resisting bony wall of the spinal cavity. The cavities are curious in their size and shape and dis-

tribution. They spread lengthwise in the cord, really forming tubes, and one may branch into two or even more. This gives rise to considerable variation in the appearance of transverse sections at different levels; thus a section at one level may show the existence of a single cavity, a section a little lower may show two cavities almost side by side, while a section still lower may show a single cavity again, one of the branches not having penetrated so low down. These cavities have no epithelial lining, but a wall is not infrequently formed for them by an agglomeration of the cellular elements along not only the vertical extent of the canal but also its lateral branches. The cavities and the cellular growth invade any part of the cord and interfere greatly with its functions, and the correlated clinical course of the disease is much more rapid than in the other form of syringomyelia.

The naked-eye appearance of the spinal cord in a case of syringomyelia is very striking. In the cases in which the central cavity is involved, and if this cavity is of considerable size, the cord as it lies in the spinal canal looks larger than usual, and a feeling of fluctuation is imparted to the hand by it. During its removal much fluid will probably escape, and the cord collapses into a flat ribbon-like structure. This flatness and limpness is very striking after it has been removed, and on section either at the time or after hardening the large slit or irregular elliptical cavity, surrounded by apparently thin walls, explains the flatness and limpness which are present. In a case of new growth, on the other hand, the cord is enormously enlarged and seems to fill up the whole spinal canal. This is especially the case in the cervical region, and after its removal, in spite of the presence of cavities, it retains as a rule its rounded contour and solid consistence. The explanation of the difference in the two classes is that in the former the cavity is much larger in proportion to the cord substance than in the normal cord, and in the latter case, on the other hand, the cavities form but a small part of the very much enlarged and infiltrated cord. It is usually the case that when the

central canal is involved, and as has been explained a large cavity is present, this will extend upwards, and will be associated with enlargement, through dilatation, of the iter and even of the lateral ventricles. An infiltrating growth also may be found to extend into the medulla.

*Diagnosis.*—The diagnosis of the condition of syringomyelia when one has become familiar with the ordinary clinical appearance and the usual signs does not present any very great difficulties. The presence of muscular atrophy, of the curious and almost characteristic peculiarities of sensory impairment already alluded to, of the vaso-motor phenomena, and of nystagmus and lateral curvature, makes the diagnosis certain. In many cases, however, one or more of these signs or symptoms may be absent, and while in any case in which such an aggregation of signs co-exists the diagnosis is easy, in others in which the clinical picture is incomplete in minor details, doubts as to the condition will naturally arise, and it is well to consider a few of the conditions which more or less closely resemble that with which we are at present most concerned. The ordinary form of progressive muscular atrophy with the wasting of the small muscles of the hand is probably the condition in reference to which the question of syringomyelia most frequently arises. There can be little doubt that many of the earlier cases described as of the so-called Aran-Duchenne type of progressive muscular atrophy were really cases of syringomyelia. At least, it is very striking nowadays to notice how rarely one sees such cases of simple muscular atrophy without signs of lateral sclerosis. In these cases the absence of sensory symptoms will, however, usually distinguish them from cases of syringomyelia. But occasionally a case may be met with in which there is the characteristic claw hand, nystagmus, lateral curvature, and extensor plantar response, and yet no disturbance of sensation be detectable. The diagnosis of this condition from amyotrophic lateral sclerosis may also present difficulties. In the latter condition, however, we have merely the affection of the upper

and lower motor neurons without any sensory disturbance or modification, and without any nystagmus or spinal deformity. Cervical pachymeningitis is a condition which closely simulates syringomyelia. Indeed, it may occasionally be impossible to make a definite distinction. A case presenting wasting of both hands and upper limbs with distinct sensory modification might be referred to either class. If, however, the dissociation of sensation is distinct, and if, besides, any such conditions as lateral curvature and nystagmus be present, syringomyelia will be probable. If, on the other hand, neither of these signs be present, then cervical pachymeningitis is the more likely. Of course, the history will also be of significance, for in the latter the onset is, as a rule at least, much more acute. The question of the relation of leprosy to syringomyelia is one which has been much discussed. The discussion arose first with reference to cases described by Morvan of an interesting morbid condition met with in Brittany, exhibiting chiefly muscular atrophy, vaso-motor phenomena with marked trophic conditions, especially the occurrence of painless whitlows, and of actual mutilations as a result of these. It is now generally agreed that these are cases of syringomyelia—in other words, that Morvan's disease is merely a variety of syringomyelia. Yet the resemblance to leprosy may be very marked and the distinction may not be easy. In the latter disease, however, the areas of anaesthesia are not so extensive, being the result of peripheral nerve disturbance; the mutilations are, as a rule, more severe, the disturbances are more symmetrical, and there is no lateral curvature. Further, nodular swellings on the nerves, especially the ulnar, can frequently be felt.

No difficulty is usually experienced in distinguishing syringomyelia from peripheral neuritis. In the latter, of course, there is muscular wasting, and there is frequently nystagmus, but the pain in the nerves, the diminution or loss of the reflexes, and the history of toxæmia from alcohol, arsenic, etc., will usually be obtained. The question of diagnosis between syringomyelia and hysteria should not arise

except through ignorance, or from the gross neglect of signs and symptoms incompatible with mere functional derangements.

*Prognosis.*—The prognosis in undoubted cases of syringomyelia is necessarily bad, so far at least as recovery is concerned. But it is remarkable how in certain cases the disease seems to remain quite stationary, and even to undergo some degree of amelioration. The case in Fig. 45, with marked and undoubted signs of syringomyelia, has altered little, if at all, in the last ten years. On the other hand, in many cases the weakness, wasting, and other symptoms rapidly increase and lead to a fatal issue in a few years—in one case actually in eighteen months. In that case the condition was one of infiltrating gliomatosis, and in such cases the disease runs a much more rapid course. In the other class of cases, however, in which there is a central cavity, probably congenital, with enveloping sclerotic tissue, possibly undeveloped or embryonic neuroglia, the course is a much longer one, and the clinical condition may persist unchanged during a long course of years. While this is no doubt true in many cases, it yet seems as if in others there existed a peculiar liability to pulmonary troubles. It may be that the vaso-motor mechanism of the lungs is liable to be disordered—a supposition quite compatible with the vaso-motor disturbances which are visibly present, and it is undoubtedly true that some even of these chronic, slowly progressive cases succumb to an insidious form of bronchitis or broncho-pneumonia. In the somewhat rare cases in which the vocal cords are affected, and in which the other structures subserved by the medulla are involved, the danger of accidental complication is much increased. One also has to remember that in these cases the disease is situated in close proximity to structures which are essential to life, and that in these uncertainty in regard to prognosis and a guarded forecast of the future are inevitable.

*Treatment.*—No form of actual medicinal treatment is conceivable which can check the progress of such a disease

as this, a disease which is in the nature either of a degenerative tendency imposed on certain structures and actually present at birth, or of a process of infiltrating new growth, as vague and indefinite in its origin as other conditions of neoplasm. But at least one drug has the power of modifying the nutrition of the structures chiefly morbid, viz. the anterior horn-cells of the cord, and the administration of this drug, strychnia, has in many cases seemed to be followed by improvement. If it is given, it is best given hypodermically, and the treatment should be commenced with a dose of  $\frac{1}{100}$  of a grain twice a day, and the quantity increased up to  $\frac{1}{30}$  or even  $\frac{1}{20}$ . It may be, of course, that the effect of the drug is to get out of the cells all of which they are capable. Yet in some cases the effect has been so distinct and so prolonged as almost to exclude such an explanation. So it is to be assumed that the drug has an active influence in favouring and promoting the nutrition of the affected cells. It should be given a trial in every case. The general environment of the patient is, of course, of the utmost importance. Physical and mental strain are to be carefully avoided, for there is reason for supposing that, especially in diseased conditions of the anterior horn-cells, such strain is apt to affect these structures. Avoidance of cold and chills is also essential, for, as already stated, there seems to exist, even in the most slowly progressive cases, a tendency to pulmonary complications. Except for such measures, the disease must be allowed to follow its own course of rapid deterioration, or slower change, but it must not be forgotten that in some cases the tendency to advance seems to be almost, if not quite, non-existent. Trophic and other disturbances must, of course, be treated as they arise.



## DIPHThERIC PARALYSIS

No one now doubts the occurrence of paralysis after diphtheria in certain cases. Opinions vary much as to its frequency, and it is not unlikely that it varies in different epidemics just as the death-rate also probably does. The nature of the sequela, if we may so call it, also necessarily introduces a difficulty, for it occurs at such varying intervals after the acute disease, that it is seldom, if ever, possible to retain a large number of cases under observation long enough to ascertain accurately the frequency with which it occurs. Some observers have been strongly of opinion that paralysis as a sequel of diphtheria is much more common in young subjects; others have expressed a firm conviction that it is more common in adults. It is probably equally common or uncommon in the two classes, and Sannes' figure of 1 in 9, arrived at from a consideration of 1382 cases, probably represents the ordinary frequency of its occurrence. It is not unlikely that the occurrence is a little more frequent, if cases in which there is evidence of slight neuritis, as shown, *e.g.*, by absence of knee-jerk, are to be included among cases of post-diphtheritic paralysis. One interesting fact has been pointed out by Goodall, viz. that 82 per cent. of paralysed cases had albuminuria, and 30 per cent. of non-paralysed cases. This fact may indicate either that a large dose of toxin is present, producing the kidney affection and the paralysis, or that, the kidney affection being present, the toxin is not so readily excreted, and so remains to affect the nervous structures. Some observers have been of opinion that mild cases of diphtheria are more frequently followed by paralysis; others have held quite the contrary opinion. There is probably not much in either observation, for it is

undoubtedly true that many very mild cases are followed by severe and widespread paralysis, and it is equally true that many severe cases of paralysis succeed to extremely severe throat affection.

*Symptoms.*—The time at which symptoms of paralysis occur after an attack of diphtheria varies very much. They may appear before the local affection has disappeared: they may be delayed as long as ten weeks after the faucial affection. The commonest time of onset is from three to four weeks after the throat affection. There may be a short period of lassitude and general weakness. The patient has apparently convalesced satisfactorily for a certain time, when some weakness is manifested. This is usually succeeded by a change in the voice, which assumes a nasal character. This is due to paralysis of the palate, and is almost invariable. Palate affection was present in 102 out of 125 of Goodall's cases, and in 83 of these was the earliest symptom. Occasionally the palate escapes altogether, and it would appear that this is most likely to occur in cases in which the faucial affection has been slight. There is in the ordinary case, at the time the palate has become affected, some feeling of tingling or numbness in the extremities and considerable weakness, especially of the lower limbs. Usually the knee-jerk is now found to be absent. Occasionally it is present and unusually active, but disappears later. The paralysis may be restricted to the throat, the palate being immobile, and there may be regurgitation of fluids through the nose. A characteristic toneless cough is also sometimes present—the result of paralysis of the adductors of the vocal cords. Such a condition is usually associated with defective closure of the glottis, and this will facilitate the accidental entrance of food material into the larynx, and may result in severe and troublesome coughing. As a rule, however, symptoms of paralysis elsewhere are superadded, and the commonest is that already alluded to, viz. loss of the knee-jerks and associated weakness of the lower limbs. No other symptom may be present, although it is not uncommon for the patient at this time

to complain of inability to read—the result of paralysis of the ciliary muscle. Other ocular muscles may also be affected; of these probably the most common is one or both of the external recti. The muscles supplied by the third nerve may also be affected, inducing a condition of very marked and troublesome diplopia. These are doubtless the most common symptoms in diphtheritic paralysis, viz., affection of the palate and vocal cords, affection of the lower limbs, and ocular paralysis. The upper limbs may also be involved in the paralysis—not usually so markedly as the lower. Where all these symptoms are present, and even if they be not all present, the condition of the heart has to be carefully watched. Weakness and irregularity are very apt to occur, and sudden death from heart-failure is not uncommon. But even short of this, alarming irregularity and weakness may occur, necessitating the utmost care. The intercostals also may suffer, and the diaphragm as well. Should any lung complications now occur the patient's condition, on account of the weakness of the respiratory muscles and of the heart, is most precarious. But even in very serious cases, in which all the complications mentioned have been present, recovery has occurred, and the almost invariable rule is that when recovery does take place it is complete.

