

adults as interchangeable, may cling mechanically to a specific person, or may use the parent's hand to obtain desired objects without ever making eye contact (as if it were the hand rather than the person that is relevant). Over the course of development, the child may become more willing to be passively engaged in social interaction and may even become more interested in social interaction. However, even in such instances, the child tends to treat other people in unusual ways (e.g., expecting other people to answer ritualized questions in specific ways, having little sense of other people's boundaries, and being inappropriately intrusive in social interaction). In older individuals, tasks involving long-term memory (e.g., train timetables, historical dates, chemical formulas, or recall of the exact words of songs heard years before) may be excellent, but the information tends to be repeated over and over again, regardless of the appropriateness of the information to the social context. Rates of the disorder are four to five times higher in males than in females. Females with the disorder are more likely, however, to exhibit more severe Mental Retardation.

Prevalence

The median rate of Autistic Disorder in epidemiological studies is 5 cases per 10,000 individuals, with reported rates ranging from 2 to 20 cases per 10,000 individuals. It remains unclear whether the higher reported rates reflect differences in methodology or an increased frequency of the condition.

Course

By definition, the onset of Autistic Disorder is prior to age 3 years. In some instances, parents will report that they have been worried about the child since birth or shortly afterward because of the child's lack of interest in social interaction. Manifestations of the disorder in infancy are more subtle and difficult to define than those seen after age 2 years. In a minority of cases, the child may be reported to have developed normally for the first year (or even 2 years) of life. Autistic Disorder follows a continuous course. In school-age children and adolescents, developmental gains in some areas are common (e.g., increased interest in social functioning as the child reaches school age). Some individuals deteriorate behaviorally during adolescence, whereas others improve. Language skills (e.g., presence of communicative speech) and overall intellectual level are the strongest factors related to ultimate prognosis. Available follow-up studies suggest that only a small percentage of individuals with the disorder go on as adults to live and work independently. In about one-third of cases, some degree of partial independence is possible. The highest functioning adults with Autistic Disorder typically continue to exhibit problems in social interaction and communication along with markedly restricted interests and activities.

Familial Pattern

There is an increased risk of Autistic Disorder among siblings of individuals with the disorder, with approximately 5% of siblings also exhibiting the condition. There also appears to be risk for various developmental difficulties in affected siblings.

Differential Diagnosis

Periods of developmental regression may be observed in normal development, but these are neither as severe or as prolonged as in Autistic Disorder. Autistic Disorder must be differentiated from **other Pervasive Developmental Disorders**. **Rett's Disorder** differs from Autistic Disorder in its characteristic sex ratio and pattern of deficits. Rett's Disorder has been diagnosed only in females, whereas Autistic Disorder occurs much more frequently in males. In Rett's Disorder, there is a characteristic pattern of head growth deceleration, loss of previously acquired purposeful hand skills, and the appearance of poorly coordinated gait or trunk movements. Particularly during the preschool years, individuals with Rett's Disorder may exhibit difficulties in social interaction similar to those observed in Autistic Disorder, but these tend to be transient. Autistic Disorder differs from **Childhood Disintegrative Disorder**, which has a distinctive pattern of severe developmental regression in multiple areas of functioning following at least 2 years of normal development. In Autistic Disorder, developmental abnormalities are usually noted within the first year of life. When information on early development is unavailable or when it is not possible to document the required period of normal development, the diagnosis of Autistic Disorder should be made. **Asperger's Disorder** can be distinguished from Autistic Disorder by the lack of delay or deviance in early language development. Asperger's Disorder is not diagnosed if criteria are met for Autistic Disorder.

Schizophrenia with childhood onset usually develops after years of normal, or near normal, development. An additional diagnosis of Schizophrenia can be made if an individual with Autistic Disorder develops the characteristic features of Schizophrenia (see p. 298) with active-phase symptoms of prominent delusions or hallucinations that last for at least 1 month. In **Selective Mutism**, the child usually exhibits appropriate communication skills in certain contexts and does not have the severe impairment in social interaction and the restricted patterns of behavior associated with Autistic Disorder. In **Expressive Language Disorder** and **Mixed Receptive-Expressive Language Disorder**, there is a language impairment, but it is not associated with the presence of a qualitative impairment in social interaction and restricted, repetitive, and stereotyped patterns of behavior. It is sometimes difficult to determine whether an additional diagnosis of Autistic Disorder is warranted in an individual with **Mental Retardation**, especially if the Mental Retardation is Severe or Profound. An additional diagnosis of Autistic Disorder is reserved for those situations in which there are qualitative deficits in social and communicative skills and the specific behaviors characteristic of Autistic Disorder are present. Motor stereotypies are characteristic of Autistic Disorder; an additional diagnosis of **Stereotypic Movement Disorder** is not given when these are better accounted for as part of the presentation of Autistic Disorder. Symptoms of overactivity and inattention are frequent in Autistic Disorder, but a diagnosis of **Attention-Deficit/Hyperactivity Disorder** is not made if Autistic Disorder is present.

Diagnostic criteria for 299.00 Autistic Disorder

- A. A total of six (or more) items from (1), (2), and (3), with at least two from (1), and one each from (2) and (3):
- (1) qualitative impairment in social interaction, as manifested by at least two of the following:
 - (a) marked impairment in the use of multiple nonverbal behaviors such as eye-to-eye gaze, facial expression, body postures, and gestures to regulate social interaction
 - (b) failure to develop peer relationships appropriate to developmental level
 - (c) a lack of spontaneous seeking to share enjoyment, interests, or achievements with other people (e.g., by a lack of showing, bringing, or pointing out objects of interest)
 - (d) lack of social or emotional reciprocity
 - (2) qualitative impairments in communication as manifested by at least one of the following:
 - (a) delay in, or total lack of, the development of spoken language (not accompanied by an attempt to compensate through alternative modes of communication such as gesture or mime)
 - (b) in individuals with adequate speech, marked impairment in the ability to initiate or sustain a conversation with others
 - (c) stereotyped and repetitive use of language or idiosyncratic language
 - (d) lack of varied, spontaneous make-believe play or social imitative play appropriate to developmental level
 - (3) restricted repetitive and stereotyped patterns of behavior, interests, and activities, as manifested by at least one of the following:
 - (a) encompassing preoccupation with one or more stereotyped and restricted patterns of interest that is abnormal either in intensity or focus
 - (b) apparently inflexible adherence to specific, nonfunctional routines or rituals
 - (c) stereotyped and repetitive motor mannerisms (e.g., hand or finger flapping or twisting, or complex whole-body movements)
 - (d) persistent preoccupation with parts of objects
- B. Delays or abnormal functioning in at least one of the following areas, with onset prior to age 3 years: (1) social interaction, (2) language as used in social communication, or (3) symbolic or imaginative play.
- C. The disturbance is not better accounted for by Rett's Disorder or Childhood Disintegrative Disorder.
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299.80 Rett's Disorder

Diagnostic Features

The essential feature of Rett's Disorder is the development of multiple specific deficits following a period of normal functioning after birth. Individuals have an apparently normal prenatal and perinatal period (Criterion A1) with normal psychomotor development through the first 5 months of life (Criterion A2). Head circumference at birth is also within normal limits (Criterion A3). Between ages 5 and 48 months, head growth decelerates (Criterion B1). There is a loss of previously acquired purposeful hand skills between ages 5 and 30 months, with the subsequent development of characteristic stereotyped hand movements resembling hand-wringing or hand washing (Criterion B2). Interest in the social environment diminishes in the first few years after the onset of the disorder (Criterion B3), although social interaction may often develop later in the course. Problems develop in the coordination of gait or trunk movements (Criterion B4). There is also severe impairment in expressive and receptive language development, with severe psychomotor retardation (Criterion B5).

Associated Features and Disorders

Rett's Disorder is typically associated with Severe or Profound Mental Retardation, which, if present, should be coded on Axis II. There are no specific laboratory findings associated with the disorder. There may be an increased frequency of EEG abnormalities and seizure disorder in individuals with Rett's Disorder. Nonspecific abnormalities on brain imaging have been reported. Preliminary data suggest that a genetic mutation is the cause of some cases of Rett's Disorder.

Prevalence

Data are limited to mostly case series, and it appears that Rett's Disorder is much less common than Autistic Disorder. This disorder has been reported only in females.

Course

The pattern of developmental regression is highly distinctive. Rett's Disorder has its onset prior to age 4 years, usually in the first or second year of life. The duration of the disorder is lifelong, and the loss of skills is generally persistent and progressive. In most instances, recovery is quite limited, although some very modest developmental gains may be made and interest in social interaction may be observed as individuals enter later childhood or adolescence. The communicative and behavioral difficulties usually remain relatively constant throughout life.

Differential Diagnosis

Periods of developmental regression may be observed in normal development, but these are neither as severe or as prolonged as in Rett's Disorder. For the differential

diagnosis between Rett's Disorder and Autistic Disorder, see p. 74. Rett's Disorder differs from Childhood Disintegrative Disorder and Asperger's Disorder in its characteristic sex ratio, onset, and pattern of deficits. Rett's Disorder has been diagnosed only in females, whereas Childhood Disintegrative Disorder and Asperger's Disorder appear to be more common in males. The onset of symptoms in Rett's Disorder can begin as early as age 5 months, whereas in Childhood Disintegrative Disorder the period of normal development is typically more prolonged (i.e., at least until age 2 years). In Rett's Disorder, there is a characteristic pattern of head growth deceleration, loss of previously acquired purposeful hand skills, and the appearance of poorly coordinated gait or trunk movements. In contrast to Asperger's Disorder, Rett's Disorder is characterized by a severe impairment in expressive and receptive language development.

Diagnostic criteria for 299.80 Rett's Disorder

- A. All of the following:
 - (1) apparently normal prenatal and perinatal development
 - (2) apparently normal psychomotor development through the first 5 months after birth
 - (3) normal head circumference at birth
 - B. Onset of all of the following after the period of normal development:
 - (1) deceleration of head growth between ages 5 and 48 months
 - (2) loss of previously acquired purposeful hand skills between ages 5 and 30 months with the subsequent development of stereotyped hand movements (e.g., hand-wringing or hand washing)
 - (3) loss of social engagement early in the course (although often social interaction develops later)
 - (4) appearance of poorly coordinated gait or trunk movements
 - (5) severely impaired expressive and receptive language development with severe psychomotor retardation
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299.10 Childhood Disintegrative Disorder

Diagnostic Features

The essential feature of Childhood Disintegrative Disorder is a marked regression in multiple areas of functioning following a period of at least 2 years of apparently normal development (Criterion A). Apparently normal development is reflected in age-appropriate verbal and nonverbal communication, social relationships, play, and adaptive behavior. After the first 2 years of life (but before age 10 years), the child has a clinically significant loss of previously acquired skills in at least two of the following areas: expressive or receptive language, social skills or adaptive behavior, bowel or bladder control, play, or motor skills (Criterion B). Most typically, acquired skills are lost in almost all areas.

Individuals with this disorder exhibit the social and communicative deficits and behavioral features generally observed in Autistic Disorder (see p. 70). There is qualitative impairment in social interaction (Criterion C1) and in communication (Criterion C2), and restricted, repetitive, and stereotyped patterns of behavior, interests, and activities (Criterion C3). The disturbance is not better accounted for by another specific Pervasive Developmental Disorder or by Schizophrenia (Criterion D). This condition has also been termed *Heller's syndrome*, *dementia infantilis*, or *disintegrative psychosis*.

Associated Features and Disorders

Childhood Disintegrative Disorder is usually associated with Severe Mental Retardation, which, if present, should be coded on Axis II. Various nonspecific neurological symptoms or signs may be noted. There seems to be an increased frequency of EEG abnormalities and seizure disorder. Although it appears likely that the condition is the result of some insult to the developing central nervous system, no precise mechanism has been identified. The condition is occasionally observed in association with a general medical condition (e.g., metachromatic leukodystrophy, Schilder's disease) that might account for the developmental regression. In most instances, however, extensive investigation does not reveal such a condition. If a neurological or other general medical condition is associated with the disorder, it should be recorded on Axis III. The laboratory findings will reflect any associated general medical conditions.

Prevalence

Epidemiological data are limited, but Childhood Disintegrative Disorder appears to be very rare and much less common than Autistic Disorder, although the condition is likely underdiagnosed. Although initial studies suggested an equal sex ratio, the most recent data suggest that the condition is more common among males.

Course

By definition, Childhood Disintegrative Disorder can only be diagnosed if the symptoms are preceded by at least 2 years of normal development and the onset is prior to age 10 years. When the period of normal development has been quite prolonged (5 or more years), it is particularly important to conduct a thorough physical and neurological examination to assess for the presence of a general medical condition. In most cases, the onset is between ages 3 and 4 years and may be insidious or abrupt. Premonitory signs can include increased activity levels, irritability, and anxiety followed by a loss of speech and other skills. During this time, the child may also lose interest in the environment. Usually the loss of skills reaches a plateau, after which some limited improvement may occur, although improvement is rarely marked. In other instances, especially when the disorder is associated with a progressive neurological condition, the loss of skills is progressive. This disorder follows a continuous course, and in the majority of cases, the duration is lifelong. The social, communicative, and behavioral difficulties remain relatively constant throughout life.

Differential Diagnosis

Periods of regression may be observed in normal development, but these are neither as severe or as prolonged as in Childhood Disintegrative Disorder. Childhood Disintegrative Disorder must be differentiated from **other Pervasive Developmental Disorders**. For the differential diagnosis with **Autistic Disorder**, see p. 74. For the differential diagnosis with **Rett's Disorder**, see p. 76. In contrast to **Asperger's Disorder**, Childhood Disintegrative Disorder is characterized by a clinically significant loss in previously acquired skills and a greater likelihood of Mental Retardation. In Asperger's Disorder, there is no delay in language development and no marked loss of developmental skills.

Childhood Disintegrative Disorder must be differentiated from a **dementia** with onset during infancy or childhood. Dementia occurs as a consequence of the direct physiological effects of a general medical condition (e.g., head trauma), whereas Childhood Disintegrative Disorder typically occurs in the absence of an associated general medical condition.

Diagnostic criteria for 299.10 Childhood Disintegrative Disorder

- A. Apparently normal development for at least the first 2 years after birth as manifested by the presence of age-appropriate verbal and nonverbal communication, social relationships, play, and adaptive behavior.
 - B. Clinically significant loss of previously acquired skills (before age 10 years) in at least two of the following areas:
 - (1) expressive or receptive language
 - (2) social skills or adaptive behavior
 - (3) bowel or bladder control
 - (4) play
 - (5) motor skills
 - C. Abnormalities of functioning in at least two of the following areas:
 - (1) qualitative impairment in social interaction (e.g., impairment in nonverbal behaviors, failure to develop peer relationships, lack of social or emotional reciprocity)
 - (2) qualitative impairments in communication (e.g., delay or lack of spoken language, inability to initiate or sustain a conversation, stereotyped and repetitive use of language, lack of varied make-believe play)
 - (3) restricted, repetitive, and stereotyped patterns of behavior, interests, and activities, including motor stereotypies and mannerisms
 - D. The disturbance is not better accounted for by another specific Pervasive Developmental Disorder or by Schizophrenia.
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299.80 Asperger's Disorder

Diagnostic Features

The essential features of Asperger's Disorder are severe and sustained impairment in social interaction (Criterion A) and the development of restricted, repetitive patterns of behavior, interests, and activities (Criterion B). The disturbance must cause clinically significant impairment in social, occupational, or other important areas of functioning (Criterion C). In contrast to Autistic Disorder, there are no clinically significant delays or deviance in language acquisition (e.g., single non-echoed words are used communicatively by age 2 years, and spontaneous communicative phrases are used by age 3 years) (Criterion D), although more subtle aspects of social communication (e.g., typical give-and-take in conversation) may be affected. In addition, during the first 3 years of life, there are no clinically significant delays in cognitive development as manifested by expressing normal curiosity about the environment or in the acquisition of age-appropriate learning skills and adaptive behaviors (other than in social interaction) (Criterion E). Finally, the criteria are not met for another specific Pervasive Developmental Disorder or for Schizophrenia (Criterion F). This condition is also termed Asperger's syndrome.

The impairment in reciprocal social interaction is gross and sustained. There may be marked impairment in the use of multiple nonverbal behaviors (e.g., eye-to-eye gaze, facial expression, body postures and gestures) to regulate social interaction and communication (Criterion A1). There may be failure to develop peer relationships appropriate to developmental level (Criterion A2) that may take different forms at different ages. Younger individuals may have little or no interest in establishing friendships. Older individuals may have an interest in friendship but lack understanding of the conventions of social interaction. There may be a lack of spontaneous seeking to share enjoyment, interests, or achievements with other people (e.g., not showing, bringing, or pointing out objects they find interesting) (Criterion A3). Lack of social or emotional reciprocity may be present (e.g., not actively participating in simple social play or games, preferring solitary activities, or involving others in activities only as tools or "mechanical" aids) (Criterion A4). Although the social deficit in Asperger's Disorder is severe and is defined in the same way as in Autistic Disorder, the lack of social reciprocity is more typically manifest by an eccentric and one-sided social approach to others (e.g., pursuing a conversational topic regardless of others' reactions) rather than social and emotional indifference.

As in Autistic Disorder, restricted, repetitive patterns of behavior, interests, and activities are present (Criterion B). Often these are primarily manifest in the development of encompassing preoccupations about a circumscribed topic or interest, about which the individual can amass a great deal of facts and information (Criterion B1). These interests and activities are pursued with great intensity often to the exclusion of other activities.

The disturbance must cause clinically significant impairment in social adaptation, which in turn may have a significant impact on self-sufficiency or on occupational or other important areas of functioning (Criterion C). The social deficits and restricted patterns of interests, activities, and behavior are the source of considerable disability.

In contrast to Autistic Disorder, there are no clinically significant delays in early language (e.g., single words are used by age 2, communicative phrases are used by age 3) (Criterion D). Subsequent language may be unusual in terms of the individual's preoccupation with certain topics and his or her verbosity. Difficulties in communication may result from social dysfunction and the failure to appreciate and utilize conventional rules of conversation, failure to appreciate nonverbal cues, and limited capacities for self-monitoring.

Individuals with Asperger's Disorder do not have clinically significant delays in cognitive development or in age-appropriate self-help skills, adaptive behavior (other than in social interaction), and curiosity about the environment in childhood (Criterion E). Because early language and cognitive skills are within normal limits in the first 3 years of life, parents or caregivers are not usually concerned about the child's development during that time, although upon detailed interviewing they may recall unusual behaviors. The child may be described as talking before walking, and indeed parents may believe the child to be precocious (e.g., with a rich or "adult" vocabulary). Although subtle social problems may exist, parents or caregivers often are not concerned until the child begins to attend a preschool or is exposed to same-age children; at this point the child's social difficulties with same-age peers may become apparent.

By definition the diagnosis is not given if the criteria are met for any other specific Pervasive Developmental Disorder or for Schizophrenia (although the diagnoses of Asperger's Disorder and Schizophrenia may coexist if the onset of the Asperger's Disorder clearly preceded the onset of Schizophrenia) (Criterion F).

Associated Features and Disorders

In contrast to Autistic Disorder, Mental Retardation is not usually observed in Asperger's Disorder, although occasional cases in which Mild Mental Retardation is present have been noted (e.g., when the Mental Retardation becomes apparent only in the school years, with no apparent cognitive or language delay in the first years of life). Variability of cognitive functioning may be observed, often with strengths in areas of verbal ability (e.g., vocabulary, rote auditory memory) and weaknesses in nonverbal areas (e.g., visual-motor and visual-spatial skills). Motor clumsiness and awkwardness may be present but usually are relatively mild, although motor difficulties may contribute to peer rejection and social isolation (e.g., inability to participate in group sports). Symptoms of overactivity and inattention are frequent in Asperger's Disorder, and indeed many individuals with this condition receive a diagnosis of Attention-Deficit/Hyperactivity Disorder prior to the diagnosis of Asperger's Disorder. Asperger's Disorder has been reported to be associated with a number of other mental disorders, including Depressive Disorders.

Specific Age and Gender Features

The clinical picture may present differently at different ages. Often the social disability of individuals with the disorder becomes more striking over time. By adolescence some individuals with the disorder may learn to use areas of strength (e.g., rote verbal abilities) to compensate for areas of weakness. Individuals with Asperger's Dis-

order may experience victimization by others; this, and feelings of social isolation and an increasing capacity for self-awareness, may contribute to the development of depression and anxiety in adolescence and young adulthood. The disorder is diagnosed much more frequently (at least five times) in males than in females

Prevalence

Definitive data regarding the prevalence of Asperger's Disorder are lacking.

Course

Asperger's Disorder is a continuous and lifelong disorder. In school-age children, good verbal abilities may, to some extent, mask the severity of the child's social dysfunction and may also mislead caregivers and teachers—that is, caregivers and teachers may focus on the child's good verbal skills but be insufficiently aware of problems in other areas (particularly social adjustment). The child's relatively good verbal skills may also lead teachers and caregivers to erroneously attribute behavioral difficulties to willfulness or stubbornness in the child. Interest in forming social relationships may increase in adolescence as the individuals learn some ways of responding more adaptively to their difficulties—for example, the individual may learn to apply explicit verbal rules or routines in certain stressful situations. Older individuals may have an interest in friendship but lack understanding of the conventions of social interaction and may more likely make relationships with individuals much older or younger than themselves. The prognosis appears significantly better than in Autistic Disorder, as follow-up studies suggest that, as adults, many individuals are capable of gainful employment and personal self-sufficiency.

Familial Pattern

Although the available data are limited, there appears to be an increased frequency of Asperger's Disorder among family members of individuals who have the disorder. There may also be an increased risk for Autistic Disorder as well as more general social difficulties.

Differential Diagnosis

Asperger's Disorder must be distinguished from the other **Pervasive Developmental Disorders**, all of which are characterized by problems in social interaction. It differs from **Autistic Disorder** in several ways. In Autistic Disorder there are, by definition, significant abnormalities in the areas of social interaction, language, and play, whereas in Asperger's Disorder early cognitive and language skills are not delayed significantly. Furthermore, in Autistic Disorder, restricted, repetitive, and stereotyped interests and activities are often characterized by the presence of motor mannerisms, preoccupation with parts of objects, rituals, and marked distress in change, whereas in Asperger's Disorder these are primarily observed in the all-encompassing pursuit of a circumscribed interest involving a topic to which the individual devotes inordinate amounts of time amassing information and facts. Differentiation of the two con-

ditions can be problematic in some cases. In Autistic Disorder, typical social interaction patterns are marked by self-isolation or markedly rigid social approaches, whereas in Asperger's Disorder there may appear to be motivation for approaching others even though this is then done in a highly eccentric, one-sided, verbose, and insensitive manner.

Asperger's Disorder must also be differentiated from Pervasive Developmental Disorders other than Autistic Disorder. **Rett's Disorder** differs from Asperger's Disorder in its characteristic sex ratio and pattern of deficits. Rett's Disorder has been diagnosed only in females, whereas Asperger's Disorder occurs much more frequently in males. In Rett's Disorder, there is a characteristic pattern of head growth deceleration, loss of previously acquired purposeful hand skills, and the appearance of poorly coordinated gait or trunk movements. Rett's Disorder is also associated with marked degrees of Mental Retardation and gross impairments in language and communication.

Asperger's Disorder differs from **Childhood Disintegrative Disorder**, which has a distinctive pattern of developmental regression following at least 2 years of normal development. Children with Childhood Disintegrative Disorder also display marked degrees of Mental Retardation and language impairment. In contrast, in Asperger's Disorder there is no pattern of developmental regression and, by definition, no significant cognitive or language delays.

Schizophrenia of childhood onset usually develops after years of normal, or near normal, development, and characteristic features of the disorder, including hallucinations, delusions, and disorganized speech, are present. In **Selective Mutism**, the child usually exhibits appropriate communication skills in certain contexts and does not have the severe impairment in social interaction and the restricted patterns of behavior associated with Asperger's Disorder. Conversely, individuals with Asperger's Disorder are typically verbose. In **Expressive Language Disorder** and **Mixed Receptive-Expressive Language Disorder**, there is language impairment but no associated qualitative impairment in social interaction and restricted, repetitive, and stereotyped patterns of behavior. Some individuals with Asperger's Disorder may exhibit behavioral patterns suggesting **Obsessive-Compulsive Disorder**, although special clinical attention should be given to the differentiation between preoccupations and activities in Asperger's Disorder and obsessions and compulsions in Obsessive-Compulsive Disorder. In Asperger's Disorder these interests are the source of some apparent pleasure or comfort, whereas in Obsessive-Compulsive Disorder they are the source of anxiety. Furthermore, Obsessive-Compulsive Disorder is typically not associated with the level of impairment in social interaction and social communication seen in Asperger's Disorder.

The relationship between Asperger's Disorder and **Schizoid Personality Disorder** is unclear. In general, the social difficulties in Asperger's Disorder are more severe and of earlier onset. Although some individuals with Asperger's Disorder may experience heightened and debilitating anxiety in social settings as in **Social Phobia** or **other Anxiety Disorders**, the latter conditions are not characterized by pervasive impairments in social development or by the circumscribed interests typical of Asperger's Disorder. Asperger's Disorder must be distinguished from **normal social awkwardness** and **normal age-appropriate interests and hobbies**. In Asperger's Disorder, the social deficits are quite severe and the preoccupations are all-encompassing and interfere with the acquisition of basic skills.

Diagnostic criteria for 299.80 Asperger's Disorder

- A. Qualitative impairment in social interaction, as manifested by at least two of the following:
 - (1) marked impairment in the use of multiple nonverbal behaviors such as eye-to-eye gaze, facial expression, body postures, and gestures to regulate social interaction
 - (2) failure to develop peer relationships appropriate to developmental level
 - (3) a lack of spontaneous seeking to share enjoyment, interests, or achievements with other people (e.g., by a lack of showing, bringing, or pointing out objects of interest to other people)
 - (4) lack of social or emotional reciprocity
 - B. Restricted repetitive and stereotyped patterns of behavior, interests, and activities, as manifested by at least one of the following:
 - (1) encompassing preoccupation with one or more stereotyped and restricted patterns of interest that is abnormal either in intensity or focus
 - (2) apparently inflexible adherence to specific, nonfunctional routines or rituals
 - (3) stereotyped and repetitive motor mannerisms (e.g., hand or finger flapping or twisting, or complex whole-body movements)
 - (4) persistent preoccupation with parts of objects
 - C. The disturbance causes clinically significant impairment in social, occupational, or other important areas of functioning.
 - D. There is no clinically significant general delay in language (e.g., single words used by age 2 years, communicative phrases used by age 3 years).
 - E. There is no clinically significant delay in cognitive development or in the development of age-appropriate self-help skills, adaptive behavior (other than in social interaction), and curiosity about the environment in childhood.
 - F. Criteria are not met for another specific Pervasive Developmental Disorder or Schizophrenia.
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299.80 Pervasive Developmental Disorder Not Otherwise Specified (Including Atypical Autism)

This category should be used when there is a severe and pervasive impairment in the development of reciprocal social interaction associated with impairment in either verbal or nonverbal communication skills or with the presence of stereotyped behavior, interests, and activities, but the criteria are not met for a specific Pervasive Developmental Disorder, Schizophrenia, Schizotypal Personality Disorder, or Avoidant Personality Disorder. For example, this category includes "atypical autism"—presentations that do not meet the criteria for Autistic Disorder because of late age at onset, atypical symptomatology, or subthreshold symptomatology, or all of these.

Attention-Deficit and Disruptive Behavior Disorders

Attention-Deficit/Hyperactivity Disorder

Diagnostic Features

The essential feature of Attention-Deficit/Hyperactivity Disorder is a persistent pattern of inattention and/or hyperactivity-impulsivity that is more frequently displayed and more severe than is typically observed in individuals at a comparable level of development (Criterion A). Some hyperactive-impulsive or inattentive symptoms that cause impairment must have been present before age 7 years, although many individuals are diagnosed after the symptoms have been present for a number of years, especially in the case of individuals with the Predominantly Inattentive Type (Criterion B). Some impairment from the symptoms must be present in at least two settings (e.g., at home and at school or work) (Criterion C). There must be clear evidence of interference with developmentally appropriate social, academic, or occupational functioning (Criterion D). The disturbance does not occur exclusively during the course of a Pervasive Developmental Disorder, Schizophrenia, or other Psychotic Disorder and is not better accounted for by another mental disorder (e.g., a Mood Disorder, Anxiety Disorder, Dissociative Disorder, or Personality Disorder) (Criterion E).

