WHO/MNH/96.3
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Distr. General

# ICD-10 GUIDE FOR MENTAL RETARDATION

DIVISION OF MENTAL HEALTH AND PREVENTION OF SUBSTANCE ABUSE

WORLD HEALTH ORGANIZATION

GENEVA

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# ICD-10 GUIDE FOR MENTAL RETARDATION

This guide has been prepared to assist those working with mentally retarded patients to make the best use of the 10th edition of the International Classification of diseases

People with mental retardation usually have multiple problems. To describe these problems adequately it is usually necessary to use several diagnoses taken from different parts of the classification. It is necessary to record the degree of mental retardation and the presence of associated physical and mental disorders; to record the degree of psychosocial disability; and to note relevant abnormal psychosocial situations. These factors can be recorded in a systematic and orderly way by using a multiaxial system. The axes in a multiaxial system are not axes in the sense of statistically divided axes; they are means of recording different kinds of features of the case, the following axes form the structure of the ICD scheme of classification for the mentally retarded.

Axis I Severity of retardation and problem behaviours
Axis II Associated medical conditions
Axis III Associated psychiatric disorders
Axis IV Global assessment of psychosocial disability
Axis V Associated abnormal psychosocial situations

The diagnostic codes to be recorded on each of these axes are part contained in the International Classification of Diseases. However most are scattered through the main volume and some are difficult to treat. Also some of the advice on diagnosis provided for psychiatric disorders in the accompanying volume of Clinical Descriptions and Diagnostic Guidelines for Mental and Behavioural Disorders (the 'Blue Book') are not easy to apply to patients with mental retardation. For this reason, this guide contains additional advice on certain mental and behavioural disorders. It also lists the diagnoses of physical disease that are most often used for patients with mental retardation, together with the codes for abnormal situations (Z codes) most likely to be relevant to their patients.

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1996

# CONTENTS

# Axis I
# Severity of Retardation and Problem Behaviours

This axis is used to record codes from Section F7 of the classification. Severity of retardation is recorded with a second digit code (F70 to F79). It is possible to record problem behaviours by using a decimal place code. In ICD-10 abnormal behaviour associated with mental retardation can be specified in only three ways as: none or minimal (x.0); significant, requiring attention or treatment (x.1); or 'other' (x.8). In this guide 6 additional second decimal codes have been provided to record the type of behaviour when x.1 (significant) is employed. These additional codes are described at the end of this section. The codings for degree of mental retardation are:

| | |
|---|---|
| F70 | Mild Mental Retardation |
| F71 | Moderate Mental Retardation |
| F72 | Severe Mental Retardation |
| F73 | Profound Mental Retardation |
| F78 | Other Mental Retardation |
| F79 | Unspecified Mental Retardation. |

## Diagnostic guidelines

Mental retardation is a condition of arrested or incomplete development of the mind, which is especially characterized by impairment of skills manifested during the developmental period, which contribute to the overall level of intelligence, i.e. cognitive, language, motor, and social abilities. Retardation can occur with or without any other mental or physical disorder. However, mentally retarded individuals can experience the full range of mental disorders, and the prevalence of other mental disorders is at least three to four times greater in this population than in the general population. In addition, mentally retarded individuals are at greater risk of exploitation and physical/sexual abuse. Adaptive behaviour is always impaired, but in protected social environments where support is available this impairment may not be at all obvious in subjects with mild mental retardation.

Intelligence is not a unitary characteristic but is assessed on the basis of a large number of different, more or less specific skills. Although the general tendency is for all these skills to develop to a similar level in each individual, there can be large discrepancies, especially in persons who are mentally retarded. Such people may show severe impairment in on particular area (e.g. language), or may have a particular area of higher skill (e.g. in simple visuo-spatial tasks) against a background of severe mental retardation. This presents problems when determining the diagnostic category in which a retarded person should be classified. The assessment of intellectual level should be based on whatever information is available, including clinical findings, adaptive behaviour (judged in relation to the individual's cultural background), and psychometric test performance.

For a definite diagnosis of mental retardation, there should be reduced level of intellectual functioning resulting in diminished ability to adapt to the daily demands of the normal social environment. Associated mental or physical disorders have a major influence on the clinical picture and the use made of any skills. *The diagnostic category chosen should therefore be based on global assessments of ability not on any single area of specific impairment or skill.* The IQ levels given are provided as a guide and

should not be applied rigidly in view of the problems of cross-cultural validity. The categories given below are arbitrary divisions of a complex continuum, and cannot be defined with absolute precision. The IQ should be determined from standardized, individually administered intelligence tests for which local cultural norms have been determined, and the test selected should be appropriate to the individual's level of functioning and additional specific handicapping conditions, e.g. expressive language problems, hearing impairment, physical involvement. Scales of social maturity and adaptation, again locally standardized, should be completed if at all possible by interviewing a parent or care-provider who is familiar with the individual's skills in everyday life. Without the use of standardized procedures, the diagnosis must be regarded as provisional.

Reference should be made to WHO guidelines on "Assessment of People with Mental Retardation" (WHO 1992), to the American Association for Mental Retardation guidelines "Mental Retardation, Definition Classification and Systems of Support" (AAMR 1992) both of which give details of standardized instruments which may be used to assess mental retardation and functional adaptation. *A short list of appropriate tests is given in the Appendix to this guide (p. 75).*

## F70 Mild mental retardation

### Clinical description

Mildly retarded people acquire language with some delay but most achieve the ability to use speech for everyday purposes, to hold conversations, and to engage in the clinical interview. Most of them also achieve full independence in self-care (eating, washing, dressing, bowel and bladder control) and in practical and domestic skills, even if the rate of development is considerably slower than normal. The main difficulties are usually seen in academic school work, and many have particular problems in reading and writing. However, mildly retarded people can be greatly helped by education designed to develop their skills and compensate for their handicaps. Most of those in the higher ranges of mental retardation are potentially capable of work demanding practical rather than academic abilities, including unskilled or semiskilled manual labour. In a sociocultural context requiring little academic achievement, some degree of mild retardation may not itself represent a problem. However, if there is also a noticeable emotional and social immaturity, the consequences of the handicap, e.g. inability to cope with the demands of marriage or child-rearing, or difficulty fitting in with cultural traditions and expectations, will be apparent.

In general the behavioural, emotional, and social difficulties of the mildly mentally retarded, and the needs for treatment are more closely akin to those found in people of normal intelligence than to the specific problems of the moderately and severely retarded. An organic aetiology is being identified in increasing proportions of patients, although not yet in the majority.

### Diagnostic guidelines

If the proper standardized IQ tests are used, the range 50 to 69 is indicative of mild mental retardation. Understanding and use of language tend to be delayed to a varying degree, and executive speech problems that interfere with the development of independence may persist into adult life. An organic aetiology is identifiable in only a minority of subjects. Associated conditions such as autism, other developmental disorders, epilepsy, conduct disorders, or physical disability are found in varying proportion. If such disorders are present, they should be coded independently.

*Includes*: feeble mindedness
mild mental subnormality

mild oligophrenia
moron

## F71 Moderate mental retardation

Clinical Description

Individuals in this category are slow in developing comprehension and use of language, and their eventual achievement in this area is limited. Achievement of self-care and motor skills is also retarded, and some need supervision throughout life. Progress in school work is limited, but a proportion of these individuals learn the basic skills needed for reading, writing, and counting. Educational programmes can provide opportunities for them to develop their limited potential and to acquire some basic skills; such programmes are appropriate for slow learners with a low ceiling of achievement. As adults, moderately retarded people are usually able to do simple practical work, if the tasks are carefully structured and skilled supervision is provided. Completely independent living in adult life is rarely achieved. Generally, however, such people are fully mobile and physically active and the majority show evidence of social development in their ability to establish contact, to communicate with others, and, to engage in simple social activities.

Diagnostic guidelines

The IQ is usually in the range 35 to 49. Discrepant profiles of abilities are common in this group, with some individuals achieving higher levels in visuo-spatial skills than in tasks dependant on language, while others are markedly clumsy but enjoy social interaction and simple conversation. The level of development of language is variable: some of those affected can take part in simple conversations while others have only enough language to communicate their basic needs. Some never learn language, though they may understand simple instructions and may learn to use manual signs to compensate to some extent for their speech disabilities. An organic etiology can be identified in the majority of moderately mentally retarded people. Childhood autism or other pervasive developmental disorders are present in a substantial minority, and have a major effect upon the clinical picture and the type of management needed. Epilepsy, and neurological and physical disabilities are also common, although most moderately retarded people are able to walk without assistance. It is sometimes possible to identify other psychiatric conditions, but the limited level of language development may make diagnosis difficult and dependent upon information obtained from others who are familiar with the individual. Any such associated disorders should be coded independently.