The account which has just been given of the various forms which paralysis after diphtheria may assume practically gives its symptomatology. The prominent symptoms, however, may be re-enumerated. Lassitude and *malaise* in a patient, even apart from diphtheria, should always excite suspicion, especially if these are combined with albuminuria. The absence of knee-jerks is most significant, but their presence, and even their unusual activity, does not negative the diagnosis, as they may disappear later. The nasal voice, toneless cough, and still more, regurgitation of fluids through the nose are almost pathognomonic. Difficulty with near vision, indicating paralysis of the ciliary muscle, is very common, and strabismus, a result of paralysis of the ocular muscles, is not rare. Weakness

of the upper limbs and of the respiratory muscles, including the diaphragm, only occurs in severe cases.

Undoubtedly the symptoms of gravest significance are those to be ascribed to interference with the innervation of the heart, and it is very important to recognise this, as sudden failure of the heart's action is a common cause of death. Any strain on this organ, such as is implied in sudden change of position, especially from the recumbent to the sitting posture, must therefore be most carefully avoided.

Among the symptoms of diphtheritic paralysis of great importance must be mentioned what have been called "bulbar crises." It is in children the subjects of diphtheritic paralysis presenting such symptoms as those enumerated above that bulbar crises are apt to arise. Dr. Leonard Guthrie, in a now classical paper on this subject, enumerates these symptoms as follows: (1) Marked listlessness and apathy; (2) a weak, hoarse, and nasal voice; (3) irregular and sighing inspiration; (4) a loose, weak, and almost noiseless cough with accumulation of mucus in the air passages; (5) a rapid pulse. The crises are sudden acute exacerbations of the symptoms just enumerated. Their exciting cause may be a fit of passion or other strong emotion, sometimes physical exertion, or they may occur without any obvious cause. During a crisis there is complete and sudden paralysis of deglutition, complete aphonia, and the most alarming dyspnoea. Instead of lethargy there is extreme restlessness. The head is thrown back, the *alae nasi* work, the month is open, the pupils are dilated, and the face is bluish, or pallid, and sweating. Inspiration becomes more sudden or gasping, expiration more feeble and irregular, and sighing more common. There is often paralysis of the diaphragm. Saliva and mucus accumulate in the fauces, and trickle through the nose and half-open month, and moist râles are audible all over the chest. The temperature rises suddenly to  $102^{\circ}$  or  $103^{\circ}$ , the pulse-rate increases to 140 or 150, or even higher, yet there is no loss of consciousness, and the child, unable to speak, points significantly to its larynx or epigastrium when asked if it is in pain. Violent

vomiting may now ensue, and, although Dr. Gee has recorded nine cases of violent vomiting after diphtheria, of which no less than eight were fatal, Dr. Guthrie regards the act of vomiting under such conditions as those described as beneficial, although, at the same time, a sign that grave danger exists. The dyspnoea is often relieved by the vomiting, and, although the patient lies exhausted and apparently at the point of death, death rarely occurs in a first attack, and the crisis is at an end for the time. Within a few minutes, or it may be a few hours, similar attacks recur, and during or after these death may occur from exhaustion, syncope, cardiac thrombosis, or even from actual suffocation, the result of accumulation of mucus in the air-passages.

The duration of the crises varies from a few minutes to a few hours. They may be expected at any time within six weeks of the appearance of paralytic symptoms, and the critical period during which they may recur is very variable. Even if forty-eight hours have passed without a renewal of the crises, a further attack may occur and prove fatal. Nor does the completeness of the remission afford any guidance to prognosis.

Vomiting in the course of diphtheritic paralysis, as already stated, is mentioned by Gee. It is also mentioned by Hector Mackenzie, who records 72 cases. He came to the conclusion that it is not uræmic, as it occurs without albuminuria, and that it is probably due to implication of the vagus, as it is associated with heart failure and other symptoms of vagus affection.

Any affection of sphincters is most uncommon, although transient incontinence of urine may occur, probably the result of muscular weakness and general asthenia, not of affection of lumbar centres.

*Etiology.*—There is now general agreement as to the close connection between the bacillus described by Löffler in 1884 and the vast majority of cases of diphtheria. It may be granted that there are some cases which clinically cannot be regarded as anything but diphtheria, in which the presence of the bacillus cannot be demonstrated. This

bacillus has its habitat in the false membrane, and is not found in the blood. Diphtheritic paralysis is the most important sequel of diphtheria, and it is to be ascribed to the action of certain toxic substances resulting from the action probably of the micro-organism. These toxic substances have a profound effect on certain nervous structures, and give rise by their action to paralysis varying in extent from slight palatal weakness to almost universal powerlessness. Albuminuria is another common sequel to diphtheria. Trousseau long ago noted an increase in the albuminuria coincident with the onset of the paralysis, and Goodall, as already stated, has recently noted that 82 per cent. of the paralytic cases suffered also from albuminuria, while only 30 per cent. of the non-paralytic cases had this complication. Such an observation suggests that, the kidney being probably one of the great excretory channels for the toxic substances to which paralysis is due, in the condition of that organ which gives rise to albuminuria the excretion may not be so completely or satisfactorily performed, and so the toxic substances giving rise to paralysis are retained in the blood.

What the nature of this toxic substance is is still open to doubt. Roux and Yersin demonstrated that the inoculation of poisonous products precipitated from broth cultures caused paresis—an observation confirmed by other experimenters. Sidney Martin thinks the bacilli set free a ferment which converts albumin into albumoses which act as poisons. Uschinsky and Büchner, on the other hand, regard the bacterial poisons as the direct products of the bacterial cells, and Kauthack has stated that we cannot yet decide whether the toxins are themselves the poison, or whether they are poison-producing ferments; nor can it be decided on what the prolonged slow action of the toxins under special conditions depends. Any explanation to be satisfactory must also account for these not very unusual cases of relapse in diphtheritic paralysis. It would seem as if in such cases the toxic substance had been stored up somewhere and suddenly released.

As regards the *pathological anatomy* of the nervous system, the most obvious changes are in the peripheral nerves, and are usually described as those of "peripheral neuritis." But the change in the fibres is really a parenchymatous one. The white substance of the medullated fibres is broken up, the primitive sheath remaining intact. The axis cylinders are frequently ruptured. Sensory as well as motor fibres suffer, and the sympathetic fibres may also be affected. The cells of the anterior cornua also suffer in a varying degree. Some observers consider these changes primary, regarding those of the nerve as secondary. It is more likely that the affection of the different structures is simultaneous and to a certain degree unequal and independent, for in some cases changes in the nerves have been described without changes in the cells, and conversely the cells have been described as much changed while the nerves have remained normal. The corresponding muscular fibres are of course affected, undergoing fatty degeneration to a varying extent. The cardiac muscle is found in most fatal cases to have undergone extensive fatty degeneration.

*Diagnosis.*—The diagnosis of diphtheritic paralysis presents occasional difficulty, especially in the cases in which the history of throat or other diphtheritic affection is indefinite or absent. It must be remembered that paralysis may follow other diphtheritic affections than the faucial, and the writer has seen a case in no way distinguishable from one of diphtheritic paralysis, which occurred in a child who had had no throat affection, but had for several weeks suffered from a large ulcerating sore on the arm, the result of a severe burn. A case has also been described by Bourges as the result of a streptococcal infection. No mistake is likely to be made in the case of children. In adults several conditions may occur which are not easy to distinguish. Any form of peripheral neuritis may present analogous symptoms. In diphtheritic paralysis, however, the throat affection is almost constant. Of course, the most common form of peripheral neuritis is alcoholic in origin, and the history in such cases,

although often difficult to obtain, is very significant. Tenderness in the nerve trunks and loss of memory are usually present, and the time that has been taken to develop the paralysis is usually much longer. Landry's paralysis has been shown by Farquhar Buzzard to be a form of toxic disease, and it is very similar in its effects to diphtheritic paralysis. Indeed, in some instances it is impossible from the clinical conditions to determine whether the patient is suffering from one or the other disease, and in such circumstances the history is all-important. The ordinary form of bulbar paralysis is not likely to be mistaken for diphtheritic paralysis. The morbid process is slower and more insidious, and is rarely complicated by affection of the ocular muscles. Acute anterior poliomyelitis of very extensive distribution may also affect the structures in the bulb, but this disease in such an extensive form is rare. Here again the history must be relied upon. The signs to be chiefly sought for as evidence of diphtheritic paralysis are the faucial affection and the affection of the ciliary muscle. If these occur, combined with weakness of limbs and of ocular movements, they are almost certainly the result of diphtheria. The resemblances of certain cases of myasthenia to diphtheritic paralysis are referred to in the article on that disease.

*Prognosis.*—It is always exceedingly difficult to foretell what is likely to be the result in any case of diphtheritic paralysis in a child. It is rarely that death occurs in the case of an adult, but the disease is very often fatal in the case of children. Even cases apparently slight not infrequently undergo sudden and rapid change for the worse, especially if widespread and profound paralysis appear, and death may occur from cardiac failure or from some pulmonary complication. Even without the appearance of more extensive or severe paralysis, a patient apparently slightly ill may die suddenly from cardiac failure. Such a result is very apt to occur from any sudden change of posture, and the possibility of such an accident emphasizes the necessity, to which reference will be made when the treatment is dis-



cussed, of the need for the most extreme care to keep the child in the recumbent posture. Or a patient apparently only slightly ill may gradually but steadily become weaker and die simply of asthenia. The danger of the so-called "crises," the result probably of affection of the vagus nerve or its nucleus, will have been sufficiently evident from the description of these conditions, and how fatal the complication is will be evident from a consideration of Dr. Guthrie's nine cases, of which no fewer than seven died. But while, on the one hand, the grave danger to life present in every case of diphtheritic paralysis must be recognised, it can, on the other hand, be stated that no case of diphtheritic paralysis, however grave it may appear, is necessarily hopeless. It is often surprising, and in the highest degree gratifying, to watch how a child paralysed in all four limbs, with even the intercostals paralysed and broncho-pneumonia present, will gradually fight its way back to convalescence and complete recovery. The gravest danger is undoubtedly from cardiac failure, and the risks of this are much increased if any pulmonary complications are present.

*Treatment.*—As soon as the affection is recognised, no matter how slight it may be, the patient should be put to bed. The recumbent posture is absolutely necessary, and it should be maintained until it is evident that all danger of the paralysis becoming more extensive or severe has passed away. Even sitting up in bed should not be allowed, and in the case of children it is well to ensure the recumbent posture by some light but effective yoke. It is in the slight cases only that such a thing is necessary. In the more severe cases the paralysis itself is so disabling that it ensures the most safe posture. It may, however, in such a case also be necessary when slight improvement has taken place and there is some temptation to try to sit up. The danger of sitting up or even of being lifted up, is a very real one, and the writer has known of a case in which an inexperienced nurse was horrified to see a child whom she simply raised fall back

dead. Excessive care, therefore, is necessary in the nursing and general hygiene of this disease. Of course frequent and regular feeding with wholesome, stimulating, and easily digestible food is indispensable. Alcohol is extremely useful, and of other drugs to be used strychnia is invaluable. In severe cases it is often necessary to administer it hypodermically, in doses which will vary with the age of the patient. The combination of atropin with the strychnia has also been recommended, especially when the "crises" are present. Even after the patient is no longer dangerously ill it is wiser to continue the strychnia and to combine it with other tonics, such as quinine and iron. In the severe cases in which pulmonary complications may have ensued, the inhalation of oxygen affords great comfort and not uncommonly distinct relief, and every case of this paralysis is to be regarded as one which has to be tided over a certain time until the poison has exhausted itself, and at the same time to be carefully guarded from the conditions likely to set up any complications.