Inattention may be manifest in academic, occupational, or social situations. Individuals with this disorder may fail to give close attention to details or may make careless mistakes in schoolwork or other tasks (Criterion A1a). Work is often messy and performed carelessly and without considered thought. Individuals often have difficulty sustaining attention in tasks or play activities and often find it hard to persist with tasks until completion (Criterion A1b). They often appear as if their mind is elsewhere or as if they are not listening or did not hear what has just been said (Criterion A1c). There may be frequent shifts from one uncompleted activity to another. Individuals diagnosed with this disorder may begin a task, move on to another, then turn to yet something else, prior to completing any one task. They often do not follow through on requests or instructions and fail to complete schoolwork, chores, or other duties (Criterion A1d). Failure to complete tasks should be considered in making this diagnosis only if it is due to inattention as opposed to other possible reasons (e.g., failure to understand instructions, defiance). These individuals often have difficulties organizing tasks and activities (Criterion A1e). Tasks that require sustained mental effort are experienced as unpleasant and markedly aversive. As a result, these individuals typically avoid or have a strong dislike for activities that demand sustained self-application and mental effort or that require organizational demands or close concentration (e.g., homework or paperwork) (Criterion A1f). This avoidance must be due to the person's difficulties with attention and not due to a primary oppositional attitude, although secondary oppositionalism may also occur. Work habits are often disorganized and the materials necessary for doing the task are often scattered, lost, or carelessly handled and damaged (Criterion A1g). Individuals with this disorder

der are easily distracted by irrelevant stimuli and frequently interrupt ongoing tasks to attend to trivial noises or events that are usually and easily ignored by others (e.g., a car honking, a background conversation) (Criterion A1h). They are often forgetful in daily activities (e.g., missing appointments, forgetting to bring lunch) (Criterion A1i). In social situations, inattention may be expressed as frequent shifts in conversation, not listening to others, not keeping one's mind on conversations, and not following details or rules of games or activities.

Hyperactivity may be manifested by fidgetiness or squirming in one's seat (Criterion A2a), by not remaining seated when expected to do so (Criterion A2b), by excessive running or climbing in situations where it is inappropriate (Criterion A2c), by having difficulty playing or engaging quietly in leisure activities (Criterion A2d), by appearing to be often "on the go" or as if "driven by a motor" (Criterion A2e), or by talking excessively (Criterion A2f). Hyperactivity may vary with the individual's age and developmental level, and the diagnosis should be made cautiously in young children. Toddlers and preschoolers with this disorder differ from normally active young children by being constantly on the go and into everything; they dart back and forth, are "out of the door before their coat is on," jump or climb on furniture, run through the house, and have difficulty participating in sedentary group activities in preschool classes (e.g., listening to a story). School-age children display similar behaviors but usually with less frequency or intensity than toddlers and preschoolers. They have difficulty remaining seated, get up frequently, and squirm in, or hang on to the edge of, their seat. They fidget with objects, tap their hands, and shake their feet or legs excessively. They often get up from the table during meals, while watching television, or while doing homework; they talk excessively; and they make excessive noise during quiet activities. In adolescents and adults, symptoms of hyperactivity take the form of feelings of restlessness and difficulty engaging in quiet sedentary activities.

Impulsivity manifests itself as impatience, difficulty in delaying responses, blurting out answers before questions have been completed (Criterion A2g), difficulty awaiting one's turn (Criterion A2h), and frequently interrupting or intruding on others to the point of causing difficulties in social, academic, or occupational settings (Criterion A2i). Others may complain that they cannot get a word in edgewise. Individuals with this disorder typically make comments out of turn, fail to listen to directions, initiate conversations at inappropriate times, interrupt others excessively, intrude on others, grab objects from others, touch things they are not supposed to touch, and clown around. Impulsivity may lead to accidents (e.g., knocking over objects, banging into people, grabbing a hot pan) and to engagement in potentially dangerous activities without consideration of possible consequences (e.g., repeatedly climbing to precarious positions or riding a skateboard over extremely rough terrain).

Attentional and behavioral manifestations usually appear in multiple contexts, including home, school, work, and social situations. To make the diagnosis, some impairment must be present in at least two settings (Criterion C). It is very unusual for an individual to display the same level of dysfunction in all settings or within the same setting at all times. Symptoms typically worsen in situations that require sustained attention or mental effort or that lack intrinsic appeal or novelty (e.g., listening to classroom teachers, doing class assignments, listening to or reading lengthy materials, or working on monotonous, repetitive tasks). Signs of the disorder may be minimal or absent when the person is receiving frequent rewards for appropriate

behavior, is under close supervision, is in a novel setting, is engaged in especially interesting activities, or is in a one-to-one situation (e.g., the clinician's office). The symptoms are more likely to occur in group situations (e.g., in playgroups, classrooms, or work environments). The clinician should therefore gather information from multiple sources (e.g., parents, teachers) and inquire about the individual's behavior in a variety of situations within each setting (e.g., doing homework, having meals).

Subtypes

Although many individuals present with symptoms of both inattention and hyperactivity-impulsivity, there are individuals in whom one or the other pattern is predominant. The appropriate subtype (for a current diagnosis) should be indicated based on the predominant symptom pattern for the past 6 months.

314.01 Attention-Deficit/Hyperactivity Disorder, Combined Type. This subtype should be used if six (or more) symptoms of inattention and six (or more) symptoms of hyperactivity-impulsivity have persisted for at least 6 months. Most children and adolescents with the disorder have the Combined Type. It is not known whether the same is true of adults with the disorder.

314.00 Attention-Deficit/Hyperactivity Disorder, Predominantly Inattentive Type. This subtype should be used if six (or more) symptoms of inattention (but fewer than six symptoms of hyperactivity-impulsivity) have persisted for at least 6 months. Hyperactivity may still be a significant clinical feature in many such cases, whereas other cases are more purely inattentive.

314.01 Attention-Deficit/Hyperactivity Disorder, Predominantly Hyperactive-Impulsive Type. This subtype should be used if six (or more) symptoms of hyperactivity-impulsivity (but fewer than six symptoms of inattention) have persisted for at least 6 months. Inattention may often still be a significant clinical feature in such cases.

Recording Procedures

Individuals who at an earlier stage of the disorder had the Predominantly Inattentive Type or the Predominantly Hyperactive-Impulsive Type may go on to develop the Combined Type and vice versa. The appropriate subtype (for a current diagnosis) should be indicated on the basis of the predominant symptom pattern for the past 6 months. If clinically significant symptoms remain but criteria are no longer met for any of the subtypes, the appropriate diagnosis is Attention-Deficit/Hyperactivity Disorder, In Partial Remission. When an individual's symptoms do not currently meet full criteria for the disorder and it is unclear whether criteria for the disorder have previously been met, Attention-Deficit/Hyperactivity Disorder Not Otherwise Specified should be diagnosed.

Associated Features and Disorders

Associated descriptive features and mental disorders. Associated features vary depending on age and developmental stage and may include low frustration tolerance, temper outbursts, bossiness, stubbornness, excessive and frequent insistence

that requests be met, mood lability, demoralization, dysphoria, rejection by peers, and poor self-esteem. Academic achievement is often markedly impaired and devalued, typically leading to conflict with the family and with school authorities. Inadequate self-application to tasks that require sustained effort is often interpreted by others as indicating laziness, a poor sense of responsibility, and oppositional behavior. Family relationships are often characterized by resentment and antagonism, especially because variability in the individual's symptomatic status often leads others to believe that all the troublesome behavior is willful. Family discord and negative parent-child interactions are often present. Such negative interactions often diminish with successful treatment. On average, individuals with Attention-Deficit/Hyperactivity Disorder obtain less schooling than their peers and have poorer vocational achievement. Also, on average, intellectual level, as assessed by individual IQ tests, is several points lower in children with this disorder compared with peers. At the same time, great variability in IQ is evidenced: individuals with Attention-Deficit/Hyperactivity Disorder may show intellectual development in the above-average or gifted range. In its severe form, the disorder is markedly impairing, affecting social, familial, and scholastic adjustment. All three subtypes are associated with significant impairment. Academic deficits and school-related problems tend to be most pronounced in the types marked by inattention (Predominantly Inattentive and Combined Types), whereas peer rejection and, to a lesser extent, accidental injury are most salient in the types marked by hyperactivity and impulsivity (Predominantly Hyperactive-Impulsive and Combined Types). Individuals with the Predominantly Inattentive Type tend to be socially passive and appear to be neglected, rather than rejected, by peers.

A substantial proportion (approximately half) of clinic-referred children with Attention-Deficit/Hyperactivity Disorder also have Oppositional Defiant Disorder or Conduct Disorder. The rates of co-occurrence of Attention-Deficit/Hyperactivity Disorder with these other Disruptive Behavior Disorders are higher than with other mental disorders, and this co-occurrence is most likely in the two subtypes marked by hyperactivity-impulsivity (Hyperactive-Impulsive and Combined Types). Other associated disorders include Mood Disorders, Anxiety Disorders, Learning Disorders, and Communication Disorders in children with Attention-Deficit/Hyperactivity Disorder. Although Attention-Deficit/Hyperactivity Disorder appears in at least 50% of clinic-referred individuals with Tourette's Disorder, most individuals with Attention-Deficit/Hyperactivity Disorder do not have accompanying Tourette's Disorder. When the two disorders coexist, the onset of the Attention-Deficit/Hyperactivity Disorder often precedes the onset of the Tourette's Disorder.

There may be a history of child abuse or neglect, multiple foster placements, neurotoxin exposure (e.g., lead poisoning), infections (e.g., encephalitis), drug exposure in utero, or Mental Retardation. Although low birth weight may sometimes be associated with Attention-Deficit/Hyperactivity Disorder, most children with low birth weight do not develop Attention-Deficit/Hyperactivity Disorder, and most children with Attention-Deficit/Hyperactivity Disorder do not have a history of low birth weight.

Associated laboratory findings. There are no laboratory tests, neurological assessments, or attentional assessments that have been established as diagnostic in the clin-

ical assessment of Attention-Deficit/Hyperactivity Disorder. Tests that require effortful mental processing have been noted to be abnormal in groups of individuals with Attention-Deficit/Hyperactivity Disorder compared with peers, but these tests are not of demonstrated utility when one is trying to determine whether a particular individual has the disorder. It is not yet known what fundamental cognitive deficits are responsible for such group differences.

Associated physical examination findings and general medical conditions.

There are no specific physical features associated with Attention-Deficit/Hyperactivity Disorder, although minor physical anomalies (e.g., hypertelorism, highly arched palate, low-set ears) may occur at a higher rate than in the general population. There may also be a higher rate of accidental physical injury.

Specific Culture, Age, and Gender Features

Attention-Deficit/Hyperactivity Disorder is known to occur in various cultures, with variations in reported prevalence among Western countries probably arising more from different diagnostic practices than from differences in clinical presentation.

It is difficult to establish this diagnosis in children younger than age 4 or 5 years, because their characteristic behavior is much more variable than that of older children and may include features that are similar to symptoms of Attention-Deficit/Hyperactivity Disorder. Furthermore, symptoms of inattention in toddlers or preschool children are often not readily observed because young children typically experience few demands for sustained attention. However, even the attention of toddlers can be held in a variety of situations (e.g., the average 2- or 3-year-old child can typically sit with an adult looking through picture books). Young children with Attention-Deficit/Hyperactivity Disorder move excessively and typically are difficult to contain. Inquiring about a wide variety of behaviors in a young child may be helpful in ensuring that a full clinical picture has been obtained. Substantial impairment has been demonstrated in preschool-age children with Attention-Deficit/Hyperactivity Disorder. In school-age children, symptoms of inattention affect classroom work and academic performance. Impulsive symptoms may also lead to the breaking of familial, interpersonal, and educational rules. Symptoms of Attention-Deficit/Hyperactivity Disorder are typically at their most prominent during the elementary grades. As children mature, symptoms usually become less conspicuous. By late childhood and early adolescence, signs of excessive gross motor activity (e.g., excessive running and climbing, not remaining seated) are less common, and hyperactivity symptoms may be confined to fidgetiness or an inner feeling of jitteriness or restlessness. In adulthood, restlessness may lead to difficulty in participating in sedentary activities and to avoiding pastimes or occupations that provide limited opportunity for spontaneous movement (e.g., desk jobs). Social dysfunction in adults appears to be especially likely in those who had additional concurrent diagnoses in childhood. Caution should be exercised in making the diagnosis of Attention-Deficit/Hyperactivity Disorder in adults solely on the basis of the adult's recall of being inattentive or hyperactive as a child, because the validity of such retrospective data is often problematic. Although supporting information may not always be available, corroborating information from other informants (including prior school records) is helpful for improving the accuracy of the diagnosis.

The disorder is more frequent in males than in females, with male-to-female ratios ranging from 2:1 to 9:1, depending on the type (i.e., the Predominantly Inattentive Type may have a gender ratio that is less pronounced) and setting (i.e., clinic-referred children are more likely to be male).

Prevalence

The prevalence of Attention-Deficit/Hyperactivity Disorder has been estimated at 3%–7% in school-age children. These reported rates vary depending on the nature of the population sampled and the method of ascertainment. Data on prevalence in adolescence and adulthood are limited. Evidence suggests that the prevalence of Attention-Deficit/Hyperactivity Disorder as defined in DSM-IV may be somewhat greater than the prevalence of the disorder based on DSM-III-R criteria because of the inclusion of the Predominantly Hyperactive-Impulsive and Predominantly Inattentive Types (which would have been diagnosed as Attention-Deficit Hyperactivity Disorder Not Otherwise Specified in DSM-III-R).

Course

Most parents first observe excessive motor activity when the children are toddlers, frequently coinciding with the development of independent locomotion. However, because many overactive toddlers will not go on to develop Attention-Deficit/Hyperactivity Disorder, special attention should be paid to differentiating normal overactivity from the hyperactivity characteristic of Attention-Deficit/Hyperactivity Disorder before making this diagnosis in early years. Usually, the disorder is first diagnosed during elementary school years, when school adjustment is compromised. Some children with the Predominantly Inattentive Type may not come to clinical attention until late childhood. In the majority of cases seen in clinical settings, the disorder is relatively stable through early adolescence. In most individuals, symptoms (particularly motor hyperactivity) attenuate during late adolescence and adulthood, although a minority experience the full complement of symptoms of Attention-Deficit/Hyperactivity Disorder into mid-adulthood. Other adults may retain only some of the symptoms, in which case the diagnosis of Attention-Deficit/Hyperactivity Disorder, In Partial Remission, should be used. The latter diagnosis applies to individuals who no longer have the full disorder but still retain some symptoms that cause functional impairment.

Familial Pattern

Attention-Deficit/Hyperactivity Disorder has been found to be more common in the first-degree biological relatives of children with Attention-Deficit/Hyperactivity Disorder than in the general population. Considerable evidence attests to the strong influence of genetic factors on levels of hyperactivity, impulsivity, and inattention as measured dimensionally. However, family, school, and peer influences are also crucial in determining the extent of impairments and comorbidity. Studies also suggest that there is a higher prevalence of Mood and Anxiety Disorders, Learning Disorders, Substance-Related Disorders, and Antisocial Personality Disorder in family members of individuals with Attention-Deficit/Hyperactivity Disorder.

Differential Diagnosis

In early childhood, it may be difficult to distinguish symptoms of Attention-Deficit/Hyperactivity Disorder from **age-appropriate behaviors in active children** (e.g., running around or being noisy).

Symptoms of inattention are common among children with low IQ who are placed in academic settings that are inappropriate to their intellectual ability. These behaviors must be distinguished from similar signs in children with Attention-Deficit/Hyperactivity Disorder. In children with **Mental Retardation**, an additional diagnosis of Attention-Deficit/Hyperactivity Disorder should be made only if the symptoms of inattention or hyperactivity are excessive for the child's mental age. Inattention in the classroom may also occur when children with high intelligence are placed in academically **understimulating environments**. Attention-Deficit/Hyperactivity Disorder must also be distinguished from difficulty in goal-directed behavior in children from inadequate, disorganized, or chaotic environments. Thorough histories of symptom pattern obtained from multiple informants (e.g., baby-sitters, grandparents, or parents of playmates) are helpful in providing a confluence of observations concerning the child's inattention, hyperactivity, and capacity for developmentally appropriate self-regulation in various settings.

Individuals with **oppositional behavior** may resist work or school tasks that require self-application because of an unwillingness to conform to others' demands. These symptoms must be differentiated from the avoidance of school tasks seen in individuals with Attention-Deficit/Hyperactivity Disorder. Complicating the differential diagnosis is the fact that some individuals with Attention-Deficit/Hyperactivity Disorder develop secondary oppositional attitudes toward such tasks and devalue their importance, often as a rationalization for their failure.

The increased motor activity that may occur in Attention-Deficit/Hyperactivity Disorder must be distinguished from the repetitive motor behavior that characterizes **Stereotypic Movement Disorder**. In Stereotypic Movement Disorder, the motor behavior is generally focused and fixed (e.g., body rocking, self-biting), whereas the fidgetiness and restlessness in Attention-Deficit/Hyperactivity Disorder are more typically generalized. Furthermore, individuals with Stereotypic Movement Disorder are not generally overactive; aside from the stereotypy, they may be underactive.

Attention-Deficit/Hyperactivity Disorder is not diagnosed if the symptoms are better accounted for by **another mental disorder** (e.g., Mood Disorder [especially Bipolar Disorder], Anxiety Disorder, Dissociative Disorder, Personality Disorder, Personality Change Due to a General Medical Condition, or a Substance-Related Disorder). In all these disorders, the symptoms of inattention typically have an onset after age 7 years, and the childhood history of school adjustment generally is not characterized by disruptive behavior or teacher complaints concerning inattentive, hyperactive, or impulsive behavior. When a Mood Disorder or Anxiety Disorder co-occurs with Attention-Deficit/Hyperactivity Disorder, each should be diagnosed. Attention-Deficit/Hyperactivity Disorder is not diagnosed if the symptoms of inattention and hyperactivity occur exclusively during the course of a **Pervasive Developmental Disorder** or a **Psychotic Disorder**. Symptoms of inattention, hyperactivity, or impulsivity related to the use of medication (e.g., bronchodilators, isoniazid, akathisia from neuroleptics) in children before age 7 years are not diagnosed as Attention-Deficit/Hyperactivity Disorder but instead are diagnosed as **Other Substance-Related Disorder Not Otherwise Specified**.

Diagnostic criteria for Attention-Deficit/Hyperactivity Disorder

A. Either (1) or (2):

- (1) six (or more) of the following symptoms of **inattention** have persisted for at least 6 months to a degree that is maladaptive and inconsistent with developmental level:

Inattention

- (a) often fails to give close attention to details or makes careless mistakes in schoolwork, work, or other activities
- (b) often has difficulty sustaining attention in tasks or play activities
- (c) often does not seem to listen when spoken to directly
- (d) often does not follow through on instructions and fails to finish schoolwork, chores, or duties in the workplace (not due to oppositional behavior or failure to understand instructions)
- (e) often has difficulty organizing tasks and activities
- (f) often avoids, dislikes, or is reluctant to engage in tasks that require sustained mental effort (such as schoolwork or homework)
- (g) often loses things necessary for tasks or activities (e.g., toys, school assignments, pencils, books, or tools)
- (h) is often easily distracted by extraneous stimuli
- (i) is often forgetful in daily activities

- (2) six (or more) of the following symptoms of **hyperactivity-impulsivity** have persisted for at least 6 months to a degree that is maladaptive and inconsistent with developmental level:

Hyperactivity

- (a) often fidgets with hands or feet or squirms in seat
- (b) often leaves seat in classroom or in other situations in which remaining seated is expected
- (c) often runs about or climbs excessively in situations in which it is inappropriate (in adolescents or adults, may be limited to subjective feelings of restlessness)
- (d) often has difficulty playing or engaging in leisure activities quietly
- (e) is often "on the go" or often acts as if "driven by a motor"
- (f) often talks excessively

Impulsivity

- (g) often blurts out answers before questions have been completed
- (h) often has difficulty awaiting turn
- (i) often interrupts or intrudes on others (e.g., butts into conversations or games)

- B. Some hyperactive-impulsive or inattentive symptoms that caused impairment were present before age 7 years.
- C. Some impairment from the symptoms is present in two or more settings (e.g., at school [or work] and at home).

Diagnostic criteria for Attention-Deficit/Hyperactivity Disorder (*continued*)

- D. There must be clear evidence of clinically significant impairment in social, academic, or occupational functioning.
- E. The symptoms do not occur exclusively during the course of a Pervasive Developmental Disorder, Schizophrenia, or other Psychotic Disorder and are not better accounted for by another mental disorder (e.g., Mood Disorder, Anxiety Disorder, Dissociative Disorder, or a Personality Disorder).

Code based on type:

314.01 Attention-Deficit/Hyperactivity Disorder, Combined Type:

if both Criteria A1 and A2 are met for the past 6 months

314.00 Attention-Deficit/Hyperactivity Disorder, Predominantly

Inattentive Type: if Criterion A1 is met but Criterion A2 is not met for the past 6 months

314.01 Attention-Deficit/Hyperactivity Disorder, Predominantly

Hyperactive-Impulsive Type: if Criterion A2 is met but Criterion A1 is not met for the past 6 months

Coding note: For individuals (especially adolescents and adults) who currently have symptoms that no longer meet full criteria, "In Partial Remission" should be specified.

314.9 Attention-Deficit/Hyperactivity Disorder Not Otherwise Specified

This category is for disorders with prominent symptoms of inattention or hyperactivity-impulsivity that do not meet criteria for Attention-Deficit/Hyperactivity Disorder. Examples include

1. Individuals whose symptoms and impairment meet the criteria for Attention-Deficit/Hyperactivity Disorder, Predominantly Inattentive Type but whose age at onset is 7 years or after
2. Individuals with clinically significant impairment who present with inattention and whose symptom pattern does not meet the full criteria for the disorder but have a behavioral pattern marked by sluggishness, daydreaming, and hypoactivity

Conduct Disorder

Diagnostic Features

The essential feature of Conduct Disorder is a repetitive and persistent pattern of behavior in which the basic rights of others or major age-appropriate societal norms or rules are violated (Criterion A). These behaviors fall into four main groupings:

aggressive conduct that causes or threatens physical harm to other people or animals (Criteria A1–A7), nonaggressive conduct that causes property loss or damage (Criteria A8–A9), deceitfulness or theft (Criteria A10–A12), and serious violations of rules (Criteria A13–A15). Three (or more) characteristic behaviors must have been present during the past 12 months, with at least one behavior present in the past 6 months. The disturbance in behavior causes clinically significant impairment in social, academic, or occupational functioning (Criterion B). Conduct Disorder may be diagnosed in individuals who are older than age 18 years, but only if the criteria for Antisocial Personality Disorder are not met (Criterion C). The behavior pattern is usually present in a variety of settings such as home, school, or the community. Because individuals with Conduct Disorder are likely to minimize their conduct problems, the clinician often must rely on additional informants. However, the informant's knowledge of the child's conduct problems may be limited by inadequate supervision or by the child's not having revealed them.

Children or adolescents with this disorder often initiate aggressive behavior and react aggressively to others. They may display bullying, threatening, or intimidating behavior (Criterion A1); initiate frequent physical fights (Criterion A2); use a weapon that can cause serious physical harm (e.g., a bat, brick, broken bottle, knife, or gun) (Criterion A3); be physically cruel to people (Criterion A4) or animals (Criterion A5); steal while confronting a victim (e.g., mugging, purse snatching, extortion, or armed robbery) (Criterion A6); or force someone into sexual activity (Criterion A7). Physical violence may take the form of rape, assault, or, in rare cases, homicide.

Deliberate destruction of others' property may include deliberate fire setting with the intention of causing serious damage (Criterion A8) or deliberately destroying other people's property in other ways (e.g., smashing car windows, school vandalism) (Criterion A9).

Acts of deceitfulness or theft may include breaking into someone else's house, building, or car (Criterion A10); frequently lying or breaking promises to obtain goods or favors or to avoid debts or obligations (e.g., "conning" other people) (Criterion A11); or stealing items of nontrivial value without confronting the victim (e.g., shoplifting, forgery) (Criterion A12).

There may also be serious violations of rules (e.g., school, parental) by individuals with this disorder. Children with this disorder often have a pattern, beginning before age 13 years, of staying out late at night despite parental prohibitions (Criterion A13). There may be a pattern of running away from home overnight (Criterion A14). To be considered a symptom of Conduct Disorder, the running away must have occurred at least twice (or only once if the individual did not return for a lengthy period). Run-away episodes that occur as a direct consequence of physical or sexual abuse do not typically qualify for this criterion. Children with this disorder may often be truant from school, beginning prior to age 13 years (Criterion A15). In older individuals, this behavior is manifested by often being absent from work without good reason.

Subtypes

Two subtypes of Conduct Disorder are provided based on the age at onset of the disorder (i.e., Childhood-Onset Type and Adolescent-Onset Type). The subtypes differ in regard to the characteristic nature of the presenting conduct problems, develop-

mental course and prognosis, and gender ratio. Both subtypes can occur in a mild, moderate, or severe form. In assessing the age at onset, information should preferably be obtained from the youth and from caregiver(s). Because many of the behaviors may be concealed, caregivers may underreport symptoms and overestimate the age at onset.

312.81 Childhood-Onset Type. This subtype is defined by the onset of at least one criterion characteristic of Conduct Disorder prior to age 10 years. Individuals with Childhood-Onset Type are usually male, frequently display physical aggression toward others, have disturbed peer relationships, may have had Oppositional Defiant Disorder during early childhood, and usually have symptoms that meet full criteria for Conduct Disorder prior to puberty. Many children with this subtype also have concurrent Attention-Deficit/Hyperactivity Disorder. Individuals with Childhood-Onset Type are more likely to have persistent Conduct Disorder and to develop adult Antisocial Personality Disorder than are those with Adolescent-Onset Type.

312.82 Adolescent-Onset Type. This subtype is defined by the absence of any criteria characteristic of Conduct Disorder prior to age 10 years. Compared with those with the Childhood-Onset Type, these individuals are less likely to display aggressive behaviors and tend to have more normative peer relationships (although they often display conduct problems in the company of others). These individuals are less likely to have persistent Conduct Disorder or to develop adult Antisocial Personality Disorder. The ratio of males to females with Conduct Disorder is lower for the Adolescent-Onset Type than for the Childhood-Onset Type.

312.89 Unspecified Onset. This subtype is used if the age at onset of Conduct Disorder is unknown.

Severity Specifiers

Mild. Few if any conduct problems in excess of those required to make the diagnosis are present, and conduct problems cause relatively minor harm to others (e.g., lying, truancy, staying out after dark without permission).

Moderate. The number of conduct problems and the effect on others are intermediate between "mild" and "severe" (e.g., stealing without confronting a victim, vandalism).

Severe. Many conduct problems in excess of those required to make the diagnosis are present, or conduct problems cause considerable harm to others (e.g., forced sex, physical cruelty, use of a weapon, stealing while confronting a victim, breaking and entering).

Associated Features and Disorders

Associated descriptive features and mental disorders. Individuals with Conduct Disorder may have little empathy and little concern for the feelings, wishes, and well-being of others. Especially in ambiguous situations, aggressive individuals with this disorder frequently misperceive the intentions of others as more hostile and threatening than is the case and respond with aggression that they then feel is reasonable and

justified. They may be callous and lack appropriate feelings of guilt or remorse. It can be difficult to evaluate whether displayed remorse is genuine because some of these individuals learn that expressing guilt may reduce or prevent punishment. Individuals with this disorder may readily inform on their companions and try to blame others for their own misdeeds. Self-esteem may be low, despite a projected image of "toughness." For other individuals, measured self-esteem may be overly inflated. Poor frustration tolerance, irritability, temper outbursts, and recklessness are frequent associated features. Accident rates appear to be higher in individuals with Conduct Disorder than in those without it.