*Includes*: imbecility
moderate mental subnormality
moderate oligophrenia

## F72 Severe mental retardation

Clinical description

This category is broadly similar to that of moderate mental retardation in terms of the clinical picture, the presence of an organic aetiology, and the associated conditions. The lower levels of achievement mentioned under F71 are also the most common in this group. Most people in this category suffer from a marked degree of motor impairment or other associated deficits, indicating the presence of clinically significant damage to or maldevelopment of the central nervous system.

Diagnostic guidelines

The IQ is usually in the range 20 to 34.

*Includes*: severe mental subnormality
severe oligophrenia

## F73 Profound mental retardation

Clinical description

The IQ in this category is estimated to be under 20, which means in practice that affected individuals are severely limited in their ability to understand or comply with requests or instructions. Most such individuals are immobile or severely restricted in mobility, incontinent, and capable at most of only very rudimentary forms of nonverbal communication. They possess little or no ability to care for their own basic needs, and require constant help and supervision.

Diagnostic guidelines

The IQ is under 20. Comprehension and use of language is limited to, at best, understanding basic commands and making simple requests. The most basic and simple visuo-spatial skills of sorting and matching may be acquired, and the affected person may be able with appropriate supervision and guidance to take a small part in domestic and practical tasks. An organic aetiology can be identified in most cases. Severe neurological or other physical disabilities affecting mobility are common, as are epilepsy and visual and hearing impairments. Pervasive developmental disorders in their most severe form, especially atypical autism, are particularly frequent, especially in those who are mobile.

*Includes*: idiocy
profound mental subnormality
profound oligophrenia

## F78 Other mental retardation

This category should be used only when assessment of the degree of intellectual retardation by means of the usual procedures is rendered particularly difficult or impossible by associated sensory or physical impairments, as in blind, deaf-mute, and severely behaviourally disturbed or physically disabled people.

## F79 Unspecified mental retardation

There is evidence of mental retardation, but insufficient information is available to assign the patient to one of the above categories.

*Includes*: mental deficiency NOS
mental subnormality NOS
oligophrenia NOS

# Diagnostic codes for recording abnormal behaviour

Note that the codes are not used when abnormal behaviour is a recognised part of another disorder (for example autism) which has been recorded on Axis III.

(a) The *degree* of impairment of behaviour is coded with a first decimal place:

F7 x .0 Non or minimal impairment of behaviour
F7 x .1 Significant impairment of behaviour requiring attention or treatment
F7 x .8 Other impairments of behaviour
F7 x .9 Without mention of impairment of behaviour

(b) The *type* of the behaviour recorded above is recorded with a second decimal place.

F7 x .11 *Repetitive self injury*
This category is for repeated self-injury among adults sufficient to cause tissue damage. It includes self injury which would cause tissue damage if protection or restrain were not used. Repeated self injurious behaviour includes head banging, face slapping, eye poking and biting of hands, lips or other body parts.
*Excludes* Stereotyped movement disorders of childhood F98.4, nailbiting and thumbsucking F98.8

F7 x .12 *Pica*
This category is for pica defined as in F98.3 but persisting into adult life

F7 x .13 *Hyperkinesis*
This category is for hyperkinetic behaviour defined as for F90 but persisting into adult life

F7 x .14 *Wandering and absconding*
This category excludes F91 conduct disorder of childhood

F7 x .15 *Aggression towards other*
This category excludes F91 conduct disorder of childhood

F7 x .16 *Hair pulling*
This category is for repeated hair pulling which does not fulfil the criteria for trichotillomania (F63.3)

# Axis II
# Associated Medical Conditions

This axis is for associated medical conditions whether or not they are judged to be causative of the mental retardation. Those certainly causative are classified as such by dividing the parts IIa definitely causative; IIb other. The conditions listed here are among those encountered most frequently whether causative or not. A glossary is provided to aid the diagnosis of certain common conditions.

The listing follows the order in ICD-10, not the order of frequency of occurrence.

## Infections and parasitic diseases

| | |
|---|---|
| A17 | Tuberculosis of the nervous system |
| A50 | Congenital syphilis |
| A81.1 | Subacute sclerosing panencephalitis |
| A83 | Mosquito-borne viral encephalitis |
| A83.0 | Japanese encephalitis |
| A85 | Other viral encephalitis, not elsewhere classified |
| B69 | Cysticercosis of the central nervous system |
| B94.8 | Sequelae of other specified infectious and parasitic diseases, two particularly relevant types are classified with the additional codes:<br>P37.1 Congenital toxoplasmosis or<br>B58.2 Toxoplasma meningoencephalitis |

## Benign neoplasms

| | |
|---|---|
| D18 | Haemangioma and lymphangioma, any site |
| D32 | Benign neoplasm of meninges |
| D33 | Benign neoplasm of brain and other parts of central nervous system |

## Diseases of the blood

| | |
|---|---|
| D50 | Iron deficiency anaemia |
| D51 | Vitamin $B_{12}$ deficiency anaemia |
| D52 | Folate deficiency anaemia |
| D53 | Other nutritional anaemias |
| D56 | Thalassaemia |
| D57 | Sickle-cell disorders |

## Endocrine nutritional and metabolic disorders

| | |
|---|---|
| E00 | Congenital iodine-deficiency syndrome |
| E01 | Iodine-deficiency-related thyroid disorders and allied conditions |
| E03 | Other hypothyroidism |
| E70 | Disorders of aromatic amino-acid metabolism (includes phenylketonuria) |
| E71 | Disorders of branched-chain amino-acid metabolism and fatty acid metabolism |
| E72 | Other disorders of amino-acid metabolism |
| E73 | Lactose intolerance |

| | |
|---|---|
| E74 | Other disorders of carbohydrate metabolism |
| E75 | Disorders of sphingolipid metabolism and other lipid storage disorders |
| E76 | Disorders of glycosaminoglycan metabolism |
| E77 | Disorders of glycoprotein metabolism |
| E78 | Disorders of lipoprotein metabolism and other lipidaemias |
| E79 | Disorders of purine and pyrimidine metabolism |
| E80 | Disorders of porphyrin and bilirubin metabolism |
| E88 | Other metabolic disorders |

## Diseases of the nervous system

| | |
|---|---|
| G04 | Encephalitis, myelitis and encephalomyelitis |
| G05 | Encephalitis, myelitis and encephalomyelitis in diseases classified elsewhere |
| G05.0 | Encephalitis, myelitis and encephalomyelitis in bacterial diseases classified elsewhere |
| G23 | Other degenerative diseases of basal ganglia |
| G31 | Other degenerative diseases of the nervous system, not elsewhere classified |
| G37 | Other demyelinating diseases of central nervous system |
| G40 | Epilepsy |
| G41 | Status epilepticus |
| G80 | Infantile cerebral palsy |
| G81 | Hemiplegia |
| G82 | Paraplegia and tetraplegia |
| G91 | Hydrocephalus |
| G92 | Toxic encephalopathy |

## Diseases of the eyes

| | |
|---|---|
| H53 | Visual disturbance |
| H54 | Blindness and low vision |
| H55 | Nystagmus and other irregular eye movements |

## Diseases of the circulatory system

| | |
|---|---|
| I69.0 | Sequelae of subarachnoid haemorrhage |
| I69.1 | Sequelae of intracerebral haemorrhage |
| I69.2 | Sequelae of other nontraumatic intracranial haemorrhage<br>With additional code from I60-I67 or P52 Intracranial nontraumatic haemorrhage of fetus and newborn |