## FAMILY PERIODIC PARALYSIS

In 1874 a case was published by Hartwig under the title of "Intermittent Spinal Paralysis," in which the prominent clinical phenomena were recurring attacks of almost complete paralysis in a patient who between the attacks was apparently quite well. The attacks of paralysis were accompanied with complete or almost complete loss of electrical excitability in the muscles, whereas between the attacks the muscles responded normally to each current. The details of the family history were not given, and it was not until 1882, when Schachnowitsch published an account of a patient whose father had been similarly affected, that it was recognised that the condition, besides being periodic, was also apt to occur as a family disease. In 1885 Westphal gave an exceedingly complete account of the clinical phenomena of the disease, noting the paralysis of muscles, the order of affection of muscles, and the temporary dilatation of the heart which sometimes occur during the attack. In the following year Cournot published the first case in which the family nature of the affliction was very striking. Since that time little has been added to our knowledge of the disease, but interesting and valuable examples have been published by Oppenheim and Goldflam in Germany, and Taylor, Mitchell, Putnam, and Crafts in America. In this country Dr. Singer in 1901 published in 'Brain' a valuable and carefully studied case of Dr. Tooth's, from the Queen Square Hospital. The phenomena were very exhaustively observed, and the interest and value of the case considerably enhanced by the careful examination of the excreta by Dr. Goodbody. Dr. Singer also furnished an excellent critical digest of the various pub-

lished memoranda on the disease. In the same year Dr. Parquhar Buzzard gave a very interesting account of three cases of the disease in two generations of a family, and suggested an explanation of its phenomena, to which reference will be made later.

*Clinical manifestations of the disease.*—These essentially consist in the onset of attacks of flaccid paralysis in an individual otherwise healthy. The paralysis is of the motor structures only. No paralysis of afferent structures has ever been described, although subjective phenomena of tingling and numbness have been frequently complained of. One of Buzzard's cases, on being tested with a Faradic current, volunteered the information that the battery produced the sensation which he frequently had at the commencement of an attack. The attack of paralysis is gradual in its onset. It affects the proximal muscles of the limbs before the terminal parts, and the trunk muscles also suffer. The respiratory muscles are involved. The diaphragm is never paralysed, although there is occasionally evidence that its action is impaired. The heart is affected in some degree, and marked increase of the cardiac area of dulness, indicating dilatation, has been noticed by different observers since Westphal first described it. The cranial nerves are not seriously affected, although ptosis has been occasionally observed, and also a difficulty in opening the mouth and in swallowing. Articulation also is occasionally modified, and a slight emotional condition has been described when an attack is impending. When an attack is at its height the patient lies, breathing with some difficulty and considerable effort, with the limbs in a condition of flaccid paralysis, unable to raise the head, but without any severe pain. The pulse rate is slowed and the pulse may be irregular. The deep reflexes are abolished, and no response can be obtained from the muscles with either a constant or interrupted current. The first sound of the heart is often impure, and a definite murmur of mitral regurgitation has been present. The paralysis lasts for a varying time, sometimes only five or six hours, sometimes

as long as 70 hours. Even if an attack passes off in a few hours it may be several days before the patient regains his accustomed facility in moving about.

The age at which the attacks commence varies greatly. Sometimes they may supervene as late as 24. In Buzzard's cases they were certainly present as early as 2, although Singer had stated 6 to be the earliest age at which an attack had been noted when his paper was published. The sexes seem to suffer with equal frequency. The relation of the attacks to exercise seems to be fairly definite. They have usually succeeded exercise, but after an interval of rest, and they most often occur during rest in bed. In some cases, however, there has been no obvious relation to exercise. In Singer's case the attacks occurred more frequently if the patient remained in bed. An ancestor of one of Mitchell's patients had sometimes to get off his horse and "have a paralysis." Besides the severe attacks, there are also slighter, apparently abortive attacks, which in some instances have been capable of being arrested by taking exercise. Thus one patient thought he could "walk off" an impending attack.

The intervals between the attacks vary in duration. They may at first amount to months and gradually diminish to hours. In some cases, on the other hand, there has been a definite lengthening of the intervals between the attacks as life advanced.

*Etiology.*—Heredity seems to play an undoubted part in the etiology of this disease. It is true, as in most cases of familial disease, that isolated cases occur in which no trace of a similar tendency can be verified in any other member of the family. Yet that in the majority of cases a family tendency is present can scarcely be doubted. Thus in the four families of the cases recorded by Consot, Goldflam, Taylor and Mitchell, there were in all 42 cases. Goldflam and Taylor were able to trace the disease through five generations. The families in which this disease occurs do not show a proclivity to any other nervous disorder.

No previous disease or illness seems to induce a tendency

to suffer from this form of paralysis. The suggestion that malaria has something to do with it was probably only suggested by the periodicity of the attacks. The relation of physical over-exertion, or even of physical exertion, to the attacks seems to be a close one. In most cases attacks succeeded such exertion, usually after an interval of rest. In Buzzard's cases attacks could be brought on by exercise. Yet in some cases the absence of any such relationship has been striking. Gastro-intestinal disturbances and indiscretions in food and drink have been blamed in some cases as the exciting causes of attacks, but apparently without sufficient cause. Disturbed emotional states, on the other hand, seem to have been the precursors of attacks in more than one instance.

*Pathology.*—No patient, so far as is known, has died of this disease, so that anatomical investigations have been restricted to examination of the blood and of portions of muscle excised during the paralysed condition. As regards the blood, although slight degrees of lymphocytosis have been described by different observers, these have not been sufficiently marked or sufficiently constant to have any significance.

The muscle has been examined by Goldflam, Crafts, and Singer. All these observers found marked vacuolation and fissuring. Singer is inclined to regard this as in part at least artificial, and due to the mode of preparation. Yet, as he says, this peculiar behaviour of the muscle under preparation may indicate some abnormality in it, rendering it more liable to shrinkage and alteration.

Even if it be granted, however, that these slight changes in the muscles exist, we are only driven back a step further and have to ask, What is the cause of them, and do they explain the curious phenomena of the disease? Ever since Westphal first hazarded the suggestion, a toxic origin for the disease has been assumed. Search has been made for some toxic substance which might by its action induce such a condition as we have described. Crafts claims to have obtained from the excreta a substance which pro-

duced temporary paralysis in guinea-pigs, but this observation so far has not been corroborated. Goldflam and Goodbody have found an increased toxicity of the urine during and just after attacks, but apparently this toxicity was simply an increase in the normal toxicity and not the effect of any special toxin. It has therefore been suggested that there is, on account of muscular work, some excessive production of normal substances which inhibit the action of certain motor structures—nervous or muscular. In the cases in which the attacks of paralysis occur during rest in bed, a storing up of these toxic substances, which produces the same effect, must be supposed to take place, on account of the absence of muscular effort and consequent elimination. The copious sweating which frequently occurs during an attack and the strikingly beneficial effect of active diuresis in Dr. Singer's case certainly favour such a view. In reference to the inquiry, On which structures would such a substance exert its effects? Buzzard points out that muscles exhausted as a result of work cease to respond to volitional stimuli long before they cease to respond to electrical stimuli. Waller and others have also pointed out that in such circumstances indirect excitability to electrical stimulation disappears, whilst direct excitability remains. In family periodic paralysis, however, this is not the case: the excitability of muscles to volitional, mechanical, galvanic or faradic stimulation disappears gradually and equally step by step. So that the phenomena do not resemble those due to affections of the lower neuron. Nor are they analogous to the effects produced by curara, for Donath and Lukaš have shown that the paralysed muscles in a curarised animal are still responsive to electrical stimulation. Buzzard therefore hazards the view that the muscle substance itself is the structure affected; he doubts whether the changes described in muscles are altogether artificial, as Singer suggests, and he regards the abnormal, possibly toxic condition of the lymph as the probable source of the changes in the muscle plasma leading to periodic paralysis. It must be acknowledged that in the

absence of any other satisfactory explanation of the phenomena of this disease, such an hypothesis deserves careful examination.

*Diagnosis.*—The diagnosis of such a disease has usually to be made in the interval between attacks, but if the history of the periodic attacks is clear, and especially if it is possible to see an attack, no difficulty can arise to any one who knows of the existence of the disease, even if he has never previously seen a case. In some respects it resembles and might suggest myasthenia: in others it is not unlike myotonia, but a mistake is scarcely possible. The occurrence of similar attacks in more than one member of the family will only confirm the diagnosis. The absence of such a history, however, will not by any means negative it.

*Prognosis.*—So far as we know at present no patient has died of this disease. Yet it must be remembered that the great severity of some of the attacks, and especially the respiratory and cardiac phenomena in these, indicate an element of danger which it is wise not to overlook. Although in some cases, up to a certain age, the attacks may show a tendency to increase in severity and frequency, the general tendency is probably towards a diminution both in the number and duration of attacks.

*Treatment.*—It may be said at once that the treatment of this disease is not satisfactory. Galvanism, massage, and various drugs have been tried, with no effect on the attacks. Dr. Singer, acting on the supposition that the exciting cause of an attack may be the failure to remove the toxic products of metabolism from the body, attempted treatment calculated to remove these products by diuresis. The attempt in his case was strikingly successful. He administered large doses of soda-water and imperial drink, in addition to the ordinary fluid taken during meals, and simultaneously small doses of acetate of potassium and digitalis. Previous to the treatment the patient had been having at least one attack every week. In the three weeks during which he was under this treatment he had no attacks. Similarly satisfactory results attended the resump-



tion of the treatment after a period during which it had been stopped and the attacks had recurred. Buzzard, however, carrying out treatment on the same lines, failed to obtain any benefit in his patients. It should be observed, however, that Buzzard's cases were distinctly family cases, and therefore presumably had the tendency to attacks more ingrained. Singer's case was an isolated one without any family history of similar attacks, and in cases of that class exhaustive treatment on his lines is certainly to be strongly urged. Of course, moderate exercise, simple food, and good hygienic conditions are most desirable, and, as Singer suggests, an occasional Turkish bath may be beneficial.

## TETANY

TETANY, or tetanilla, is a name that has been applied to a clinical condition which is characterised by contraction of certain muscles, especially the muscles of the extremities. These contractions have been, in the adult, associated with various causal or exciting conditions. Especially have they been present after removal of the thyroid gland, and in association with gastric dilatation, and occasionally with other abnormal abdominal conditions. They have also frequently been noted during lactation, or in the puerperal state. With those conditions as they occur in the adult, we are not particularly concerned, except in so far as they may indicate the cause of the condition in these patients, and incidentally throw light upon the nature and etiology of the condition as it occurs in children. The symptoms of tetany in children are present nearly always in association with rickets. Gastric dilatation is frequently present, possibly the result of fermentative changes which take place from using unsuitable food. Diarrhoea is also frequently associated with the condition, and laryngismus stridulus is not uncommon. A distinction has been unnecessarily drawn between the condition which we know as tetany, and the so-called carpo-pedal contractions which occur characteristically in rickets. But the distinction is really an artificial one, and there is little doubt that tetany and carpo-pedal contractions which occur in rickets are manifestations, in different degrees, of an identical condition.

*Symptoms.*—The symptom which specially characterises tetany is spasm, especially in the small muscles of the hands and feet. The resulting position of the hand is one

in which the phalanges are extended, but in which flexion has taken place at the metacarpo-phalangeal joint. The thumb is usually turned in to the palm of the hand, so that the hand is drawn up into the so-called "obstetrical position," the position which the accoucheur assumes in the exercise of the movements which are especially associated with his work. In the feet, the contraction which takes place is similarly of the small muscles leading to flexion



FIG. 59.—Case of tetany showing carpo-pedal contractions. From a photograph by Dr. Farquhar Buzzard.

of the toes into the sole. As regards the rest of the limbs, it may be generally stated that the position assumed is one of extension. And when the spasm affects the trunk muscles, the muscles on the ventral aspect are contracted. There is rarely spasm in the dorsal muscles. Retention of urine may occur from spasm of the sphincter of the bladder. Spasm may also occur in the face muscles, and vaso-motor phenomena, redness and coldness, and even œdema, of the extremities may show themselves. There is a curious condition of mechanical irritability of the

nerve trunks. This is especially seen in the facial nerve in many cases, and is known as "Chvostek's sign." This mechanical irritability may also show itself in other nerve-trunks, but it is not constant. There is a curious condition of electrical irritability also, this being characterised by the fact that the opening contractions, and especially the anodal opening contractions, are in excess of the closing contractions of a galvanic circuit. There is no change in the reflexes.