Conduct Disorder is often associated with an early onset of sexual behavior, drinking, smoking, use of illegal substances, and reckless and risk-taking acts. Illegal drug use may increase the risk that Conduct Disorder will persist. Conduct Disorder behaviors may lead to school suspension or expulsion, problems in work adjustment, legal difficulties, sexually transmitted diseases, unplanned pregnancy, and physical injury from accidents or fights. These problems may preclude attendance in ordinary schools or living in a parental or foster home. Suicidal ideation, suicide attempts, and completed suicide occur at a higher-than-expected rate. Conduct Disorder may be associated with lower-than-average intelligence, particularly with regard to verbal IQ. Academic achievement, particularly in reading and other verbal skills, is often below the level expected on the basis of age and intelligence and may justify the additional diagnosis of a Learning or Communication Disorder. Attention-Deficit/Hyperactivity Disorder is common in children with Conduct Disorder. Conduct Disorder may also be associated with one or more of the following mental disorders: Learning Disorders, Anxiety Disorders, Mood Disorders, and Substance-Related Disorders. The following factors may predispose the individual to the development of Conduct Disorder: parental rejection and neglect, difficult infant temperament, inconsistent child-rearing practices with harsh discipline, physical or sexual abuse, lack of supervision, early institutional living, frequent changes of caregivers, large family size, history of maternal smoking during pregnancy, peer rejection, association with a delinquent peer group, neighborhood exposure to violence, and certain kinds of familial psychopathology (e.g., Antisocial Personality Disorder, Substance Dependence or Abuse).

Associated laboratory findings. In some studies, lower heart rate and lower skin conductance have been noted in individuals with Conduct Disorder compared with those without the disorder. However, levels of physiological arousal are not diagnostic of the disorder.

Specific Culture, Age, and Gender Features

Concerns have been raised that the Conduct Disorder diagnosis may at times be misapplied to individuals in settings where patterns of undesirable behavior are sometimes viewed as protective (e.g., threatening, impoverished, high-crime). Consistent with the DSM-IV definition of mental disorder, the Conduct Disorder diagnosis should be applied only when the behavior in question is symptomatic of an underlying dysfunction within the individual and not simply a reaction to the immediate social context. Moreover, immigrant youth from war-ravaged countries who have a history of aggressive behaviors that may have been necessary for their survival in that

context would not necessarily warrant a diagnosis of Conduct Disorder. It may be helpful for the clinician to consider the social and economic context in which the undesirable behaviors have occurred.

Symptoms of the disorder vary with age as the individual develops increased physical strength, cognitive abilities, and sexual maturity. Less severe behaviors (e.g., lying, shoplifting, physical fighting) tend to emerge first, whereas others (e.g., burglary) tend to emerge later. Typically, the most severe conduct problems (e.g., rape, theft while confronting a victim) tend to emerge last. However, there are wide differences among individuals, with some engaging in the more damaging behaviors at an early age (which is predictive of a worse prognosis).

Conduct Disorder, especially the Childhood-Onset Type, is more common in males. Gender differences are also found in specific types of conduct problems. Males with a diagnosis of Conduct Disorder frequently exhibit fighting, stealing, vandalism, and school discipline problems. Females with a diagnosis of Conduct Disorder are more likely to exhibit lying, truancy, running away, substance use, and prostitution. Whereas confrontational aggression is more often displayed by males, females tend to use more nonconfrontational behaviors.

Prevalence

The prevalence of Conduct Disorder appears to have increased over the last decades and may be higher in urban than in rural settings. Rates vary widely depending on the nature of the population sampled and methods of ascertainment. General population studies report rates ranging from less than 1% to more than 10%. Prevalence rates are higher among males than females. Conduct Disorder is one of the most frequently diagnosed conditions in outpatient and inpatient mental health facilities for children.

Course

The onset of Conduct Disorder may occur as early as the preschool years, but the first significant symptoms usually emerge during the period from middle childhood through middle adolescence. Oppositional Defiant Disorder is a common precursor to the Childhood-Onset Type of Conduct Disorder. Onset is rare after age 16 years. The course of Conduct Disorder is variable. In a majority of individuals, the disorder remits by adulthood. However, a substantial proportion continue to show behaviors in adulthood that meet criteria for Antisocial Personality Disorder. Many individuals with Conduct Disorder, particularly those with Adolescent-Onset Type and those with few and milder symptoms, achieve adequate social and occupational adjustment as adults. Early onset predicts a worse prognosis and an increased risk in adult life for Antisocial Personality Disorder and Substance-Related Disorders. Individuals with Conduct Disorder are at risk for later Mood Disorders, Anxiety Disorders, Somatoform Disorders, and Substance-Related Disorders.

Familial Pattern

Estimates from twin and adoption studies show that Conduct Disorder is influenced by both genetic and environmental factors. The risk for Conduct Disorder is increased

in children with a biological or adoptive parent with Antisocial Personality Disorder or a sibling with Conduct Disorder. The disorder also appears to be more common in children of biological parents with Alcohol Dependence, Mood Disorders, or Schizophrenia or biological parents who have a history of Attention-Deficit/Hyperactivity Disorder or Conduct Disorder.

Differential Diagnosis

Although **Oppositional Defiant Disorder** includes some of the features observed in Conduct Disorder (e.g., disobedience and opposition to authority figures), it does not include the persistent pattern of the more serious forms of behavior in which either the basic rights of others or age-appropriate societal norms or rules are violated. When the individual's pattern of behavior meets the criteria for both Conduct Disorder and Oppositional Defiant Disorder, the diagnosis of Conduct Disorder takes precedence and Oppositional Defiant Disorder is not diagnosed.

Although children with **Attention-Deficit/Hyperactivity Disorder** often exhibit hyperactive and impulsive behavior that may be disruptive, this behavior does not by itself violate age-appropriate societal norms and therefore does not usually meet criteria for Conduct Disorder. When criteria are met for both Attention-Deficit/Hyperactivity Disorder and Conduct Disorder, both diagnoses should be given.

Irritability and conduct problems often occur in children or adolescents with a **Mood Disorder**. These can usually be distinguished from the pattern of conduct problems seen in Conduct Disorder based on the episodic course and accompanying symptoms characteristic of the Mood Disorder. If criteria for both are met, diagnoses of both Conduct Disorder and the Mood Disorder can be given.

The diagnosis of **Adjustment Disorder** (With Disturbance of Conduct or With Mixed Disturbance of Emotions and Conduct) should be considered if clinically significant conduct problems that do not meet the criteria for another specific disorder develop in clear association with the onset of a psychosocial stressor. Isolated conduct problems that do not meet criteria for Conduct Disorder or Adjustment Disorder may be coded as **Child or Adolescent Antisocial Behavior** (see "Additional Conditions That May Be a Focus of Clinical Attention," p. 740). Conduct Disorder is diagnosed only if the conduct problems represent a repetitive and persistent pattern that is associated with impairment in social, academic, or occupational functioning.

Diagnostic criteria for Conduct Disorder

- A. A repetitive and persistent pattern of behavior in which the basic rights of others or major age-appropriate societal norms or rules are violated, as manifested by the presence of three (or more) of the following criteria in the past 12 months, with at least one criterion present in the past 6 months:

Aggression to people and animals

- (1) often bullies, threatens, or intimidates others
- (2) often initiates physical fights
- (3) has used a weapon that can cause serious physical harm to others (e.g., a bat, brick, broken bottle, knife, gun)

Diagnostic criteria for Conduct Disorder (continued)

- (4) has been physically cruel to people
- (5) has been physically cruel to animals
- (6) has stolen while confronting a victim (e.g., mugging, purse snatching, extortion, armed robbery)
- (7) has forced someone into sexual activity

Destruction of property

- (8) has deliberately engaged in fire setting with the intention of causing serious damage
- (9) has deliberately destroyed others' property (other than by fire setting)

Deceitfulness or theft

- (10) has broken into someone else's house, building, or car
- (11) often lies to obtain goods or favors or to avoid obligations (i.e., "cons" others)
- (12) has stolen items of nontrivial value without confronting a victim (e.g., shoplifting, but without breaking and entering; forgery)

Serious violations of rules

- (13) often stays out at night despite parental prohibitions, beginning before age 13 years
 - (14) has run away from home overnight at least twice while living in parental or parental surrogate home (or once without returning for a lengthy period)
 - (15) is often truant from school, beginning before age 13 years
- B. The disturbance in behavior causes clinically significant impairment in social, academic, or occupational functioning.
- C. If the individual is age 18 years or older, criteria are not met for Antisocial Personality Disorder.

Code based on age at onset:

312.81 Conduct Disorder, Childhood-Onset Type: onset of at least one criterion characteristic of Conduct Disorder prior to age 10 years

312.82 Conduct Disorder, Adolescent-Onset Type: absence of any criteria characteristic of Conduct Disorder prior to age 10 years

312.89 Conduct Disorder, Unspecified Onset: age at onset is not known

Specify severity:

Mild: few if any conduct problems in excess of those required to make the diagnosis **and** conduct problems cause only minor harm to others

Moderate: number of conduct problems and effect on others intermediate between "mild" and "severe"

Severe: many conduct problems in excess of those required to make the diagnosis **or** conduct problems cause considerable harm to others

For individuals over age 18 years, a diagnosis of Conduct Disorder can be given only if the criteria are not also met for **Antisocial Personality Disorder**. The diagnosis of Antisocial Personality Disorder cannot be given to individuals under age 18 years.

313.81 Oppositional Defiant Disorder

Diagnostic Features

The essential feature of Oppositional Defiant Disorder is a recurrent pattern of negativistic, defiant, disobedient, and hostile behavior toward authority figures that persists for at least 6 months (Criterion A) and is characterized by the frequent occurrence of at least four of the following behaviors: losing temper (Criterion A1), arguing with adults (Criterion A2), actively defying or refusing to comply with the requests or rules of adults (Criterion A3), deliberately doing things that will annoy other people (Criterion A4), blaming others for his or her own mistakes or misbehavior (Criterion A5), being touchy or easily annoyed by others (Criterion A6), being angry and resentful (Criterion A7), or being spiteful or vindictive (Criterion A8). To qualify for Oppositional Defiant Disorder, the behaviors must occur more frequently than is typically observed in individuals of comparable age and developmental level and must lead to significant impairment in social, academic, or occupational functioning (Criterion B). The diagnosis is not made if the disturbance in behavior occurs exclusively during the course of a Psychotic or Mood Disorder (Criterion C) or if criteria are met for Conduct Disorder or Antisocial Personality Disorder (in an individual over age 18 years).

Negativistic and defiant behaviors are expressed by persistent stubbornness, resistance to directions, and unwillingness to compromise, give in, or negotiate with adults or peers. Defiance may also include deliberate or persistent testing of limits, usually by ignoring orders, arguing, and failing to accept blame for misdeeds. Hostility can be directed at adults or peers and is shown by deliberately annoying others or by verbal aggression (usually without the more serious physical aggression seen in Conduct Disorder). Manifestations of the disorder are almost invariably present in the home setting, but may not be evident at school or in the community. Symptoms of the disorder are typically more evident in interactions with adults or peers whom the individual knows well, and thus may not be apparent during clinical examination. Usually individuals with this disorder do not regard themselves as oppositional or defiant, but justify their behavior as a response to unreasonable demands or circumstances.

Associated Features and Disorders

Associated features and disorders vary as a function of the individual's age and the severity of the Oppositional Defiant Disorder. In males, the disorder has been shown to be more prevalent among those who, in the preschool years, have problematic temperaments (e.g., high reactivity, difficulty being soothed) or high motor activity. During the school years, there may be low self-esteem (or overly inflated self-esteem), mood lability, low frustration tolerance, swearing, and the precocious use of alcohol, tobacco, or illicit drugs. There are often conflicts with parents, teachers, and peers. There may be a vicious cycle in which the parent and child bring out the worst in each other. Oppositional Defiant Disorder is more prevalent in families in which child care is disrupted by a succession of different caregivers or in families in which harsh,

inconsistent, or neglectful child-rearing practices are common. Attention-Deficit/Hyperactivity Disorder is common in children with Oppositional Defiant Disorder. Learning Disorders and Communication Disorders also tend to be associated with Oppositional Defiant Disorder.

Specific Age and Gender Features

Because transient oppositional behavior is very common in preschool children and in adolescents, caution should be exercised in making the diagnosis of Oppositional Defiant Disorder especially during these developmental periods. The number of oppositional symptoms tends to increase with age. The disorder is more prevalent in males than in females before puberty, but the rates appear to be equal after puberty. Symptoms are generally similar in each gender, except that males may have more confrontational behavior and more persistent symptoms.

Prevalence

Rates of Oppositional Defiant Disorder from 2% to 16% have been reported, depending on the nature of the population sample and methods of ascertainment.

Course

Oppositional Defiant Disorder usually becomes evident before age 8 years and usually not later than early adolescence. The oppositional symptoms often emerge in the home setting but over time may appear in other settings as well. Onset is typically gradual, usually occurring over the course of months or years. In a significant proportion of cases, Oppositional Defiant Disorder is a developmental antecedent to Conduct Disorder. Although Conduct Disorder, Childhood-Onset Type is often preceded by Oppositional Defiant Disorder, many children with Oppositional Defiant Disorder do not subsequently develop Conduct Disorder.

Familial Pattern

Oppositional Defiant Disorder appears to be more common in families in which at least one parent has a history of a Mood Disorder, Oppositional Defiant Disorder, Conduct Disorder, Attention-Deficit/Hyperactivity Disorder, Antisocial Personality Disorder, or a Substance-Related Disorder. In addition, some studies suggest that mothers with a Depressive Disorder are more likely to have children with oppositional behavior, but it is unclear to what extent maternal depression results from or causes oppositional behavior in children. Oppositional Defiant Disorder is more common in families in which there is serious marital discord.

Differential Diagnosis

The disruptive behaviors of individuals with Oppositional Defiant Disorder are of a less severe nature than those of individuals with Conduct Disorder and typically do not include aggression toward people or animals, destruction of property, or a pat-

tern of theft or deceit. Because all of the features of Oppositional Defiant Disorder are usually present in **Conduct Disorder**, Oppositional Defiant Disorder is not diagnosed if the criteria are met for Conduct Disorder. Oppositional behavior is a common associated feature of **Mood Disorders** and **Psychotic Disorders** presenting in children and adolescents and should not be diagnosed separately if the symptoms occur exclusively during the course of a Mood or Psychotic Disorder. Oppositional behaviors must also be distinguished from the disruptive behavior resulting from inattention and impulsivity in **Attention-Deficit/Hyperactivity Disorder**. When the two disorders co-occur, both diagnoses should be made. In individuals with **Mental Retardation**, a diagnosis of Oppositional Defiant Disorder is given only if the oppositional behavior is markedly greater than is commonly observed among individuals of comparable age, gender, and severity of Mental Retardation. Oppositional Defiant Disorder must also be distinguished from a failure to follow directions that is the result of **impaired language comprehension** (e.g., hearing loss, Mixed Receptive-Expressive Language Disorder). Oppositional behavior is a **typical feature of certain developmental stages** (e.g., early childhood and adolescence). A diagnosis of Oppositional Defiant Disorder should be considered only if the behaviors occur more frequently and have more serious consequences than is typically observed in other individuals of comparable developmental stage and lead to significant impairment in social, academic, or occupational functioning. New onset of oppositional behaviors in adolescence may be due to the process of normal individuation.

Diagnostic criteria for 313.81 Oppositional Defiant Disorder

- A. A pattern of negativistic, hostile, and defiant behavior lasting at least 6 months, during which four (or more) of the following are present:
 - (1) often loses temper
 - (2) often argues with adults
 - (3) often actively defies or refuses to comply with adults' requests or rules
 - (4) often deliberately annoys people
 - (5) often blames others for his or her mistakes or misbehavior
 - (6) is often touchy or easily annoyed by others
 - (7) is often angry and resentful
 - (8) is often spiteful or vindictive
 - Note:** Consider a criterion met only if the behavior occurs more frequently than is typically observed in individuals of comparable age and developmental level.
 - B. The disturbance in behavior causes clinically significant impairment in social, academic, or occupational functioning.
 - C. The behaviors do not occur exclusively during the course of a Psychotic or Mood Disorder.
 - D. Criteria are not met for Conduct Disorder, and, if the individual is age 18 years or older, criteria are not met for Antisocial Personality Disorder.
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312.9 Disruptive Behavior Disorder Not Otherwise Specified

This category is for disorders characterized by conduct or oppositional defiant behaviors that do not meet the criteria for Conduct Disorder or Oppositional Defiant Disorder. For example, include clinical presentations that do not meet full criteria either for Oppositional Defiant Disorder or Conduct Disorder, but in which there is clinically significant impairment.

Feeding and Eating Disorders of Infancy or Early Childhood

The Feeding and Eating Disorders of Infancy or Early Childhood are characterized by persistent feeding and eating disturbances. The specific disorders included are Pica, Rumination Disorder, and Feeding Disorder of Infancy or Early Childhood. Note that Anorexia Nervosa and Bulimia Nervosa are included in the "Eating Disorders" section (see p. 583).

307.52 Pica

Diagnostic Features

The essential feature of Pica is the eating of one or more nonnutritive substances on a persistent basis for a period of at least 1 month (Criterion A). The typical substances ingested tend to vary with age. Infants and younger children typically eat paint, plaster, string, hair, or cloth. Older children may eat animal droppings, sand, insects, leaves, or pebbles. Adolescents and adults may consume clay or soil. There is no aversion to food. This behavior must be developmentally inappropriate (Criterion B) and not part of a culturally sanctioned practice (Criterion C). The eating of nonnutritive substances is an associated feature of other mental disorders (e.g., Pervasive Developmental Disorder, Mental Retardation). If the eating behavior occurs exclusively during the course of another mental disorder, a separate diagnosis of Pica should be made only if the eating behavior is sufficiently severe to warrant independent clinical attention (Criterion D).

Associated Features and Disorders

Pica is frequently associated with Mental Retardation and Pervasive Developmental Disorders. Although vitamin or mineral deficiencies (e.g., zinc) have been reported in some instances, usually no specific biological abnormalities are found. In some cases, Pica comes to clinical attention only following general medical complications (e.g., lead poisoning as a result of ingesting paint or paint-soaked plaster, mechanical bowel problems, intestinal obstruction as a result of hair ball tumors, intestinal perforation,

or infections such as toxoplasmosis and toxocariasis as a result of ingesting feces or dirt). Poverty, neglect, lack of parental supervision, and developmental delay increase the risk for the condition.

Specific Culture, Age, and Gender Features

In some cultures, the eating of dirt or other seemingly nonnutritive substances is believed to be of value. Pica is more commonly seen in young children and occasionally in pregnant females.

Prevalence

Epidemiological data on Pica are limited. Among individuals with Mental Retardation, the prevalence of the disorder appears to increase with the severity of the retardation (e.g., it has been reported to be as high as 15% in adults with Severe Mental Retardation).

Course

Pica may have its onset in infancy. In most instances, the disorder appears to last for several months and then remits. It may occasionally continue into adolescence or, less frequently, into adulthood. In individuals with Mental Retardation, the behavior may diminish during adulthood.

Differential Diagnosis

Before approximately ages 18–24 months, mouthing and sometimes eating of nonnutritive substances are relatively common and do not imply the presence of Pica. Pica is diagnosed only when the behavior is judged to be persistent (i.e., present for at least 1 month) and inappropriate given the individual's developmental level. Eating of nonnutritive substances may occur during the course of other mental disorders (e.g., in a **Pervasive Developmental Disorder**, in **Schizophrenia** as a result of delusional beliefs, and in **Kleine-Levin syndrome**). In such instances, an additional diagnosis of Pica should be given only if the eating behavior is sufficiently severe to warrant independent clinical attention. Pica can be distinguished from **other eating disorders** (e.g., Ruminant Disorder, Feeding Disorder of Infancy or Early Childhood, Anorexia Nervosa, and Bulimia Nervosa) by the consumption of nonnutritive substances.

Diagnostic criteria for 307.52 Pica

- A. Persistent eating of nonnutritive substances for a period of at least 1 month.
 - B. The eating of nonnutritive substances is inappropriate to the developmental level.
 - C. The eating behavior is not part of a culturally sanctioned practice.
 - D. If the eating behavior occurs exclusively during the course of another mental disorder (e.g., Mental Retardation, Pervasive Developmental Disorder, Schizophrenia), it is sufficiently severe to warrant independent clinical attention.
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307.53 Rumination Disorder**Diagnostic Features**

The essential feature of Rumination Disorder is the repeated regurgitation and re-chewing of food occurring after feeding that develops in an infant or child after a period of normal functioning and lasts for at least 1 month (Criterion A). Partially digested food is brought up into the mouth without apparent nausea, retching, disgust, or associated gastrointestinal disorder. The food is then either ejected from the mouth or, more frequently, chewed and reswallowed. The symptoms are not due to an associated gastrointestinal or other general medical condition (e.g., Sandifer's syndrome, esophageal reflux) (Criterion B) and do not occur exclusively during the course of Anorexia Nervosa or Bulimia Nervosa. If the symptoms occur exclusively during the course of Mental Retardation or a Pervasive Developmental Disorder, they must be sufficiently severe to warrant independent clinical attention (Criterion C). The disorder is most commonly observed in infants but may be seen in older individuals, particularly those who also have Mental Retardation. Infants with the disorder display a characteristic position of straining and arching the back with the head held back, make sucking movements with their tongues, and give the impression of gaining satisfaction from the activity.

Associated Features and Disorders

Infants with Rumination Disorder are generally irritable and hungry between episodes of regurgitation. Although the infant is apparently hungry and ingests large amounts of food, malnutrition may occur because regurgitation immediately follows the feedings. Weight loss, failure to make expected weight gains, and even death can result (with mortality rates as high as 25% reported). Malnutrition appears to be less likely in older children and adults in whom the disorder may be either continuous or episodic. Psychosocial problems such as lack of stimulation, neglect, stressful life situations, and problems in the parent-child relationship may be predisposing factors. Understimulation of the infant may result if the caregiver becomes discouraged and alienated because of the unsuccessful feeding experiences or the noxious odor of the

regurgitated material. In some instances, Feeding Disorder of Infancy or Early Childhood may also develop. In older children and adults, Mental Retardation is a predisposing factor.

Prevalence

Rumination Disorder appears to be uncommon. It may occur more often in males than in females.

Course

The onset of Rumination Disorder may occur in the context of developmental delays. The age at onset is between ages 3 and 12 months, except in individuals with Mental Retardation in whom the disorder may occur at a somewhat later developmental stage. In infants, the disorder frequently remits spontaneously. In some severe cases, however, the course is continuous.

Differential Diagnosis

In infants, **congenital anomalies** (e.g., pyloric stenosis or gastroesophageal reflux) or **other general medical conditions** (e.g., infections of the gastrointestinal system) can cause regurgitation of food and should be ruled out by appropriate physical examinations and laboratory tests. Rumination can be distinguished from **normal vomiting of early infancy** by the apparently voluntary nature of the rumination (e.g., observation of characteristic preparatory movements followed by regurgitation and sucking or chewing movements that appear to be pleasurable). Rumination Disorder is not diagnosed if the symptoms occur exclusively during the course of **Anorexia Nervosa** or **Bulimia Nervosa**.

Diagnostic criteria for 307.53 Rumination Disorder

- A. Repeated regurgitation and rechewing of food for a period of at least 1 month following a period of normal functioning.
 - B. The behavior is not due to an associated gastrointestinal or other general medical condition (e.g., esophageal reflux).
 - C. The behavior does not occur exclusively during the course of Anorexia Nervosa or Bulimia Nervosa. If the symptoms occur exclusively during the course of Mental Retardation or a Pervasive Developmental Disorder, they are sufficiently severe to warrant independent clinical attention.
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307.59 Feeding Disorder of Infancy or Early Childhood

Diagnostic Features

The essential feature of Feeding Disorder of Infancy or Early Childhood is the persistent failure to eat adequately, as reflected in significant failure to gain weight or significant weight loss over at least 1 month (Criterion A). There is no gastrointestinal or other general medical condition (e.g., esophageal reflux) severe enough to account for the feeding disturbance (Criterion B). The feeding disturbance is also not better accounted for by another Mental Disorder (e.g., Rumination Disorder) or by lack of available food (Criterion C). The onset of the disorder must be before age 6 years (Criterion D).

Associated Features and Disorders

Associated descriptive features and mental disorders. Infants with feeding disorders may be more irritable and difficult to console during feeding than infants without feeding disorders. They may appear apathetic and withdrawn and may also exhibit developmental delays. In some instances, parent-child interaction problems may contribute to or exacerbate the infant's feeding problem (e.g., presenting food inappropriately or responding to the infant's food refusal as if it were an act of aggression or rejection). Inadequate caloric intake may exacerbate the associated features (e.g., irritability, developmental lags) and further contribute to feeding difficulties. Factors in the infant that may be associated with the condition include temperamental characteristics or intrauterine growth retardation and preexisting developmental impairments that make the infant less responsive. Other factors that may be associated with the condition include parental psychopathology and child abuse or neglect.

Associated laboratory findings. There may be nonspecific findings associated with the malnutrition that is sometimes seen with Feeding Disorder of Infancy or Early Childhood (e.g., anemia and low serum albumin and total protein).

Associated physical examination findings and general medical conditions. There may be malnutrition that, in severe cases, can be life threatening in Feeding Disorder of Infancy or Early Childhood.

Specific Age and Gender Features

A later onset (e.g., age 2 or 3 years rather than infancy) is associated with lesser degrees of developmental delay and malnutrition, although growth retardation may be observed. Feeding Disorder of Infancy or Early Childhood is equally common in males and females.

Prevalence

Of all pediatric hospital admissions, 1%–5% are for failure to thrive, and up to one-half of these may reflect feeding disturbances without any apparent predisposing

general medical condition. Data from community samples suggest a point prevalence of around 3% for failure to thrive.

Course

Feeding Disorder of Infancy or Early Childhood commonly has its onset in the first year of life, but may have an onset in children ages 2–3 years. The majority of children have improved growth after variable lengths of time, although they remain shorter and lighter up through adolescence compared with children who did not experience growth failure.

Differential Diagnosis

Minor problems in feeding are common in infancy. The diagnosis of Feeding Disorder of Infancy or Early Childhood should be made only if the eating problem results in significant failure to gain weight or loss of weight.

This disorder is not diagnosed if the feeding disturbances are fully explained by a **gastrointestinal, endocrinological, or neurological condition**. Children with an underlying general medical condition may be more difficult to feed, and the diagnosis of Feeding Disorder of Infancy or Early Childhood should not be made in such cases unless the degree of disturbance is of greater severity than would be expected on the basis of the general medical condition alone. The diagnosis is suggested if there is improvement in feeding and weight gain in response to changing caregivers.

Diagnostic criteria for

307.59 Feeding Disorder of Infancy or Early Childhood

- A. Feeding disturbance as manifested by persistent failure to eat adequately with significant failure to gain weight or significant loss of weight over at least 1 month.
 - B. The disturbance is not due to an associated gastrointestinal or other general medical condition (e.g., esophageal reflux).
 - C. The disturbance is not better accounted for by another mental disorder (e.g., Rumination Disorder) or by lack of available food.
 - D. The onset is before age 6 years.
-

Tic Disorders

Four disorders are included in this section: Tourette's Disorder, Chronic Motor or Vocal Tic Disorder, Transient Tic Disorder, and Tic Disorder Not Otherwise Specified. A *tic* is a sudden, rapid, recurrent, nonrhythmic, stereotyped motor movement or vocalization. Motor and vocal tics may be simple (involving only a few muscles or

simple sounds) or complex (involving multiple groups of muscles recruited in orchestrated bouts or words and sentences). Examples of *simple motor tics* are eye blinking, nose wrinkling, neck jerking, shoulder shrugging, facial grimacing, and abdominal tensing. These tics usually last less than several hundred milliseconds. *Complex motor tics* include hand gestures, jumping, touching, pressing, stomping, facial contortions, repeatedly smelling an object, squatting, deep knee bends, retracing steps, twirling when walking, and assuming and holding unusual postures (including "dystonic tics" such as holding the neck in a particular tensed position). These tics are longer in duration, lasting seconds or longer. Copropraxia (a sudden, tic-like vulgar, sexual, or obscene gesture) and mirror phenomena such as echopraxia (involuntary, spontaneous imitation of someone else's movements) are complex motor tics.

