*Clinician’s Toolkit*

**ICD-10 Diagnostic categories**

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**F84 Pervasive developmental disorders**

This group of disorders is characterized by qualitative abnormalities in reciprocal social interactions and in patterns of communication, and by restricted stereotyped, repetitive repertoire of interests and activities. These qualitative abnormalities are a pervasive feature of the individual’s functioning in all situation, although they may vary in degree. In most cases, development is abnormal from infancy and, only a few exceptions, the conditions become manifest during the first 5 years of life. It is usual, but not invariable, for there to be some degree of general cognitive impairment but the disorders are defined in terms of *behaviour* that is deviant in relation to mental age (whether the individual is retarded or not). There is some disagreement on the subdivision of this overall group of pervasive development disorders.

In some cases the disordered are associated with, and presumably due to, some medical condition, of which infantile spasms, congenital rubella, tuberous sclerosis, cerebral lipidosis, and the fragile X chromosome anomaly are amongst the common. However, the disorder should be diagnosed on the basis of the behavioural features, irrespective of the presence or absence of any associated medical condition; any such associated condition must, nevertheless, be separately coded. If mental retardation is present, it is important that it too should be separately coded, under F70 – F79, because it is not a universal feature of the pervasive development disorders.

**F84.0 Childhood autism**

A pervasive developmental disorder defined by the presence of abnormal and/or impaired development that is manifest before the age of 3 years, and by the characteristic type of abnormal function in all three areas of social interaction, communication and restricted, repetitive behaviour. The disorder occurs in boys three to four times more often than in girls.

***Diagnostic guidelines***

Usually there is no prior period of unequivocally normal development but, if there is, abnormalities become apparent before the age of 3 years. There are always qualitative impairments in reciprocal social interaction. These take the form of an inadequate appreciation of socio-emotional cues, as shown by a lack of responses to other people's emotions and/or a lack of responses to other people's emotions and/or a lack of modulation of behaviour according to social context; poor use of social signals and a weak integration of social, emotional, and communicative behaviours; and, especially, a lack of socio-emotional reciprocity. Similarly, qualitative impairments in communication are universal. These take the form of a lack of social usage of whatever language skills are present; impairment in make-believe and social imitative play; poor synchrony and lack of reciprocity in conversational interchange; poor flexibility in language expression and a relative lack of creativity and fantasy in thought processes; lack of emotional response to other people's verbal and nonverbal overtures; impaired use of variations in cadence or emphasis to reflect communicative modulation; and a similar lack of accompanying gesture to provide emphasis or aid meaning in spoken communication.

The condition is also characterized by restricted, repetitive, and stereotyped patterns of behaviour, interests and activities. These take the form of a tendency to impose rigidity and routine on a wide range of aspects of day-to-day functioning; this usually applies to novel activities as well as to familiar habits and play patterns. In early childhood particularly, there may be specific attachment to unusual, typically non-soft objects. The children may insist on the performance of particular routines in rituals of a non-functional character; there may be stereotyped preoccupations with interests such as dates, routes or timetables; often there are motor stereotypies; a specific interest in non-functional elements of objects (such as their smell or feel) is common; and there may be a resistance to changes in routine or in details of the personal environment (such as the movement of ornaments or furniture in the family home).

In addition to these specific diagnostic features, it is frequent for children with autism to show a range of other nonspecific problems such as fear/phobias, sleeping and eating disturbances, temper tantrums and aggression. Self-injury (e.g. by wrist-biting) is fairly common, especially when there is associated severe mental retardation. Most individuals with autism lack spontaneity, initiative, and creativity in the organisation of their leisure time and have difficulty applying conceptualizations in decision-making in work (even when the tasks themselves are well within their capacity). The specific manifestations of deficits characteristic of autism change as the children grow older, but the deficits continue into and though adult life with a broadly similar pattern of problems socialization, communication, and interest patterns. Developmental abnormalities must have been present in the first 3 years for the diagnosis to be made, but the syndrome can be diagnosed in all age groups.

All levels of IQ can occur in association with autism, but there is significant mental retardation in some three-quarters of cases.