*Pathology.*—There seems to be very little doubt, looking at the occurrence of the condition in adults in association with removal of the thyroid gland, and with gastric or abdominal disturbance, that the phenomena of tetany are the results of some form of intoxication. It is probable that the part affected by such toxin as may be present is the lower neuron. Were the seat of the infection at a higher level, it would scarcely be likely that the electric phenomena to which reference has just been made would be present. It would seem, at all events in children, as if the rachitic condition which is present, interfered with the development of the higher centres and the control by them of the lower ones, leading to the unrestrained activity of the latter—that is, that we have in tetany a condition in the nervous system similar to that which we have conceived to be present in that system in connection with the convulsions of rickets, a condition of interference with development, so that the higher centres have not developed sufficiently to control the lower centres, and that these go on exercising their reflex and automatic actions in an excessive and uncontrolled degree. The morbid changes, slight in character, which have been found present in the cells of the anterior horns of the spinal cord, indicating interference with their structure and functions, bear out such a view as is here expressed of the pathology.

The *diagnosis* of the condition, when one is familiar with its appearance, is not by any means difficult. It is hardly possible to mistake a condition such as this for tetanus; for in tetanus we have the condition of trismus

well marked, we have opisthotonos, and a condition generally which is obviously much more serious and threatening than ever the condition is in tetany. As has already been stated, the distinction between tetany and carpo-pedal contractions seems to be a purely artificial one, and they are to be regarded as probably degrees of the same illness.

*Prognosis.*—The prognosis in tetany is good. It is rarely that death supervenes—at all events in the disease as it occurs in children. Occasionally, no doubt, the condition of rickets, and possibly of gastric dilatation and the intestinal disorder with which the tetany is associated, are sufficiently severe to cause great impairment of health, and perhaps actually to interfere with life. But such an event is rare, and in spite of the obscurity of the disease, and the severe condition of illness which may sometimes be present, the prognosis in tetany is by no means bad.

The *treatment* resolves itself into the treatment of the conditions with which we have indicated tetany to be associated. Rickets, of course, calls for good hygienic regulation, suitable food and the absence of anything likely to lead to fermentative changes in the stomach or intestinal canal. The gastric condition itself must be treated, as far as that can be done in a child, by means of food easily digested and not too abundant. It may be necessary occasionally to wash out the stomach. It may also be desirable to give intestinal antiseptics, such as salol, or grey powder, or small doses of calomel, and when such treatment as this has been carried out, the symptoms of tetany may be trusted to disappear entirely.

## MYASTHENIA

ALTHOUGH myasthenia is a disease which affects adults probably far more frequently than children, yet certain cases of the affection have been described in young adults, and even in quite young children, and it is desirable to set out briefly the main features of the condition. The full name, which is given to it by Jolly, is *myasthenia gravis pseudo-paralytica*, but this is usually shortened to *myasthenia*, or *myasthenia gravis*. It is characterised, broadly speaking, by great muscular weakness, affecting most strikingly the muscles which are supplied from the medulla oblongata, and are, therefore, closely related to the maintenance of the vital functions. So far, no definite changes in those nervous structures from which the affected muscles are supplied have been found, and it is interesting to note that so long ago as 1877 Dr. (now Sir Samuel) Wilks, published a case of bulbar paralysis without any anatomical changes—a case which, in all probability, is to be regarded as the earliest case of myasthenia gravis of which there is any record. Much interesting and important work on the subject has been recently contributed in this country by Dr. Buzzard, Dr. Edwin Bramwell, Dr. Harry Campbell, and others.

*Symptoms and clinical history.*—The first sign of weakness in this disease usually occurs in the levatores palpebrarum, giving rise to ptosis. Sometimes this ptosis is equal on the two sides; occasionally it is more marked on one side than on the other. It is frequently associated with weakness of the ocular muscles, and the effort to overcome the ptosis by means of the action of the frontales

manifests also some weakness in those muscles. In order, therefore, to get rid of the effect of the ptosis and enable him to see, the patient throws his head backwards, and, in doing so, is confronted by weakness of the neck muscles, so that he has to maintain his head in a position in which it is almost impossible for him to see on account of the drooping of the eyelids. Diplopia of varying character is often present, the variation in its character depending upon the weakness of different ocular muscles. And this weakness in the ocular muscles varies very much at different times, there being days on which a patient with this affection has no obvious signs of weakness, either of the external ocular muscles or of the levatores. Nystagmoid movements are occasionally present, these also being evidence of weak muscular action. Although the external ocular muscles are so frequently, and almost constantly, affected, it is extremely rare to have any pupil changes. Difficulty in masticating is also frequently present, the masseter muscles especially becoming easily tired, and the patient not uncommonly during eating supporting the lower jaw with one or other hand. This difficulty in masticating is often more marked in the later part of the day. The muscles about the mouth are also weak, so that there is a difficulty in whistling or in blowing out a candle. Dribbling also may occur. Swallowing is difficult, fluids are more easily taken than solids, although there may be occasional regurgitation, and the pharyngeal reflex is often absent. Palate weakness is also frequent, giving rise to a nasal voice and inability to blow out the cheeks. The laryngeal muscles themselves are rarely affected, but aching or stiffness in the tongue is frequently complained of, and the patient may be unable to protrude his tongue or to move it freely from side to side. One very striking feature relating to the weakness of those structures about the mouth or connected with articulation is the character of the speech. When the patient begins to speak articulation may be clear, and the voice good. As he proceeds, the weakness becomes marked, the tone becomes lower, a nasal

character is imparted to the articulation, and the patient finally becomes both breathless and voiceless.

The weakness in the muscles of the neck has already been referred to. The respiratory muscles are also affected, so that there is interference with the amplitude of movements during respiration. There is dyspnoea on the slightest exertion, and, as has already been said, when the patient speaks for a little time there is marked breathlessness, associated with the voicelessness already referred to. This dyspnoea constitutes an element of considerable danger, because weakness of the structures which are associated with breathing gives rise to actual crises of dyspnoea, in which the patient may die, and in several of the cases in which death has occurred it has supervened apparently as the result of an acute crisis of dyspnoea.

Besides the trunk muscles, those of the extremities are also affected. Thus, the arms may become very easily tired; a female patient, for example, finds an increasing difficulty in carrying out the movements which are necessary for doing her hair in the morning. Similarly, also, a house-painter found that after working for a little time at white-washing, with his arms at a higher level than his head, the arms drooped, and it was impossible to get them up to the same level. The hands, also, may become affected, and the handwriting may show the weakness of the hand muscles. In the lower extremities the quadriceps and the ilio-psoas group seem to suffer most. Walking far, as a rule, is impossible, and the patient usually becomes tired after walking a very short distance. Going up more than a few steps of stairs is quite beyond his power, and a sudden giving way of the legs, causing somewhat severe falls, is not unusual.

Such are the broad features of the illness. But there are various conditions which exert considerable influence upon the myasthenic weakness. Thus, emotional conditions undoubtedly intensify the weakness in a very material way. Cold in some cases acts similarly, and women affected with this disease are always worse at the menstrual periods. Persistence in a movement soon exhausts a



muscle, yet even in regard to this point it must be remembered that there are very great variations in muscular weakness, and that a movement which can be repeated only two or three times on one occasion can be repeated as often as twenty or thirty times on another. It is also noteworthy that the patient is frequently very much better in the early morning, and becomes gradually worse and weaker as the day proceeds, so that movements which can be carried out with ease and facility in the morning are quite impossible in the evening.

The electrical reactions in a case of myasthenia are of very great interest, and the reaction in muscles is characteristic. It is found that after repeated stimulation by a faradic current the response becomes more and more feeble, so that in time it disappears entirely, the response to the constant current remaining. Atrophy of muscles has sometimes been present, but is by no means the rule. The reflexes are usually very active; but in some cases, probably those in which the weakness has reached an extreme degree, it has been possible after a few taps to completely exhaust the knee-jerk, the muscle thus reacting to mechanical stimulation in the same way as it does to stimulation by faradic electricity.

*Etiology.*—Males and females are found to suffer with about equal frequency, and enough cases have now been published to permit such statistical evidence to be regarded as approximately accurate. The oldest case so far published is 55, the youngest 12; but the writer has a patient at present under his care apparently suffering from this condition who is only 8. It is not improbable that as our knowledge and observation of the condition extend, the limit of age may be increased in each direction. Occupation seems to have little connection with the disease, although the majority of the cases described have occurred in manual workers. In many instances the symptoms have followed some acute illness, such as influenza, scarlet fever, typhoid, severe diarrhoea; and pregnancy, menstruation, emotional strain, cold baths, and over-exertion have been invoked in

different cases as exciting causes. In some instances more than one member of a family has suffered, and occasionally the condition has been superinduced on some form of weakness already existing, such as lead palsy. More than this as to the possible etiology it is impossible to say at present.

*Morbid anatomy and pathology.*—As regards morbid anatomy, so far, the cases examined have shown no definite signs of change in the nervous structures which supply the affected muscles, nor have the muscles themselves shown any material alteration, except in the rare cases in which a slight degree of atrophy has been present. In a few cases enlargement of the thymus has been present, and thymus tumours have been found present in the spleen. Multiple tumours have also been found in certain cases—sometimes collections of lymphoid tissue in muscles themselves. But in other cases equally definite from the clinical standpoint no such change has been discovered. It is impossible to say what, if any, relation such a condition has to the onset of the disease, but it is possible that in such tumours or collections of lymphoid tissue some poison is generated which has a toxic effect on the muscles or their nerve-terminations. The pathology must, therefore, to a certain extent, be one of surmise. It would certainly seem as if some form of toxic substance were produced in the body itself, possibly as a result of muscular contraction, which poisons the motor nerve endings in the muscles themselves, or which perhaps actually poisons the muscle substance. In the meantime we must be content with clinical observation and examination of all organs as well as careful histological and possibly chemical examination of the blood. So far, however, these means of investigation have given us no definite clue to the real nature of the disease.

*Diagnosis.*—The diagnosis of this condition, when one has become familiar with the type of disease, is not very difficult. Undoubtedly the closest analogy which it offers is to those cases of nuclear palsy in which the cells of the nuclei related to the bulbar and the ocular muscles undergo gradual, and, in time, complete wasting. In many

cases it may be impossible to say to which class of cases a given one belongs. It can only be by careful and repeated examination, by noticing the variation in the conditions, and by recognising the associated weakness and easy exhaustion of the trunk muscles and of those of the extremities, that one can be convinced that the condition is one of myasthenia, and not of nuclear disease. In reference to diphtheritic paralysis, as will at once be recognised, the analogy is very close. Similar structures are affected, they are affected in a similar way, and often in myasthenia the onset of the trouble is almost as rapid as it is in diphtheritic paralysis. In the latter class of cases, however, there will usually be the history of sore throat, etc., to go upon; there will be the absence of the knee-jerk usually, as contrasted with its activity in myasthenia, and there will be the comparatively early clearing up of the condition under appropriate treatment. Polio-encephalitis of a chronic character, which is really the degenerative nuclear palsy to which we have referred already, may also simulate the condition; and indeed in some instances it will be impossible to make the distinction. An acute polio-encephalitis is scarcely likely to be mistaken for myasthenia, the onset being so much more sudden. Hysteria is a name which has frequently been given to cases of myasthenia, and the curious variation in the condition, especially in the early stages, when the patient is sometimes apparently quite well, and at other times, even in the same day, profoundly weak and ill, may seem to some extent to justify the mistake. But a careful examination of the weakness of muscles, especially the weakness of ocular muscles, which it is almost impossible for a patient to simulate, will usually convince one that the condition is not a functional one.