## Diseases of the musculoskeletal system

| | |
|---|---|
| M40 | Kyphosis and lordosis |
| M41 | Scoliosis |

## Congenital abnormalities, deformation & chromosomal

## disorders

P00 Fetus and newborn affected by maternal conditions that may be unrelated to present pregnancy
P01 Fetus and newborn affected by maternal complications of pregnancy
P02 Fetus and newborn affected by complications of placenta, cord and membranes
P03 Fetus and newborn affected by other complications of labour and delivery
P04 Fetus and newborn affected by noxious influences transmitted via placenta or breast milk
P05 Slow fetal growth and fetal malnutrition
P08 Disorders related to long gestation and high birth weight
P35.0 Congenital rubella syndrome
P35.1 Congenital cytomegalovirus infection
P36 Bacterial sepsis of newborn
P37.1 Congenital toxoplasmosis
P55 Rh isoimmunization of fetus and newborn
P57 Kernicterus
Q00 Anencephaly and similar malformations
Q01 Encephalocele
Q02 Microcephaly
Q03 Congenital hydrocephalus
Q04 Other congenital malformations of the brain
Q04.0 Congenital malformation of corpus callosum
Q05 Spina bifida
Q06 Other congenital malformations of the spinal cord
Q07 Other congenital malformations of the nervous system
Q11 Anophthalmos, microphthalmos and macrophthalmos
Q12 Congenital lens malformations
Q13 Congenital malformations of anterior segment of eye
Q14 Congenital malformations of posterior segment of eye
Q15 Other congenital malformations of eye
Q16 Congenital malformations of the ear causing impairment of hearing
Q85.0 Neurofibromatosis
Q85.1 Tuberose sclerosis
Q85.8 Other phakomatoses, not elsewhere classified
- Peutz-Jeghers syndrome
- Sturge-Weber syndrome
- Von Hippel-Lindou syndrome

Q86.0 Foetal alcohol syndrome (dysmorphic)
Q87.1 Congenital malformation syndromes predominantly associated with short stature
- Prader-Willi syndrome
- de Lange syndrome

Q87.2 Congenital malformation syndromes predominantly involving limbs
- Rubinstein-Taybi syndrome

Q87.8 Other specified congenital malformations
Q90.0 Down's syndrome
Q90.0 Trisomy 21, meiotic nondisjunction
Q90.1 Trisomy 21, mosaicism (mitotic nondisjunction)
Q90.2 Trisomy 21, translocation
Q91 Edward's syndrome and Patau's syndrome
Q93.4 Deletion of short arm of chromosome 5; cri-du-chat syndrome
Q96 Turner's syndrome
Q99.2 Fragile X syndrome

## Symptoms and signs not elsewhere classified

| | |
|---|---|
| R25 | Abnormal involuntary movements |
| R26 | Abnormalities of gait and mobility |
| R27 | Other lack of coordination |
| S06 | Intracranial injury |
| T74.0 | Neglect or abandonment |
| T74.1 | Physical abuse |
| T74.2 | Sexual abuse |
| T74.3 | Psychological abuse |
| T90.5 | Sequelae of intracranial injury<br>With additional code from S06 -Intracranial injury or P10 intracranial laceration and haemorrhage due to birth injury |

## Notes on selected diagnoses from Axis II

The order follows that in ICD-10, and does not reflect relative importance.

### A50 Congenital syphilis

Congenital syphilis has been known as a cause of mental handicap for many years.

The spirochaete responsible may produce abortion, stillbirth or a very large variety of disorders in surviving children depending on the stage of development at which the infection has taken place and the nature of the treatment given.

The protean manifestations of this condition include failure to thrive, rashes, jaundice, hepatosplenomegaly, anaemia, inflammation of the joints and nerve deafness.

The brain is involved in about 60% of cases and some of those so affected will have mental handicap of varying degree.

The general decline of syphilis in many countries has lead to a great reduction of mental handicap due to this cause.

The most effective form of prevention is the serological detection and immediate treatment of syphilitic infection in women as soon as they are known to be pregnant.

### E00-03 Hypothyroidism

Underfunctioning of the thyroid gland has been associated with mental handicap for many years. The thyroid hormones are essential for protein synthesis and any condition which interferes with the production of these hormones may affect both the development and the function of the brain.

The synthesis of thyroxine and triiodothyronine may be interfered with in many ways. Severe iodine deficiency during pregnancy may produce Endemic Cretinism which covers a wide range of severity

and may result in spastic diplegia, deafness and motor disorders in addition to mental handicap.

Hypothyroidism, which may also be cause by auto-immune mechanisms, goitrogenic agents, drugs, congenital partial absence of the thyroid gland and genetic disorders may cause mental handicap or mental slowing, often associated with mental disorders depending upon the age of onset.

Hypothyroidism is particularly associated with Down's syndrome and since the symptoms of mental and physical slowing and increase in weight are often found in Down's syndrome without thyroid disorder and since the onset is usually insidious the diagnosis may be overlooked. It is therefore, necessary to test thyroid function periodically in order not to miss the diagnosis.

The diagnosis of hyperthyroidism is relatively easy to make and the treatment which is pure replacement therapy by the administration of oral thyroxine, is both inexpensive and completely effective.

## E70 Phenylketonuria

This disorder, when untreated, is characterised by severe mental handicap, epilepsy, behaviour disorders, light pigmentation, poor co-ordination and autistic-like behaviour.

It is an autosomal recessive disorder in which an increase in cellular concentrations of the amino acide phenylalanine hydroxilase, which is necessary for the conversion of this amino acide to tyrosine. The resulting deficiency of tyrosine leads to a shortage of melanin and this, in turn, to a lack of this pigment in the skin and hair.

The diagnosis is established by measuring the concentrations of phenylalanine in the urine or blood.

The treatment is based on providing an artificial diet containing reduced amounts of phenylalanine. In order to ensure the best result this should be instituted from birth and accompanied by other supportive measures.

## G04-05 Cerebral infections

A very large number of infections of the brain may cause damage of a nature and extent sufficient to cause mental handicap of any degree of severity. The main emphasis in this group of disorders lies in primary prevention with the use of suitable hygiene measures and vaccines. Once an infection has become established the outcome depends very largely on early diagnosis and the nature of the infecting organism. While the treatment of infections by viruses remains basically supportive, infections due to bacteria tend to respond to appropriate treatment with antibiotics.

Infection of the foetus with the rubella virus tends to result in serious neurological damage often associated with blindness, deafness, self-injurious behaviour and severe mental impairment. The prevention of this condition by immunisation is safe, simple and inexpensive.

## G40 Epilepsy

The incidence of epilepsy in people with mental handicap correlates approximately with the degree of cognitive impairment covering a range from 1% in the mildly handicapped to more than 45%

in those with severe mental and other handicaps. The correlation stems from the fact that the increase in the degree of mental handicap is highly correlated with the extent of associated brain damage.

Virtually every known type of epilepsy is found but mixed types of epilepsy are relatively commoner. Temporal lobe disorders are also relatively common and may give rise to serious difficulties in diagnosis since they may result in behaviours which closely resemble conventional psychiatric and behavioural disorders.

Epilepsy should always be considered in the assessment of any handicapped person with any type of disturbance of consciousness, movement disorder or unusual mental state. Treatment is conventional but is frequently very difficult because of the higher incidence of multifocal forms of epilepsy.

## G80 Cerebral Palsy

This condition is difficult to define but is conventionally described as a group of motor disorders with or without associated sensory and intellectual deficits where the cause is injury to the immature brain. Such damage to the brain has been associated with oxygen deprivation for any reason, trauma and infection as well as many metabolic disorders. However the cause of cerebral palsy is still unknown in a significant number of cases. The types of movement disorder are generally classified in the following five categories; hypotonic, ataxic, spastic, athetoid, and tremulous. The ataxic form shows involuntary movements, disorders of balance and co-ordination and abnormal muscle tone. Deep tendon reflexes are usually increased. The hypotonic form shows a combination of decreased resistance to movement, less spontaneous movement and poor balance.

In the athetoid type there is hyperextension of the torso, open mouth, drooling and writhing movements of the limbs but no increase in the stretch reflexes.

The tremulous form is characterised by a regular tremor which may increase in frequence and amplitude with emotion.

The limbs may be affected by pareses of varying degree and in varying combinations (hemiplegia, paraplegia, quadriplegia) and a large number of other neurological deficits might be found depending upon the site and type of the brain damage.

The association between cerebral palsy and mental handicap depends upon the site and extent of brain damage and this accounts for the fact that the range of intelligence found in people with cerebral palsy cover the entire range of abilities.

## P35.1 Congenital cytomegalovirus

This condition is caused by infection of the foetus by the cytomegalovirus. It is a Herpes virus and may lie dormant for variable periods becoming reactivated from time to time; mostly in the cervix or uterus. The incidence of antibodies in females of child-bearing age varies from 40% to 100% in different populations and the incidence of congenital infections varies from 1 in 50 to 1 in 500. Damage to the foetus of varying degrees may be produced at any stage of gestation.