*Includes:* autistic disorder

 infantile autism

 infantile psychosis

 Kanner's syndrome

*Differential diagnosis.* Apart from the other varieties of pervasive developmental disorder it is important to consider: specific developmental disorder of receptive language (F80.2) with secondary socio-emotional problems; reactive attachment disorder (F94.1) or disinhibited attachment disorder (F94.2); mental retardation (F70-F79) with some associated emotional/behaviour disorder; schizophrenia (F20.-) of unusually early onset; and Rett's syndrome (F84.2).

*Excludes:* autistic psychopathy (F84.5)

**F84.1 Atypical autism**

A pervasive developmental disorder that differs from autism in terms *either* of age of onset *or* of failure to fulfil all three sets of diagnostic criteria. Thus, abnormal and/or impaired development becomes manifest for the first time only after age 3 years; and/or there are insufficient demonstrable abnormalities in one or two of the three areas of psychopathology required for the diagnosis of autism; it also occurs in individuals with a severe specific developmental disorder of receptive language. Atypical autism thus constitutes a meaningfully separate condition from autism.

*Includes:* atypical childhood psychosis

 mental retardation with autistic features

**F 84.2 Rett's syndrome**

A condition of unknown cause, so far only reported in girls, which has been differentiated on the basis of a characteristic onset, course, and pattern of symptomatology. Typically, apparently normal or near-normal early development is followed by partial or complete loss of acquired hand skills and of speech, together with deceleration in head growth, usually with an onset between 7 and 24 months of age. Hand-wringing stereotypies, hyperventilation and loss of purposive hand movements are particularly characteristic. Social and play development are arrested in the first 2 or 3 years, but social interest tends to be maintained. During middle childhood, trunk ataxia and apraxia, associated with scoliosis or hyphoscoliosis tend to develop and sometimes there are choreoathetoid movements. Severe mental handicap invariably results. Fits frequently develop during early or middle childhood.

***Diagnostic guidelines***

In most cases onset is between 7 and 24 months of age. The most characteristic feature is a loss of purposive hand movements and acquired fine motor manipulative skills. This is accompanied by loss, partial loss or lack of development of language; distinctive stereotyped tortuous wringing or “hand-washing” movements, with the arms flexed in front of the chest or chin; stereotypic wetting of the hands with saliva; lack of proper chewing of food; often episodes of hyperventilation; almost always a failure to gain bowel and bladder control; often excessive drooling and protrusion of the tongue; and a loss of social engagement. Typically, the children retain a kind of “social smile”, looking at or “through” people, but not interacting socially with them in early childhood (although social interaction often develops later). The stance and gait tend to become broad-based, the muscles are hypotonic, trunk movements usually become poorly coordinated, and scoliosis or kyphoscoliosis usually develops. Spinal atrophies, with severe motor disability, develop in adolescence or adulthood in about half the cases. Later, rigid spasticity may become manifest, and is usually more pronounced in the lower than in the upper limbs. Epileptic fits, usually involving some type of minor attack, and with an onset generally before the age of 8 years, occur in the majority of cases. In contrast to autism, both deliberate self-injury and complex stereotyped preoccupations or routines are rare.

*Differential diagnosis*. Initially, Rett's syndrome is differentiated primarily on the basis of the lack of purposive hand movements, deceleration of head growth, ataxia, stereotypic “hand-washing” movements, and lack of proper chewing. The course of the disorder, in terms of progressive motor deterioration, confirms the diagnosis.

**F84.3 Other childhood disintegrative disorder**

A pervasive developmental disorder (other than Rett's syndrome) this is defined by a period of normal development before onset, and by a definite loss, over the course of a few months, of previously acquired skills in at least several areas of development, together with the onset of characteristic abnormalities of social communicative, and behavioural functioning. Often there is a prodromic period of vague illness; the child becomes restive, irritable, anxious and overactive. This is followed by impoverishment and then loss of speech and language, accompanied by behavioural disintegration. In some cases the loss of skills is persistently progressive (usually when the disorder is associated with a progressive diagnosable neurological condition), but more often the decline over a period of some months is followed by a plateau and then a limited improvement. The prognosis is usually very poor, and most individuals are left with severe mental retardation. There is uncertainty about the extent to which this condition differs from autism. In some cases the disorder can be shown to be due to some associated encephalophathy, but the diagnosis should be made on the behavioural features. Any associated neurological condition should be separately coded.