*Prognosis.*—The prognosis in this disease is always somewhat grave. It is, no doubt, true that certain patients remain in the same curiously variable condition during many years. A patient was recently seen with very marked ptosis, almost complete paralysis of all the ocular movements, some weakness of the trunk muscles, and also of

the extremities, in whom the condition had been present, with very little change, except such variations as we have alluded to as incidental to the disease, during so long a period as 14 years. Yet in other cases there is very little doubt that the disease is a much more serious one, and much more rapidly progressive, and may lead to sudden and somewhat unexpected death. A patient was recently seen whose first symptoms occurred so recently as six months ago. She had become extremely weak and died of respiratory failure. The danger usually lies in an affection of the respiratory apparatus, and patients have frequently died from what may be described as "respiratory crises." In one instance a patient was admitted to hospital one afternoon at 3 o'clock. She became suddenly dyspnoic about eight, tracheotomy was performed, without affording her very much relief, and she died at two o'clock in the morning. These sudden and ominous changes in a patient apparently well a few hours before, stamp the disease as one to be looked upon always as possibly serious. Yet in some cases recovery apparently takes place, and it is possible that this is occasionally permanent.

*Treatment.*—Treatment, so far, has not resulted in any modification of the disease. There is no doubt that strychnia, especially if given hypodermically, has a good influence. Thyroid treatment and various other forms of treatment by organic extracts have been tried without any benefit; and in the meantime all that we can say in regard to treatment is, that it must be carried out on general principles—good food, fresh air, absence of exhausting, tiring, disturbing conditions, and the use of drugs like strychnia. The food should be liquid or semi-solid, and the use of a tube for feeding should never be resorted to, for in some cases in which it has been used sudden death has followed almost at once. The presence of collections of lymphoid tissue in different parts of the body suggests a possible pathology, as already stated. It also suggests that, should it be found that such tissue is in some way related to the disease, perhaps as the source of a muscle poison, it may be possible to find an antitoxin for it.

## MUSCULAR DYSTROPHY

*History.*—Some years ago it was the custom to describe what we now know as cases of muscular dystrophy, in several varieties as cases of myopathy. The most striking and most familiar variety was that known as pseudo-hypertrophic paralysis. The next most frequent was the so-called idiopathic muscular atrophy; and a third variety was described which was known as the facio-scapulo-humeral, or Landouzy-Déjérine variety. All these varieties were characterised by the occurrence of a diseased condition in the muscles, leading to their disability. It was supposed to be unassociated with any disease of the spinal cord or the nervous system, and its origin was obscure. It is now considered more convenient to describe the disease under the name of “muscular dystrophy,” as it indicates the essential nature, at all events so far as we know at present, of the disease, and it does not tie us down to a description of types which are by no means definite, even in the same family. The so-called pseudo-hypertrophic variety has been recognised under this name ever since its graphic description by Duchenne. Its features were muscular weakness, associated with enlargement of the muscles, sometimes a few, sometimes many, and atrophy of others. The most noteworthy addition to our knowledge, after Duchenne’s description of these cases, was in the important lecture of Gowers on pseudo-hypertrophic paralysis. In this a full description of the disease was given, numerous clinical observations and important facts were detailed, and all that was known regarding its pathological anatomy was clearly set out. In this lecture, also, Gowers

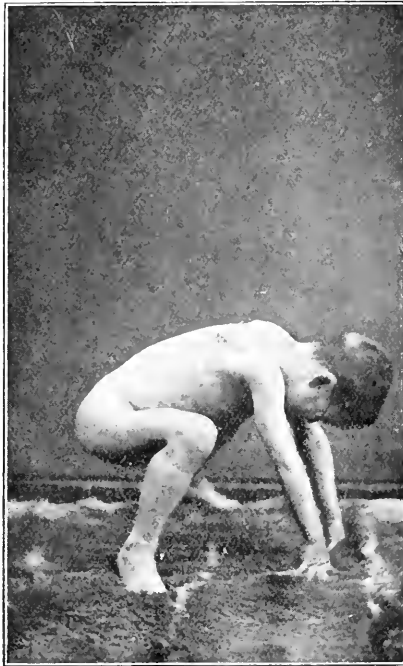
clearly recognised that hypertrophy, or seeming hypertrophy, and atrophy may be combined in different proportions, and that there are cases which connect the two extreme conditions of enlargement of many muscles and wasting in all. In this we get, as it were, a glimpse of the group which Erb afterwards clearly outlined under the name of the juvenile form of progressive muscular atrophy. This form was characterised by its occurrence in young people under the age of 20, and by the fact that there was enlargement of certain muscles, with wasting, especially of those about the shoulders. From a consideration of the characters and histories of these cases, Erb concluded that the three varieties—his own juvenile form, or idiopathic atrophy, a form described by Leyden, in which the hereditary tendency was strikingly marked, and pseudo-hypertrophic paralysis—were but examples of one condition, for which he proposed the name that now threatens to become general, *dystrophia muscularis progressiva*. The next step in the evolution of our knowledge of this interesting affection came from Landouzy and Déjérine, who, in 1885, published an exhaustive paper on what they called *myopathie atrophique*. It described observations on an exceedingly interesting group, the essential characteristics of which were widespread muscular atrophy, commencing in the face, without any hypertrophy. These cases were really identical with a disease which had been described by Duchenne under the name of *atrophie musculaire progressive de l'enfance*. Duchenne, however, was under the impression that those cases described by him with wasting of the muscles of the face as well as of the limbs, and a tendency to occur in several members of a family, were identical with progressive muscular atrophy as it occurred in adults. Landouzy and Déjérine, however, completed Duchenne's original description by discovering from *post-mortem* examination that the disease did not depend upon, and was not associated with, changes in the spinal cord—that it was, in short, a muscular affection without any detectable nervous lesion. They therefore regarded the

condition as one of myopathy; but relying upon the commencement in the face and the absence of hypertrophy, they looked upon the variety as a distinct and separate one, refusing to recognise the essential identity of these cases with those described by Erb, in spite of the fact that in one of their quoted cases the face was not affected, and considerable enlargement of the calves, if it was not present at the time, had been a few years before. The most important later contribution to the literature of this disease is again the work of Erb. In this he submits to examination numerous clinical records of cases of different kinds of myopathy, and makes it clear by observations on cases of different types amongst members of the same family, by the description of cases which form distinct connecting links between the different varieties, and finally, by observations on the histological conditions in the affected muscles, that all the varieties to which we have referred—pseudo-hypertrophic paralysis, the hereditary form described by Leyden, the juvenile form of idiopathic atrophy of Erb, and the Landouzy-Déjérine type, identical with the *atrophie musculaire progressive de l'enfance* of Duchenne—are essentially examples of the same disease.

*Clinical condition.*—In spite of the fact that the division into varieties is by no means a satisfactory one, and is, perhaps, somewhat unnatural, it is most convenient to describe this disease, to some extent at least, under different types. Undoubtedly the most common, the most characteristic, and the most striking type is that known as *pseudo-hypertrophic paralysis*. In this variety of the disease the onset usually occurs in early life. It is noticed, for example, that a child becomes a little less smart and quick in walking than usual, that there is a tendency to assume a mode of progression in which the heels are not brought down to the ground, and that walking upstairs becomes extremely difficult, while the ability to walk, or even run downstairs, is still retained. Associated with this there is frequently noted a difficulty in rising from a chair, especially from a

very low chair, and this difficulty is seen at its greatest if the child attempts to rise from the floor. If a patient suffering in this way is carefully examined, it is usually found that the apparent muscular condition is very much better than one would be led to suppose from the obvious muscular weakness. The lower limbs are usually well developed and firm; the calves, however, are found to be

FIG. 60.



unusually hard, and are frequently enlarged. Associated with this enlargement of the calf, there is frequently a slight degree of talipes equinus, the result of decay of muscular fibre and its replacement by fibrous tissue, and the contraction to which such a modification gives rise. The thigh is usually fairly well developed, but on careful examination will usually be recognised as being thinner and smaller, especially having regard to the bulk of the



calf. At this stage the knee-jerk may be present. It is not invariably present, but it is by no means invariably absent, as has been stated. As a matter of fact, the presence, or the diminution, or the absence, will depend upon the degree to which the vastus internus is involved in the muscular disease. As regards the trunk, the muscles about the pelvis and back have usually become somewhat

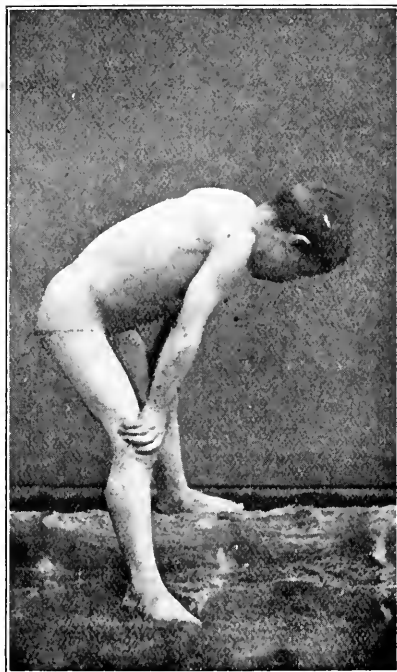
FIG. 61.



weak. As a consequence of this, there is marked lordosis, the shoulders are thrown very far back, so that a line dropped from the shoulder-blade perpendicularly falls well behind the hips. The muscles about the shoulder-blades are altered. They may be wasted, but it is more frequent at such an early stage as this to find a certain degree of enlargement, especially of the infraspinatus. And Gowers has stated that, next to the calf muscles, the infraspinati

are most generally enlarged. It may be found, also, that there is some enlargement of the deltoid. The pectorals are usually small, the latissimus dorsi may be exceedingly small, or almost undetectable. It has been said that it is sometimes congenitally absent. The upper arm muscles are small and weak; the forearm may be firm and fairly strong. In some cases, however, the unusual firmness

FIG. 62.



suggests a similar condition of pseudo-hypertrophy to that present in the calves. It is very rare to have the small muscles of the hand affected, although the writer has known a definite degree of enlargement to occur in the first dorsal interosseous muscle. This enlargement, in the case referred to, was associated with considerable enlargement also of the tongue; but the tongue is enlarged in other cases apart altogether from any change in the small

muscles of the hand. Wasting of the small muscles of the hand is rare, but does occur in this disease.

Such, then, is a case of ordinary pseudo-hypertrophic paralysis as it occurs in its most frequent form, and in a child. But it must be remembered that the disease is one which is by no means confined to children; and although it most frequently does occur in early life, it does not neces-

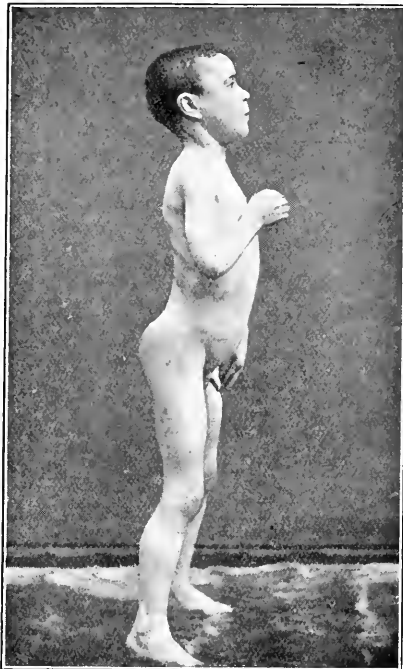
FIG. 63.



sarily begin at that time. There is undoubtedly a considerable number of cases in which the disease does not begin until after the age of puberty. It has been stated that there is always some defect in walking in such cases, dating from early life. This is not the case; and, as illustrating the extreme degrees in which the disease may occur, two cases may be mentioned that were long under observation. One was that of a man who, when first seen, was aged 45

years. In every typical aspect he was a case of pseudo-hypertrophic paralysis, with a feeble walk, slight talipes equinus, lordosis, enlarged calves, and some enlargement about the infraspinatus and the deltoid, and weakness of the pectorales and latissimus dorsi. This patient had been able

FIG. 64.