There are many abnormalities associated with this condition the commonest being hepatomegaly, splenomegaly, microcephaly, cerebral palsy, intracranial calcification, epilepsy and conductive deafness.

If the brain is involved 90% of those so affected will be mentally handicapped.

## P57 Kernicterus

High levels of bilirubin in the blood attack nervous tissue and may lead to brain damage in the newborn. Elevated levels of bilirubin may result from any cause of excessive intravascular haemolysis or impaired hepatic conjugation.

Among the causes are neonatal sepsis, prematurity, a variety of drugs, haemolytic disease of the newborn and glucose-6-phosphase dehydrogenase deficiency.

If the blood levels of bilirubin are low or adequately treated there may be no resulting brain damage but prolonged high levels may result in mental handicap, movement disorders and sensorineural hearing loss.

The diagnosis is made by measuring blood level of bilirubin and treatment methods include exchange-transfusion and hyperimmune Rh gammaglobulin to prevent maternal Rh sensitisation.

## Q02 Microcephaly

This condition, which may be defined as having a head smaller than 3 standard deviations below the mean for the particular age, is mostly secondary to more than 50 conditions where brain growth is inhibited although there appear to be some primary causes which are genetically determined. Appropriate investigation will usually reveal the cause of the anomaly.

## Q05 Spina Bifida

Spina Bifida is an example of one of the many disorders which result from defects of midline closure of the central nervous system (spinal dysraphism).

This group includes hydrocephalus, meningomyelocele, spina bifida and variations on all these conditions.

The clinical picture includes all types of pareses of limbs. Brain damage may be absent or of varying degrees of severity. A variable percentage of people with this condition have mental handicap the severity of which covers a wide range. Diagnosis is made by clinical examination and X-ray.

Ultrasound of the foetus together with the measurement of maternal blood alphafoetoprotein (AFP) levels help in diagnosing this condition in the foetus.

## Q85.1 Tuberose Sclerosis

Tuberose Sclerosis is one of the most important examples of conditions in which haematomata are prominent features. Haematosis implies an organisational defect producing an abnormal mixture of tissues.

Because skin and brain are often involved these conditions are important in mental handicap.

Tuberose Sclerosis, also known as adenoma sebaceum, is an autosomal dominant condition with very variable expressivity and is found in about 0.5% of people with profound handicap. Most people with this condition have it as a result of a new mutation from unaffected parents. The brain almost always shows areas of calcification and particularly in periventricular areas as well as local distortion by abnormal masses of neural tissue.

Fibro-angiomatous lesions, the so-called adenoma sebaceum develop in the nasolatial fold and elsewhere on the face. Cafe au lait spots, oval, pale skin patches are very common.

Those who have the mild form of the condition may live perfectly normal lives and the diagnosis may only come to light when the diagnosis of the severe form comes to light in one of their children.

In the severe form many organ systems may be affected and this is characterised by developmental delay, epileptic seizures and severe behaviour disorders.

## Q87.1 De Lange Syndrome
## Cornelia de Lange Syndrome
## Brachmann de Lange Syndrome

This syndrome is characterised by mental retardation, congenital short stature, bushy eyebrows which are fused in the midline, hirsutism, small nose with anteverted nostrils, thin lips with small 'beak' on upper lip, downward curving mouth, microcephaly and many other abnormalities.

The associated behavioural disorders include feeding problems, absence of speech and self injurious actions. They tend to avoid physical contact and social interaction and typically exhibit difficult, unpredictable behaviour which is very difficult to manage.

The cause is unknown and most cases are sporadic.

## Q87.1 Prader-Willi Syndrome

The Prader-Willi Syndrome is characterised by obesity, cryptorchidism, excessive appetite, behaviour problems, short stature and varying degrees of mental handicap.

They tend to be floppy babies who have feeding problems and show developmental delay. At about two years of age persistent appetite develops and this is accompanied by excessive weight gain. Absence of the prepubertal growth spurt results in small stature. Many of those who become very obese develop non-insulin dependent diabetes.

The associated mental handicap covers a wide range of severity but is mostly in the mild to moderate range.

The very obese may suffer from impaired breather and hypoxia resulting in sleepiness, cyanosis

and heart failure. The main behaviour abnormalities are excessive appetite which may be associated with pica, scavenging and food stealing. Severe temper tantrums and aggressive behaviour is often associated with the frustration of not obtaining food. They are also prone to pick and scratch their skin. When not showing these behaviours they are generally cheerful and good natured.

The disorder is associated with a defective gene on chromosome number 15. It is an example of genetic imprinting in that both copies of chromosome 15 need to come from the mother in this condition.

## Q87.2 Rubenstein-Taybi Syndrome

This syndrome is characterised by broad fingers and toes (approximately 12 other conditions also have this feature), downward sloping palpebral fissures, underdeveloped maxillae with narrow palate, small head and sometimes malformed ears, short stature, squint and cryptorchidism.

The level of intelligence covers a wide range, the most frequent lying in the 40-50 range. The physical findings are very variable and there may be other anatomical disorders. Respiratory infections and feeding disorders are common.

## Q90.0 Down's Syndrome

This syndrome is caused by trisomy of the distal part of chromosome 21; the commonest form [95%] being trisomy of the whole of this chromosome.

It is the commonest cause of severe mental handicap with an incidence of 1 in 650 live births in virtually all countries.

People with Down's syndrome are characterised by short stature, flat faces, hypotonia and hyperflexibility of the joints, epicanthic folds, small ears, short fingers and toes and fine, soft sparse hair.

They are prone to infections because of an immunological defect and are particularly prone to problems of vision and hearing. Fifty percent are born with cardiac disorders of varying severity with a cumulative mortality of 40%.

The range of abilities covered by this condition is very wide. 10% are severely multiply handicapped, a few are sufficiently able to lead independent lives while the majority have a severe mental handicap and will need care all their lives.

The incidence of Down's syndrome at birth is closely related to maternal age; the likelihood of giving birth to a child with Down's syndrome at age 30 is 1 in 900, at 37 years is 1 in 250, at 40 years 1 in 100 and 1 in 40 at 44 years of age.

## Q99.2 Fragile X syndrome

This condition is the most common cause of inherited mental handicap with an estimated prevalence of 1 in every 1000 males and 1 case of generally milder mental handicap in every 200 females. In males it is characterized by moderate to severe mental handicap, long thin faces with prominent jaws, large protuberant ears, macroorchidism and autistic features. The responsible gene, FMR-1 includes a repeated CGG triplet sequence that constitutes the Fragile X mutation.

All mothers of children with Fragile X are carriers and have a small increase in CGG repeats while everyone with a full mutation (a large increase in CGG repeats is mentally retarded).

## S06 Intracranial injury

Any trauma to the brain, depending upon the site and extent, may affect intellectual function. Common causes in children are associated with birth trauma and road traffic accidents while non-accidental injury and domestic accidents account for a minority. This latter cause of mental handicap is often overlooked any may become evident only on careful history taking. Sequelae of intracranial injury can be coded as T90.5. Whilst this axis can be used to record the physical injury and sequelae, the psychological and behavioural effects are recorded on the other axes.

## T74 Non-accidental injury

Although reliable data on non-accidental injury is not available there is evidence that a substantial proportion of children injured in this way are subjected to head injuries as well. The effect of this trauma ranges from the trivial to lesions which may result in death and an unknown proportion may develop mental handicap. Such handicap may be of nay degree of severity but is non-progressive. Its importance lies in recognising it as a diagnostic category in order to prevent recurrence in the same child or any of its siblings.

# Axis III
# Associated Psychiatric Disorders

## Diagnosis and coding

Precedence should be given to the **most important or relevant** diagnoses. There may be doubt about whether a patient's disorder has an organic aetiology. A non-organic diagnosis should usually be made unless there is firm evidence of an organic aetiology (see discussion of the terms organic and non-organic in the introduction to F0).