Simple vocal tics are meaningless sounds such as throat clearing, grunting, sniffing, snorting, and chirping. *Complex vocal tics* more clearly involve speech and language and include the sudden, spontaneous expression of single words or phrases; speech blocking; sudden and meaningless changes in the pitch, emphasis, or volume of speech; palilalia (repeating one's own sounds or words); and echolalia (repeating the last-heard sound, word, or phrase). Coprolalia is the sudden, inappropriate expression of a socially unacceptable word or phrase and may include obscenities as well as specific ethnic, racial, or religious slurs. Coprolalia is found in fewer than 10% of individuals with a tic disorder.

Tics generally are experienced as irresistible but can be suppressed for varying lengths of time. Some children (and an occasional adult) are not aware of their tics. However, with development, many (but not all) persons with tics experience a premonitory urge—a rising tension or somatic sensation in a part of the body that precedes the motor or vocal tic, and a feeling of relief or tension reduction following the expression of the tic. Individuals with tics may feel that the tic is between "voluntary" and "involuntary" in that it is often experienced as a giving in to a mounting tension or bodily need, similar to the tension that precedes a sneeze or the almost irresistible need to scratch an itch. An individual may feel the need to perform a complex tic in a specific way or repeatedly until he or she achieves the feeling that the tic has been done "just right." Only then will the individual experience a reduction in the anxiety or tension.

Tics are often emitted in bouts of one or several tics; the bouts are separated by periods of non-tic behavior lasting from seconds to hours. Tics generally change in severity (frequency of tics, forcefulness, and the degree tics disrupt ongoing behavior) during the course of hours and over the course of a day. Tics may vary in frequency and disruptiveness in different contexts. For example, children and adults may be better able to suppress tics when in school, at work, or in the physician's office than at home. Tics generally decrease or stop during sleep, although some individuals have occasional tics while asleep or awaken suddenly with a tic. Tics are often more frequent when a person relaxes in private (e.g., when watching television) and are decreased when the individual engages in directed, effortful activity (e.g., reading or sewing). Tics may be exacerbated during periods of stress, such as when there are heightened work pressures or during examinations.

Differential Diagnosis

Tic Disorders must be distinguished from other types of **abnormal movements** that may accompany **general medical conditions** (e.g., Huntington's disease, stroke, Lesch-Nyhan syndrome, Wilson's disease, Sydenham's chorea, multiple sclerosis, postviral encephalitis, head injury) and from abnormal movements that are due to the **direct effects of a substance** (e.g., a neuroleptic medication). Taking account of the medical and family history, movement morphology, rhythm, and modifying influences can assist in making a correct diagnosis. **Chorea** typically is a simple, random, irregular, and nonstereotyped movement that has no premonitory component and increases when the person is distracted. **Dystonic movements** are slow, protracted twisting movements interspersed with prolonged states of muscular tension. **Athetoid movements** are slow, irregular, writhing movements, most frequently in the fingers and toes but often involving the neck. **Myoclonic movements** are brief, simple, shocklike muscle contractions that may affect parts of muscles or muscle groups. Unlike tics, myoclonic movements may continue during sleep. **Hemiballismic movements** are intermittent, coarse, large-amplitude, unilateral movements of the limbs. **Spasms** are stereotypic, prolonged movements involving the same groups of muscles that are usually slower but are sometimes more rapid than tics. **Hemifacial spasm** consists of irregular, repetitive, unilateral jerks of facial muscles. **Synkinesis** is an involuntary movement that is concurrent exclusively with a specific voluntary act (e.g., movement of the corner of the mouth when the person intends to close the eye).

When the tics are a direct physiological consequence of medication use, a **Medication-Induced Movement Disorder Not Otherwise Specified** would be diagnosed instead of a Tic Disorder (e.g., in the clearest case, when the tics occur with the use of a medication and stop when the medication is discontinued). In some cases, certain medications (e.g., stimulants) may exacerbate a preexisting Tic Disorder, in which case no additional diagnosis of a medication-induced disorder is necessary.

Tics must also be distinguished from **Stereotypic Movement Disorder** and stereotypies in **Pervasive Developmental Disorders**. Differentiating simple tics (e.g., eye blinking) from the complex movements characteristic of stereotyped movements is relatively straightforward. The distinction between complex motor tics and stereotyped movements is less clear-cut. In general, stereotyped movements appear to be more driven, rhythmic, self-stimulating or soothing, and intentional, whereas tics have a more involuntary quality (although some individuals describe tics as having a voluntary component) and generally occur in temporal bouts or clusters. Complex tics may be difficult to distinguish from **compulsions** (as in Obsessive-Compulsive Disorder); making this distinction is all the more challenging because Obsessive-Compulsive Disorder is common in individuals with Tic Disorders. Compulsions are performed in response to an obsession (e.g., hand washing to allay a concern about germs) or according to rules that must be applied rigidly (e.g., the need to line things up in a specific order). Compulsions typically are more elaborate than tics and are more likely to resemble "normal" behavior. Whereas compulsions are often, though not always, preceded by a persistent worry or concern, tics are more likely to be preceded by a transient "physical" tension in a part of the body (e.g., in the nose or shoulder muscles or in the throat) that is reduced by the tic. When individuals manifest

symptoms of both Obsessive-Compulsive Disorder and a Tic Disorder (especially Tourette's Disorder), both diagnoses may be warranted. Certain vocal or motor tics (e.g., barking, echolalia, palilalia) must be distinguished from disorganized or catatonic behavior in **Schizophrenia**.

The Tic Disorders can be distinguished from one another based on duration and variety of tics and age at onset. **Transient Tic Disorder** includes motor and/or vocal tics lasting for at least 4 weeks but for no longer than 12 consecutive months. **Tourette's Disorder** and **Chronic Motor or Vocal Tic Disorder** each have a duration of more than 12 months, but Tourette's Disorder requires multiple motor tics and at least one vocal tic during part of this time. Often, the diagnosis may change over time during the natural history of a Tic Disorder. For example, during the first months, a child may be diagnosed as having a Transient Tic Disorder. After a year, with further tics and longer duration, the diagnosis may become Tourette's Disorder. **Tic Disorder Not Otherwise Specified** would be appropriate for clinically significant presentations lasting less than 4 weeks, for presentations with an age at onset above age 18 years, and for the unusual case of an individual with only one motor tic and only one vocal tic.

307.23 Tourette's Disorder

Diagnostic Features

The essential features of Tourette's Disorder are multiple motor tics and one or more vocal tics (Criterion A). These may appear simultaneously or at different periods during the illness. The tics occur many times a day, recurrently throughout a period of more than 1 year. During this period, there is never a tic-free period of more than 3 consecutive months (Criterion B). The onset of the disorder is before age 18 years (Criterion C). The tics are not due to the direct physiological effects of a substance (e.g., stimulants) or a general medical condition (e.g., Huntington's disease or post-viral encephalitis) (Criterion D).

The anatomical location, number, frequency, complexity, and severity of the tics change over time. Simple and complex motor tics may affect any part of the body, including the face, head, torso, and upper and lower limbs. Simple motor tics are rapid, meaningless contractions of one or a few muscles, such as eye blinking. Complex motor tics involving touching, squatting, deep knee bends, retracing steps, and twirling when walking may be present. The vocal tics include various words or sounds such as clicks, grunts, yelps, barks, sniffs, snorts, and coughs. Coprolalia, a complex vocal tic involving the uttering of obscenities, is present in only a small minority of individuals (less than 10%) and is not required for a diagnosis of Tourette's Disorder.

In approximately one-half of the individuals with this disorder, the first symptoms to appear are bouts of a single tic, most frequently eye blinking. Less frequently, initial tics involve another part of the face or the body, such as facial grimacing, head jerking, tongue protrusion, sniffing, hopping, skipping, throat clearing, stuttering-like block in speech fluency, or uttering sounds or words. Sometimes this disorder begins with multiple symptoms starting at the same time.

Associated Features and Disorders

The most common associated symptoms of Tourette's Disorder are obsessions and compulsions. Hyperactivity, distractibility, and impulsivity are also relatively common. Social discomfort, shame, self-consciousness, and demoralization and sadness frequently occur. Persistent motor and vocal tics may cause a broad range of distress and impairment, ranging from none to severe. Younger children, in particular, may be unaware of their tics, suffer no distress, and show no impairment in any area of functioning. A high percentage of children, adolescents, and adults with Tourette's Disorder do not seek medical attention for their tics. At the other end of the spectrum, there are individuals with Tourette's Disorder who are burdened by intrusive, recurrent, forceful, and socially stigmatizing motor and vocal tics.

Social, academic, and occupational functioning may be impaired because of rejection by others or anxiety about having tics in social situations. Chronic tic symptoms can cause considerable distress and can lead to social isolation and personality changes. In severe cases of Tourette's Disorder, the tics may directly interfere with daily activities (e.g., conversing, reading, or writing). Rare complications of Tourette's Disorder include physical injury, such as blindness due to self-inflicted eye injury (from head banging or eye gouging), orthopedic problems (from knee bending, neck jerking, or head turning), skin problems (from picking or lip licking), and neurological sequelae (e.g., from disc disease related to many years of forceful neck movements). The severity of the tics may be exacerbated by administration of central nervous system stimulants, such as those used in the treatment of Attention-Deficit/Hyperactivity Disorder, although some individuals can tolerate these medications without an exacerbation of their tics or may even have a reduction in tics. Obsessive-Compulsive Disorder and Attention-Deficit/Hyperactivity Disorder often co-occur in individuals with Tourette's Disorder. Attentional problems or obsessive symptomatology may precede or follow the onset of tics. Obsessive-compulsive symptoms found in individuals with Tourette's Disorder may constitute a specific subtype of Obsessive-Compulsive Disorder. This subtype appears to be characterized by an earlier age at onset, male preponderance, higher frequency of certain obsessive-compulsive symptoms (more aggressive symptoms and less concern about contamination and hoarding), and poorer response to pharmacotherapy with selective serotonin reuptake inhibitors. Disruptive behavior, impulsiveness, and social immaturity are prominent features in those children and adolescents who also have Attention-Deficit/Hyperactivity Disorder. These clinical features may interfere with academic progress and interpersonal relationships and lead to greater impairment than that caused by the Tourette's Disorder.

Specific Culture and Gender Features

Tourette's Disorder has been widely reported in diverse racial and ethnic groups. Although in clinical settings the disorder is diagnosed approximately three to five times more often in males than in females, the gender ratio is perhaps as low as 2:1 in community samples.

Prevalence

The prevalence of Tourette's Disorder is related to age. Many more children (5–30 per 10,000) are affected than adults (1–2 per 10,000).

Course

The age at onset of Tourette's Disorder may be as early as age 2 years, is usually during childhood or early adolescence, and is by definition before age 18 years. The median age at onset for motor tics is about 6–7 years. The duration of the disorder may be lifelong, though periods of remission lasting from weeks to years may occur. In most cases, the severity, frequency, disruptiveness, and variability of the symptoms diminish during adolescence and adulthood. In other cases, the symptoms actually disappear entirely, usually by early adulthood. In a few cases, the symptoms may worsen in adulthood. The predictors of this course are not known.

Familial Pattern

The vulnerability to Tourette's Disorder and related disorders is transmitted within families and appears to be genetic. The mode of genetic transmission, however, is not known. Pedigree studies suggest that there are genes of major effect. Although some early studies suggested a pattern of transmission that is consistent with an autosomal dominant pattern, other studies suggest a more complex mode of transmission. "Vulnerability" implies that the child receives the genetic or constitutional basis for developing a Tic Disorder; the precise type or severity of disorder may be different from one generation to another and is modified by nongenetic factors. Not everyone who inherits the genetic vulnerability will express symptoms of a Tic Disorder. The range of forms in which the vulnerability may be expressed includes Tourette's Disorder, Chronic Motor or Vocal Tic Disorder, and some forms of Obsessive-Compulsive Disorder. It also appears that individuals with Tourette's Disorder are at greater risk for Attention-Deficit/Hyperactivity Disorder. In some individuals with Tourette's Disorder, there is no evidence of a familial pattern.

Differential Diagnosis

Refer to the "Differential Diagnosis" section for Tic Disorders (p. 110).

Diagnostic criteria for 307.23 Tourette's Disorder

- A. Both multiple motor and one or more vocal tics have been present at some time during the illness, although not necessarily concurrently. (A *tic* is a sudden, rapid, recurrent, nonrhythmic, stereotyped motor movement or vocalization.)
 - B. The tics occur many times a day (usually in bouts) nearly every day or intermittently throughout a period of more than 1 year, and during this period there was never a tic-free period of more than 3 consecutive months.
 - C. The onset is before age 18 years.
 - D. The disturbance is not due to the direct physiological effects of a substance (e.g., stimulants) or a general medical condition (e.g., Huntington's disease or postviral encephalitis).
-

307.22 Chronic Motor or Vocal Tic Disorder**Diagnostic Features**

The essential feature of Chronic Motor or Vocal Tic Disorder is the presence of either motor tics *or* vocal tics, but *not both* (Criterion A). This differs from Tourette's Disorder in which there must be both multiple motor and one or more vocal tics. The other essential features (Criteria B, C, and D) are the same as for Tourette's Disorder. A diagnosis of Chronic Motor or Vocal Tic Disorder cannot be made if the criteria for Tourette's Disorder have ever been met (Criterion E). The other characteristics of Chronic Motor or Vocal Tic Disorder are generally the same as for Tourette's Disorder (see p. 111), except that the severity of the symptoms and the functional impairment are usually much less. It appears that Chronic Motor or Vocal Tic Disorder and Tourette's Disorder may be genetically related because they often occur in the same families.

Differential Diagnosis

Refer to the "Differential Diagnosis" section for Tic Disorders (p. 110).

Diagnostic criteria for 307.22 Chronic Motor or Vocal Tic Disorder

- A. Single or multiple motor or vocal tics (i.e., sudden, rapid, recurrent, nonrhythmic, stereotyped motor movements or vocalizations), but not both, have been present at some time during the illness.
 - B. The tics occur many times a day nearly every day or intermittently throughout a period of more than 1 year, and during this period there was never a tic-free period of more than 3 consecutive months.
 - C. The onset is before age 18 years.
 - D. The disturbance is not due to the direct physiological effects of a substance (e.g., stimulants) or a general medical condition (e.g., Huntington's disease or postviral encephalitis).
 - E. Criteria have never been met for Tourette's Disorder.
-

307.21 Transient Tic Disorder

Diagnostic Features

The essential feature of Transient Tic Disorder is the presence of single or multiple motor tics and/or vocal tics (Criterion A). The tics occur many times a day, nearly every day for at least 4 weeks, but for no longer than 12 consecutive months (Criterion B). The other essential features (Criteria C and D) are the same as for Tourette's Disorder. Transient Tic Disorder is not diagnosed if the criteria for Tourette's Disorder or Chronic Motor or Vocal Tic Disorder (both of which require a duration of at least 1 year) have ever been met (Criterion E). The other characteristics of the disorder are generally the same as for Tourette's Disorder (see p. 111), except that the severity of the symptoms and the functional impairment are usually much less.

Specifiers

The course of Transient Tic Disorder may be indicated by specifying **Single Episode** or **Recurrent**.

Differential Diagnosis

Refer to the "Differential Diagnosis" section for Tic Disorders (p. 110).

Diagnostic criteria for 307.21 Transient Tic Disorder

- A. Single or multiple motor and/or vocal tics (i.e., sudden, rapid, recurrent, nonrhythmic, stereotyped motor movements or vocalizations)
- B. The tics occur many times a day, nearly every day for at least 4 weeks, but for no longer than 12 consecutive months.
- C. The onset is before age 18 years.
- D. The disturbance is not due to the direct physiological effects of a substance (e.g., stimulants) or a general medical condition (e.g., Huntington's disease or postviral encephalitis).
- E. Criteria have never been met for Tourette's Disorder or Chronic Motor or Vocal Tic Disorder.

Specify if:

Single Episode or Recurrent

307.20 Tic Disorder Not Otherwise Specified

This category is for disorders characterized by tics that do not meet criteria for a specific Tic Disorder. Examples include tics lasting less than 4 weeks or tics with an onset after age 18 years.

Elimination Disorders

Encopresis**Diagnostic Features**

The essential feature of Encopresis is repeated passage of feces into inappropriate places (e.g., clothing or floor) (Criterion A). Most often this is involuntary but occasionally may be intentional. The event must occur at least once a month for at least 3 months (Criterion B), and the chronological age of the child must be at least 4 years (or for children with developmental delays, a mental age of at least 4 years) (Criterion C). The fecal incontinence must not be due exclusively to the direct physiological effects of a substance (e.g., laxatives) or a general medical condition except through a mechanism involving constipation (Criterion D).

When the passage of feces is involuntary rather than intentional, it is often related to constipation, impaction, and retention with subsequent overflow. The constipation may develop for psychological reasons (e.g., anxiety about defecating in a particular place or a more general pattern of anxious or oppositional behavior) leading to avoid-

ance of defecation. Physiological predispositions to constipation include ineffectual straining or paradoxical defecation dynamics, with contraction rather than relaxation of the external sphincter or pelvic floor during straining for defecation. Dehydration associated with a febrile illness, hypothyroidism, or a medication side effect can also induce constipation. Once constipation has developed, it may be complicated by an anal fissure, painful defecation, and further fecal retention. The consistency of the stool may vary. In some individuals it may be of normal or near-normal consistency. It may be liquid in other individuals who have overflow incontinence secondary to fecal retention.

Subtypes

Encopresis is coded according to the subtype that characterizes the presentation:

787.6 With Constipation and Overflow Incontinence. There is evidence of constipation on physical examination (i.e., the presence of a large stool mass on abdominal or rectal examination) or a history of a stool frequency of less than three per week. Feces in overflow incontinence are characteristically (but not invariably) poorly formed, and leakage can be infrequent to continuous, occurring mostly during the day and rarely during sleep. Only part of the feces is passed during toileting, and the incontinence resolves after treatment of the constipation.

307.7 Without Constipation and Overflow Incontinence. There is no evidence of constipation on physical examination or by history. Feces are likely to be of normal form and consistency, and soiling is intermittent. Feces may be deposited in a prominent location. This is usually associated with the presence of Oppositional Defiant Disorder or Conduct Disorder or may be the consequence of anal masturbation. Soiling without constipation appears to be less common than soiling with constipation.

Associated Features and Disorders

The child with Encopresis often feels ashamed and may wish to avoid situations (e.g., camp or school) that might lead to embarrassment. The amount of impairment is a function of the effect on the child's self-esteem, the degree of social ostracism by peers, and the anger, punishment, and rejection on the part of caregivers. Smearing feces may be deliberate or accidental resulting from the child's attempt to clean or hide feces that were passed involuntarily. When the incontinence is clearly deliberate, features of Oppositional Defiant Disorder or Conduct Disorder may also be present. Many children with Encopresis and chronic constipation are enuretic and may have associated vesico-ureteric reflux and chronic urinary tract infections that may remit with treatment.

Prevalence

It is estimated that approximately 1% of 5-year-olds have Encopresis, and the disorder is more common in males than in females.

Course

Encopresis is not diagnosed until a child has reached a chronological age of at least 4 years (or for children with developmental delays, a mental age of at least 4 years). Inadequate, inconsistent toilet training and psychosocial stress (e.g., entering school or the birth of a sibling) may be predisposing factors. Two types of course have been described: a "primary" type in which the individual has never established fecal continence, and a "secondary" type in which the disturbance develops after a period of established fecal continence. Encopresis can persist with intermittent exacerbations for years.

Differential Diagnosis

A diagnosis of Encopresis in the presence of a general medical condition is appropriate only if the mechanism involves functional constipation. Fecal incontinence related to other general medical conditions (e.g., chronic diarrhea, spina bifida, anal stenosis) would not warrant a DSM-IV diagnosis of Encopresis.

Diagnostic criteria for Encopresis

- A. Repeated passage of feces into inappropriate places (e.g., clothing or floor) whether involuntary or intentional.
- B. At least one such event a month for at least 3 months.
- C. Chronological age is at least 4 years (or equivalent developmental level).
- D. The behavior is not due exclusively to the direct physiological effects of a substance (e.g., laxatives) or a general medical condition except through a mechanism involving constipation.

Code as follows:

787.6 With Constipation and Overflow Incontinence

307.7 Without Constipation and Overflow Incontinence

307.6 Enuresis (Not Due to a General Medical Condition)

Diagnostic Features

The essential feature of Enuresis is repeated voiding of urine during the day or at night into bed or clothes (Criterion A). Most often this is involuntary but occasionally may be intentional. To qualify for a diagnosis of Enuresis, the voiding of urine must occur at least twice per week for at least 3 months or else must cause clinically significant distress or impairment in social, academic (occupational), or other important areas of functioning (Criterion B). The individual must have reached an age at which

continence is expected (i.e., the chronological age of the child must be at least 5 years, or, for children with developmental delays, a mental age of at least 5 years) (Criterion C). The urinary incontinence is not due exclusively to the direct physiological effects of a substance (e.g., diuretics) or a general medical condition (e.g., diabetes, spina bifida, a seizure disorder) (Criterion D).

Subtypes

The situation in which the Enuresis occurs may be noted by one of the following subtypes:

Nocturnal Only. This is the most common subtype and is defined as passage of urine only during nighttime sleep. The enuretic event typically occurs during the first one-third of the night. Occasionally the voiding takes place during the rapid eye movement (REM) stage of sleep, and the child may recall a dream that involved the act of urinating.

Diurnal Only. This subtype is defined as the passage of urine only during waking hours. Diurnal Enuresis is more common in females than in males and is uncommon after age 9 years. Individuals with diurnal Enuresis can be divided into two groups. One group with "urge incontinence" has Enuresis characterized by sudden urge symptoms and detrusor instability on cystometry. Another group with "voiding postponement" consciously defer micturition urges until incontinence results, with the deferral sometimes due to a reluctance to use the toilet because of social anxiety or a preoccupation with school or play activity. This latter group has a high rate of symptoms of disruptive behavior. The enuretic event most commonly occurs in the early afternoon on school days.

Nocturnal and Diurnal. This subtype is defined as a combination of the two subtypes above.

Associated Features and Disorders

The amount of impairment associated with Enuresis is a function of the limitation on the child's social activities (e.g., ineligibility for sleep-away camp) or its effect on the child's self-esteem, the degree of social ostracism by peers, and the anger, punishment, and rejection on the part of caregivers. Although most children with Enuresis do not have a coexisting mental disorder, the prevalence of coexisting behavioral symptoms is higher in children with Enuresis than in children without Enuresis. Developmental delays, including speech, language, learning, and motor skills delays, are also present in a portion of children with Enuresis. Encopresis, Sleepwalking Disorder, and Sleep Terror Disorder may be present. Urinary tract infections are more common in children with Enuresis, especially the Diurnal Type, than in those who are continent. The Enuresis commonly persists after appropriate treatment of an associated infection. A number of predisposing factors have been suggested, including delayed or lax toilet training, psychosocial stress, delays in the development of normal circadian rhythms of urine production with resulting nocturnal polyuria or abnormalities of central vasopressin receptor sensitivity, and reduced functional bladder capacities with bladder hyperreactivity (unstable bladder syndrome).

Prevalence

The prevalence of Enuresis is around 5%–10% among 5-year-olds, 3%–5% among 10-year-olds, and around 1% among individuals age 15 years or older.

Course

Two types of course of Enuresis have been described: a “primary” type in which the individual has never established urinary continence, and a “secondary” type in which the disturbance develops after a period of established urinary continence. By definition, primary Enuresis begins at age 5 years. The most common time for the onset of secondary Enuresis is between the ages of 5 and 8 years, but it may occur at any time. After age 5 years, the rate of spontaneous remission is between 5% and 10% per year. Most children with the disorder become continent by adolescence, but in approximately 1% of cases the disorder continues into adulthood.

Familial Pattern

Approximately 75% of all children with Enuresis have a first-degree biological relative who has had the disorder. The risk of Enuresis is five- to sevenfold greater in the offspring of a parent who had a history of Enuresis. The concordance for the disorder is greater in monozygotic twins than in dizygotic twins. Although molecular genetic analyses have detected links to several chromosomes, no significant associations between linkage to a chromosome interval and type of enuresis have been identified.

Differential Diagnosis

The diagnosis of Enuresis is not made in the presence of a **neurogenic bladder** or the presence of a **general medical condition that causes polyuria or urgency** (e.g., untreated diabetes mellitus or diabetes insipidus) or during an **acute urinary tract infection**. However, a diagnosis of Enuresis is compatible with such conditions if urinary incontinence was regularly present prior to the development of the general medical condition or if it persists after the institution of appropriate treatment.

Diagnostic criteria for 307.6 Enuresis

- A. Repeated voiding of urine into bed or clothes (whether involuntary or intentional).
- B. The behavior is clinically significant as manifested by either a frequency of twice a week for at least 3 consecutive months or the presence of clinically significant distress or impairment in social, academic (occupational), or other important areas of functioning.
- C. Chronological age is at least 5 years (or equivalent developmental level).
- D. The behavior is not due exclusively to the direct physiological effect of a substance (e.g., a diuretic) or a general medical condition (e.g., diabetes, spina bifida, a seizure disorder).

Specify type:

Nocturnal Only

Diurnal Only

Nocturnal and Diurnal

**Other Disorders of Infancy,
Childhood, or Adolescence**

309.21 Separation Anxiety Disorder

Diagnostic Features

The essential feature of Separation Anxiety Disorder is excessive anxiety concerning separation from the home or from those to whom the person is attached (Criterion A). This anxiety is beyond that which is expected for the individual's developmental level. The disturbance must last for a period of at least 4 weeks (Criterion B), begin before age 18 years (Criterion C), and cause clinically significant distress or impairment in social, academic (occupational), or other important areas of functioning (Criterion D). The diagnosis is not made if the anxiety occurs exclusively during the course of a Pervasive Developmental Disorder, Schizophrenia, or other Psychotic Disorder or, in adolescents or adults, if it is better accounted for by Panic Disorder With Agoraphobia (Criterion E).

Individuals with this disorder may experience recurrent excessive distress on separation from home or major attachment figures (Criterion A1). When separated from attachment figures, they often need to know their whereabouts and need to stay in touch with them (e.g., by telephone calls). Some individuals become extremely homesick and uncomfortable to the point of misery when away from home. They may yearn to return home and be preoccupied with reunion fantasies. When separated

from major attachment figures, these individuals are often preoccupied with fears that accidents or illness will befall the attachment figures or themselves (Criterion A2). Children with this disorder often express fear of being lost and never being reunited with their parents (Criterion A3). They are often uncomfortable when traveling independently away from the house or from other familiar areas and may avoid going places by themselves. They may be reluctant or refuse to attend school or camp, to visit or sleep at friends' homes, or to go on errands (Criterion A4). These children may be unable to stay or go in a room by themselves and may display "clinging" behavior, staying close to and "shadowing" the parent around the house or requiring someone to be with them when going to another room in the house (Criterion A5).

Children with this disorder often have difficulty at bedtime and may insist that someone stay with them until they fall asleep (Criterion A6). During the night, they may make their way to their parents' bed (or that of another significant person, such as a sibling); if entry to the parental bedroom is barred, they may sleep outside the parents' door. There may be nightmares whose content expresses the individual's fears (e.g., destruction of the family through fire, murder, or other catastrophe) (Criterion A7). Physical complaints, such as stomachaches, headaches, nausea, and vomiting are common when separation occurs or is anticipated (Criterion A8). Cardiovascular symptoms such as palpitations, dizziness, and feeling faint are rare in younger children but may occur in older individuals.

Specifier

Early Onset. This specifier may be used to indicate onset of the disorder before age 6 years.

Associated Features and Mental Disorders

Children with Separation Anxiety Disorder tend to come from families that are close-knit. When separated from home or major attachment figures, they may recurrently exhibit social withdrawal, apathy, sadness, or difficulty concentrating on work or play. Depending on their age, individuals may have fears of animals, monsters, the dark, muggers, burglars, kidnappers, car accidents, plane travel, and other situations that are perceived as presenting danger to the integrity of the family or themselves. Concerns about death and dying are common. School refusal may lead to academic difficulties and social avoidance. Children may complain that no one loves them or cares about them and that they wish they were dead. When extremely upset at the prospect of separation, they may show anger or occasionally hit out at someone who is forcing separation. When alone, especially in the evening or the dark, young children may report unusual perceptual experiences (e.g., seeing people peering into their room, scary creatures reaching for them, feeling eyes staring at them). Children with this disorder are often described as demanding, intrusive, and in need of constant attention. The child's excessive demands often become a source of parental frustration, leading to resentment and conflict in the family. Sometimes, children with the disorder are described as unusually conscientious, compliant, and eager to please. The children may have somatic complaints that result in physical examinations and medical procedures. Depressed mood is frequently present and may become more per-

sistent over time, justifying an additional diagnosis of Dysthymic Disorder or Major Depressive Disorder in some cases. The disorder may precede the development of Panic Disorder With Agoraphobia. Comorbidity with other Anxiety Disorders may be common, especially in clinical settings.