Figs. 60 to 64.—These figures illustrate the successive attitudes assumed in rising from the ground by a patient suffering from muscular dystrophy (pseudo-hypertrophic type). From photographs kindly lent by Dr. J. H. Crocker.

to get about all his life. He had never been able to run, and from his earliest childhood had walked, according to his brother's account, in exactly the way in which he did when first seen. This patient lived for ten years, and during those ten years, when he was under frequent observation, he did not alter in any material respect. The other extreme case was that of a man who was first seen

when he was aged 20 years. He came on account of difficulty in walking, which had been increasing for over a year. When he came first, he walked with his shoulders well thrown back, very marked lordosis, some degree of talipes equinus, and great feebleness. He was unable to get up from the floor, and had great difficulty in even getting up from his chair. The history was that he had been a perfectly healthy, strong man all his life until a year before. Not only had he been able to walk and run like other people, but he had actually been an athlete, and had won prizes for racing. The disease, unfortunately, progressed very rapidly, and, after a short time, led to complete inability to walk. He has now been, for five years, completely bedridden, the condition of helplessness having supervened within three years of the onset of the first symptoms of his illness.

Such cases as these show the extreme variation both in the progress of the disease and the curiously dissimilar conditions attending its onset as well as its progress, and they have been related in order to emphasise the fact that although the great majority of cases of pseudo-hypertrophic paralysis, recognisable as such, occur in children, yet adults are by no means exempt, and that the disease sometimes actually commences after adult life has been attained.

#### IDIOPATHIC MUSCULAR ATROPHY.

Although, as has been already stated, these types of muscular dystrophy are by no means definite, it may be well to mention the salient points of cases in which the atrophy is the predominant feature, cases which differ but little in their mode of onset from cases of pseudo-hypertrophic paralysis, yet which commence more frequently in young adult life. They are characterised by the same weakness, the same difficulty in walking, especially the similar difficulty in going upstairs, with the ease and facility in descending. And they differ chiefly in the fact that in the majority of cases no enlargement of muscles is

present, but in most there is wasting of nearly all, and especially of those which have to do with locomotion. The thighs and the legs may be exceedingly small and thin. Even, however, when the calves are small there is a hardness about them which is almost characteristic. The muscles about the shoulders are also wasted, and so are those of the upper limbs. The pectorals are exceedingly small, and so is the *latissimus dorsi*, which may actually be absent. As a rule, such cases are not so rapid in their progress towards complete paralysis as are cases of pseudo-hypertrophic paralysis. Yet that the diseases are essentially the same is shown by the fact that cases which may be at one time described as cases of pseudo-hypertrophic paralysis, at a later stage may show all the signs of the idiopathic variety, and that in the same family cases of the two different varieties may occur.

The *juvenile type of Erb*, so far as it follows a definite clinical course, is characterised by the occurrence of wasting, especially about the muscles of the shoulder girdle, occurring in a young adult otherwise healthy. This wasting about the shoulders may be unassociated at first with any affection of the pelvic girdle or of the legs. As a rule, however, after the atrophy about the shoulders has become marked, weakness and atrophy in the lower limbs frequently occur. Enlargement of muscles, so-called hypertrophy, in this variety is not a marked feature. Occasionally, however, it is present, especially in the deltoid and in the *infraspinatus*; and there may be that curious disproportion already referred to in considering pseudo-hypertrophic paralysis, between the small size of the upper arm and the well-developed firm character of the forearm. There is very little doubt that the affection of the face frequently occurs in association with this weakness about the shoulders, and although Landouzy and Déjérine have described the so-called *facio-scapulo-humeral type* as being characterised by the commencement of weakness in the muscles of the face, it is scarcely to be doubted that the affection of the face may be superadded to a pre-existing

affection of the shoulder girdle. So far as the facial affection is concerned, it consists essentially of weakness of the orbicularis palpebrarum and of the orbicularis oris,



FIG. 65.—Muscular dystrophy. Face in the Landouzy-Déjérine type, showing weakness of the orbicularis palpebrarum and orbicularis oris. From a photograph by Dr. Collier.

so that the patient is quite unable to completely close his eyes, and is also unable to pucker up the mouth so as to whistle, or blow out a candle.

There is no doubt that the so-called juvenile type of Erb, which, as just stated, essentially consists in wasting of

muscles about the shoulder girdle, exists in patients who have no affection of the face at all. It is also, we believe,



FIG. 66.—Muscular dystrophy, Landouzy-Déjérine type, showing great wasting about shoulders, etc.

the case that the affection of the face may be superadded to an affection of the shoulder girdle—that is, that the shoulder-girdle muscles may be first affected and the facial muscles



subsequently suffer. But it is also probable that certain cases of so-called double facial paralysis which have been found in early life, and which have been characterised by a



FIG. 67.—Muscular dystrophy. Lambouzy-Dejerme type, showing marked lordosis and wasting, especially above and below knee and above elbows.

gradual onset of weakness of the orbicularis palpebrarum and orbicularis oris, are really cases of myopathy, in which in an early stage only an affection of the face is obvious, to which may subsequently be superadded some affection of

the muscles around the shoulder girdle. The clinical condition in all those diseases is almost sufficiently indicated by what has already been stated. The weakness of the muscles, apparently proportionate to their size in the cases in which atrophy is distinct, and disproportionate to their bulk when there is pseudo-hypertrophy present; the absence of fibrillary twitchings, the diminution or suppression of reflexes, are points which are thoroughly dis-

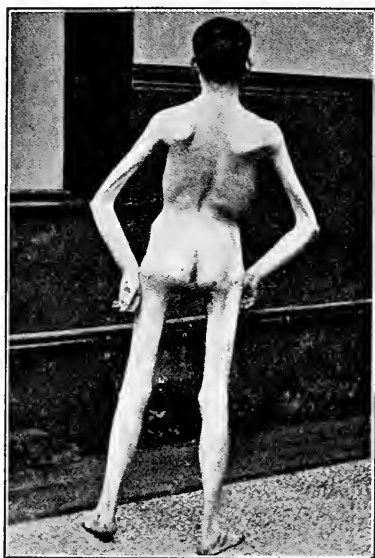


FIG. 68.—Muscular dystrophy, Landouzy-Déjérine type, showing great wasting of limbs and trunk.

tinctive of the condition. The response of the muscles to electrical currents is diminished, but there is no characteristic reaction of degeneration. Contractures frequently take place, as has already been stated, on account of the substitution of fibrous tissue for muscular tissue. The organic reflexes are never affected, and sensation remains unimpaired, even when the muscular weakness has gone on to an extreme degree.

*Mental state.*—There seems to be no constant mental change associated with the muscular weakness. There is

very little doubt that many patients suffering from pseudo-hypertrophic paralysis are very backward mentally, and are almost imbecile. It has been stated that this may depend upon the absence of the ordinary advantages of education and of association with children of the same age; but in many cases the mental weakness is far too extreme to be accounted for in this way. In other cases the patient may be bright and sensible; but it must be confessed that in the majority of cases of pseudo-hypertrophic paralysis, especially when the disease dates from quite early life, the mental condition is one of considerable backwardness. In the other varieties of the disease, especially in those in which the onset occurs in early adult life, the mental condition is quite unaffected.

*Course of the disease.*—In the vast majority of cases the weakness of muscles gradually progresses until a condition of complete helplessness is reached. In some cases, undoubtedly, the progress is exceedingly slow, and in such a case as that related above, of the man who first came under observation at the age of 45, it would almost seem as if the disease had been quite at a standstill for very many years. No doubt the same is true in a less degree of other cases—that is, that the progress is exceedingly slow, so slow, indeed, as to be almost imperceptible. But these exceptional conditions, although they may possibly encourage a more hopeful prognosis if any tendency to an extremely slow progress has been present, do not affect the general rule that the progress is steady towards complete helplessness. In many instances while the legs still remain fairly strong and vigorous the patient becomes unable to walk on account of the talipes equinus which the contraction of the calf muscles sets up. In some cases of this nature it has been possible, by means of tenotomy, to so restore the condition of the lower limbs that locomotion, which had become impossible, was re-established. This, of course, must be of somewhat unusual occurrence; but it is a fact to be borne in mind, and may be especially helpful in reference to the cases in which the

deformity caused by contractures of muscles may seem to be out of proportion to the weakness of the limbs, as suggesting that correction of the deformity may, for a time at least, restore the usefulness of the limbs.

*Etiology.*—One thing stands out very clearly with reference to the etiology of this condition—that is, that in so many cases it is a family disease, transmitted from one generation to another, and frequently affecting several members of the same family. But there seems very little doubt that it does occasionally arise either as an isolated condition or as affecting more than one member in a family in which no trace of any hereditary weakness can be discovered. It is difficult to know whether consanguinity of parents plays any part in the etiology. The writer has known, through the kindness of Sir William Gowers, a family of ten, nine of whom were affected with the disease. The parents were double first consins, *i.e.* two brothers had married two sisters, and the son of one marriage had married the daughter of the other; and they were in such a social position that it was possible to trace the family history back through several generations, and no trace of any similar illness could be discovered. Such a fact is of great significance. One curious point in reference to the disease is that it so much more frequently affects males than females. Females by no means escape, but the incidence of the disease is certainly much greater amongst males. Yet, in spite of this fact, the transmission of the disease is frequently through a female who herself has shown no signs of it. Thus, in a family of three—two males and one female—the two brothers were affected, but the sister escaped. She married, however, and had a family of three, two of whom were affected with pseudo-hypertrophic paralysis. The broad fact in reference to the etiology of the disease seems to be that there is inherited in those who suffer from it some condition, apparently of the muscular tissue itself, which leads to its early decay and perhaps its death. What relation this disease of the muscles has to the nervous system, or whether it has any such relation, is still uncertain.

*Morbid anatomy.*—In the few cases in which opportunity for examination of the central nervous system has been offered no change which can be brought into definite relation with the disease has been found in the spinal cord or in the nerves. It is, however, conceivable that the methods either now in use or which may afterwards be devised may reveal some change. The only histological change which has been discovered so far is the change in the muscles. The change essentially consists in a wasting of muscular fibre and its replacement by fibrous and fatty tissue. The muscle-fibres themselves seem to be altered, but no change so far has been found in the muscle-spindles. It may be—as Gowers has suggested—that the disease is essentially one of the embryonic muscular tissue; that the muscle is, so to speak, in an undeveloped condition—in a condition in which it cannot undergo development, but in which it changes and atrophies, and is replaced by fibrous tissue. Certain changes have been described in the spinal cord, such as the presence of cavities and fissures; and it is possible that these changes may have some bearing upon the etiology of the condition, as showing in the nervous system a tendency similar to that which is present in the muscular tissue, namely, towards the persistence of an embryonic condition. But any definite relation of such changes in the cord to the muscular condition has not been demonstrated.

*Diagnosis.*—The diagnosis of this disease, when one is familiar with the different varieties and the different forms in which it occurs, does not, as a rule, present any very great difficulties. The absence of fibrillary twitchings, the rarity with which the small muscles of the hand become affected, and the age at which it usually occurs, sufficiently distinguish it from the progressive muscular atrophy due to degeneration of the spinal cord. The presence also of a history of a similar condition occurring in more than one member of the same family—a by no means uncommon experience—will also be another point to distinguish it from the condition just referred to. In some instances a

doubt has arisen as to whether a patient suffering from muscular dystrophy might not be the subject of an affection of the spinal column leading to pressure upon the spinal cord and giving rise to paraplegia. This doubt could only arise in those somewhat unusual cases in which the angular curvature associated with caries of the spine is not distinct. But the history of the onset, the limitation of the weakness to the lower limbs, the common affection at least to a slight degree of the sphincters, the state of the reflexes, and the possible association of some degree of anæsthesia in pressure paraplegia will be sufficient usually to enable the distinction to be made. Peripheral neuritis is scarcely to be mistaken for this disease, the tenderness of the nerve-trunks and the history of some toxic condition being usually sufficient to set one upon the right track. One condition has been met with which closely simulated pseudo-hypertrophic paralysis in a child. The child came under observation on account of a certain amount of difficulty in walking, which had come on slowly and gradually in the previous few weeks. There was no pain, no enlargement of muscles, and no wasting. The muscles also all responded quite readily to the faradic current. But when the boy was put on the floor he got up in what we recognise as the characteristic method which a patient assumes who is suffering from pseudo-hypertrophic paralysis (see Figs. 60 to 64). This little boy died some weeks later from tubercular meningitis, and he was found to have double hip-joint disease of tuberculous origin. It is very interesting that such an interference with both hip-joints should have given rise to an assumption of the curious attitudes in getting up from the floor with which most of us are familiar, and which are so constant in pseudo-hypertrophic paralysis as almost to be regarded as characteristic.