***Diagnostic guidelines***

Diagnosis is based on an apparently normal development up to the age of at least 2 years, followed by a definite loss of previously acquired skills; this is accompanied by qualitatively abnormal social functioning. It is usual for there to be a profound regression in, or loss of, language, a regression in the level of play, social skills, and adaptive behaviour, and often a loss of bowel or bladder control, sometimes with a deteriorating motor control. Typically, this is accompanied by a general loss of interest in the environment, by stereotyped, repetitive motor mannerisms, and by an autistic-like impairment of social interaction and communication. In some respects, the syndrome resembles dementia in adult life, but it differs in three key respects: there is usually no evidence of any identifiable organic disease or damage (although organic brain dysfunction of some type is usually inferred); the loss of skills may be followed by a degree of recovery; and the impairment in socialization and communication has deviant qualities typical of autism rather than of intellectual decline. For all these reasons the syndrome is included here rather than under F00-F09.

*Includes:* dementia infantilis

 disintegrative psychosis

 Heller's syndrome

 symbiotic psychosis

*Excludes:* acquired aphasia with epilepsy (F80.3)

 elective mutism (F94.0)

 Rett's syndrome (F84.2)

 schizophrenia (F20.-)

**F84.4 Overactive disorder associated with mental retardation and stereotyped movements**

This is an ill-defined disorder of uncertain nosological validity. The category is included here because of the evidence that children with moderate to severe mental retardation (IQ below 50) who exhibit major problems in hyperactivity and inattention frequently show stereotyped behaviours; such children tend not to benefit from stimulant drugs (unlike those with an IQ in the normal range) and may exhibit a severe dysphoric reaction (sometimes with psychomotor retardation) when given stimulants; in adolescence the over activity tends to be replaced by underactivity (a pattern that is *not* usual in hyperkinetic children with normal intelligence). It is also common for the syndrome to be associated with a variety of developmental delays, either specific or global.

The extent to which the behavioural pattern is a function of low IQ or of organic brain damage is not known, neither is it clear whether the disorders in children with mild mental retardation who show the hyperkinetic syndrome would be better classified here or under F90.-, at present they are included in F90-.

***Diagnostic guidelines***

Diagnosis depends on the combination of developmentally inappropriate severe over activity, motor stereotypies, and moderate to severe mental retardation, all three must be present for the diagnosis. If the diagnostic criteria for F84.0, F84.1 or F84.2 are met, that condition should be diagnosed instead.

**F84.5 Asperger's syndrome**

A disorder of uncertain nosological validity, characterized by the same kind of qualitative abnormalities of reciprocal social interaction that typify autism, together with a restricted, stereotyped, repetitive repertoire of interests and activities. The disorder differs from autism primarily in that there is no general delay or retardation in language or cognitive development. Most individuals are of normal general intelligence but it is common for them to be markedly clumsy; the condition occurs predominantly in boys (in a ratio of about eight boys to one girl). It seems highly likely that at least some cases represent mild varieties of autism, but it is uncertain whether or not that is so for all. There is a strong tendency for the abnormalities to persist into adolescence and adult life and it seems that they represent individual characteristics that are not greatly affected by environmental influences. Psychotic episodes occasionally occur in early adult life.

***Diagnostic guidelines***

Diagnosis is based on the combination of a lack of any clinically significant general delay in language or cognitive development plus, as with autism, the presence of qualitative deficiencies in reciprocal social interaction and restricted, repetitive, stereotyped patterns of behaviour, interests, and activities. There may or may not be problems in communication similar to those associated with autism, but significant language retardation would rule out the diagnosis.

*Includes:* autistic psychopathy

 schizoid disorder of childhood

*Excludes:* anankastic personality disorder (F60.5)

 attachment disorders of childhood (F94.1, F94.2)

 obsessive-compulsive disorder (F42.-)

 schizotypal disorder (F21)

 simple schizophrenia (F20.6)

**F84.8 Other pervasive developmental disorders**

**F84.9 Pervasive developmental disorder, unspecified**

This is a residual diagnostic category that should be used for disorders which fit the general description for pervasive developmental disorders but in which a lack of adequate information, or contradictory findings, means that the criteria for any of the other F84 codes cannot be met.