It is usually possible to establish the diagnosis of a psychiatric disorder for people with mild mental retardation in the same way as for the non-retarded population. This may be much more difficult or impossible for people with severe and profound retardation. Factors influencing the ability to establish diagnoses include language and communication problems, the effect of low intelligence on the capacity to describe complex subjective experiences, and the presence of additional disorders or impairments (such as epilepsy, sensory impairments or autism). These may have a pathoplastic effect on psychopathology, may prevent the description of cardinal features of a disorder, or may result in behaviour which resembles that seen in psychotic disorders. Some additional disorders (such as epilepsy) amy predispose to psychiatric disorder.

When making psychiatric diagnoses, greater weight may have to be given to informant's accounts, and to observed behaviour, and less to the patient's specific description of subjective experiences. If a patient has a severe cognitive impairment, or communication disorder, the clinician may be unable to make some diagnoses, (for example of schizophrenia). In such cases, less specific diagnoses may have to be used, (for example F28 other non-organic psychotic disorders).

## Coding disorders of organic aetiology

Psychotic disorders due to epilepsy or other organic conditions can be coded by using codes within F0 (for example F06.0, Organic Hallucinosis; F06.2 Organic Delusional (Schizophrenia-like) Disorder). Other codes within F0 may be relevant to the coding of behavioural and psychiatric disorders among people with mental retardation (for example F07.0 Organic Personality Disorder).

It should be noted that code F54 (Psychological and behavioural factors associated with disorders or diseases classified elsewhere) refers to *psychological or behavioural influences thought to have played a major part in the etiology of physical disorders* and not to behaviour which results from a physical disorder. Behavioural phenotypes may be coded using F07.8 *if clinically relevant.* The disorder itself, (such as Fragile-X syndrome) should be recorded on Axis II.

### F00-09 Organic, including symptomatic mental disorders

In the majority of cases, the recording of a diagnosis of any one of the disorders in this block will require the use of two codes: one for the psychopathological syndrome and another for the underlying disorder. The aetiological code should be recorded on Axis II.

Dementia

Dementia is a syndrome due to disease of the brain, usually of a chronic or progressive nature, in which there is disturbance of multiple higher cortical functions, including memory, thinking, orientation, comprehension, calculation, learning capacity, language and judgement. Consciousness is not clouded. Impairments of cognitive functioning are commonly accompanied, and occasionally preceded, by deterioration in emotional control, social behaviour, or motivation. This syndrome occurs in Alzheimer's disease, in cerebrovascular disease, and in other conditions primarily or secondarily affecting the brain.

Dementia is more than dysmnesia; there is also impairment of thinking and of reasoning capacity, and a reduction in the flow of ideas. The processing of incoming information is impaired, in that the individual finds it increasingly difficult to attend to more than one stimulus at a time, such as taking part in a conversation with several persons, and to shift the focus of attention from one topic to another. If dementia is the sole diagnosis, evidence of clear consciousness is required. However delirium superimposed upon dementia is common (F05.1). The above symptoms and impairments should have been evident *for at least six months* for a confident clinical diagnosis of dementia to be made.

Dementia and mental retardation

When dementia occurs in mentally retarded people it produces an appreciable decline in the previously low level of intellectual functioning, and usually some interference with personal activities of daily living, such as washing, dressing, eating, personal hygiene, excretory and toilet activities. How such a decline manifests itself will depend largely on the social and cultural setting in which the patient lives.

Care should be taken to avoid false positive identification. Other conditions producing a decline in functioning include depressive disorders, delirium (which may be induced by medication) and disorders resulting in motor slowness (such as hypothyroidism or depression). Depressive disorders may be difficult to distinguish from dementias, and the prescription of antidepressant medication may be justified when the diagnosis is uncertain despite careful investigation. Dementia and depression may co-exist. Hypothyroidism and other endocrine or metabolic disorders may result in dementia (F02) if untreated.

Cognitive decline resulting from a dementing process must be distinguished from cognitive impairment arising during the developmental period.

It is important not to make assumptions about premorbid levels of cognitive functioning (or to employ assessment instruments which make such assumptions). The diagnosis must be based on clear evidence of a *decline* in functioning within several domains of cognitive ability, and social behaviour, and daily living skills.

ICD-10 allows some dementias with known aetiologies to be coded separately. When such a dementia is diagnosed, the underlying physical disorder should be coded. In many instances aetiological diagnosis is only possible after post-mortem examination or brain biopsy.

## F00 Dementia in Alzheimer's disease

Alzheimer's disease is a primary degenerative cerebral disease of unknown aetiology, with characteristic neuropathological and neurochemical features. It is usually insidious in onset and develops slowly but steadily over a number of years. This period can be as short as 2 or 3 years, but can occasionally be considerably longer. The onset can be in middle adult life or even earlier (Alzheimer's

disease of presenile onset), but the incidence is higher in later life (Alzheimer's disease of senile onset). In cases with onset before the ages of 65-70, there is a likelihood of a family history of a similar temporal or parietal lobe damage, including dysphasia or dyspraxia. In cases with a later onset, the course tends to be slower and to be characterized by more general impairment of higher cortical functions. The clinical features may not progress in parallel with the above brain changes: one may be present with only minimal evidence of the other. It is often possible to make the diagnosis on clinical grounds alone, but aetiological diagnosis can only be confirmed by autopsy or brain biopsy. The following are essential for a definite diagnosis:

(a) Presence of dementia.
(b) Insidious onset with slow deterioration (but see note about Down's syndrome).
(c) Absence of clinical or other evidence that the mental state may be due to other systemic or brain disease
(d) Absence of a sudden, apoplectic onset, or of focal neurological signs as hemiparesis or sensory loss (although these phenomena may be superimposed later).

Down's syndrome and Alzheimer's disease

The prevalence of histopathological changes of the type characteristic of Alzheimer's disease is high among people with Down's syndrome in middle and late life. Clinical Alzheimer's disease affects a minority of people with Down's syndrome but becomes more prevalent among this population with increasing age. Progression is sometimes more rapid than in the non Down's syndrome population.

## F01 Vascular Dementia

Vascular dementia may be distinguished from dementia in Alzheimer's disease by its history, clinical features, and course. There is typically a history of transient ischaemic attacks. The dementia may follow a series of cerebrovascular accidents or a major stroke. Infarcts are usually small but cumulative in their effect. A dementia is present, but insight and judgement may be relatively well preserved. An abrupt onset or stepwise deterioration, and the presence of focal neurological signs, increases the probability of the diagnosis. Associated features include hypertension, carotid bruit, emotional lability or transient depressive mood or explosive laughter, and transient episodes of clouded consciousness or delirium. Vascular dementia may coexist with Alzheimer's disease (code F00.2).

## F02 Dementia in other diseases classified elsewhere

This code allows dementias associated with disorders such as Pick's disease, Parkinson's disease, Creutzfeldt-Jakob disease, Huntington's disease and human immunodeficiency virus disease to be coded. Dementia can occur as a manifestation or consequence of a variety of cerebral and somatic conditions, some of which may also be associated with mental retardation (such as Aicardi syndrome or hypothyroidism). To specify the etiology, the ICD-10 code for the underlying condition should be added.

## F03 Unspecified Dementia

## F05 Delirium, not induced by alcohol and other psychoactive substances

This is an etiologically nonspecific syndrome characterised by concurrent disturbances of consciousness and attention, perception, thinking, memory, psychomotor behaviour, emotion and the sleep-

walk cycle. A delirious state may be superimposed on, or progress to, dementia. The category should *not* be used for states of delirium associated with the use of psychoactive drugs specified in F10-9. Delirious states due to *prescribed* medication should be coded here, and the medication concerned be recorded on axis II using a T code from Chapter XIX.

Delirious states occur relatively commonly among people with mental retardation, possibly because underlying cerebral disorders predispose to a delirious response to infections, medications, systemic diseases etc.

For definite diagnosis, symptoms should be present in each one of the following areas, which should constitute a *change* from the person's usual pattern of functioning and emotional responses.

(a) An impairment of consciousness and attention (on a continuum from clouding to coma).
(b) Global disturbance of cognition (perceptual distortions, illusions and hallucinations - often visual; impairment of thinking and comprehension, typically with some degree of incoherence; impairment of recent memory with relatively intact unpredictable shifts from one to the other; increased reaction time; enhanced startle reaction; increased or decreased flow of speech.
(c) Disturbance of the sleep-wake cycle (insomnia; sleep loss; reversal of the cycle; daytime drowsiness; nocturnal worsening of symptoms; disturbing dreams or nightmares which may continue as hallucinations from awakening;
(d) Emotional disturbances such as depression, anxiety or fear, irritability, euphoria, apathy or perplexity.