*Prognosis.*—The prognosis of this disease is essentially bad. There is no likelihood of any recovery being effected, and it is only too probable that the progress will be steadily downwards. Yet the occurrence of such a case as has already been alluded to more than once, rare as it may

he, shows that a downward progress is not invariable. There is, however, very little doubt that in practically all the cases the tendency is for the muscular weakness to become greater and greater and to become more and more widespread. Of course, in a man who has been strong and well up to early adult life the danger of complications, pulmonary and otherwise, must be much less than in a child in whom the muscular weakness dates from quite early life.

*Treatment.*—From what has already been said it will be obvious that treatment is not likely to be of very much avail. Fresh air, good food, and an encouragement to get about so long as it is at all possible, are really the only measures that suggest themselves as likely to minimise the danger and the discomforts of such an illness. Various organic extracts—thyroid, thymus, pituitary, and others—have been tried, but so far without any good effect. Gymnastic exercises, also massage and electrical treatment, although they may to a certain extent nourish the muscles, and keep up their ability to act longer than would otherwise be the case, seem to be quite ineffective in modifying the progress of the disease.

## PERONEAL FORM OF MUSCULAR ATROPHY

THE variety of muscular atrophy to which this name has been applied was described almost simultaneously by Charcot and Marie in France, and by Tooth in this country in 1886. A very important contribution to the symptomatology and general characteristics of the disease, both clinical and pathological, was made by Hoffmann three years later, under the designation of "Neurotische Muskelatrophie." Ormerod, Sachs, Siemerling, and others have also written on the same subject, and the disease has gradually acquired more definite outlines, although neither in its symptomatology nor in its morbid anatomy and etiology is it yet quite as definite as it is likely to become. Nor are its relations quite clear on the one hand to cases of myopathy, and on the other hand to cases of muscular atrophy dependent upon nervous disease.

The disease is essentially one of children and young adults, and is characterised by the early affection of the peroneal group of muscles. From them the affection may spread so as to involve the anterior tibial group and frequently in a slighter degree the calf muscles. It is nearly always symmetrical, although the affection may be slightly less severe in one leg than in the other. At this stage a patient suffering from the disease may present a condition in other respects quite normal, but the leg below the knee is much wasted, the movements of the toes and at the ankle, especially those of dorsiflexion and eversion, may be extremely slight or altogether absent, and no response to a faradic current of even painful intensity can be evoked. In short, the leg below the knee has more the character of a rigid stilt than of a freely movable limb.



At this stage no other phenomenon may be present. The thigh may be well developed—may indeed appear to be immensely large and full, the muscles respond to a weak faradic current, and the knee-jerk may be active. The part which usually suffers next in order is the ball of the

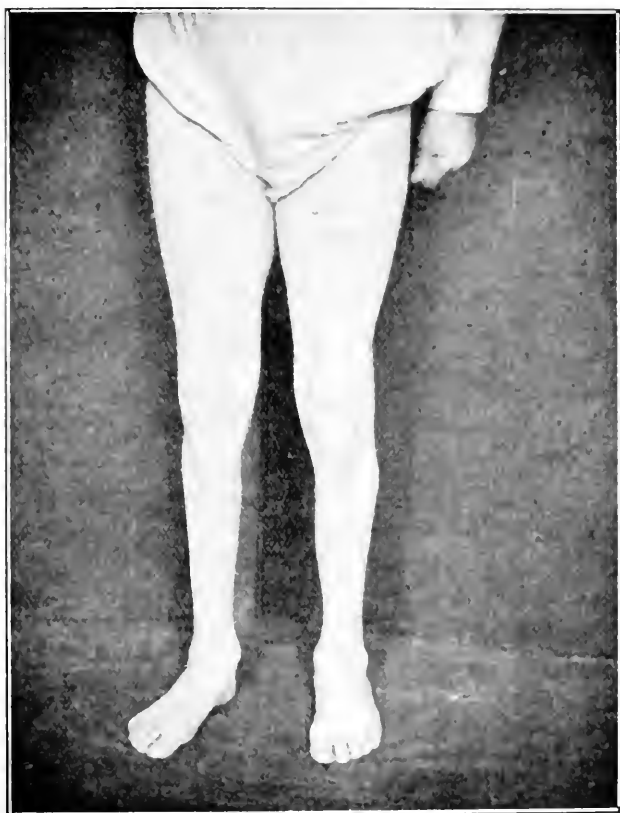


FIG. 60.—Peroneal type of muscular atrophy showing marked wasting below the knees. From a photograph by Dr. Collier

thumb, and the wasting here is of the same character as that below the knee. This may in turn spread so as to involve the forearm muscles, and the picture presented at this stage is one in which wasting of the muscles below the knees and below the elbows is the chief characteristic.

The wasting is, of course, associated with much loss of power. The weakness and wasting may subsequently spread so as to involve the thighs, the upper arms, and even the pelvic and shoulder girdle muscles, and the wasting usually occurs in the same way, viz. a gradual



FIG. 70.—Peroneal type of muscular atrophy, from Dr. Allen Starr's work on 'Organic Nervous Diseases.'

disappearance of the muscular fibres and a gradual extinction of the faradic contractility. There is never any hypertrophy of muscles, and fibrillary twitching is frequent but not constant. The facial muscles do not suffer. From the nature of the disease it can be understood that the occurrence of deformities is common. Double club foot is

especially apt to occur, and it may be said that the presence of acquired double talipes varus should always suggest this form of muscular atrophy. Some impairment of sensibility has been described in certain cases, especially in the legs, but this is certainly not constant.

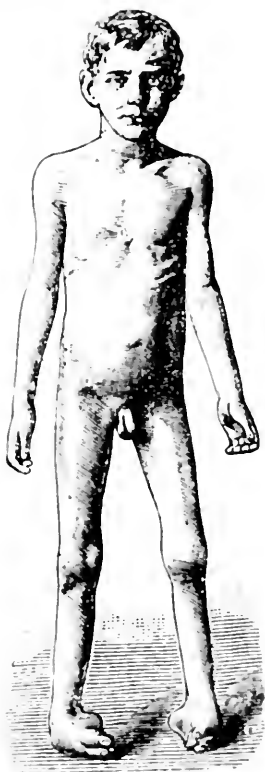


FIG. 71.—“Peroneal type” of muscular atrophy. From Hoffmann's article on “Neurotische Muskelatrophie” in the ‘Archiv für Psychiatrie,’ vol. xx, showing wasting and deformity.

The *course* of the disease is very variable. In some cases it reaches a stage at which there is marked wasting of both legs below the knee, and some wasting of the hands, and then it ceases to advance apparently for a long time—sometimes several years. In some instances the arrest of the disease seems to be permanent, in others it

seems to make a fresh start after a period of quiescence and then the progress if slow is usually steady. In other cases there is a more or less gradual increase in the affection, both in the extent of the disease and in the intensity with which it affects different muscles. But it must always be remembered that arrest may take place at any stage, although it cannot be said that it is to be expected.

The *etiology* and *pathology* of the condition are very obscure. It is undoubtedly a family disease, frequently affecting several members of the same family and in several successive generations yet apparently not transmitted in the female line especially. But, as in all family diseases, isolated cases are frequently met with, occurring in a family otherwise untainted. The relation of the onset of the symptoms to any preceding illness is not constant. Sometimes the first symptoms are manifested after an acute illness such as measles; in other cases the occurrence of the early symptoms does not seem to be determined by any special exciting cause.

*Morbid anatomy.*—In the affected muscles the wasting is a simple atrophy and a replacement of the bulky muscular tissue by the less voluminous fibrous tissue. There is no change other than this in the muscular fibres—no hypertrophy of any or such a condition as is found in muscular dystrophy. A degenerative neuritis has been described in the related nerves, the sclerotic changes have also been found in the posterior columns of the spinal cord. Slighter changes have also been described in the lateral columns, but the relation of these changes to the disease is extremely obscure. It may indeed be stated that the disease is one of the most obscure, according to our present knowledge, and one of the most anomalous, with which we have to deal.

The *diagnosis* is not very difficult when one really grasps the main facts in the history and the peculiar distribution of the paralysis. The gradual onset distinguishes it from acute infantile paralysis. It offers many points of resemblance to progressive muscular atrophy—the wasting, fibrillary twitching, etc.—but the age of onset, the distri-

bution of the atrophy, and the resulting deformities are sufficient as a rule to distinguish it. With myopathic diseases or muscular dystrophies it has much in common; and it is not impossible that subsequent investigation may indicate an even closer relationship than is at present obvious. Yet the distribution of the wasting, the frequent presence of fibrillary twitching, and the affection of the small muscles of the hand are points of definite difference from any of the varieties of muscular dystrophy which are known. Peripheral neuritis from alcohol or arsenic offers many points of resemblance; but the history of the illness, the age of the patient, and the absence of those poisons will be sufficient as a rule to mark the distinction.

The *prognosis* is necessarily bad so far as any hope of complete cure is concerned. Yet it is not uncommon to find a case in which the disease advances to a certain point and is then arrested—perhaps permanently. In some cases the arrest, however, is not permanent, and in most the progress of the morbid process is slow but steady. It cannot be asserted that any *treatment* so far tried has been effective in any striking degree—namely, massage, galvanism, and various hydropathic measures, which have all been well tried. Massage and galvanism have seemed in some cases to have a definitely beneficial effect. Drugs, unfortunately, are not very effective. Strychnine—especially if given hypodermically—has seemed in some cases to do good.

## PARALYSIS IN CONNECTION WITH JOINT AFFECTIONS

THERE are various diseased conditions of joints which give rise to immobility of the related limb or part of a limb, closely simulating paralysis which results from disease of the nervous system. Attention has already been directed to paralysis arising in connection with caries of the spine; but although this disease frequently commences with a change in the intervertebral joints, the cause of the paralysis in such a condition is the effect directly produced upon nervous structures by the inflammatory results of the disease in the bone. So that this paralysis must not be regarded as of the nature of immobility connected with joint disease. In other conditions, however, to which reference is about to be made, the joint condition which gives rise to the immobility is not associated necessarily with any diseased condition of the nervous system.

Occasionally it happens that in acute hip-joint disease, especially if the affection is bilateral, a condition of difficulty in moving may be induced, which closely simulates the difficulty present almost characteristically in the condition of pseudo-hypertrophic paralysis. Thus, in one such case, the patient was a boy admitted to hospital on account of difficulty in walking and in getting up from the ground or from a chair. He walked slowly and deliberately, balancing himself first on one foot and slowly swinging forward the other, resting in turn upon that, and swinging the other one forward in its turn. There was also a certain degree of lordosis present, and when it is stated that in rising from the floor he assumed all the varied attitudes which characterise the attempts of a patient with pseudo-

hypertrophic paralysis going through the same movements, it can be seen how closely the whole condition simulated that met with in pseudo-hypertrophic paralysis. The case presented great difficulty in diagnosis for some weeks, until undoubted symptoms of tubercular meningitis declared themselves, and at the *post-mortem* examination both hip-joints were found to be in a state of acute tuberculous inflammation. This case is related as being an extreme example of diseases of joints simulating conditions of paralysis. The disability produced by the diseased joints resulted, apparently, in an impairment of the function of structures which are intimately connected with mobility. An affection of a single joint may produce a similar condition in one leg, and the possibility of such an explanation in any obscure case of apparent paralysis should always be borne in mind. In the case to which reference has just been made, the normal reaction of the muscles to a faradic current and the recent onset of the weakness were the factors which made it possible to exclude the diagnosis of pseudo-hypertrophic paralysis.