Specific Culture, Age, and Gender Features

There are cultural variations in the degree to which it is considered desirable to tolerate separation. It is important to differentiate Separation Anxiety Disorder from the high value some cultures place on strong interdependence among family members.

The manifestations of the disorder may vary with age. Younger children may not express specific fears of definite threats to parents, home, or themselves. As children get older, worries or fears are often of specific dangers (e.g., kidnapping, mugging). Anxiety and anticipation of separation become manifest in mid-childhood. Although adolescents with this disorder, especially males, may deny anxiety about separation, it may be reflected in their limited independent activity and reluctance to leave home. In older individuals, the disorder may limit the person's ability to handle changes in circumstances (e.g., moving, getting married). Adults with the disorder are typically overconcerned about their offspring and spouses and experience marked discomfort when separated from them. In clinical samples, the disorder is apparently equally common in males and females. In epidemiological samples, the disorder is more frequent in females.

Prevalence

Separation Anxiety Disorder is not uncommon; prevalence estimates average about 4% in children and young adolescents. Separation Anxiety Disorder decreases in prevalence from childhood through adolescence.

Course

Separation Anxiety Disorder may develop after some life stress (e.g., the death of a relative or pet, an illness of the child or a relative, a change of schools, a move to a new neighborhood, or immigration). Onset may be as early as preschool age and may occur at any time before age 18 years, but onset as late as adolescence is uncommon. Typically there are periods of exacerbation and remission. In some cases, both the anxiety about possible separation and the avoidance of situations involving separation (e.g., going away to college) may persist for many years. However, the majority of children with Separation Anxiety Disorder are free of impairing Anxiety Disorders at extended follow-up.

Familial Pattern

Separation Anxiety Disorder is more common in first-degree biological relatives than in the general population and is relatively more frequent in children of mothers with Panic Disorder.

Differential Diagnosis

Separation anxiety can be an associated feature of **Pervasive Developmental Disorders, Schizophrenia, or other Psychotic Disorders**. If the symptoms of Separation Anxiety Disorder occur exclusively during the course of one of these disorders, a separate diagnosis of Separation Anxiety Disorder is not given. Separation Anxiety Disorder is distinguished from **Generalized Anxiety Disorder** in that the anxiety predominantly concerns separation from home and attachment figures. In children or adolescents with Separation Anxiety Disorder, threats of separation may lead to extreme anxiety and even a Panic Attack. In contrast to Panic Disorder, the anxiety concerns separation from attachment figures or from home rather than being incapacitated by an unexpected Panic Attack. In adults, Separation Anxiety Disorder is rare and should not be given as an additional diagnosis if the separation fears are better accounted for by Agoraphobia in **Panic Disorder With Agoraphobia or Agoraphobia Without History of Panic Disorder**. Truancy is common in **Conduct Disorder**, but anxiety about separation is not responsible for school absences and the child usually stays away from, rather than returns to, the home. Some cases of school refusal, especially in adolescence, are due to **Social Phobia or Mood Disorders** rather than separation anxiety. Children with Separation Anxiety Disorder may be oppositional in the context of being forced to separate from attachment figures. **Oppositional Defiant Disorder** should be diagnosed only if there is oppositional behavior at times other than at times of separation or when separation is anticipated. Similarly, children with Separation Anxiety Disorder may become depressed while being separated or in anticipation of separation. A **Depressive Disorder** should be diagnosed only if the depression occurs at other times.

Unlike the hallucinations in **Psychotic Disorders**, the unusual perceptual experiences in Separation Anxiety Disorder are usually based on a misperception of an actual stimulus, occur only in certain situations (e.g., nighttime), and are reversed by the presence of an attachment figure. Clinical judgment must be used in distinguishing **developmentally appropriate levels of separation anxiety** from the clinically significant concerns about separation seen in Separation Anxiety Disorder.

Diagnostic criteria for 309.21 Separation Anxiety Disorder

- A. Developmentally inappropriate and excessive anxiety concerning separation from home or from those to whom the individual is attached, as evidenced by three (or more) of the following:
- (1) recurrent excessive distress when separation from home or major attachment figures occurs or is anticipated
 - (2) persistent and excessive worry about losing, or about possible harm befalling, major attachment figures
 - (3) persistent and excessive worry that an untoward event will lead to separation from a major attachment figure (e.g., getting lost or being kidnapped)
 - (4) persistent reluctance or refusal to go to school or elsewhere because of fear of separation
 - (5) persistently and excessively fearful or reluctant to be alone or without major attachment figures at home or without significant adults in other settings
 - (6) persistent reluctance or refusal to go to sleep without being near a major attachment figure or to sleep away from home
 - (7) repeated nightmares involving the theme of separation
 - (8) repeated complaints of physical symptoms (such as headaches, stomachaches, nausea, or vomiting) when separation from major attachment figures occurs or is anticipated
- B. The duration of the disturbance is at least 4 weeks.
- C. The onset is before age 18 years.
- D. The disturbance causes clinically significant distress or impairment in social, academic (occupational), or other important areas of functioning.
- E. The disturbance does not occur exclusively during the course of a Pervasive Developmental Disorder, Schizophrenia, or other Psychotic Disorder and, in adolescents and adults, is not better accounted for by Panic Disorder With Agoraphobia.

Specify if:

Early Onset: if onset occurs before age 6 years

313.23 Selective Mutism (formerly Elective Mutism)

Diagnostic Features

The essential feature of Selective Mutism is the persistent failure to speak in specific social situations (e.g., school, with playmates) where speaking is expected, despite speaking in other situations (Criterion A). The disturbance interferes with educational or occupational achievement or with social communication (Criterion B). The disturbance must last for at least 1 month and is not limited to the first month of school (during which many children may be shy and reluctant to speak) (Criterion C). Selec-

tive Mutism should not be diagnosed if the individual's failure to speak is due solely to a lack of knowledge of, or comfort with, the spoken language required in the social situation (Criterion D). It is also not diagnosed if the disturbance is better accounted for by embarrassment related to having a Communication Disorder (e.g., Stuttering) or if it occurs exclusively during a Pervasive Developmental Disorder, Schizophrenia, or other Psychotic Disorder (Criterion E). Instead of communicating by standard verbalization, children with this disorder may communicate by gestures, nodding or shaking the head, or pulling or pushing, or, in some cases, by monosyllabic, short, or monotone utterances, or in an altered voice.

Associated Features and Disorders

Associated features of Selective Mutism may include excessive shyness, fear of social embarrassment, social isolation and withdrawal, clinging, compulsive traits, negativism, temper tantrums, or controlling or oppositional behavior, particularly at home. There may be severe impairment in social and school functioning. Teasing or scapegoating by peers is common. Although children with this disorder generally have normal language skills, there may occasionally be an associated Communication Disorder (e.g., Phonological Disorder, Expressive Language Disorder, or Mixed Receptive-Expressive Language Disorder) or a general medical condition that causes abnormalities of articulation. Mental Retardation, hospitalization, or extreme psychosocial stressors may be associated with the disorder. In addition, in clinical settings, children with Selective Mutism are almost always given an additional diagnosis of an Anxiety Disorder (especially Social Phobia).

Specific Culture and Gender Features

Immigrant children who are unfamiliar with or uncomfortable in the official language of their new host country may refuse to speak to strangers in their new environment. This behavior should not be diagnosed as Selective Mutism. Selective Mutism is slightly more common in females than in males.

Prevalence

Selective Mutism is apparently rare and is found in fewer than 1% of individuals seen in mental health settings.

Course

Onset of Selective Mutism is usually before age 5 years, but the disturbance may not come to clinical attention until entry into school. The degree of persistence of the disorder is variable. It may persist for only a few months or may continue for several years. In some cases, particularly in those with severe Social Phobia, anxiety symptoms may become chronic.

Differential Diagnosis

Selective Mutism should be distinguished from speech disturbances that are better accounted for by a **Communication Disorder**, such as **Phonological Disorder**, **Expressive Language Disorder**, **Mixed Receptive-Expressive Language Disorder**, or **Stuttering**. Unlike Selective Mutism, the speech disturbance in these conditions is not restricted to a specific social situation. Children in families who have immigrated to a country where a different language is spoken may refuse to speak the new language because of **lack of knowledge of the language**. If comprehension of the new language is adequate, but refusal to speak persists, a diagnosis of Selective Mutism may be warranted. Individuals with a **Pervasive Developmental Disorder**, **Schizophrenia** or other **Psychotic Disorder**, or **severe Mental Retardation** may have problems in social communication and be unable to speak appropriately in social situations. In contrast, Selective Mutism should only be diagnosed in a child who has an established capacity to speak in some social situations (e.g., typically at home). The social anxiety and social avoidance in **Social Phobia** may be associated with Selective Mutism. In such cases, both diagnoses may be given.

Diagnostic criteria for 313.23 Selective Mutism

- A. Consistent failure to speak in specific social situations (in which there is an expectation for speaking, e.g., at school) despite speaking in other situations.
 - B. The disturbance interferes with educational or occupational achievement or with social communication.
 - C. The duration of the disturbance is at least 1 month (not limited to the first month of school).
 - D. The failure to speak is not due to a lack of knowledge of, or comfort with, the spoken language required in the social situation.
 - E. The disturbance is not better accounted for by a Communication Disorder (e.g., Stuttering) and does not occur exclusively during the course of a Pervasive Developmental Disorder, Schizophrenia, or other Psychotic Disorder.
-

313.89 Reactive Attachment Disorder of Infancy or Early Childhood

Diagnostic Features

The essential feature of Reactive Attachment Disorder is markedly disturbed and developmentally inappropriate social relatedness in most contexts that begins before age 5 years and is associated with grossly pathological care (Criterion A). There are two types of presentations. In the Inhibited Type, the child persistently fails to initiate and to respond to most social interactions in a developmentally appropriate way. The

child shows a pattern of excessively inhibited, hypervigilant, or highly ambivalent responses (e.g., frozen watchfulness, resistance to comfort, or a mixture of approach and avoidance) (Criterion A1). In the Disinhibited Type, there is a pattern of diffuse attachments. The child exhibits indiscriminate sociability or a lack of selectivity in the choice of attachment figures (Criterion A2). The disturbance is not accounted for solely by developmental delay (e.g., as in Mental Retardation) and does not meet criteria for Pervasive Developmental Disorder (Criterion B). By definition, the condition is associated with grossly pathological care that may take the form of persistent disregard of the child's basic emotional needs for comfort, stimulation, and affection (Criterion C1); persistent disregard of the child's basic physical needs (Criterion C2); or repeated changes of primary caregiver that prevent formation of stable attachments (e.g., frequent changes in foster care) (Criterion C3). The pathological care is presumed to be responsible for the disturbed social relatedness (Criterion D).

Subtypes

The predominant type of disturbance in social relatedness may be indicated by specifying one of the following subtypes for Reactive Attachment Disorder:

Inhibited Type. In this subtype, the predominant disturbance in social relatedness is the persistent failure to initiate and to respond to most social interactions in a developmentally appropriate way.

Disinhibited Type. This subtype is used if the predominant disturbance in social relatedness is indiscriminate sociability or a lack of selectivity in the choice of attachment figures.

Associated Features and Disorders

Associated descriptive features and mental disorders. Certain situations (e.g., prolonged hospitalization of the child, extreme poverty, or parental inexperience) may predispose to the development of pathological care. However, grossly pathological care does not always result in the development of Reactive Attachment Disorder; some children may form stable attachments and social relationships even in the face of marked neglect or abuse. Extreme neglect—and especially institutional care with limited opportunities to form selective attachments—increases risk of developing the disorder. Reactive Attachment Disorder may be associated with developmental delays, Feeding Disorder of Infancy or Early Childhood, Pica, or Rumination Disorder.

Associated laboratory findings. Laboratory findings consistent with malnutrition may be present.

Associated physical examination findings and general medical conditions. Physical examination may document associated general medical conditions that are associated with extreme neglect (e.g., growth delay, evidence of physical abuse, malnutrition, vitamin deficiencies, or infectious diseases).

Prevalence

Epidemiological data are limited, but Reactive Attachment Disorder appears to be very uncommon.

Course

The onset of Reactive Attachment Disorder is usually in the first several years of life and, by definition, begins before age 5 years. The course appears to vary depending on individual factors in child and caregivers, the severity and duration of associated psychosocial deprivation, and the nature of intervention. Considerable improvement or remission may occur if an appropriately supportive environment is provided. Otherwise, the disorder follows a continuous course. Indiscriminate sociability may persist even after the child has developed selective attachments.

Differential Diagnosis

In **Mental Retardation**, appropriate attachments to caregivers usually develop consistent with the child's general developmental level, and these attachments are clearly present by the time a child has a mental age of 10 months. However, some infants and young children with Severe Mental Retardation may present particular problems for caregivers and exhibit symptoms characteristic of Reactive Attachment Disorder. Reactive Attachment Disorder should be diagnosed only if it is clear that the characteristic problems in formation of selective attachments are not a function of the retardation.

Reactive Attachment Disorder must be differentiated from **Autistic Disorder** and **other Pervasive Developmental Disorders**. In the Pervasive Developmental Disorders, selective attachments either fail to develop or are highly deviant, but this usually occurs in the face of a reasonably supportive psychosocial environment. Autistic Disorder and other Pervasive Developmental Disorders are also characterized by the presence of a qualitative impairment in communication and restricted, repetitive, and stereotyped patterns of behavior. Reactive Attachment Disorder is not diagnosed if the criteria are met for a Pervasive Developmental Disorder.

The Inhibited Type of Reactive Attachment Disorder must be distinguished from **Social Phobia**. In Social Phobia, the social inhibition is apparent in social settings or in anticipation of social encounters but does not occur with familiar caregivers in familiar settings. Socially deviant behavior in Reactive Attachment Disorder, including inhibition, is apparent across social contexts.

The Disinhibited Type must be distinguished from the impulsive or hyperactive behavior characteristic of **Attention-Deficit/Hyperactivity Disorder**. In contrast to Attention-Deficit/Hyperactivity Disorder, the disinhibited behavior in Reactive Attachment Disorder is characteristically associated with being overly familiar with or seeking comfort from an unfamiliar adult caregiver rather than with generally impulsive behavior.

Reactive Attachment Disorder should also be differentiated from Disruptive Behavior Disorders such as **Conduct Disorder** and **Oppositional Defiant Disorder**. The term "affectionless psychopath" has been used to describe children who were

raised in settings that limited opportunities for the child to develop selective attachments (e.g., institutions) and who exhibited a pattern of antisocial and aggressive behavior, inability to form lasting relationships with adults, and miscellaneous symptoms such as enuresis and stereotypies. Nevertheless, no direct link between Reactive Attachment Disorder and "affectionless psychopathy" has been established. Disturbances of attachment in the early years may increase the risk for antisocial behaviors in later childhood and adolescence, but antisocial behaviors are not necessarily signs of Reactive Attachment Disorder. Grossly pathogenic care is a defining feature of Reactive Attachment Disorder. An additional notation of Child Abuse, Child Neglect, or Parent-Child Relational Problem may be warranted. When grossly pathogenic care does not result in marked disturbances in social relatedness, Child Neglect or Parent-Child Relational Problem may be noted rather than Reactive Attachment Disorder.

Diagnostic criteria for 313.89 Reactive Attachment Disorder of Infancy or Early Childhood

- A. Markedly disturbed and developmentally inappropriate social relatedness in most contexts, beginning before age 5 years, as evidenced by either (1) or (2):
 - (1) persistent failure to initiate or respond in a developmentally appropriate fashion to most social interactions, as manifest by excessively inhibited, hypervigilant, or highly ambivalent and contradictory responses (e.g., the child may respond to caregivers with a mixture of approach, avoidance, and resistance to comforting, or may exhibit frozen watchfulness)
 - (2) diffuse attachments as manifest by indiscriminate sociability with marked inability to exhibit appropriate selective attachments (e.g., excessive familiarity with relative strangers or lack of selectivity in choice of attachment figures)
- B. The disturbance in Criterion A is not accounted for solely by developmental delay (as in Mental Retardation) and does not meet criteria for a Pervasive Developmental Disorder.
- C. Pathogenic care as evidenced by at least one of the following:
 - (1) persistent disregard of the child's basic emotional needs for comfort, stimulation, and affection
 - (2) persistent disregard of the child's basic physical needs
 - (3) repeated changes of primary caregiver that prevent formation of stable attachments (e.g., frequent changes in foster care)
- D. There is a presumption that the care in Criterion C is responsible for the disturbed behavior in Criterion A (e.g., the disturbances in Criterion A began following the pathogenic care in Criterion C).

Specify type:

Inhibited Type: if Criterion A1 predominates in the clinical presentation

Disinhibited Type: if Criterion A2 predominates in the clinical presentation

307.3 Stereotypic Movement Disorder (formerly Stereotypy/Habit Disorder)

Diagnostic Features

The essential feature of Stereotypic Movement Disorder is motor behavior that is repetitive, often seemingly driven, and nonfunctional (Criterion A). This motor behavior markedly interferes with normal activities or results in self-inflicted bodily injury that is significant enough to require medical treatment (or would result in such injury if protective measures were not used) (Criterion B). If Mental Retardation is present, the stereotypic or self-injurious behavior is sufficiently severe to become a focus of treatment (Criterion C). The behavior is not better accounted for by a compulsion (as in Obsessive-Compulsive Disorder), a tic (as in the Tic Disorders), a stereotypy that is part of a Pervasive Developmental Disorder, or hair pulling (as in Trichotillomania) (Criterion D). The behavior is also not due to the direct physiological effects of a substance or a general medical condition (Criterion E). The motor behaviors must persist for at least 4 weeks (Criterion F).

The stereotypic movements may include hand waving, rocking, playing with hands, fiddling with fingers, twirling objects, head banging, self-biting, or hitting various parts of one's own body. Sometimes the individual uses an object in performing these behaviors. The behaviors may cause permanent and disabling tissue damage and may sometimes be life-threatening. For instance, severe head banging or hitting may lead to cuts, bleeding, infection, retinal detachment, and blindness.

Specifiers

The clinician may specify **With Self-Injurious Behavior** if the behavior results in bodily damage that requires specific treatment (or that would result in bodily damage if protective measures were not used).

Associated Features and Disorders

Associated descriptive features and mental disorders. The individual (especially an individual with Lesch-Nyhan syndrome) may develop methods of self-restraint (e.g., holding hands inside shirts, trousers, or in pockets) to attempt to control the self-injurious behaviors. When the self-restraint is interfered with, the behaviors return. If the behaviors are extreme or repulsive to others, there may be psychosocial complications due to the individual's exclusion from social and community activities. Stereotypic Movement Disorder occurs most commonly in association with Mental Retardation. The more severe the retardation, the higher the risk for self-injurious behaviors. The disorder can also occur in non-developmentally delayed populations (e.g., individuals with body rocking associated with Generalized Anxiety Disorder).

This disorder may also occur in association with severe sensory deficits (blindness and deafness) and may be more common in institutional environments in which the individual receives insufficient stimulation. Self-injurious behaviors occur in certain general medical conditions associated with Mental Retardation (e.g., fragile X syn-

drome, Down syndrome, de Lange syndrome, and especially Lesch-Nyhan syndrome, which is characterized by severe self-biting).

Associated laboratory findings. If there is self-injury, the laboratory findings will reflect its nature and severity (e.g., anemia may be present if there is a chronic blood loss from self-inflicted rectal bleeding).

Associated physical examination findings and general medical conditions. Signs of chronic tissue damage may be present (e.g., bruises, bite marks, cuts, scratches, skin infections, rectal fissures, foreign bodies in bodily orifices, visual impairment due to eye gouging or traumatic cataract, and fractures or deformed bones). In less severe cases, there may be a chronic skin irritation or calluses from biting, pinching, scratching, or saliva smearing.

Specific Age and Gender Features

Self-injurious behaviors occur in individuals of all ages. There are indications that head banging is more prevalent in males (with about a 3:1 ratio), and self-biting may be more prevalent in females.

Prevalence

There is limited information on the prevalence of Stereotypic Movement Disorder. The estimates of prevalence of self-injurious behaviors in individuals with Mental Retardation vary from 2% and 3% in children and adolescents living in the community to approximately 25% in adults with severe or profound Mental Retardation living in institutions.

Course

There is no typical age at onset or pattern of onset for Stereotypic Movement Disorder. The onset may follow a stressful environmental event. In nonverbal individuals with Severe Mental Retardation, stereotypic movements may be triggered by a painful general medical condition (e.g., a middle ear infection leading to head banging). The stereotypic movements often peak in adolescence and then may gradually decline. However, especially in individuals with Severe or Profound Mental Retardation, the movements may persist for years. The focus of these behaviors often changes (e.g., a person may engage in hand biting that may then subside and head hitting may emerge).

Differential Diagnosis

Stereotypic movements may be associated with **Mental Retardation**, especially for individuals in nonstimulating environments. Stereotypic Movement Disorder should be diagnosed only in individuals in whom the stereotypic or self-injurious behavior is of sufficient severity to become a focus of treatment. Repetitive stereotyped movements are a characteristic feature of **Pervasive Developmental Disorders**. Stereotyp-

ic Movement Disorder is not diagnosed if the stereotypies are better accounted for by a Pervasive Developmental Disorder. Compulsions in **Obsessive-Compulsive Disorder** are generally more complex and ritualistic and are performed in response to an obsession or according to rules that must be applied rigidly. Differentiating the complex movements characteristic of Stereotypic Movement Disorder from **simple tics** (e.g., eye blinking) is relatively straightforward. However, differentiating Stereotypic Movement Disorder from **complex motor tics** can be quite difficult, given the similarities between the two in terms of intentionality, rhythmicity, and drivenness.

In **Trichotillomania**, by definition, the repetitive behavior is limited to hair pulling. The self-induced injuries in Stereotypic Movement Disorder should be distinguished from **Factitious Disorder With Predominantly Physical Signs and Symptoms**, in which the motivation of the self-injury is to assume the sick role. **Self-mutilation associated with certain Psychotic Disorders and Personality Disorders** is premeditated, complex, and sporadic and has a meaning for the individual within the context of the underlying, severe mental disorder (e.g., is the result of delusional thinking). **Involuntary movements associated with neurological conditions** (such as Huntington's disease) usually follow a typical pattern, and the signs and symptoms of the neurological condition are present. **Tardive Dyskinesia** usually results from chronic neuroleptic use and consists of characteristic orofacial dyskinesias or, less commonly, irregular truncal or limb movements. In addition, these types of movements do not result in direct self-injury.

Developmentally appropriate self-stimulatory behaviors in young children (e.g., thumb sucking, rocking, and head banging) are usually self-limited and rarely result in tissue damage requiring treatment. **Self-directed behaviors in individuals with sensory deficits** (e.g., blindness) are repetitive and stereotyped but usually do not result in dysfunction or in self-injury.

Many people engage in **repetitive behaviors for various reasons** (practicing to improve a motor skill, culturally sanctioned practices). In contrast to Stereotypic Movement Disorder, these behaviors do not interfere with normal activities nor do they result in self-injury.

Diagnostic criteria for 307.3 Stereotypic Movement Disorder

- A. Repetitive, seemingly driven, and nonfunctional motor behavior (e.g., hand shaking or waving, body rocking, head banging, mouthing of objects, self-biting, picking at skin or bodily orifices, hitting own body).
- B. The behavior markedly interferes with normal activities or results in self-inflicted bodily injury that requires medical treatment (or would result in an injury if preventive measures were not used).
- C. If Mental Retardation is present, the stereotypic or self-injurious behavior is of sufficient severity to become a focus of treatment.
- D. The behavior is not better accounted for by a compulsion (as in Obsessive-Compulsive Disorder), a tic (as in Tic Disorder), a stereotypy that is part of a Pervasive Developmental Disorder, or hair pulling (as in Trichotillomania).
- E. The behavior is not due to the direct physiological effects of a substance or a general medical condition.
- F. The behavior persists for 4 weeks or longer.

Specify if:

With Self-Injurious Behavior: if the behavior results in bodily damage that requires specific treatment (or that would result in bodily damage if protective measures were not used)

313.9 Disorder of Infancy, Childhood, or Adolescence Not Otherwise Specified

This category is a residual category for disorders with onset in infancy, childhood, or adolescence that do not meet criteria for any specific disorder in the Classification.

Delirium, Dementia, and Amnestic and Other Cognitive Disorders

This section includes Delirium, Dementia, Amnestic Disorders, and Cognitive Disorder Not Otherwise Specified. The predominant disturbance is a clinically significant deficit in cognition that represents a significant change from a previous level of functioning. For each disorder in this section, the etiology is either a general medical condition (although the specific general medical condition may not be identifiable) or a substance (i.e., a drug of abuse, medication, or toxin), or a combination of these factors.

In DSM-III-R, these disorders were placed in a section titled "Organic Mental Syndromes and Disorders." The term *organic mental disorder* is no longer used in DSM-IV because it incorrectly implies that "nonorganic" mental disorders do not have a biological basis. In DSM-IV, disorders formerly called "organic mental disorders" have been grouped into three sections: 1) Delirium, Dementia, and Amnestic and Other Cognitive Disorders; 2) Mental Disorders Due to a General Medical Condition; and 3) Substance-Related Disorders.

A **delirium** is characterized by a disturbance of consciousness and a change in cognition that develop over a short period of time. The disorders included in the "Delirium" section are listed according to presumed etiology: Delirium Due to a General Medical Condition, Substance-Induced Delirium (i.e., due to a drug of abuse, a medication, or toxin exposure), Delirium Due to Multiple Etiologies, or Delirium Not Otherwise Specified (if the etiology is indeterminate).

A **dementia** is characterized by multiple cognitive deficits that include impairment in memory. The dementias are also listed according to presumed etiology: Dementia of the Alzheimer's Type, Vascular Dementia, Dementia Due to Other General Medical Conditions (e.g., human immunodeficiency virus [HIV] disease, head trauma, Parkinson's disease, Huntington's disease), Substance-Induced Persisting Dementia (i.e., due to a drug of abuse, a medication, or toxin exposure), Dementia Due to Multiple Etiologies, or Dementia Not Otherwise Specified (if the etiology is indeterminate).

An **amnestic disorder** is characterized by memory impairment in the absence of other significant accompanying cognitive impairments. The disorders in the "Amnestic Disorders" section also are listed according to presumed etiology: Amnestic Disorder Due to a General Medical Condition, Substance-Induced Persisting Amnestic Disorder, or Amnestic Disorder Not Otherwise Specified.

Cognitive Disorder Not Otherwise Specified is for presentations that are characterized by cognitive dysfunction presumed to be due to either a general medical condition or substance use that do not meet criteria for any of the disorders listed elsewhere in this section.

Introductory text is provided that discusses the general features for each group of disorders, regardless of etiology. This is followed by text and criteria for each disorder with specific etiology.

Delirium

The disorders in the "Delirium" section share a common symptom presentation of a disturbance in consciousness and cognition, but are differentiated based on etiology: **Delirium Due to a General Medical Condition**, **Substance-Induced Delirium** (including medication side effects), and **Delirium Due to Multiple Etiologies**. In addition, **Delirium Not Otherwise Specified** is included in this section for presentations in which the clinician is unable to determine a specific etiology for the delirium.

Diagnostic Features

The essential feature of a delirium is a disturbance of consciousness that is accompanied by a change in cognition that cannot be better accounted for by a preexisting or evolving dementia. The disturbance develops over a short period of time, usually hours to days, and tends to fluctuate during the course of the day. There is evidence from the history, physical examination, or laboratory tests that the delirium is a direct physiological consequence of a general medical condition, Substance Intoxication or Withdrawal, use of a medication, or toxin exposure, or a combination of these factors.