The onset is usually rapid, the course diurnally fluctuating and the total duration of the condition less than six months. Evidence of cerebral dysfunction which *was not previously present* such as an abnormal electroencephalogram with slowing of the background activity may be helpful if the diagnosis is uncertain. Delirium superimposed on dementia is coded F.05.1

## F06 Other mental disorders due to brain damage and dysfunction and to physical disease

Psychoses and other psychiatric disorders which are etiologically linked epilepsy or other brain disorders or physical diseases may be coded here.

F06.0 Organic hallucinosis
F06.1 Organic catatonic disorder
F06.2 Organic delusional (schizophrenia-like) disorder
F06.3 Organic mood (affective) disorder
F06.4 Organic anxiety disorder
F06.5 Organic dissociative disorder
F06.6 Organic emotionally labile (asthenic) disorder
F06.7 Mild cognitive disorder
F06.8 Other specified mental disorders due to brain damage and dysfunction and to physical disease. This category may be used to code specified psychiatric disorders due to brain damage of dysfunction or to physical disease other than those listed above (F06.0-F06.7).
F06.9 Unspecified mental disorders due to brain damage and dysfunction and to physical disease. This code should only be used where a mental disorder is due to brain damage and dysfunction or to physical disease, and it is not possible to code it elsewhere.

## F07 Personality and behavioural disorders due to brain disease, damage and dysfunction

Alteration of personality and behaviour can be a residual or concomitant disorder of brain disease, damage or dysfunction. The underlying etiology should be sought and recorded.

### F07.0 Organic personality disorder

This disorder is characterized by a significant alteration of the habitual patterns of premorbid behaviour. The expression of emotions, needs, and impulses is affected. This syndrome occurs with frontal lobe lesions and circumscribed lesions in other brain areas. In addition to a history or other evidence of brain disease, damage or dysfunction, the diagnosis requires the presence of:

(a) consistently reduced ability to persevere with goal-directed activities (especially those involving longer periods of time and postponed gratification).

(b) altered emotional behaviour (lability, shallow and unwarranted cheerfulness, euphoria, inappropriate jocularity), the easy change to irritability, or short lived anger or aggression; apathy may be a prominent feature.

(c) expression of needs and impulses without regard to consequences or social convention (stealing, inappropriate sexual advances, etc.).

(d) cognitive disturbances such as suspiciousness and/or excessive preoccupation with a single (usually abstract) theme such as religion or "right" and "wrong".

(e) marked alteration in the rate and flow of language production, with features such as circumstantiality, over-inclusiveness, viscosity and hyperphagia.

(f) altered sexual behaviour.

### F07.1 Postencephalitic syndrome

### F07.2 Postconcussional syndrome

### F07.8 Other organic personality and behavioural disorders due to brain disease, damage and dysfunction

This category may be used to code specified personality and behavioural disorders due to brain damage of dysfunction or to physical disease other than those listed above (F07.0-F07.2). It may be applicable to behavioural phenotypes in disorders such as Fragile-X syndrome, Prader-Willi syndrome and Williams syndrome if the resulting behaviour is clinically significant. The physical disorder should also be coded using axis II.

### F07.9 Unspecified mental disorders due to brain damage, damage and dysfunction and to physical disease

## F10-19 Mental and behavioural disorders due to psychoactive

## substance use

These disorders are not common among people with mental retardation at present. They may become more prevalent in countries where policies of deinstitutionalisation and normalisation are pursued, or where contact with psychoactive substances is socially sanctioned. The clinical features are not markedly altered by pre-existing mental retardation, but may be affected by other disorders or disabilities associated with mental retardation. For example, seizures related to substance use of withdrawal may be more likely among people with a pre-existing seizure disorder.

The substance involved is indicated by means of the second and third characters (ie. the first two digits after the letter F), and the fourth and fifth characters specify the clinical states. To save space, all the psychoactive substances are listed first, followed by the four-character codes; these should be used, as required, for each substances specified, but it should be noted that not all four-character codes are applicable to all substances. It is always advisable to seek corroboration from more than one source of evidence relating to substance use. Objective analyses provide the most compelling evidence of present or recent use, though these data have limitations with regard to past use and current levels of use. Many drug users take more than one type of drug, but the diagnosis of the disorder should be classified, whenever possible, according to the most important single substance (or class of substances) used. When in doubt, code the drug or type of drug most frequently misused, particularly in those cases involving continuous or daily use. Misuse of non-psychoactive substances, such as laxatives or aspirin should be coded by means of F55. - (abuse of non-dependence-producing substances), with a fourth character to specify the type of substance involved.

F10 Mental and behavioural disorders due to use of alcohol
F11 Mental and behavioural disorders due to use of opioids
F12 Mental and behavioural disorders due to use of cannabinoids
F13 Mental and behavioural disorders due to use of sedatives or hypnotic
F19 Mental and behavioural disorders due to multiple drug use and use of other psychoactive substances

Four character categories may be used to specify clinical conditions including:

F1x.0 Acute intoxication
F1x.1 Harmful use
F1x.2 Dependence syndrome
F1.x3 Withdrawal state
F1x.4 Withdrawal state with delirium
F1x.5 Psychotic disorder
F1x.6 Amnesic syndrome
F1x.7 Residual and late-onset psychotic disorder
F1x.8 Other mental and behavioural disorders
F1x.9 Unspecified mental and behavioural disorder

A fifth character can be used to detail the clinical characteristics. eg F1x.51 psychotic disorder predominantly delusional but this level of detail can seldom be achieved in diagnosis among mentally retarded people except for codes that specify the presence of convulsions, these are:

F1x.31 Withdrawal state with convulsions
F1x.41 Withdrawal state with convulsions and delirium

## F20-F29 Schizophrenia, schizotypal and delusional disorders

## F20 Schizophrenia

The schizophrenic disorders are characterised in general by fundamental and characteristic distortions of thinking and perception, and by inappropriate or blunted affect. Clear consciousness and intellectual capacity are usually maintained, although certain cognitive deficits may evolve in the course of time. The disturbance involves the most basic functions that give the normal person a feeling of individuality, uniqueness, and self direction. The most intimate thoughts, feelings, and acts are often felt to be known to or shared by others, and explanatory delusions may develop, to the effect that natural or supernatural forces are at work to influence the afflicted individual's thoughts and actions in ways that are often bizarre. The individual may see himself or herself as the pivot of all that happens. Hallucinations, especially auditory, are common and may comment on the individual's behaviour and thoughts. Perception is frequently disturbed in other ways: colours or sounds may seem unduly vivid or altered in quality, and irrelevant features or ordinary things may appear more important than the whole object or situation. Perplexity is also common early on and frequently leads to a belief that everyday situations possess a special, usually sinister, meaning intended uniquely for the individual. In the characteristic schizophrenic disturbance of thinking, peripheral and irrelevant features of a total concept, which are inhibited in normal directed mental activity, are brought to the fore and utilised in place of those that are relevant appropriate to the situation. Thus thinking becomes vague, elliptical, and obscure, and its expression in speech sometimes incomprehensible. Breaks and interpolations in the train of thought are frequent, and thoughts may seem to be withdrawn by some outside agency. Mood is characteristically shallow, capricious, or incongruous. Ambivalence and disturbance of volition may appear as inertia, negativism, or stupor. Catatonia may be present. The onset may be acute, with seriously disturbed behaviour, or insidious, with a gradual development of odd ideas and conduct. The course of disorder shows equally great variation and is by no means inevitably chronic or deteriorating (the course is specified by five-character categories). In a proportion of cases, which may vary in different cultures and populations, the outcome is complete, or nearly complete, recovery. The sexes are approximately equally affected but the onset tends to be late in women.