Occasionally a child is brought to hospital on account of difficulty in moving the leg. Examination reveals considerable swelling and pain in one hip or in one knee, the condition being really the result of an attack of acute rheumatism. Occasionally the same condition affects the shoulder, and a similar state of apparent paralysis is produced in the arm. It also sometimes happens that a rheumatic affection of the cervical spine may be mistaken for cervical caries, and the writer has known a case in which a rigid support was actually ordered for a child, but before the support had arrived the acute inflammatory condition, which was really the result of rheumatism, had entirely disappeared. In all such rheumatic cases the joint affection is, as a rule, quickly removed by treatment. Occasionally, however, although such an experience is more common in adult life, the onset of weakness and pain in the arm is more gradual, is accompanied with feverishness, and does not clear away with anti-rheumatic remedies.

In such a condition we are dealing with an arthritis, obscure in origin, which we vaguely, however, characterise as gonty or rhenmatic. Occasionally the patient does not seek advice until considerable adhesions have taken place, and then he is found to have a weak arm, with limited and restricted movement and considerable muscular wasting, affecting especially the deltoid, but involving also all the muscles about the shoulder, and including the biceps and triceps. Examination of the joint by fixing the scapula with one hand on the shoulder, while the other is employed to move the arm away from the side and to rotate the head of the humerus in its cavity, reveals the presence of adhesions of some strength, and the true nature of the condition becomes obvious. In some cases the nerve-sheaths are involved in the inflammation, and a condition of brachial neuritis is established.

It must, however, also be remembered that such adhesions as are here described are especially apt to occur in the shoulder of the affected side in cases of hemiplegia,—especially, as has been pointed out by Dr. Hughlings Jackson, in cases of left hemiplegia—and the mistake must not be made of regarding the arthritic condition as the only one in such cases.

The *diagnosis* of such a condition is sufficiently considered in describing it. It may at first sight resemble a condition of anterior poliomyelitis, but the history will help to exclude this, and the fixation at the shoulder, contrasted with the undue mobility in anterior poliomyelitis, will be sufficient to mark the difference. The normal reaction of the muscles also to faradism is in marked contrast to the great impairment or actual loss of faradic reaction in anterior poliomyelitis.

The *prognosis*, especially in young subjects, is quite favourable, and if the condition is recognised early, before the adhesions are fairly fixed, recovery may be rapid if proper treatment be adopted.

The *treatment* may be considered as of two parts: (1) treatment in the early stage, and (2) treatment later, when



the adhesions are formed and the joint is fixed. In the early stage, hot sponging and douching locally, and the use of small doses of mercury internally (*e. g.* gr. j of blue pill thrice daily) are the lines of treatment found to be most successful. Massage and passive movements are also useful, although it is possible to do harm at this stage by the too energetic use of such measures. Gentle movements, hot applications, and the use of mercury at this stage comprise all the measures necessary. In the later stage attempts must be made to get rid of the adhesions. In most cases massage and passive movements are sufficient for this. The use of the constant current also seems to help, possibly by influencing the nerves of the brachial plexus, the sheaths of which are frequently involved secondarily to the joint affection. Occasionally in severe cases it is necessary to give gas and forcibly break down the adhesions. In such cases the after-treatment by means of massage and passive movement is at first difficult to carry out because of the pain, but they must be persisted with, and the result, as a rule, is quite satisfactory, so far as the mobility of the limb and the restoration of its function are concerned.

In another condition of joint deformity a peculiarity of gait and appearance arises which is such as to lead to occasional mistakes in diagnosis. The writer has known the condition of congenital hip dislocation more than once diagnosed as pseudo-hypertrophic paralysis. And when the peculiarity in the walk, the lordosis, and the deformity are considered, it will be acknowledged that even a good observer may occasionally fall into this error. Whatever may be the conditions determining congenital hip dislocation, a case of this character comes before us on account of the peculiarity in walking, or the malposition in standing, or the unusual degree of tiredness attending either act. As the patient walks, there is a roll or lurch from one side to the other, usually more distinct to one side, for the deformity is rarely equal on the two sides. Examination reveals an undue mobility at the hip-joint, and it is usually found that the head of the femur seems to slip from the

acetabulum on to the dorsum of the ilium. Occasionally the slipping is on to the ischium; but whether it is on to the ilium or the ischium, it results in a shortening of the limb, and a consequent impairment of that uniformity in

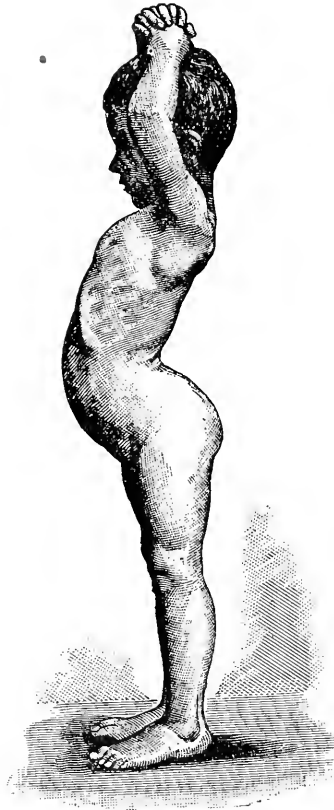


FIG. 72.—Congenital hip dislocation showing lordosis, after Professor Lorenz.

gait which is the rule in the normal person. When the patient stands there is a tilting of the pelvis and a consequent lordosis which, still more than the gait, suggest pseudo-hypertrophic paralysis (Fig. 72).

The *diagnosis* of such a condition is not always easy. The points of most importance which seem to distinguish it

from pseudo-hypertrophic paralysis are—the normal condition and bulk of the muscles and the absolutely normal reaction of those to faradism, and the natural condition of the reflexes.

The *prognosis*, as regards life, is of course good, and there is no likelihood of the lameness and difficulty with locomotion becoming worse.



FIG. 73.—Coxa vara in a child, showing great degree of mobility at the hip joints. Dr. G. A. Sutherland's case.

The *treatment* is a matter for the surgeon, but the writer was much struck a few years ago in discussing this subject with an extremely intelligent medical man (who himself suffered from the condition, and who had seen the results obtained on the Continent by various forms of surgical treatment), by his remark that he was very glad his parents had had no operative measures tried in his case.

The condition of *coxa vara* should be referred to, especi-

ally in reference to congenital hip dislocation. This condition is dependent upon a rachitic twisting of the neck of the femur. It may be unilateral, but is more frequently bilateral. It results in an undue mobility of the joint in certain directions. Eversion and abduction are, as a rule, much more extensive than normal. So is adduction. Inversion is often impossible to any degree, although exceptionally the natural attitude assumed is one of inward rotation.

In many cases it is exceedingly difficult to make the distinction between coxa vara and congenital hip dislocation. In the latter, the mobility of the head of the femur and the descent of the trochanter on traction are points which help in diagnosis. But frequently the help of the X rays is necessary to allow of a definite decision being arrived at. The gait may closely simulate that in congenital hip dislocation, and that, as has been stated, may strongly suggest pseudo-hypertrophic paralysis. But the great mobility of the limb at the hip-joint in coxa vara, and the comparatively slight lordosis, are points which help in distinguishing it from congenital hip dislocation.

It will be recognised that various other conditions of inflammation or of other changes in or near joints, such as periosteal hæmorrhages in scurvy rickets, etc., may give rise to immobility as a protective measure, and such immobility may be easily mistaken for some form of paralysis, especially as muscular wasting may be marked in these conditions. In such cases, however, the recognition of the change, however slight, in the joints or bone, and the normal reactions of the muscles and the unaltered reflexes will help to reveal the true nature of the case, and sharply distinguish it from any condition resulting from a lesion of the nervous system.

## FUNCTIONAL PARAPLEGIA

It is not uncommon to find a child—usually after an illness of a more or less indefinite character—suffering from a condition in which inability to stand or walk is complained of. Sometimes this loss of power is not complete. The patient is able to stand, but to stand only unsteadily; and an attempt to walk is characterised not so much by unsteadiness as by a curious waviness in the line of progression, which recalls somewhat the skater's progress rather than that of the ordinary walker. In the former class of cases the condition is one of great limpness or flaccidity. On attempting to raise the child on to her feet she seems to slip through one's hands in a manner which recalls a similar flabbiness met with in patients suffering from muscular dystrophy. If she is got upon her feet they fail to afford her any support, and if left alone she may collapse in a heap on the floor. In the second class the disability varies. Sometimes the walk is of the wavy character already mentioned; sometimes one foot is dragged, sometimes both, but in a manner which is usually either a gross exaggeration of the ordinary spastic gait of organic disease or which is strikingly different from any gait met with in organic paralysis, or which offers a distinct resemblance to such different types as locomotor ataxy and paralysis agitans. The deep reflexes in such cases are active—sometimes very active; true ankle clonus is never present, and the plantar reflex is often not to be obtained. If it is present, however, it is flexor in type. Occasionally difficulty in seeing may be complained of. There may also be a tendency to retention of urine, which is much increased should a catheter be

unfortunately used, and there is occasionally, although not so frequently as in similar conditions in adults, a degree of hemi-anæsthesia and so-called "stocking" and "glove" anæsthesia, *i.e.* anæsthesia of the distribution which the names suggest.

Such a condition of functional paraplegia may be present as early as 5 years of age. It is most likely to be met with, however, between 8 and 12, and it may occur later.



FIG. 74.—Functional paraplegia, illustrating one type of gait in that condition. From a photograph by Dr. Collier.

*Etiology and pathology.*—It is not easy to account for the onset of the condition. As has already been stated, it is usually the sequel to an illness, usually of an indefinite character, and it scarcely ever occurs except in children who are over-indulged and treated generally in a way which is likely to aggravate any morbid mental tendency which may be present. That there is any actual structural change underlying these manifestations is almost incredible, especially when the rapid result of treatment to be presently referred to is considered.

*Diagnosis.*—There is no condition more difficult to diagnose unless one's eyes are open to the possibility of its occurrence. The diagnosis is rendered particularly difficult in some cases in which the initial illness has been a sore throat. The possibility of diphtheritic paralysis in any such case must be considered, but the activity of the reflexes, the absence of squinting or of difficulty in swallowing will usually be sufficient to negative such a view. The writer has also known such cases to be mistaken for spinal caries, giving rise to paraplegia. Yet the absence of ankle clonus and of extensor plantar responses is usually enough to render such a view untenable, especially if the paraplegia be severe; and the absence of any rigidity of the spinal column, and of any angular curvature is conclusive. When the gait is of the rolling character already described the possibility of cerebellar tumour has to be considered. Yet the absence of severe headache, of vomiting and of optic neuritis makes such a view highly improbable. And in any such case it has to be remembered that the diagnosis must be arrived at by a process of excluding all possible organic disease, a course fraught with much difficulty. For it is one of the most important things to remember that in the early stages of organic disease of the nervous system, *e.g.* tubercular meningitis and even cerebral tumour, it is not unusual to find these so-called functional symptoms among the early phenomena of the disease. So that one has to be careful in diagnosing functional paraplegia, lest its possible significance as an early sign of organic change should be ignored. Yet in many instances it is undoubtedly a purely functional condition undetermined by any structural change, and offering a most favourable field for a considerable therapeutic success.

*Prognosis.*—The prognosis is, of course, good. When the condition is simple, cure is certain, and often gratifyingly quick. In other cases it is slower, but in all it is ultimately complete. It should, however, be borne in mind that such functional manifestations occurring at an early age stamp

the child as neurotic in the highest degree and endow its future with a certain amount of anxiety.

*Treatment.*—If such a patient can be removed from its usually too indulgent and comfortable environment, the treatment is easy and the result certain. Admission to a nursing home or hospital, where its nurses are kind but firm, and where appropriate treatment can be used is by far the most satisfactory way. When treatment has to be carried out at home the result is never certain, and is always delayed. One sometimes has to resort to the expedient of advising the mother to go away for a holiday. In one case which the writer saw, this course resulted in a complete cure of the child before the mother's return. The line of treatment to be adopted is by means of faradic stimulation to the limbs, and, by measures calculated to improve the child's nutrition, to overcome whatever resistance there may be in the nervous structures to the transmission of voluntary impulses. No doubt such treatment has some physical effect. It is probable, however, that the suggestion which it conveys to the child's imagination is also an effective part of its action, and any adjuncts which may intensify this effect are desirable.



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