The disturbance in consciousness is manifested by a reduced clarity of awareness of the environment. The ability to focus, sustain, or shift attention is impaired (Criterion A). Questions must be repeated because the individual's attention wanders, or the individual may perseverate with an answer to a previous question rather than appropriately shift attention. The person is easily distracted by irrelevant stimuli. Because of these problems, it may be difficult (or impossible) to engage the person in conversation.

There is an accompanying change in cognition (which may include memory impairment, disorientation, or language disturbance) or development of a perceptual disturbance (Criterion B). Memory impairment is most commonly evident in recent memory and can be tested by asking the person to remember several unrelated objects or a brief sentence, and then to repeat them after a few minutes of distraction. Disorientation is usually manifested by the individual's being disoriented to time (e.g., thinking it is morning in the middle of the night) or being disoriented to place (e.g., thinking he or she is home rather than in a hospital). In mild delirium, disorientation to time may be the first symptom to appear. Disorientation to self is less common. Speech or language disturbances may be evident as dysarthria (i.e., the impaired ability to articulate), dysnomia (i.e., the impaired ability to name objects),

dysgraphia (i.e., the impaired ability to write), or even aphasia. In some cases, speech is rambling and irrelevant, in others pressured and incoherent, with unpredictable switching from subject to subject. It may be difficult for the clinician to assess for changes in cognitive function because the individual may be inattentive and incoherent. Under these circumstances, it is helpful to review carefully the individual's history and to obtain information from other informants, particularly family members.

Perceptual disturbances may include misinterpretations, illusions, or hallucinations. For example, the banging of a door may be mistaken for a gunshot (misinterpretation); the folds of the bedclothes may appear to be animate objects (illusion); or the person may "see" a group of people hovering over the bed when no one is actually there (hallucination). Although sensory misperceptions are most commonly visual, they may occur in other sensory modalities as well, such as auditory, tactile, gustatory, and olfactory. Misperceptions range from simple and uniform to highly complex. The individual may have a delusional conviction of the reality of the hallucinations and exhibit emotional and behavioral responses consistent with their content.

The disturbance develops over a short period of time and tends to fluctuate during the course of the day (Criterion C). For example, during morning hospital rounds, the person may be coherent and cooperative, but at night might insist on pulling out intravenous lines and going home to parents who died years ago.

Associated Features and Disorders

Delirium is often associated with a disturbance in the sleep-wake cycle. This disturbance can include daytime sleepiness or nighttime agitation and difficulty falling asleep or excessive sleepiness throughout the day or wakefulness throughout the night. In some cases, complete reversal of the night-day sleep-wake cycle can occur. Delirium is frequently accompanied by disturbed psychomotor behavior. Many individuals with delirium are restless or hyperactive. Manifestations of increased psychomotor activity may include groping or picking at the bedclothes, attempting to get out of bed when it is unsafe or untimely, and sudden movements. On the other hand, the individual may show decreased psychomotor activity, with sluggishness and lethargy that approach stupor. Psychomotor activity often shifts from one extreme to the other over the course of a day. While hyperactive, the individual is more likely to show evidence of hallucinations, delusions, and agitation, whereas in hypoactive states, the individual is less likely to show evidence of hallucinations, delusions, or other perceptual disturbances. Comparable levels of cognitive impairment have been observed with both the hyperactive and the hypoactive states.

The individual may exhibit emotional disturbances such as anxiety, fear, depression, irritability, anger, euphoria, and apathy. There may be rapid and unpredictable shifts from one emotional state to another, although some individuals with delirium have a constant emotional tone. Fear often accompanies threatening hallucinations or transient delusions. If fear is marked, the person may attack those who are falsely perceived as threatening. Injuries may be sustained from falling out of bed or trying to escape while attached to intravenous lines, respiratory tubes, urinary catheters, or other medical equipment. The disturbed emotional state may also be evident in calling out, screaming, cursing, muttering, moaning, or other sounds. These behaviors

are especially prevalent at night and under conditions in which stimulation and environmental cues are lacking. Impaired judgment may interfere with proper medical treatment. Depending on the etiology, delirium can be associated with a number of nonspecific neurological abnormalities, such as tremor, myoclonus, asterixis, and reflex or muscle tone changes.

In addition to laboratory findings that are characteristic of associated or etiological general medical conditions (or intoxication or withdrawal states), the EEG is typically abnormal, showing generalized slowing. Fast activity is occasionally found, for example, in some cases of Alcohol Withdrawal Delirium.

Specific Culture, Age, and Gender Features

Cultural and educational background should be taken into consideration in the evaluation of an individual's mental capacity. Individuals from certain backgrounds may not be familiar with the information used in certain tests of general knowledge (e.g., names of presidents, geographical knowledge), memory (e.g., date of birth in cultures that do not routinely celebrate birthdays), and orientation (e.g., sense of placement and location may be conceptualized differently in some cultures).

Children may be particularly susceptible to delirium compared with adults (other than the elderly), especially when it is related to febrile illnesses and certain medications (e.g., anticholinergics). This is perhaps due to their immature brain development and physiological differences. In children, delirium may be mistaken for uncooperative behavior, and eliciting the distinctive cognitive signs may be difficult. If familiar figures cannot soothe the child, this may be suggestive of delirium. The elderly are also especially susceptible to delirium compared with younger adults, perhaps because of physiological differences. Increasing age in adults is associated with higher rates of delirium even after controlling for other risk factors. Male gender appears to be an independent risk factor for delirium in elderly individuals.

Prevalence

The point prevalence of delirium in the general population is 0.4% in adults age 18 years and older and 1.1% in those age 55 and older. The point prevalence of delirium in the hospitalized medically ill ranges from 10% to 30%. In the hospitalized elderly, about 10%–15% are reported to exhibit delirium on admission, and 10%–40% may be diagnosed with delirium while in the hospital. Up to 60% of nursing home residents age 75 years and older may be delirious at any given time. As many as 25% of hospitalized cancer patients and 30%–40% of hospitalized AIDS patients develop delirium during their hospitalization. Up to 80% of those with terminal illness develop delirium near death. The rate of delirium in these populations depends greatly on the nature of their associated general medical conditions and surgical procedures.

Course

The symptoms of delirium usually develop over hours to days, although in some individuals they may begin abruptly (e.g., after a head injury). More typically, prodromal symptoms, such as restlessness, anxiety, irritability, disorientation, distracti-

bility, or sleep disturbance, progress to full-blown delirium within a 1- to 3-day period. The delirium may resolve in a few hours to days, or symptoms may persist for weeks to months, particularly in elderly individuals and individuals with coexisting dementia. If the underlying etiological factor is promptly corrected or is self-limited, recovery is more likely to be complete and more rapid. Individuals with better premorbid cognitive and physical functioning have better recovery from delirium. Those with previous episodes of delirium may be at increased risk for recurrent symptoms.

While the majority of individuals have a full recovery, delirium may progress to stupor, coma, seizures, or death, particularly if the underlying cause is untreated. Full recovery is less likely in the elderly, with estimated rates of full recovery by the time of hospital discharge varying from 4% to 40%. Many symptoms will not have resolved by 3–6 months after discharge. Persistent cognitive deficits are also common in elderly individuals recovering from delirium, although such deficits may be due to preexisting dementia that was not fully appreciated. Being admitted to a hospital from home (as opposed to from an institutional setting) is related to a higher rate of improvement in mental state.

Delirium in the medically ill is associated with significant morbidity. Medically ill individuals with delirium, particularly the elderly, have significantly increased risk of medical complications, such as pneumonia and decubitus ulcers, resulting in longer duration of hospital stays. Delirium is also associated with increased functional decline and risk of institutional placement. Hospitalized patients 65 years or older with delirium have three times the risk of nursing home placement and about three times the functional decline as hospitalized patients without delirium at both discharge and 3 months postdischarge. In postoperative patients, delirium is a harbinger of limited recovery and poor long-term outcome and is often associated with increased risk for postoperative complications, longer postoperative recuperation periods, longer hospital stays, and increased long-term disability.

Delirium in the medically ill is also associated with increased mortality. Elderly individuals who develop delirium during a hospitalization may have up to a 20%–75% chance of dying during that hospitalization. Patients who develop delirium during a hospitalization also have a very high death rate during the months following discharge. Up to 15% of elderly patients with delirium die within a 1-month period and up to 25% die within a 6-month period after discharge. Other risk factors, such as type of illness, illness severity, preexisting cognitive impairment, and age, contribute significantly to this association. Patients with malignancies and delirium have a particularly high mortality rate both in-hospital and after discharge compared with patients with malignancies who do not also have delirium.

Differential Diagnosis

The most common differential diagnostic issue is whether the person has a **dementia** rather than a delirium, has a delirium alone, or has a delirium superimposed on a preexisting dementia. Memory impairment is common to both a delirium and a dementia, but the person with a dementia alone is alert and does not have the disturbance in consciousness that is characteristic of a delirium. The temporal onset and course of cognitive impairments are helpful in distinguishing between delirium and dementia.

In delirium the onset of symptoms is much more rapid (i.e., usually over hours to days), whereas in dementia the onset is typically more gradual or insidious. Delirium symptom severity characteristically fluctuates during a 24-hour period, whereas dementia symptom severity generally does not. When symptoms of a delirium are present, information from family members, other caretakers, or medical records may be helpful in determining whether the symptoms of a dementia were preexisting. Coding of a delirium superimposed on the different types of dementias is discussed under "Recording Procedures" for each type of delirium.

The presumed etiology determines the specific delirium diagnosis (text and criteria for each delirium diagnosis are provided separately later in this section). If it is judged that the delirium is a consequence of the direct physiological effects of a general medical condition, then **Delirium Due to a General Medical Condition** is diagnosed. If the delirium results from the direct physiological effects of a drug of abuse, then **Substance Intoxication Delirium** or **Substance Withdrawal Delirium** is diagnosed, depending on whether the delirium occurred in association with Substance Intoxication or Substance Withdrawal. If the delirium results from medication use or toxin exposure, then **Substance-Induced Delirium** is diagnosed. It is not uncommon for the delirium to be due to both a general medical condition and substance (including medication) use. This may be seen, for example, in an elderly individual with a serious general medical condition that is being treated with multiple medications. When there is more than one etiology (e.g., both a substance and a general medical condition), **Delirium Due to Multiple Etiologies** is diagnosed. If it is not possible to establish a specific etiology (i.e., substance induced or due to a general medical condition), **Delirium Not Otherwise Specified** is diagnosed.

The diagnosis of Substance Intoxication Delirium or Substance Withdrawal Delirium is made instead of **Substance Intoxication** or **Substance Withdrawal** only if the symptoms of the delirium are in excess of those usually associated with the intoxication or withdrawal syndrome and are sufficiently severe to warrant independent clinical attention. Even in individuals with obvious signs of intoxication or withdrawal, other possible causes of the delirium (i.e., Delirium Due to a General Medical Condition) must not be overlooked. For example, a head injury that occurs as a result of falls or fighting during intoxication may be responsible for the delirium.

Delirium that is characterized by vivid hallucinations, delusions, language disturbances, and agitation must be distinguished from **Brief Psychotic Disorder**, **Schizophrenia**, **Schizophreniform Disorder**, and other **Psychotic Disorders**, as well as from **Mood Disorders With Psychotic Features**. In delirium, the psychotic symptoms are fragmented and unsystematized. Delirium that is characterized by mood changes and anxiety must also be distinguished from **Mood Disorders** and **Anxiety Disorders**. Finally, delirium associated with fear, anxiety, and dissociative symptoms such as depersonalization must be distinguished from **Acute Stress Disorder**, which is precipitated by exposure to a severely traumatic event. Psychotic, mood, anxiety, and dissociative symptoms associated with delirium typically fluctuate, occur in the context of a reduced ability to appropriately maintain and shift attention, and are usually associated with EEG abnormalities. There is often memory impairment and disorientation in delirium, but generally not in these other disorders. Finally, in delirium, the person generally shows evidence of an underlying general medical condition, Substance Intoxication or Withdrawal, or medication use.

Delirium must be distinguished from **Malingering** and from **Factitious Disorder**. This distinction is made based on the often atypical presentation in Malingering and Factitious Disorder and the absence of a general medical condition or substance that is etiologically related to the apparent cognitive disturbance.

Individuals may present with some but not all symptoms of delirium. Subsyndromal presentations need to be carefully assessed because they may be harbingers of a full-blown delirium or may signal an as yet undiagnosed underlying general medical condition. Such presentations should be coded as **Cognitive Disorder Not Otherwise Specified**.

293.0 Delirium Due to a General Medical Condition

Diagnostic and Associated Features

The descriptive features of Delirium Due to a General Medical Condition (Criteria A–C) are discussed on pp. 136–137. In addition, to diagnose Delirium Due to a General Medical Condition, there must be evidence from the history, physical examination, or laboratory findings that the cognitive disturbance is the direct physiological consequence of a general medical condition (Criterion D).

In determining whether the delirium is due to a general medical condition, the clinician must first establish the presence of a general medical condition. Further, the clinician must establish that the delirium is etiologically related to the general medical condition. A careful and comprehensive assessment of multiple factors is necessary to make this judgment. Although there are no infallible guidelines, several considerations provide some guidance in this area. One consideration is the presence of a temporal association between the onset, exacerbation, or remission of the general medical condition and that of the delirium. Evidence from the literature that suggests that there can be a direct association between the general medical condition in question and the development of a delirium can provide a useful context in the assessment of a particular situation. In addition, the clinician must also judge that the disturbance is not better accounted for by a Substance-Induced Delirium or a primary mental disorder (e.g., a Manic Episode). This determination is explained in greater detail in the “Mental Disorders Due to a General Medical Condition” section (p. 181).

Delirium can be associated with many different general medical conditions, each of which has characteristic physical examination and laboratory findings. In systemic illnesses, focal neurological signs are not usually found. Various forms of tremor may be present. Asterixis, a flapping movement of the hyperextended hands, was originally described in hepatic encephalopathy but may also be found in association with other causes of delirium. Signs of autonomic hyperactivity (e.g., tachycardia, sweating, flushed face, dilated pupils, and elevated blood pressure) commonly occur. In addition to laboratory findings that are characteristic of etiological general medical conditions (or intoxication or withdrawal states), the EEG is generally abnormal, showing either generalized slowing or fast activity.

Recording Procedures

In recording the diagnosis of Delirium Due to a General Medical Condition, the clinician should note both the delirium and the identified general medical condition judged to be causing the disturbance on Axis I (e.g., 293.0 Delirium Due to Hypoglycemia). The ICD-9-CM code for the general medical condition should also be noted on Axis III (e.g., 251.2 hypoglycemia.) (See Appendix G for a list of selected ICD-9-CM diagnostic codes for general medical conditions.) When the delirium is superimposed on a preexisting dementia, both diagnoses should be made (e.g., 294.11 Dementia of the Alzheimer's Type, With Behavioral Disturbance, and 293.0 Delirium Due to Hyponatremia). Since Alzheimer's disease is not an established etiology for delirium but only a risk factor, the etiology of any delirium superimposed on Alzheimer's disease must be determined. Because of ICD-9-CM coding requirements, delirium superimposed on Vascular Dementia is noted by coding the appropriate subtype of the dementia (e.g., 290.41 Vascular Dementia, With Delirium). In situations in which it is unclear whether the cognitive deficits are due to delirium or to dementia, it may be useful to make a provisional diagnosis of delirium and observe the person carefully while continuing efforts to identify the nature of the disturbance.

Associated General Medical Conditions

General medical conditions associated with delirium include central nervous system disorders (e.g., head trauma, ictal and postictal states, vascular diseases such as stroke and hypertensive encephalopathy, degenerative diseases such as Pick's disease, infection, brain tumor), metabolic disorders (e.g., renal or hepatic disease, fluid or electrolyte imbalance such as dehydration, sodium or potassium imbalance, anemia, hypoxia, hypercarbia, hypoglycemia, thiamine deficiency, hypoalbuminemia, endocrinopathy, acid-base imbalance), cardiopulmonary disorders (myocardial infarction, congestive heart failure, cardiac arrhythmia, shock, respiratory failure), and systemic illness or effects (e.g., infection such as septicemia, pneumonia, and urinary tract infection; neoplasm; severe trauma; sensory deprivation such as visual and hearing impairment; temperature dysregulation; postoperative state). Certain focal lesions of the right parietal lobe and inferomedial surface of the occipital lobe also may lead to a delirium.

Differential Diagnosis

See p. 139 for a general discussion of the differential diagnosis of delirium.

Diagnostic criteria for 293.0 Delirium Due to . . .
[Indicate the General Medical Condition]

- A. Disturbance of consciousness (i.e., reduced clarity of awareness of the environment) with reduced ability to focus, sustain, or shift attention.
- B. A change in cognition (such as memory deficit, disorientation, language disturbance) or the development of a perceptual disturbance that is not better accounted for by a preexisting, established, or evolving dementia.
- C. The disturbance develops over a short period of time (usually hours to days) and tends to fluctuate during the course of the day.
- D. There is evidence from the history, physical examination, or laboratory findings that the disturbance is caused by the direct physiological consequences of a general medical condition.

Coding note: If delirium is superimposed on a preexisting Vascular Dementia, indicate the delirium by coding 290.41 Vascular Dementia, With Delirium.

Coding note: Include the name of the general medical condition on Axis I, e.g., 293.0 Delirium Due to Hepatic Encephalopathy; also code the general medical condition on Axis III (see Appendix G for codes).

Substance-Induced Delirium

Diagnostic and Associated Features

The descriptive features of Substance-Induced Delirium (Criteria A–C) are discussed on pp. 136–137. In addition, to diagnose Substance-Induced Delirium, there must be evidence from the history, physical examination, or laboratory findings of Substance Intoxication or Withdrawal, medication side effects, or toxin exposure judged to be etiologically related to the delirium (Criterion D). A delirium that occurs during Substance Intoxication is diagnosed as Substance Intoxication Delirium, and a delirium that occurs during Substance Withdrawal is diagnosed as Substance Withdrawal Delirium. A delirium that is associated with medication side effects or toxin exposure is diagnosed as Substance-Induced Delirium (see criteria set for Substance Intoxication Delirium, p. 145).

Delirium that occurs during Substance Intoxication usually arises within minutes to hours after taking relatively high doses of certain drugs such as cannabis, cocaine, and hallucinogens. However, onset can also be delayed for some substances that can accumulate over time because they have long half-lives (e.g., diazepam). Usually the delirium resolves as the intoxication ends or within a few hours to days. However, the duration may be longer after intoxication with phencyclidine and might persist for longer periods for individuals with brain damage, in the elderly, and in individuals taking combinations of substances. The time between taking a substance and onset of Intoxication Delirium may be shorter in individuals having poor clearance (e.g., due to renal or hepatic disease).

Delirium that is associated with Substance Withdrawal develops as tissue and fluid concentrations of the substance decrease after reduction or termination of sustained, usually high-dose use of alcohol or sedative, hypnotic, or anxiolytic drugs. In individuals having poor clearance, experiencing drug interactions, or taking combinations of substances, Substance Withdrawal Delirium can occur after the reduction or termination of lower doses. The duration of the delirium tends to vary with the half-life of the substance involved: longer-acting substances usually are associated with more protracted withdrawal. Substance Withdrawal Delirium may continue for only a few hours or may persist for as long as 2–4 weeks.

This diagnosis should be made instead of a diagnosis of Substance Intoxication or Substance Withdrawal only when the cognitive symptoms are in excess of those usually associated with the intoxication or withdrawal syndrome and when the symptoms are sufficiently severe to warrant independent clinical attention. For a more detailed discussion of the features associated with Substance-Related Disorders, see p. 191.

Recording Procedures

A diagnosis of Substance-Induced Delirium begins with the name of the specific substance (rather than the class of substances) that is presumed to be causing the delirium (e.g., "Diazepam" rather than "Sedative, Hypnotic, or Anxiolytic"). The diagnostic code is selected from the listing of classes of substances provided in the criteria set. For substances that do not fit into any of the classes (e.g., digitalis), the code for "Other Substance" should be used. In addition, for medications prescribed at therapeutic doses, the specific medication can be indicated by listing the appropriate E-code (see Appendix G). For substances that produce intoxication or withdrawal, the name of the substance is followed by the context in which the symptoms developed (e.g., 292.81 Dextroamphetamine Intoxication Delirium; 291.0 Alcohol Withdrawal Delirium). For medication side effects and toxin exposure, the term "-Induced" is used (e.g., 292.81 Digitalis-Induced Delirium). When more than one substance is judged to play a significant role in the development of the delirium, each should be listed separately. If a substance is judged to be the etiological factor but the specific substance or class of substances is unknown, the diagnosis is 292.81 Unknown Substance-Induced Delirium.

Specific Substances

Substance Intoxication Delirium can occur with the following classes of substances: alcohol; amphetamines and related substances; cannabis; cocaine; hallucinogens; inhalants; opioids; phencyclidine and related substances; sedatives, hypnotics, and anxiolytics; and other or unknown substances. **Substance Withdrawal Delirium** can occur with the following classes of substances: alcohol (often called "delirium tremens"); sedatives, hypnotics, and anxiolytics; and other or unknown substances.

Medications reported to cause delirium include anesthetics, analgesics, antiasthmatic agents, anticonvulsants, antihistamines, antihypertensive and cardiovascular medications, antimicrobials, antiparkinsonian drugs, corticosteroids, gastrointestinal medications, histamine H₂-receptor antagonists (e.g., cimetidine), immunosuppres-

sive agents, lithium, muscle relaxants, and psychotropic medications with anticholinergic side effects. Toxins reported to cause delirium include organophosphate (anticholinesterase), insecticides, carbon monoxide, and volatile substances such as fuel or organic solvents.

Differential Diagnosis

See p. 139 for a general discussion of the differential diagnosis of delirium and p. 207 for a discussion of the differential diagnosis of Substance Intoxication and Withdrawal.

Diagnostic criteria for Substance Intoxication Delirium

- A. Disturbance of consciousness (i.e., reduced clarity of awareness of the environment) with reduced ability to focus, sustain, or shift attention.
- B. A change in cognition (such as memory deficit, disorientation, language disturbance) or the development of a perceptual disturbance that is not better accounted for by a preexisting, established, or evolving dementia.
- C. The disturbance develops over a short period of time (usually hours to days) and tends to fluctuate during the course of the day.
- D. There is evidence from the history, physical examination, or laboratory findings of either (1) or (2):
 - (1) the symptoms in Criteria A and B developed during Substance Intoxication
 - (2) medication use is etiologically related to the disturbance*

Note: This diagnosis should be made instead of a diagnosis of Substance Intoxication only when the cognitive symptoms are in excess of those usually associated with the intoxication syndrome and when the symptoms are sufficiently severe to warrant independent clinical attention.

***Note:** The diagnosis should be recorded as Substance-Induced Delirium if related to medication use. Refer to Appendix G for E-codes indicating specific medications.

Code [Specific Substance] Intoxication Delirium:

(291.0 Alcohol; 292.81 Amphetamine [or Amphetamine-Like Substance];
292.81 Cannabis; 292.81 Cocaine; 292.81 Hallucinogen; 292.81 Inhalant;
292.81 Opioid; 292.81 Phencyclidine [or Phencyclidine-Like Substance];
292.81 Sedative, Hypnotic, or Anxiolytic; 292.81 Other [or Unknown] Substance
[e.g., cimetidine, digitalis, benztropine])

Diagnostic criteria for Substance Withdrawal Delirium

- A. Disturbance of consciousness (i.e., reduced clarity of awareness of the environment) with reduced ability to focus, sustain, or shift attention.
- B. A change in cognition (such as memory deficit, disorientation, language disturbance) or the development of a perceptual disturbance that is not better accounted for by a preexisting, established, or evolving dementia.
- C. The disturbance develops over a short period of time (usually hours to days) and tends to fluctuate during the course of the day.
- D. There is evidence from the history, physical examination, or laboratory findings that the symptoms in Criteria A and B developed during, or shortly after, a withdrawal syndrome.

Note: This diagnosis should be made instead of a diagnosis of Substance Withdrawal only when the cognitive symptoms are in excess of those usually associated with the withdrawal syndrome and when the symptoms are sufficiently severe to warrant independent clinical attention.

Code [Specific Substance] Withdrawal Delirium:

(291.0 Alcohol; 292.81 Sedative, Hypnotic, or Anxiolytic; 292.81 Other [or Unknown] Substance)

Delirium Due to Multiple Etiologies

The Delirium Due to Multiple Etiologies category is included to alert clinicians to the common situation, particularly in critically ill and elderly hospitalized patients, in which the delirium has more than one etiology. For example, there may be more than one general medical condition etiologically related to the delirium (e.g., Delirium Due to Hepatic Encephalopathy, Delirium Due to Head Trauma), or the delirium may be due to the combined effects of a general medical condition (e.g., viral encephalitis) and substance use (e.g., Alcohol Withdrawal). A higher number of general medical conditions and substances is associated with a higher rate of delirium.

Recording Procedures

Delirium Due to Multiple Etiologies does not have its own separate code and should not be recorded as a diagnosis. For example, to code a delirium due to both hepatic encephalopathy and withdrawal from alcohol, the clinician would list both 293.0 Delirium Due to Hepatic Encephalopathy and 291.0 Alcohol Withdrawal Delirium on Axis I and 572.2 hepatic encephalopathy on Axis III.

Diagnostic criteria for Delirium Due to Multiple Etiologies

- A. Disturbance of consciousness (i.e., reduced clarity of awareness of the environment) with reduced ability to focus, sustain, or shift attention.
- B. A change in cognition (such as memory deficit, disorientation, language disturbance) or the development of a perceptual disturbance that is not better accounted for by a preexisting, established, or evolving dementia.
- C. The disturbance develops over a short period of time (usually hours to days) and tends to fluctuate during the course of the day.
- D. There is evidence from the history, physical examination, or laboratory findings that the delirium has more than one etiology (e.g., more than one etiological general medical condition, a general medical condition plus Substance Intoxication or medication side effect).

Coding note: Use multiple codes reflecting specific delirium and specific etiologies, e.g., 293.0 Delirium Due to Viral Encephalitis; 291.0 Alcohol Withdrawal Delirium.

780.09 Delirium Not Otherwise Specified

This category should be used to diagnose a delirium that does not meet criteria for any of the specific types of delirium described in this section.

Examples include

- 1. A clinical presentation of delirium that is suspected to be due to a general medical condition or substance use but for which there is insufficient evidence to establish a specific etiology
- 2. Delirium due to causes not listed in this section (e.g., sensory deprivation)

Dementia

The disorders in the "Dementia" section are characterized by the development of multiple cognitive deficits (including memory impairment) that are due to the direct physiological effects of a general medical condition, to the persisting effects of a substance, or to multiple etiologies (e.g., the combined effects of cerebrovascular disease and Alzheimer's disease). The disorders in this section share a common symptom presentation but are differentiated based on etiology. The diagnostic features listed in the next section pertain to **Dementia of the Alzheimer's Type**, **Vascular Dementia**, **Dementia Due to HIV Disease**, **Dementia Due to Head Trauma**, **Dementia Due to Parkinson's Disease**, **Dementia Due to Huntington's Disease**, **Dementia Due to Pick's Disease**, **Dementia Due to Creutzfeldt-Jakob Disease**, **Dementia Due to Other General Medical Conditions**, **Substance-Induced Persisting Dementia**, and **De-**

mentia Due to Multiple Etiologies. In addition, **Dementia Not Otherwise Specified** is included in this section for presentations in which the clinician is unable to determine a specific etiology for the multiple cognitive deficits.

Diagnostic Features

The essential feature of a dementia is the development of multiple cognitive deficits that include memory impairment and at least one of the following cognitive disturbances: aphasia, apraxia, agnosia, or a disturbance in executive functioning. The cognitive deficits must be sufficiently severe to cause impairment in occupational or social functioning and must represent a decline from a previously higher level of functioning. A diagnosis of a dementia should not be made if the cognitive deficits occur exclusively during the course of a delirium. However, a dementia and a delirium may both be diagnosed if the dementia is present at times when the delirium is not present. Dementia may be etiologically related to a general medical condition, to the persisting effects of substance use (including toxin exposure), or to a combination of these factors.

Memory impairment is required to make the diagnosis of a dementia and is a prominent early symptom (Criterion A1). Individuals with dementia become impaired in their ability to learn new material, or they forget previously learned material. Most individuals with dementia have both forms of memory impairment, although it is sometimes difficult to demonstrate the loss of previously learned material early in the course of the disorder. They may lose valuables like wallets and keys, forget food cooking on the stove, and become lost in unfamiliar neighborhoods. In advanced stages of dementia, memory impairment is so severe that the person forgets his or her occupation, schooling, birthday, family members, and sometimes even name.