Although no strictly pathognomonic symptoms can be identified, for practical purposes it is useful to divide the above symptoms into groups that have special importance for the diagnosis and often occur together, such as:

(a) thought echo, thought insertion or withdrawal, and thought broadcasting;
(b) delusions of control, influence, or passivity, clearly referred to body or limb movements or specific thought, actions or sensations; delusional perception;
(c) hallucinatory voices giving a running commentary on the patient's behaviour, or discussing the patient among themselves, or other types of hallucinatory voices coming from some part of the body;
(d) persistent delusions of other kinds that are culturally inappropriate and completely impossible, such as religious or political identity, or superhuman powers and abilities (eg. being able to control the weather, or being in communication with aliens from another world);
(e) persistent hallucinations in any modality, when accompanied either by fleeting or half-formed delusions without clear affective content, or by persistent over-valued ideas, or when occurring every day for weeks or months on end;
(f) breaks or interpolations in the train of thought, resulting in incoherence or irrelevant speech, or neologisms;
(g) catatonic behaviour, such as excitement, posturing, or waxy flexibility, negativism,

mutism, and stupor;

(h) "negative" symptoms such as marked apathy, paucity of speech, and blunting or incongruity of emotional responses, usually resulting in social withdrawal and lowering of social performance; it must be clear that these are not due to depression or to neuroleptic medication;

(i) a significant and consistent change in the overall quality of some aspects of personal behaviour, manifest as loss of interest, aimlessness, idleness, a self-absorbed attitude, and social withdrawal.

The normal requirement for a diagnosis of schizophrenia is that a minimum of one very clear symptom (and usually two or more if less clear-cut) belong to any one of the groups listed as (a) to (d) above, or symptoms from at least two of the groups referred to as (e) to (h), should have been clearly present for most of the time *during a period of 1 month or more.*

Conditions meeting such symptomatic requirements but of duration less than 1 month (whether treated or not) should be diagnosed in the first instance as acute schizophrenia-like psychotic disorder (23.2) and reclassified as schizophrenia if the symptoms persist for longer periods.

Viewed retrospectively, it may be clear that a prodromal phase in which symptoms and behaviour, such as loss of interest in work, social activities, and personal appearance and hygiene, together with generalised anxiety and mild degrees of depression and preoccupation, preceded the onset of psychotic symptoms by weeks or even months. Because of the difficulty in timing onset, the 1-month duration criterion applies only to the specific symptoms listed above and not to any prodromal nonpsychotic phase. The diagnosis of schizophrenia should not be made in the presence of extensive depressive or manic symptoms unless it is clear that schizophrenic symptoms antedated the affective disturbance. *Schizophrenia should not be diagnosed in the presence of overt brain disease* or during states of drug intoxication or withdrawal. Similar disorders developing in the presence of epilepsy or other brain disease should be coded under F06.2 and those induced by drugs under F1x.5.

Four subtypes are recognised (but explained below it is seldom possible to make these diagnoses in people with mental retardation).

F20.0 Paranoid schizophrenia.
F20.1 Hebephrenic schizophrenia.
F20.2 Catatonic schizophrenia.
F20.9 Schizophrenia, unspecified.

Schizophrenia and mental retardation

The diagnosis of schizophrenia and related disorders can be difficult to establish if the patient's ability to formulate ideas or communicate experiences to others is impaired. Relatively non-specific categories may have to be used, and often it will not be possible to differentiate between sub-types of schizophrenia.

Schizophrenia disorders occurring in people with mental retardation may be related to epilepsy, or organic brain disorder or to an associated pervasive developmental disorders (F84.x), or to a paranoid (F60.0) or schizoid (F60.1) personality disorder.

The course of the disorder, and the changes which occur over time, should be taken into account. A decline in social, self-care or other skills and the development of unusual or apparently irrational maladaptive behaviours may be the earliest manifestation. Symptoms which are commonly reported among people with mental retardation and schizophrenia include auditory hallucinations. It may be

difficult to assess the content or nature of auditory hallucinations. Unexplained bizarre behaviour which is out of character and *sustained in different environments,* should raise the possibility that the patient may have hallucinations or delusions. Negative symptoms are often given more weight when the diagnosis is considered for people with mental retardation. This may be appropriate, but other reasons should be excluded before these symptoms are used to support the diagnosis of schizophrenia. Autism and related disorders should not be confused with early onset schizophrenia.

## F22 Persistent delusional disorders

Delusions constitute the most conspicuous or the only clinical characteristic. They must be present for at least 3 months and be clearly personal rather than subcultural. Depressive symptoms or even a full-blown depressive episode (F32.-) may be present intermittently, or provided that the delusion persists at times when there is no disturbance of mood. There must be no evidence of brain disease, no or only occasional auditory hallucinations, and no history of schizophrenic symptoms (delusions of control, thought broadcasting, etc.).

## F23 Acute and transient psychotic disorders

The essential features of this disorder, in order of priority are:

(a) an acute onset (within 2 weeks) as the defining feature of the whole group.
(b) the presence of typical syndromes;
(c) the presence of associated acute stress.

*Acute onset* is defined as a change from a state without psychotic features to a clearly abnormal psychotic state, within a period of *2 weeks or less.* Abrupt onset is defined as a change within *48 hours* or less; such onsets may be associated with a better outcome. This period is to the onset of symptoms not the time of maximum severity. *Associated acute stress* can also be specified, with a fifth character if desired. Associated acute stress is taken to mean that the first psychotic symptoms occur within about *2 weeks* of one or more events that would be regarded as stressful to most people in similar circumstances, within the culture of the person concerned. Typical events would be bereavement, unexpected loss of partner or job, marriage, or the psychological trauma of combat, terrorism and torture. Long-standing difficulties or problems should not be included as a source of stress in this context. Complete recovery usually occurs within 2 or 3 months, often within a few weeks or even days, and only a small proportion of patients with these disorders develop persistent and disabling states.

These disorders are defined also by the *absence of organic causation* such as states of concussion, delirium or dementia.

## F24 Induced delusional disorder (Folie á Deux)

In this rare condition two people with close emotional links share the same delusions. One has a psychotic disorder, the other has induced delusions which fade rapidly when the two people are separated.

## F25 Schizoaffective disorders

These are episodic disorders in which both affective and schizophrenic symptoms are prominent

within the same episode of illness, preferably simultaneously, but at least within a few days of each other. Patients who suffer from recurrent schizoaffective episodes, particularly those whose symptoms are of the manic rather than the depressive type, usually make a full recovery and only rarely develop a defect state. A diagnosis of schizoaffective disorder should be made only when *both* definite schizophrenic and definite affective symptoms are prominent *simultaneously*, or within a few days of each other, within the same episode of illness, and when, as a consequence of this, the episode of illness does not meet criteria for either schizophrenia or a depressive or manic episode. The term should not be applied to patients who exhibit schizophrenic symptoms and affective symptoms only in different episodes of illness. It is common, for example, for a schizophrenic patient to present with depressive symptoms in the aftermath of a psychotic episode (see post schizophrenic depression (F20.4).

## F28 Other nonorganic psychotic disorders

This category is used more often in people with mental retardation because it is difficult to be certain of the exact nature of the disorder in patients who have difficulty in communicating.

## F29 Unspecified nonorganic psychosis

## F30-F39 Mood (affective) disorders

### Mood disorders and mental retardation

Care should be taken to exclude other disorders with similar symptomatology such as thyroid dysfunction (which is more prevalent among people with mental retardation) and organic mood (affective) disorders (F06.3). Diagnosis may be hindered by communication problems and greater weight may have to be given to features such as disturbances in appetite, sleep pattern and loss of interest in activities which usually give pleasure, and less weight attached to subjective experiences and descriptions. For these reasons non-specific diagnosis have often to be chosen. Frequent cycles of mood abnormality ("rapid cycling") may be a feature in some people with mental retardation.

## F30 Manic episode

This category is a single manic episode. If there are previous or subsequent affective episodes (depressive, manic, or hypomanic), the disorder should be coded under bipolar affective disorder (F31.1). Three degrees of severity are specified, sharing the common characteristics of elevated mood, and an increase in the quantity and speed of physical and mental activity.

### F30.0 Hypomania

In hypomania there are abnormalities of mood and behaviour too persistent and marked to be diagnosed as cyclothymia (F34.0) but not accompanied by hallucinations or delusions. There is a persistent mild elevation of mood (for at least several days on end), increased energy and activity, and usually marked feelings of well-being and both physical and mental efficiency. Increased sociability, talkativeness, overfamiliarity, increased sexual energy, and a decreased need for sleep are often present

but not to the extent that they lead to severe disruption of work or result in social rejection. Irritability, conceit, and boorish behaviour may take the place of the more usual euphoric sociability. Concentration and attention may be impaired, with reduced ability to settle to work or other activities, but this may not prevent the appearance of interests in quite new ventures and activities, or over-spending.