Memory may be formally tested by asking the person to register, retain, recall, and recognize information. The ability to learn new information may be assessed by asking the individual to learn a list of words. The individual is requested to repeat the words (registration), to recall the information after a delay of several minutes (retention, recall), and to recognize the words from a multiple list (recognition). Individuals with difficulty learning new information are not helped by clues or prompts (e.g., multiple-choice questions) because they did not learn the material initially. In contrast, individuals with primarily retrieval deficits can be helped by clues and prompts because their impairment is in the ability to access their memories. Remote memory may be tested by asking the individual to recall personal information or past material that the individual found of interest (e.g., politics, sports, entertainment). It is also useful to determine (from the individual and informants) the impact of the memory disturbances on the individual's functioning (e.g., ability to work, shop, cook, pay bills, return home without getting lost).

Deterioration of language function (aphasia) may be manifested by difficulty producing the names of individuals and objects (Criterion A2a). The speech of individuals with aphasia may become vague or empty, with long circumlocutory phrases and excessive use of terms of indefinite reference such as "thing" and "it." Comprehension of spoken and written language and repetition of language may also be compromised. In the advanced stages of dementia, individuals may be mute or have a deteriorated

speech pattern characterized by echolalia (i.e., echoing what is heard) or palilalia (i.e., repeating sounds or words over and over). Language is tested by asking the individual to name objects in the room (e.g., tie, dress, desk, lamp) or body parts (e.g., nose, chin, shoulder), follow commands ("Point at the door and then at the table"), or repeat phrases ("no ifs, ands, or buts").

Individuals with dementia may exhibit apraxia (i.e., impaired ability to execute motor activities despite intact motor abilities, sensory function, and comprehension of the required task) (Criterion A2b). They will be impaired in their ability to pantomime the use of objects (e.g., combing hair) or to execute known motor acts (e.g., waving goodbye). Apraxia may contribute to deficits in cooking, dressing, and drawing. Motor skill disturbances may be tested by asking the individual to execute motor functions (e.g., to show how to brush teeth, to copy intersecting pentagons, to assemble blocks, or to arrange sticks in specific designs).

Individuals with dementia may exhibit agnosia (i.e., failure to recognize or identify objects despite intact sensory function) (Criterion A2c). For example, the individual may have normal visual acuity but lose the ability to recognize objects such as chairs or pencils. Eventually they may be unable to recognize family members or even their own reflection in the mirror. Similarly, they may have normal tactile sensation, but be unable to identify objects placed in their hands by touch alone (e.g., a coin or keys).

Disturbances in executive functioning are a common manifestation of dementia (Criterion A2d) and may be related especially to disorders of the frontal lobe or associated subcortical pathways. Executive functioning involves the ability to think abstractly and to plan, initiate, sequence, monitor, and stop complex behavior. Impairment in abstract thinking may be manifested by the individual having difficulty coping with novel tasks and avoiding situations that require the processing of new and complex information. The ability to abstract can be formally assessed by asking the person to find similarities or differences between related words. Executive dysfunction is also evident in a reduced ability to shift mental sets, to generate novel verbal or nonverbal information, and to execute serial motor activities. Tests for executive function include asking the individual to count to 10, recite the alphabet, subtract serial 7s, state as many animals as possible in 1 minute, or draw a continuous line consisting of alternating m's and n's. It is also useful to determine (from the individual and informants) the impact of the disturbances in executive functioning on the individual's daily life (e.g., ability to work, plan activities, budget).

The items in both Criterion A1 (memory impairment) and Criterion A2 (aphasia, apraxia, agnosia, or disturbance in executive functioning) must be severe enough to cause significant impairment in social or occupational functioning (e.g., going to school, working, shopping, dressing, bathing, handling finances, and other activities of daily living) and must represent a decline from a previous level of functioning (Criterion B). The nature and degree of impairment are variable and often depend on the particular social setting of the individual. The same level of cognitive impairment may significantly impair an individual's ability to perform a complex job, but not a job that is less demanding. Standardized published rating scales that measure physical maintenance (e.g., personal hygiene), intellectual functioning, and the ability to use implements or tools (e.g., telephone, washing machine) can be used to measure the severity of impairment.

Dementia is not diagnosed if these symptoms occur exclusively during the course of a delirium. However, a delirium may be superimposed on a preexisting dementia, in which case both diagnoses should be given.

Associated Features and Disorders

Associated descriptive features and mental disorders. Individuals with dementia may become spatially disoriented and have difficulty with spatial tasks. Visuospatial functioning can be assessed by asking the individual to copy drawings, such as a circle, overlapping pentagons, and a cube. Poor judgment and poor insight are common in dementia. Individuals may exhibit little or no awareness of memory loss or other cognitive abnormalities. They may make unrealistic assessments of their abilities and make plans that are not congruent with their deficits and prognosis (e.g., planning to start a new business). They may underestimate the risks involved in activities (e.g., driving). Occasionally, they may harm others by becoming violent. Suicidal behavior may occur, particularly in early stages when the individual is more capable of carrying out a plan of action. Dementia is sometimes accompanied by motor disturbances of gait leading to falls. Some individuals with dementia show disinhibited behavior, including making inappropriate jokes, neglecting personal hygiene, exhibiting undue familiarity with strangers, or disregarding conventional rules of social conduct. Slurred speech may occur in dementia that is associated with subcortical pathology such as Parkinson's disease, Huntington's disease, and some cases of Vascular Dementia. The multiple cognitive impairments of dementia are often associated with anxiety, mood, and sleep disturbances. Delusions are common, especially those involving themes of persecution (e.g., that misplaced possessions have been stolen). Hallucinations can occur in all sensory modalities, but visual hallucinations are most common. Delirium is frequently superimposed on dementia because the underlying brain disease may increase susceptibility to confusional states that may be produced by medications or other concurrent general medical conditions. Individuals with dementia may be especially vulnerable to physical stressors (e.g., illness or minor surgery) and psychosocial stressors (e.g., going to the hospital, bereavement), which may exacerbate their intellectual deficits and other associated problems.

Associated laboratory findings. A discussion of associated laboratory findings that are specific to types of dementia is included in the text for each dementia. Invariably there are abnormalities in cognitive and memory functioning, which can be assessed using mental status examinations and neuropsychological testing. Neuroimaging may aid in the differential diagnosis of dementia. Computed tomography (CT) or magnetic resonance imaging (MRI) may reveal cerebral atrophy, focal brain lesions (cortical strokes, tumors, subdural hematomas), hydrocephalus, or periventricular ischemic brain injury. Functional imaging such as positron-emission tomography (PET) or single photon emission computed tomography (SPECT) are not routinely used in the evaluation of dementia, but may provide useful differential diagnostic information (e.g., parietal lobe changes in Alzheimer's disease or frontal lobe alterations in frontal lobe degenerations) in individuals without evidence of structural changes on CT or MRI scans.

Associated physical examination findings and general medical conditions. The associated physical examination findings of dementia depend on the nature, location, and stage of progression of the underlying pathology. The most common cause of dementia is Alzheimer's disease. Other frequent forms include Vascular Dementia and dementia due to other neurodegenerative processes, such as Lewy body disease (including dementia due to Parkinson's disease) and frontotemporal degeneration (including Pick's disease). Other causes are less common and include normal-pressure hydrocephalus, Huntington's disease, traumatic brain injury, brain tumors, anoxia, infectious disorders (e.g., human immunodeficiency virus [HIV], syphilis), prion diseases (e.g., Creutzfeldt-Jakob disease), endocrine conditions (e.g., hypothyroidism, hypercalcemia, hypoglycemia), vitamin deficiencies (e.g., deficiencies of thiamine or niacin), immune disorders (e.g., temporal arteritis, systemic lupus erythematosus), hepatic conditions, metabolic conditions (e.g., Kufs' disease, adrenoleukodystrophy, metachromatic leukodystrophy, and other storage diseases of adulthood and childhood), and other neurological conditions (e.g., multiple sclerosis).

Specific Culture and Age Features

Cultural and educational background should be taken into consideration in the evaluation of an individual's mental capacity. Individuals from certain backgrounds may not be familiar with the information used in certain tests of general knowledge (e.g., names of presidents, geographical knowledge), memory (e.g., date of birth in cultures that do not routinely celebrate birthdays), and orientation (e.g., sense of place and location may be conceptualized differently in some cultures). The prevalence of different causes of dementia (e.g., infections, nutritional deficiencies, traumatic brain injury, endocrine conditions, cerebrovascular diseases, seizure disorders, brain tumors, substance abuse) varies substantially across cultural groups.

The age at onset of dementia depends on the etiology, but is usually late in life, with highest prevalence above age 85 years. A significant deterioration in memory and in multiple cognitive skills, which is necessary for the diagnosis of dementia, may be difficult to document in very young children. Thus, the diagnosis of dementia may not be practical until the child is older (usually between ages 4 and 6 years). In individuals under age 18 years with Mental Retardation, an additional diagnosis of a dementia should be made only if the condition is not characterized satisfactorily by the diagnosis of Mental Retardation alone. Dementia is uncommon in children and adolescents, but can occur as a result of general medical conditions (e.g., head injury, brain tumors, HIV infection, strokes, adrenoleukodystrophies). Dementia in children may present as a deterioration in functioning (as in adults) or as a significant delay or deviation in normal development. Deteriorating school performance may be an early sign.

Prevalence

Reported prevalence of dementia varies among epidemiological studies, depending on the ages of the subjects sampled; methods of determining the presence, severity, and type of cognitive impairment; and the regions or countries studied. Community studies estimated a 1-year prospective prevalence of almost 3.0% with severe cogni-

tive impairment in the adult population. The study assessed individuals with a brief instrument that assessed current cognitive status (the Mini-Mental State Exam), which does not identify specific diagnoses. A variety of epidemiological studies have shown that the prevalence of dementia, especially Dementia of the Alzheimer's Type, increases with age. The prevalence figures range from 1.4% to 1.6% for individuals ages 65–69 years, rising to 16% to 25% for those over age 85 years.

Course

Historically, the term *dementia* implied a progressive or irreversible course. The DSM-IV definition of *dementia*, however, is based on the pattern of cognitive deficits and carries no connotation concerning prognosis. Dementia may be progressive, static, or remitting. The reversibility of a dementia is a function of the underlying pathology and of the availability and timely application of effective treatment. The mode of onset and subsequent course of dementia also depend on the underlying etiology. The level of disability depends not only on the severity of the individual's cognitive impairments but also on the available social supports. In advanced dementia, the individual may become totally oblivious to his or her surroundings and require constant care. Individuals with severe dementia are susceptible to accidents and infectious diseases, which often prove fatal.

Differential Diagnosis

Memory impairment occurs in both **delirium** and dementia. Delirium is also characterized by a reduced ability to maintain and shift attention appropriately. The clinical course can help to differentiate between delirium and dementia. Typically, symptoms in delirium fluctuate and symptoms in dementia are relatively stable. Multiple cognitive impairments that persist in an unchanged form for more than a few months suggest dementia rather than delirium. Delirium may be superimposed on a dementia, in which case both disorders are diagnosed. In situations in which it is unclear whether the cognitive deficits are due to a delirium or a dementia, it may be useful to make a provisional diagnosis of delirium and observe the person carefully while continuing efforts to identify the nature of the disturbance.

An **amnestic disorder** is characterized by severe memory impairment without other significant impairments of cognitive functioning (i.e., aphasia, apraxia, agnosia, or disturbances in executive functioning).

The presumed etiology determines the specific dementia diagnosis. If the clinician has determined that the dementia is due to **multiple etiologies**, multiple codes based on the specific dementias and their etiologies should be used (see Dementia Due to Multiple Etiologies, p. 170). In **Vascular Dementia**, focal neurological signs (e.g., exaggeration of deep tendon reflexes, extensor plantar response) and laboratory evidence of vascular disease judged to be related to the dementia are present. The clinical course of Vascular Dementia is variable and typically progresses in stepwise fashion. The presence of **Dementia Due to Other General Medical Conditions** (e.g., Pick's disease, HIV) requires evidence from the history, physical examination, and appropriate laboratory tests that a general medical condition is etiologically related to the dementia. The onset of the deterioration (gradual or sudden) and its course

(acute, subacute, or chronic) may be useful in suggesting the etiology. For example, the severity of the impairment in cognitive functioning often remains static after head injury, encephalitis, or stroke.

Multiple cognitive deficits that occur only in the context of substance use are diagnosed as **Substance Intoxication** or **Substance Withdrawal**. If the dementia results from the persisting effects of a substance (i.e., a drug of abuse, a medication, or toxin exposure), then **Substance-Induced Persisting Dementia** is diagnosed. Other causes of dementia (e.g., **Dementia Due to a General Medical Condition**) should always be considered, even in a person with Substance Dependence. For example, head injury is not infrequent during substance use and may underlie the dementia. Although researchers are seeking to develop sensitive and specific tests to confirm the diagnosis of **Dementia of the Alzheimer's Type**, it currently remains a diagnosis of exclusion, and other causes for the cognitive deficits (see above) must first be ruled out. In addition, the course is characterized by gradual onset and continuing cognitive decline. In those cases in which there is insufficient evidence to determine whether the dementia is due to a general medical condition or is substance induced, **Dementia Not Otherwise Specified** should be coded. Individuals may present with some but not all of the symptoms of dementia. Such presentations should be coded as **Cognitive Disorder Not Otherwise Specified**.

Mental Retardation is characterized by significantly subaverage current general intellectual functioning, with concurrent impairments in adaptive functioning and with an onset before age 18 years. Mental Retardation is not necessarily associated with memory impairment. In contrast, the age at onset of dementia is usually late in life. If the onset of the dementia is before age 18 years, both dementia and Mental Retardation may be diagnosed if the criteria for both disorders are met. Documenting a significant deterioration in memory and in other cognitive skills, which is necessary for the diagnosis of dementia, may be difficult in persons under age 4 years. In individuals under age 18 years, the diagnosis of dementia should be made only if the condition is not characterized satisfactorily by the diagnosis of Mental Retardation alone.

Schizophrenia can also be associated with multiple cognitive impairments and a decline in functioning, but Schizophrenia is unlike dementia in its generally earlier age at onset, its characteristic symptom pattern, and the absence of a specific etiological general medical condition or substance. Typically, the cognitive impairment associated with Schizophrenia is less severe than that seen in Dementia.

Major Depressive Disorder may be associated with complaints of memory impairment, difficulty thinking and concentrating, and an overall reduction in intellectual abilities. Individuals sometimes perform poorly on mental status examinations and neuropsychological testing. Particularly in elderly persons, it is often difficult to determine whether cognitive symptoms are better accounted for by a dementia or by a Major Depressive Episode. This differential diagnosis may be informed by a thorough medical evaluation and an evaluation of the onset of the disturbance, the temporal sequencing of depressive and cognitive symptoms, the course of illness, family history, and treatment response. The premorbid state of the individual may help to differentiate "pseudodementia" (i.e., cognitive impairments due to the Major Depressive Episode) from dementia. In dementia, there is usually a premorbid history of declining cognitive function, whereas the individual with a Major Depressive Episode is much more likely to have a relatively normal premorbid state and abrupt cognitive

decline associated with the depression. If the clinician determines that both a dementia and Major Depressive Disorder are present with independent etiologies, both should be diagnosed.

Dementia must be distinguished from **Malingering** and **Factitious Disorder**. The patterns of cognitive deficits presented in Malingering and Factitious Disorder are usually not consistent over time and are not characteristic of those typically seen in dementia. For example, individuals with Factitious Disorder or Malingering manifesting as dementia may perform calculations while keeping score during a card game, but then claim to be unable to perform similar calculations during a mental status examination.

Dementia must be distinguished from the normal decline in cognitive functioning that occurs with **aging** (as in Age-Related Cognitive Decline). The diagnosis of dementia is warranted only if there is demonstrable evidence of greater memory and other cognitive impairment than would be expected due to normal aging processes and the symptoms cause impairment in social or occupational functioning.

294.1x* Dementia of the Alzheimer's Type

Diagnostic Features

The cognitive deficits (Criterion A) and the required impairment (Criterion B) are discussed on pp. 147–150. The onset of Dementia of the Alzheimer's Type is gradual and involves continuing cognitive decline (Criterion C). Because of the difficulty of obtaining direct pathological evidence of the presence of Alzheimer's disease, the diagnosis can be made only when other etiologies for the dementia have been ruled out. Specifically, the cognitive deficits are not due to other central nervous system conditions that cause progressive deficits in memory or cognition (e.g., cerebrovascular disease, Parkinson's disease, Huntington's disease), systemic conditions that are known to cause dementia (e.g., hypothyroidism, vitamin B₁₂ deficiency, HIV infection), or the persisting effects of a substance (e.g., alcohol) (Criterion D). If there is an additional etiology (e.g., head trauma worsening a Dementia of the Alzheimer's Type), both types of dementia should be coded (see Dementia Due to Multiple Etiologies, p. 170). Dementia of the Alzheimer's Type should not be diagnosed if the symptoms occur exclusively during delirium (Criterion E). However, delirium may be superimposed on a preexisting Dementia of the Alzheimer's Type, in which case the With Delirium subtype should be indicated. Finally, the cognitive deficits are not better accounted for by another Axis I disorder (e.g., Major Depressive Disorder or Schizophrenia) (Criterion F).

Subtypes

The age at onset of Dementia of the Alzheimer's Type is indicated by the use of one of the following subtypes:

*ICD-9-CM code valid after October 1, 2000.

With Early Onset. This subtype is used if the onset of the dementia is age 65 years or under.

With Late Onset. This subtype is used if the onset of the dementia is after age 65 years.

The presence or absence of a clinically significant behavioral disturbance is indicated by using one of the following coded subtypes:

.10 Without Behavioral Disturbance. This subtype is used if the cognitive disturbance is not accompanied by any clinically significant behavioral disturbance.

.11 With Behavioral Disturbance. This subtype is used if the cognitive disturbance is accompanied by a clinically significant behavioral disturbance (e.g., wandering, agitation).

Recording Procedures

The diagnostic code depends entirely on the presence or absence of a clinically significant behavioral disturbance and not whether the dementia is of early versus late onset. Thus, the diagnostic code is 294.10 for Dementia of the Alzheimer's Type, With Early Onset, Without Behavioral Disturbance; 294.10 for Dementia of the Alzheimer's Type, With Late Onset, Without Behavioral Disturbance; 294.11 for Dementia of the Alzheimer's Type, With Early Onset, With Behavioral Disturbance; and 294.11 for Dementia of the Alzheimer's Type, With Late Onset, With Behavioral Disturbance. In addition, 331.0 Alzheimer's disease should be coded on Axis III.

Other prominent clinical features related to the Alzheimer's disease can be indicated by coding the specific additional mental disorders due to Alzheimer's disease on Axis I. For example, to indicate the presence of prominent delusions, clinically significant depressed mood, and the development of persistent aggressive behavior, 293.81 Psychotic Disorder Due to Alzheimer's Disease, With Delusions; 293.83 Mood Disorder Due to Alzheimer's Disease, With Depressive Features; and 310.1 Personality Change Due to Alzheimer's Disease, Aggressive Type, would also be coded on Axis I.

Associated Features and Disorders

Associated descriptive features and mental disorders. See p. 150 for a general discussion of features and disorders associated with dementia. The prevalence of Dementia of the Alzheimer's Type is increased in individuals with Down syndrome and in individuals with a history of head trauma. Pathological changes that are characteristic of Alzheimer's disease are present in the brains of individuals with Down syndrome by the time they are in their early 40s, although the clinical symptoms of dementia are not usually evident until later.

Associated laboratory findings. No widely accepted, sensitive, and specific biological marker is currently available that is universally accepted as diagnostic of Dementia of the Alzheimer's Type in a living individual. In the majority of cases, brain atrophy is present in Dementia of the Alzheimer's Type, with wider cortical

sulci and larger cerebral ventricles than would be expected given the normal aging process. This may be demonstrated by computed tomography (CT) or magnetic resonance imaging (MRI). Microscopic examination usually reveals histopathological changes, including senile plaques, neurofibrillary tangles, granulovascular degeneration, neuronal loss, astrocytic gliosis, and amyloid angiopathy. Lewy bodies are sometimes seen in the cortical neurons.

Associated physical examination findings and general medical conditions. In the first years of illness, few motor and sensory signs are associated with Dementia of the Alzheimer's Type. Later in the course, myoclonus and gait disorder may appear. Seizures occur in approximately 10% of individuals with the disorder.

Specific Culture, Age, and Gender Features

See p. 151 for a general discussion of culture and age features associated with dementia. Late onset (after age 65 years) of Dementia of the Alzheimer's Type is much more common than early onset. Few cases develop before age 50 years. The disorder is slightly more common in females than in males.

Prevalence

The prevalence of Dementia of the Alzheimer's Type increases dramatically with increasing age, rising from 0.6% in males and 0.8% in females at age 65 (all levels of severity) to 11% in males and 14% in females at age 85. At age 90 the prevalence rises to 21% in males and 25% in females, and by age 95 the prevalence is 36% in males and 41% in females. Moderate to severe cases make up about 40%–60% of these estimated prevalence rates.

Course

See p. 152 for a general discussion of the course of dementia. The course of Dementia of the Alzheimer's Type tends to be slowly progressive, with a loss of 3–4 points per year on a standard assessment instrument such as the Mini-Mental State Exam. Various patterns of deficits are seen. A common pattern is an insidious onset, with early deficits in recent memory followed by the development of aphasia, apraxia, and agnosia after several years. Many individuals show personality changes, increased irritability, and other behavioral signs and symptoms, starting in the early stages and becoming most pronounced in the middle stages of the disease. In the later stages of the disease, individuals may develop gait and motor disturbances and eventually become mute and bedridden. The average duration of the illness from onset of symptoms to death is 8–10 years.

Familial Pattern

Compared with the general population, first-degree biological relatives of individuals with Dementia of the Alzheimer's Type, With Early Onset, are more likely to develop the disorder. Late-onset cases may also have a genetic component. Dementia of

the Alzheimer's Type in some families has been shown to be inherited as an autosomal dominant trait with linkage to several chromosomes, including chromosomes 1, 14, and 21. However, the proportion of cases that are related to specific inherited abnormalities is not known. Individuals carrying one or both alleles coding for apolipoprotein E-4 (APOE4) on chromosome 19 bear an elevated risk for later-onset Alzheimer's disease, although this gene is not itself a cause of the disorder.

Differential Diagnosis

See p. 152 for a general discussion of the differential diagnosis of dementia.

Diagnostic criteria for 294.1x Dementia of the Alzheimer's Type

- A. The development of multiple cognitive deficits manifested by both
 - ✓(1) memory impairment (impaired ability to learn new information or to recall previously learned information)
 - (2) one (or more) of the following cognitive disturbances:
 - ✓(a) aphasia (language disturbance)
 - (b) apraxia (impaired ability to carry out motor activities despite intact motor function)
 - (c) agnosia (failure to recognize or identify objects despite intact sensory function)
 - (d) disturbance in executive functioning (i.e., planning, organizing, sequencing, abstracting)
- B. ✓ The cognitive deficits in Criteria A1 and A2 each cause significant impairment in social or occupational functioning and represent a significant decline from a previous level of functioning.
- ✓ C. The course is characterized by gradual onset and continuing cognitive decline.
- D. The cognitive deficits in Criteria A1 and A2 are not due to any of the following:
 - ✗(1) other central nervous system conditions that cause progressive deficits in memory and cognition (e.g., cerebrovascular disease, Parkinson's disease, Huntington's disease, subdural hematoma, normal-pressure hydrocephalus, brain tumor)
 - ✓(2) systemic conditions that are known to cause dementia (e.g., hypothyroidism, vitamin B₁₂ or folic acid deficiency, niacin deficiency, hypercalcemia, neurosyphilis, HIV infection)
 - ✓(3) substance-induced conditions
- ✓ E. The deficits do not occur exclusively during the course of a delirium.
- ✓ F. The disturbance is not better accounted for by another Axis I disorder (e.g., Major Depressive Disorder, Schizophrenia).

Code based on presence or absence of a clinically significant behavioral disturbance:

294.10 Without Behavioral Disturbance: if the cognitive disturbance is not accompanied by any clinically significant behavioral disturbance.

Diagnostic criteria for 294.1x Dementia of the Alzheimer's Type (*continued*)

294.11 With Behavioral Disturbance: if the cognitive disturbance is accompanied by a clinically significant behavioral disturbance (e.g., wandering, agitation).

Specify subtype:

With Early Onset: if onset is at age 65 years or below

With Late Onset: if onset is after age 65 years

Coding note: Also code 331.0 Alzheimer's disease on Axis III. Indicate other prominent clinical features related to the Alzheimer's disease on Axis I (e.g., 293.83 Mood Disorder Due to Alzheimer's Disease, With Depressive Features, and 310.1 Personality Change Due to Alzheimer's Disease, Aggressive Type).

290.4x Vascular Dementia (formerly Multi-Infarct Dementia)

Diagnostic Features

The cognitive deficits (Criterion A) and the required impairment (Criterion B) in Vascular Dementia are discussed on pp. 147–150. There must be evidence of cerebrovascular disease (i.e., focal neurological signs and symptoms or laboratory evidence) that is judged to be etiologically related to the dementia (Criterion C). The focal neurological signs and symptoms include extensor plantar response, pseudobulbar palsy, gait abnormalities, exaggeration of deep tendon reflexes, or weakness of an extremity. Computed tomography (CT) of the head and magnetic resonance imaging (MRI) usually demonstrate multiple vascular lesions of the cerebral cortex and subcortical structures. Vascular Dementia is not diagnosed if the symptoms occur exclusively during delirium (Criterion D). However, delirium may be superimposed on a pre-existing Vascular Dementia, in which case the subtype With Delirium should be indicated.

Subtypes

By ICD-9-CM convention, Vascular Dementia is the only type of dementia that employs subtypes to indicate the presence of significant associated symptoms. The following subtypes (each of which has its own separate code) must be used to indicate the predominant feature of the current clinical presentation:

With Delirium. This subtype is used if delirium is superimposed on the dementia.

With Delusions. This subtype is used if delusions are the predominant feature.

With Depressed Mood. This subtype is used if depressed mood (including presentations that meet symptom criteria for a Major Depressive Episode) is the predominant feature. A separate diagnosis of Mood Disorder Due to a General Medical Condition is not given.

Uncomplicated. This subtype is used if none of the above predominates in the current clinical presentation.

The specifier **With Behavioral Disturbance** (which cannot be coded) can also be used to indicate clinically significant behavioral disturbances (e.g., wandering).

Recording Procedures

By ICD-9-CM convention, only Vascular Dementia has codable subtypes. The diagnostic codes for Vascular Dementia depend on the subtype for predominant features: 290.41 for With Delirium, 290.42 for With Delusions, 290.43 for With Depressed Mood, 290.40 for Uncomplicated. The specifier With Behavioral Disturbance is uncoded and can be applied to each of the above subtypes (e.g., 290.43 Vascular Dementia, With Depressed Mood, With Behavioral Disturbance). In addition, the cerebrovascular condition (e.g., 436 stroke) should be coded on Axis III.

Associated Features and Disorders

Associated descriptive features and mental disorders. See p. 150 for a general discussion of features and disorders associated with dementia.

Associated laboratory findings. The extent of central nervous system lesions detected by CT and MRI in Vascular Dementia typically exceeds the extent of changes detected in the brains of healthy elderly persons (e.g., periventricular and white matter hyperintensities noted on MRI scans). Lesions often appear in both white matter and gray matter structures, including subcortical regions and nuclei. Evidence of old infarctions (e.g., focal atrophy) may be detected, as well as findings of more recent disease. EEG findings may reflect focal lesions in the brain. In addition, there may be laboratory evidence of associated cardiac and systemic vascular conditions (e.g., ECG abnormalities, laboratory evidence of renal failure).

Associated physical examination findings and general medical conditions. Common neurological signs (e.g., abnormal reflexes, weakness of an extremity, gait disturbance) are discussed in the "Diagnostic Features" section. There is often evidence of long-standing arterial hypertension (e.g., fundoscopic abnormalities, enlarged heart), valvular heart disease (e.g., abnormal heart sounds), or extracranial vascular disease that may be sources of cerebral emboli. A single stroke may cause a relatively circumscribed change in mental state (e.g., an aphasia following damage to the left hemisphere, or an amnesic disorder from infarction in the distribution of the posterior cerebral arteries), but generally does not cause Vascular Dementia, which typically results from the occurrence of multiple strokes, usually at different times.

Specific Culture, Age, and Gender Features

See p. 151 for a general discussion of culture and age features of dementia.

The onset of Vascular Dementia is typically earlier than that of Dementia of the Alzheimer's Type. The disorder is apparently more common in males than in females.

Prevalence

Vascular Dementia is reportedly much less common than Dementia of the Alzheimer's Type.

Course

See p. 152 for a general discussion of the course of dementia.

The onset of Vascular Dementia is typically abrupt, followed by a stepwise and fluctuating course that is characterized by rapid changes in functioning rather than slow progression. The course, however, may be highly variable, and an insidious onset with gradual decline is also encountered. Usually the pattern of deficits is "patchy," depending on which regions of the brain have been destroyed. Certain cognitive functions may be affected early, whereas others remain relatively unimpaired. Early treatment of hypertension and vascular disease may prevent further progression.

Differential Diagnosis

See p. 152 for a general discussion of the differential diagnosis of dementia.

Diagnostic criteria for 290.4x Vascular Dementia

- A. The development of multiple cognitive deficits manifested by both
- (1) memory impairment (impaired ability to learn new information or to recall previously learned information)
 - (2) one (or more) of the following cognitive disturbances:
 - (a) aphasia (language disturbance)
 - (b) apraxia (impaired ability to carry out motor activities despite intact motor function)
 - (c) agnosia (failure to recognize or identify objects despite intact sensory function)
 - (d) disturbance in executive functioning (i.e., planning, organizing, sequencing, abstracting)
- B. The cognitive deficits in Criteria A1 and A2 each cause significant impairment in social or occupational functioning and represent a significant decline from a previous level of functioning.
- C. Focal neurological signs and symptoms (e.g., exaggeration of deep tendon reflexes, extensor plantar response, pseudobulbar palsy, gait abnormalities, weakness of an extremity) or laboratory evidence indicative of cerebrovascular disease (e.g., multiple infarctions involving cortex and underlying white matter) that are judged to be etiologically related to the disturbance.
- D. The deficits do not occur exclusively during the course of a delirium.

Code based on predominant features:

290.41 With Delirium: if delirium is superimposed on the dementia

290.42 With Delusions: if delusions are the predominant feature

290.43 With Depressed Mood: if depressed mood (including presentations that meet full symptom criteria for a Major Depressive Episode) is the predominant feature. A separate diagnosis of Mood Disorder Due to a General Medical Condition is not given.

290.40 Uncomplicated: if none of the above predominates in the current clinical presentation

Specify if:

With Behavioral Disturbance

Coding note: Also code cerebrovascular condition on Axis III.

Dementias Due to Other General Medical Conditions

Diagnostic Features

The cognitive deficits (Criterion A) and the required impairment (Criterion B) of Dementia Due to Other General Medical Conditions are discussed on pp. 147–150. There must be evidence from the history, physical examination, or laboratory findings that a general medical condition (other than Alzheimer's disease or cerebrovascular disease) is etiologically related to the dementia (e.g., HIV infection, traumatic brain injury, Parkinson's disease, Huntington's disease, Pick's disease, Creutzfeldt-Jakob disease, normal-pressure hydrocephalus, hypothyroidism, brain tumor, or vitamin B₁₂ deficiency) (Criterion C). Dementia Due to a General Medical Condition is not diagnosed if the symptoms occur exclusively during delirium (Criterion D). However, delirium may be superimposed on a preexisting Dementia Due to a General Medical Condition, in which case both diagnoses should be given.

In determining whether the dementia is due to a general medical condition, the clinician must first establish the presence of a general medical condition. Further, the clinician must establish that the dementia is etiologically related to the general medical condition through a physiological mechanism. A careful and comprehensive assessment of multiple factors is necessary to make this judgment. Although there are no infallible guidelines for determining whether the relationship between the dementia and the general medical condition is etiological, several considerations provide some guidance in this area. One consideration is the presence of a temporal association between the onset or exacerbation of the general medical condition and that of the cognitive deficits. Evidence from the literature that suggests that there can be a direct association between the general medical condition in question and the development of a dementia can provide a useful context in the assessment of a particular situation. In addition, the clinician must also judge that the disturbance is not better accounted for by Dementia of the Alzheimer's Type, Vascular Dementia, a Substance-Induced Persisting Dementia, or another mental disorder (e.g., Major Depressive Disorder). These determinations are explained in greater detail in the "Mental Disorders Due to a General Medical Condition" section (p. 181).

See p. 150 for a general discussion of the features and disorders associated with dementia.

Subtypes

The presence or absence of a clinically significant behavioral disturbance can be indicated by using one of the following coded subtypes:

- .10 Without Behavioral Disturbance.** This subtype is used if the cognitive disturbance is not accompanied by any clinically significant behavioral disturbance.
- .11 With Behavioral Disturbance.** This subtype is used if the cognitive disturbance is accompanied by a clinically significant behavioral disturbance (e.g., wandering, agitation).

Recording Procedures

The diagnostic codes are selected depending on whether there is a clinically significant behavioral disturbance (i.e., the diagnostic code 294.10 applies when there is no clinically significant behavioral disturbance, and 294.11 applies when there is a clinically significant behavioral disturbance accompanying the cognitive deficits). The ICD-9-CM code for the etiological condition should also be noted on Axis III (e.g., 332.0 Parkinson's disease, 331.1 Pick's disease, 244.9 hypothyroidism). (See Appendix G for a list of selected ICD-9-CM diagnostic codes for general medical conditions.)

In an individual with an established history of a dementia, a superimposed Delirium Due to a General Medical Condition should be noted by coding both the dementia and the delirium on Axis I (e.g., 294.1 Dementia Due to Parkinson's Disease and 293.0 Delirium Due to Hepatic Encephalopathy). This is in contrast to Vascular Dementia, in which the With Delirium subtype is specified.

Other prominent clinical features related to the etiological general medical condition can be indicated by coding the specific additional Mental Disorder Due to General Medical Condition on Axis I. For example, to indicate the presence of prominent delusions, clinically significant depressed mood, and a change to a labile personality in an individual with Dementia Due to Parkinson's Disease, 293.81 Psychotic Disorder Due to Parkinson's Disease, With Delusions; 293.83 Mood Disorder Due to Parkinson's Disease, With Depressive Features; and 310.1 Personality Change Due to Parkinson's Disease, Labile Type would also be coded on Axis I.

294.1x* Dementia Due to HIV Disease

The essential feature of Dementia Due to HIV Disease is the presence of a dementia that is judged to be the direct pathophysiological consequence of human immunodeficiency virus (HIV) disease. Neuropathological findings most commonly involve diffuse, multifocal destruction of the white matter and subcortical structures. The spinal fluid may show normal or slightly elevated protein and a mild lymphocytosis, and HIV can usually be isolated directly from cerebrospinal fluid. Dementia that is associated with direct HIV infection of the central nervous system is typically characterized by forgetfulness, slowness, poor concentration, and difficulties with problem solving. Behavioral manifestations most commonly include apathy and social withdrawal, and occasionally these may be accompanied by delirium, delusions, or hallucinations. Tremor, impaired rapid repetitive movements, imbalance, ataxia, hypertonia, generalized hyperreflexia, positive frontal release signs, and impaired pursuit and saccadic eye movements may be present on physical examination. Children may also develop Dementia Due to HIV Disease, typically manifested by developmental delay, hypertonia, microcephaly, and basal ganglia calcification. Dementia in association with HIV infection may also result from accompanying central nervous system tumors (e.g., primary central nervous system lymphoma) and from opportunistic infections (e.g., toxoplasmosis, cytomegalovirus infection, cryptococcosis, tuberculosis, and syphilis), in which case the appropriate type of dementia should be diagnosed (e.g., 294.1 Dementia Due to Toxoplasmosis). Unusual systemic infections

*ICD-9-CM code valid after October 1, 2000.

(e.g., *Pneumocystis carinii* pneumonia) or neoplasms (e.g., Kaposi's sarcoma) may also be present.

294.1x* Dementia Due to Head Trauma

The essential feature of Dementia Due to Head Trauma is the presence of a dementia that is judged to be the direct pathophysiological consequence of head trauma. The degree and type of cognitive impairments or behavioral disturbances depend on the location and extent of the brain injury. Posttraumatic amnesia is frequently present, along with persisting memory impairment. A variety of other behavioral symptoms may be evident, with or without the presence of motor or sensory deficits. These symptoms include aphasia, attentional problems, irritability, anxiety, depression or affective lability, apathy, increased aggression, or other changes in personality. Alcohol or other Substance Intoxication is often present in individuals with acute head injuries, and concurrent Substance Abuse or Dependence may be present. Head injury occurs most often in young males and has been associated with risk-taking behaviors. When it occurs in the context of a single injury, Dementia Due to Head Trauma is usually nonprogressive, but repeated head injury (e.g., from boxing) may lead to a progressive dementia (so-called dementia pugilistica). A single head trauma that is followed by a progressive decline in cognitive function should raise the possibility of another superimposed process such as hydrocephalus or a Major Depressive Episode.

294.1x* Dementia Due to Parkinson's Disease

The essential feature of Dementia Due to Parkinson's Disease is the presence of a dementia that is judged to be the direct pathophysiological consequence of Parkinson's disease. Parkinson's disease is a slowly progressive neurological condition, characterized by tremor, rigidity, bradykinesia, and postural instability. Dementia has been reported to occur in approximately 20%–60% of individuals with Parkinson's disease and is more likely to be present in older individuals or those with more severe or advanced disease. The dementia associated with Parkinson's disease is characterized by cognitive and motoric slowing, executive dysfunction, and impairment in memory retrieval. Declining cognitive performance in individuals with Parkinson's disease is frequently exacerbated by depression. Findings on physical examination include the characteristic abnormal motor signs of resting tremor, evidence of slowness and poverty of movement (such as micrographia), or muscular rigidity and loss of associated movements. At autopsy, neuronal loss and Lewy bodies are evident in the substantia nigra. There are a number of syndromes that may manifest with dementia, parkinsonian movement disorders, and additional neurological features (e.g., progressive supranuclear palsy, olivopontocerebellar degeneration, and Vascular Dementia). Some individuals with Parkinson's disease and dementia are found at autopsy to have coexisting neuropathology indicative of Alzheimer's disease or of diffuse Lewy body disease. Dementia due to Lewy Body Disease in the absence of evidence of Parkinson's (such as tremor and cogwheel rigidity) should be diagnosed as Dementia

*ICD-9-CM code valid after October 1, 2000.

Due to Lewy body disease, one of the dementias due to other general medical conditions (see p. 167).

294.1x* Dementia Due to Huntington's Disease

The essential feature of Dementia Due to Huntington's Disease is the presence of a dementia that is judged to be the direct pathophysiological consequence of Huntington's disease. Huntington's disease is an inherited progressive degenerative disease of cognition, emotion, and movement. The disease affects men and women equally and is transmitted by a single autosomal dominant gene on the short arm of chromosome 4. The disease is usually diagnosed in the late 30s to early 40s but may begin as early as age 4 years in the juvenile form or as late as age 85 years in the late-onset form. The onset of Huntington's disease is often heralded by insidious changes in behavior and personality, including depression, irritability, and anxiety. Some individuals present with abnormalities of movement that resemble increased fidgeting and that later progress to characteristic generalized choreoathetosis. Difficulties with memory retrieval, executive functioning, and judgment are common early in the course, with more severe memory deficits occurring as the disease progresses. Disorganized speech and psychotic features are sometimes present. Late in the disease, characteristic "boxcar ventricles" may be seen on structural brain imaging due to the atrophy of the striatum. Positron-emission tomography (PET) may show striatal hypometabolism early in the disease. Offspring of individuals with Huntington's disease have a 50% chance of developing the disease. A genetic test is available to determine with relative certainty whether a given at-risk individual is likely to develop the disease; however, such testing may be best administered by centers with experience in counseling and follow-up of individuals at risk for Huntington's disease.

294.1x* Dementia Due to Pick's Disease

The essential feature of Dementia Due to Pick's Disease is the presence of a dementia that is judged to be the direct pathophysiological consequence of Pick's disease. Pick's disease is a degenerative disease of the brain that particularly affects the frontal and temporal lobes. As in other frontal lobe dementias, Pick's disease is characterized clinically by changes in personality early in the course, deterioration of social skills, emotional blunting, behavioral disinhibition, and prominent language abnormalities. Difficulties with memory, apraxia, and other features of dementia usually follow later in the course. Prominent primitive reflexes (snout, suck, grasp) may be present. As the dementia progresses, it may be accompanied by either apathy or extreme agitation. Individuals may develop such severe problems in language, attention, or behavior that it may be difficult to assess their degree of cognitive impairment. Structural brain imaging typically reveals prominent frontal and/or temporal atrophy, and functional brain imaging may localize frontotemporal hypometabolism, even in the absence of clear structural atrophy. The disorder most commonly manifests itself in individuals between ages 50 and 60 years, although it can occur among older individuals. Pick's disease is one of the pathologically distinct etiologies among the heterogeneous group of dementing processes that are associated with frontotemporal brain

*ICD-9-CM code valid after October 1, 2000.

atrophy. The specific diagnosis of a frontal lobe dementia such as Pick's disease is usually established at autopsy with the pathological finding of characteristic intraneuronal argentophilic Pick inclusion bodies. Clinically, Pick's disease often cannot be distinguished with certainty from atypical cases of Alzheimer's disease or from other dementias that affect the frontal lobes. Dementia due to frontotemporal degeneration other than Pick's disease should be diagnosed as Dementia Due to Frontotemporal Degeneration, one of the dementias due to other general medical conditions (see p. 167).

294.1x* Dementia Due to Creutzfeldt-Jakob Disease

The essential feature of Dementia Due to Creutzfeldt-Jakob Disease is the presence of a dementia that is judged to be the direct pathophysiological consequence of Creutzfeldt-Jakob disease. Creutzfeldt-Jakob disease is one of the subacute spongiform encephalopathies, a group of central nervous system diseases caused by transmissible agents known as "slow viruses" or prions. Typically, individuals with Creutzfeldt-Jakob disease manifest the clinical triad of dementia, involuntary movements (particularly myoclonus), and periodic EEG activity. However, up to 25% of individuals with the disorder may have atypical presentations, and the disease can be confirmed only by biopsy or at autopsy with the demonstration of spongiform neuropathological changes. Creutzfeldt-Jakob disease may develop at any age in adults, but most typically when they are between ages 40 and 60 years. From 5% to 15% of cases may have a familial component. Prodromal symptoms of Creutzfeldt-Jakob disease may include fatigue, anxiety, or problems with appetite, sleeping, or concentration and may be followed after several weeks by incoordination, altered vision, or abnormal gait or other movements that may be myoclonic, choreoathetoid, or ballistic, along with a rapidly progressive dementia. The disease typically progresses very rapidly over several months, although more rarely it can progress over years and appear similar in its course to other dementias. Although there are no distinctive findings on cerebrospinal fluid analysis, reliable biomarkers are being developed. Nonspecific atrophy may be apparent on neuroimaging. In most individuals, the EEG typically reveals periodic sharp, often triphasic and synchronous discharges at a rate of 0.5–2 Hz at some point during the course of the disorder. The transmissible agent thought to be responsible for Creutzfeldt-Jakob disease is resistant to boiling, formalin, alcohol, and ultraviolet radiation, but it can be inactivated by pressured autoclaving or by bleach. Transmission by corneal transplantation and human growth factor injection has been documented, and anecdotal cases of transmission to health care workers have been reported. Therefore, when neurosurgery, brain biopsy, or brain autopsy is undertaken, universal precautions should be taken with both tissue and equipment that comes in contact with tissue. Cross-species transmission of prion infections, with agents that are closely related to the human form, has now been demonstrated (e.g., the outbreak of bovine spongiform encephalopathy [mad cow disease] human variant Creutzfeldt-Jakob disease in the United Kingdom during the mid-1990s).

*ICD-9-CM code valid after October 1, 2000.

294.1x* Dementia Due to Other General Medical Conditions

In addition to the specific categories described above, a number of other general medical conditions can cause dementia. Two of the most common are due to Lewy body-related neurodegeneration ("Lewy body dementia") and to focal degeneration in the frontal and temporal lobes ("frontotemporal dementia"). Dementia Due to Parkinson's Disease is an example of the former (see p. 164), and Dementia Due to Pick's Disease is an example of the latter (see p. 165). The descriptive features, course, and etiology of dementia due to diffuse Lewy body disease without Parkinson's disease and dementia due to frontotemporal degeneration other than Pick's disease require further research. Other conditions associated with dementia include structural lesions (primary or secondary brain tumors, subdural hematoma, slowly progressive or normal-pressure hydrocephalus), endocrine conditions (hypothyroidism, hypercalcemia, hypoglycemia), nutritional conditions (deficiencies of thiamine or niacin), other infectious conditions (neurosyphilis, cryptococcosis), immune disorders (e.g., temporal arteritis, systemic lupus erythematosus), derangements of renal and hepatic function, metabolic conditions (e.g., Kufs' disease, adrenoleukodystrophy, metachromatic leukodystrophy, and other storage diseases of adulthood and childhood), and other neurological conditions such as multiple sclerosis. Unusual causes of central nervous system injury, such as electrical shock or intracranial radiation, are generally evident from the history. Rare disorders such as the childhood and adult storage diseases have a distinctive family history or clinical presentation. Associated physical examination and laboratory findings and other clinical features depend on the nature and severity of the general medical condition.

Differential Diagnosis

See p. 152 for a general discussion of the differential diagnosis of dementia.

Diagnostic criteria for 294.1x Dementia Due to Other General Medical Conditions

- A. The development of multiple cognitive deficits manifested by both
 - (1) memory impairment (impaired ability to learn new information or to recall previously learned information)
 - (2) one (or more) of the following cognitive disturbances:
 - (a) aphasia (language disturbance)
 - (b) apraxia (impaired ability to carry out motor activities despite intact motor function)
 - (c) agnosia (failure to recognize or identify objects despite intact sensory function)
 - (d) disturbance in executive functioning (i.e., planning, organizing, sequencing, abstracting)
- B. The cognitive deficits in Criteria A1 and A2 each cause significant impairment in social or occupational functioning and represent a significant decline from a previous level of functioning.
- C. There is evidence from the history, physical examination, or laboratory findings that the disturbance is the direct physiological consequence of a general medical condition other than Alzheimer's disease or cerebrovascular disease (e.g., HIV infection, traumatic brain injury, Parkinson's disease, Huntington's disease, Pick's disease, Creutzfeldt-Jakob disease, normal-pressure hydrocephalus, hypothyroidism, brain tumor, or vitamin B₁₂ deficiency).
- D. The deficits do not occur exclusively during the course of a delirium.

Code based on presence or absence of a clinically significant behavioral disturbance:

294.10 Without Behavioral Disturbance: if the cognitive disturbance is not accompanied by any clinically significant behavioral disturbance.

294.11 With Behavioral Disturbance: if the cognitive disturbance is accompanied by a clinically significant behavioral disturbance (e.g., wandering, agitation).

Coding note: Also code the general medical condition on Axis III (e.g., 042 HIV infection, 854.00 head injury, 332.0 Parkinson's disease, 333.4 Huntington's disease, 331.1 Pick's disease, 046.1 Creutzfeldt-Jakob disease; see Appendix G for additional codes).

Substance-Induced Persisting Dementia

Diagnostic and Associated Features

The cognitive deficits (Criterion A) and the required impairment (Criterion B) are discussed on pp. 147–150. Substance-Induced Persisting Dementia is diagnosed if the symptoms persist beyond the usual duration of Substance Intoxication or Withdrawal and is not diagnosed if they occur exclusively during the course of a delirium (Criterion C). However, delirium may be superimposed on a preexisting Substance-Induced Persisting Dementia, in which case both diagnoses should be given. There

must be evidence from the history, physical examination, or laboratory findings that the deficits are etiologically related to the persisting effects of substance use (e.g., a drug of abuse, a medication, toxin exposure) (Criterion D). This disorder is termed "persisting" because the dementia persists long after the individual has experienced the effects of Substance Intoxication or Substance Withdrawal.

Features that are associated with Substance-Induced Persisting Dementia are those associated with dementias generally (see p. 147). Even if currently abstinent from substance use, most individuals with this disorder have previously had a pattern of prolonged and heavy substance use that met criteria for Substance Dependence. Because these disorders persist long after use of the substance has stopped, blood or urine screens may be negative for the etiological substance. The age at onset of Substance-Induced Persisting Dementia is rarely before age 20 years. This disorder usually has an insidious onset and slow progression, typically during a period when the person qualifies for a Substance Dependence diagnosis. The deficits are usually permanent and may worsen even if the substance use stops, although some cases do show improvement.

For a more detailed discussion of the features associated with Substance-Related Disorders, see p. 191.

Recording Procedures

The name of the diagnosis begins with the specific substance (e.g., alcohol) that is presumed to have caused the dementia. The diagnostic code is selected from the listing of classes of substances provided in the criteria set. For substances that do not fit into any of the classes, the code for "Other Substance" should be used. In addition, for medications prescribed at therapeutic doses, the specific medication can be indicated by listing the appropriate E-code (see Appendix G). When more than one substance is judged to play a significant role in the development of the persisting dementia, each should be listed separately (e.g., 291.2 Alcohol-Induced Persisting Dementia; 292.82 Inhalant-Induced Persisting Dementia). If a substance is judged to be the etiological factor, but the specific substance or class of substances is unknown, the diagnosis is 292.82 Unknown Substance-Induced Persisting Dementia.

Specific Substances

Substance-Induced Persisting Dementia can occur in association with the following classes of substances: alcohol; inhalants; sedatives, hypnotics, and anxiolytics; or other or unknown substances. Medications reported to cause dementia include anticonvulsants and intrathecal methotrexate. Toxins reported to evoke symptoms of dementia include lead, mercury, carbon monoxide, organophosphate insecticides, and industrial solvents.

Differential Diagnosis

See p. 152 for a general discussion of the differential diagnosis of dementia.

Diagnostic criteria for Substance-Induced Persisting Dementia

- A. The development of multiple cognitive deficits manifested by both
 - (1) memory impairment (impaired ability to learn new information or to recall previously learned information)
 - (2) one (or more) of the following cognitive disturbances:
 - (a) aphasia (language disturbance)
 - (b) apraxia (impaired ability to carry out motor activities despite intact motor function)
 - (c) agnosia (failure to recognize or identify objects despite intact sensory function)
 - (d) disturbance in executive functioning (i.e., planning, organizing, sequencing, abstracting)
- B. The cognitive deficits in Criteria A1 and A2 each cause significant impairment in social or occupational functioning and represent a significant decline from a previous level of functioning.
- C. The deficits do not occur exclusively during the course of a delirium and persist beyond the usual duration of Substance Intoxication or Withdrawal.
- D. There is evidence from the history, physical examination, or laboratory findings that the deficits are etiologically related to the persisting effects of substance use (e.g., a drug of abuse, a medication).

Code [Specific Substance]–Induced Persisting Dementia:

(291.2 Alcohol; 292.82 Inhalant; 292.82 Sedative, Hypnotic, or Anxiolytic;
292.82 Other [or Unknown] Substance)

Dementia Due to Multiple Etiologies

The Dementia Due to Multiple Etiologies category is included to alert clinicians to the common situation in which the dementia has more than one etiology. More than one general medical condition may be etiologically related to the dementia (e.g., Dementia of the Alzheimer's Type and Dementia Due to Head Trauma), or the dementia may be due to the combined effects of a general medical condition (e.g., Parkinson's disease) and the long-term use of a substance (e.g., Alcohol-Induced Persisting Dementia).

Recording Procedures

Dementia Due to Multiple Etiologies does not have its own separate code and should not be recorded as a diagnosis. For example, both Dementia of the Alzheimer's Type and Vascular Dementia should be diagnosed for an individual with Dementia of the Alzheimer's Type, With Late Onset, Without Behavioral Disturbance, who, over the

course of several strokes, develops a significant further decline in cognitive functioning. In this example, the clinician would list both 294.10 Dementia of the Alzheimer's Type, With Late Onset, Without Behavioral Disturbance, and 290.40, Vascular Dementia, Uncomplicated, on Axis I, and 331.0 Alzheimer's Disease and 436 Stroke on Axis III.

Diagnostic criteria for Dementia Due to Multiple Etiologies

- A. The development of multiple cognitive deficits manifested by both
 - (1) memory impairment (impaired ability to learn new information or to recall previously learned information)
 - (2) one (or more) of the following cognitive disturbances:
 - (a) aphasia (language disturbance)
 - (b) apraxia (impaired ability to carry out motor activities despite intact motor function)
 - (c) agnosia (failure to recognize or identify objects despite intact sensory function)
 - (d) disturbance in executive functioning (i.e., planning, organizing, sequencing, abstracting)
- B. The cognitive deficits in Criteria A1 and A2 each cause significant impairment in social or occupational functioning and represent a significant decline from a previous level of functioning.
- C. There is evidence from the history, physical examination, or laboratory findings that the disturbance has more than one etiology (e.g., head trauma plus chronic alcohol use, Dementia of the Alzheimer's Type with the subsequent development of Vascular Dementia).
- D. The deficits do not occur exclusively during the course of a delirium.

Coding note: Use multiple codes based on specific dementias and specific etiologies, e.g., 294.10 Dementia of the Alzheimer's Type, With Late Onset, Without Behavioral Disturbance; 290.40 Vascular Dementia, Uncomplicated.

294.8 Dementia Not Otherwise Specified

This category should be used to diagnose a dementia that does not meet criteria for any of the specific types described in this section.

An example is a clinical presentation of dementia for which there is insufficient evidence to establish a specific etiology.

Amnestic Disorders

The disorders in the "Amnestic Disorders" section are characterized by a disturbance in memory that is either due to the direct physiological effects of a general medical condition or due to the persisting effects of a substance (i.e., a drug of abuse, a medication, or toxin exposure). The disorders in this section share the common symptom presentation of memory impairment, but are differentiated based on etiology. The diagnostic features listed below pertain to **Amnestic Disorder Due to a General Medical Condition** (e.g., physical trauma and vitamin deficiency) and **Substance-Induced Persisting Amnestic Disorder** (including medication side effects). In addition, **Amnestic Disorder Not Otherwise Specified** is included in this section for presentations in which the clinician is unable to determine a specific etiology for the memory disturbance. Text and criteria for Dissociative Disorders involving memory loss are not included here and instead are contained in the Dissociative Disorders section (see p. 519).

Diagnostic Features

Individuals with an amnestic disorder are impaired in their ability to learn new information or are unable to recall previously learned information or past events (Criterion A). The memory disturbance must be sufficiently severe to cause marked impairment in social or occupational functioning and must represent a significant decline from a previous level of functioning (Criterion B). The memory disturbance must not occur exclusively during the course of a delirium or a dementia (Criterion C). The ability to learn and recall new information is always affected in an amnestic disorder, whereas problems remembering previously learned information occur more variably, depending on the location and severity of brain damage. The memory deficit is most apparent on tasks that require spontaneous recall and may also be evident when the examiner provides stimuli for the person to recall at a later time. Depending on the specific area of the brain affected, deficits may be predominantly related to verbal or visual stimuli. In some forms of an amnestic disorder, the individual may remember things from the very remote past better than more recent events (e.g., a person may remember in vivid detail a hospital stay that took place a decade before the examination, but may have no idea that he or she is currently in the hospital).

The diagnosis is not made if the memory impairment occurs exclusively during the course of a delirium (i.e., occurs only in the context of reduced ability to maintain and shift attention). The ability to immediately repeat a sequential string of information (e.g., digit span) is typically not impaired in an amnestic disorder. When such impairment is evident, it suggests the presence of an attentional disturbance that may be indicative of a delirium. The diagnosis is also not made in the presence of other cognitive deficits (e.g., aphasia, apraxia, agnosia, disturbance in executive functioning) that are characteristic of a dementia. Individuals with an amnestic disorder may experience major impairment in their social and vocational functioning as a result of their memory deficits, which, at its extreme, may necessitate supervised living situations to ensure appropriate feeding and care.