## F30.1 Mania without psychotic symptoms

In this condition there is almost uncontrollable excitement, overactivity, and decreased sleep. Social inhibitions are lost and there is marked distractibility. Grandiose ideas are present.

## F30.2 Mania with psychotic symptoms

In this condition, the clinical features described under F30.1 are accompanied by grandiose or persecuting delusions and/or hallucinations which are congruent with the prevailing mood.

## F31 Bipolar affective disorder

This disorder is characterised by repeated (ie. at least two) episodes in which the patient's mood and activity levels are significantly disturbed, this disturbance consisting on some occasions of an elevation of mood and increased energy and activity (mania or hypomania) and on others of a lowering of mood and decreased energy and activity (depression). Characteristically, recovery is usually complete between episodes, and the incidence in the two sexes is more nearly equal than in other mood disorders. Patients who suffer only from repeated episodes of mania are classified as bipolar (F31.8).

## F32 Depressive episode

In typical depressive episodes of all three varieties described below (mild F32.0), moderate (F32.1) and severe (F32.2 and F32.3)), the individual usually suffers from depressed mood, loss of interest and enjoyment, and reduced energy leading to increase fatiguability and diminished activity. Marked tiredness after only slight effort is common. Other common symptoms are:

(a) reduced concentration and attention
(b) reduced self-esteem and self-confidence
(c) ideas of guilt and unworthiness (even in a mild type of episode)
(d) bleak and pessimistic views of the future
(e) ideas of acts of self-harm or suicide
(f) disturbed sleep
(g) diminished appetite

The lowered mood varies little from day to day, and is often unresponsive to circumstances, yet may show a characteristic diurnal variation. The clinical presentation shows marked individual variations, and atypical presentations are particularly common in adolescence. In some cases, anxiety, distress, and motor agitation may be more prominent at times than the depression, and the mood change may also be masked by added features such as irritability, excessive consumption of alcohol, histrionic behaviour, and exacerbation of pre-existing phobic or obsessional symptoms, or by hypochondriacal preoccupations. For depressive episodes of all three grades of severity, a duration of at least 2 weeks is usually required for diagnosis, but shorter periods may be reasonable if symptoms are unusually severe and of rapid onset. The

categories of mild (F32.0), moderate (F32.1), and severe (F32.2 and F32.3) depressive episode should be used only for a single (first) depressive episode. Further depressive episodes should be classified under one of the subdivisions of recurrent depressive disorder (F33.-).

## F32 Mild depressive episode

Depressed mood, loss of interest and enjoyment, and increased fatiguability are usually regarded as the most typical symptoms of depression, and at least two of these, plus at least two of the other symptoms described on page 119 (for F32.-) should usually be present for a definite diagnosis. None of the symptoms should be present to an intense degree. Minimum duration of the whole episode is about 2 weeks.

## F33 Recurrent depressive disorder

The disorder is characterised by repeated episodes of depression as specified in depressive episode (mild F32.0), moderate (F32.1), or severe (F32.2 and F32.3), without any history of independent episodes of mood elevation and overactivity that fulfil the criteria of mania (F30.1 and F30.2). Individual episodes last between 3 and 12 months (median duration about 6 months). Recovery is usually complete between episodes, but a minority of patients may develop a persistent depression, mainly in old age (for which this category should still be used). Individual episodes of any severity are often precipitated by stressful life events; in many cultures, both individual episodes and persistent depression are twice as common in women as in men.

## F34 Persistent mood (affective) disorder

These are persistent and usually fluctuating disorders of mood in which individual episodes are rarely if ever sufficiently severe to warrant being described as hypomanic or even mild depressive episodes. Because they last for years at a time, and sometimes for the greater part of the individual's adult life, they involve considerable subjective distress and disability.

### F34.0 Cyclothymia

### F34.1 Dysthymia

## F38 Other mood (affective) disorders

## F39 Unspecified mood (affective) disorder

This should be used as a last resort, when no other term can be used.

## F40-F48 Neurotic, stress-related and somatoform disorder

Neurotic, stress-related and somatoform disorders and mental retardation. Neurotic and related disorders among people with mental retardation, varies from one country to another. As in non-retarded

people, anxiety disorders are relatively common, and there is an overlap with depressive syndromes. Dissociative convulsions often occur in association with non-dissociative convulsions.

It is important to bear in mind the patient's opportunities and life experiences when gathering data on which to base a diagnosis of one of the disorders. Some Phobias are easily overlooked among people with mental retardation: but they may restrict the person's life, and may be treatable. Care should be taken when considering diagnoses of phobias (especially social phobias or agoraphobia) to distinguish between phobic states (ie. a change from previous pattern of behaviour) and behaviours resulting from limited opportunities or experiences, or a lack of social skills. Anxiety may reflect the presence of another disorder such in autism in which anxiety commonly develops if the environment or routine of daily life is changed. Aggressive behaviour may result, in an attempt to escape or alter the environment.

## F40 Phobic anxiety disorders

In this group of disorders, anxiety is evoked only, or predominantly, by certain well-defined situations or objects (external to the individual) which are not currently dangerous. As a result, these situations or objects are characteristically avoided or endured with dread. Phobic anxiety is indistinguishable subjectively, physiologically, and behaviourally from other types of anxiety. It may vary in severity from mild unease to terror. The individual's concern may focus on individual symptoms such as palpitations or feeling faint and is often associated with secondary fears of dying, losing control, or going mad. The anxiety is not relieved by the knowledge that other people do not regard the situation in question as dangerous or threatening. Mere contemplation of entry to the phobic situation usually generates anticipatory anxiety. There is avoidance of situations which provoke anxiety. Phobic anxiety often coexists with depression. Most phobic disorders other than social phobias are more common in women that in men.

## F40.0 Agoraphobia

All of the following criteria should be fulfilled for a definite diagnosis:

(a) the psychological or autonomic symptoms must be primarily manifestations of anxiety and not secondary to other symptoms, such as delusions or obsessional thoughts;

(b) the anxiety must be restricted to (or occur mainly in) at least two of the following situations: crowds, public places, travelling away from home, and travelling alone;

(c) avoidance of the phobic situation must be, or have been, a prominent feature

The presence or absence of panic disorder (F41.0) in the agoraphobic situation on a majority of occasions may be recorded.

## F40.1 Social phobias

Social phobias often start in adolescence and are centred around a fear of scrutiny by other people in comparatively small groups (as opposed to crowds), leading to avoidance of social situations. Unlike most other phobias, social phobias are equally common in men and women. They may be discrete (ie. restricted to eating in public, to pubic speaking, or to encounters with the opposite sex) of diffuse, involving almost all social situations outside the family circle. Avoidance is often marked, and in extreme cases may result in almost complete social isolation. All of the following criteria should be fulfilled for a definite diagnosis:

(a) the psychological, behavioural or autonomic symptoms must be primarily manifestations of anxiety and not secondary to other symptoms such as delusions or obsessional thoughts;

(b) the anxiety must be restricted to or predominate in particular social situations;
(c) avoidance of the phobic situations must be a prominent feature.

## F41 Other anxiety disorders

Manifestations of anxiety are the major symptoms of these disorders and are not restricted to any particular environmental situation. Depressive and obsessional symptoms, and even some elements of phobic anxiety, may also be present, provided that they are clearly secondary or less severe.

### F41.0 Panic disorder (episodic paroxysmal anxiety)

The essential features are recurrent attacks of severe anxiety (panic) which are not restricted to any particular situation or set of circumstances, and which are therefore unpredictable. There is also, almost invariably, a secondary fear of dying , losing contort, or going mad. Individual attacks usually last for minutes only, though sometimes longer; their frequency and the course of the disorder are both rather variable. For a definite diagnosis, several severe attacks of autonomic anxiety should have occurred within a period of about 1 month:
(a) in circumstances where there is no objective danger;
(b) without being confined to known or predictable situations; and
(c) with comparative freedom from anxiety symptoms between attacks (although anticipatory anxiety is common).

### F41.1 Generalised anxiety disorder

The essential feature is anxiety, which is generalised and persistent but not restricted to, or even strongly predominating in, any particular environmental circumstances (ie. it is "free-floating"). As in other anxiety disorders the dominant symptoms are highly variable, but complaints of continuous feelings of nervousness, trembling, muscular tension, seating, lightheadedness, palpitations, dizziness, and epigastric discomfort are common. Fears that the sufferer or a relative will shortly become ill or have an accident are often expressed, together with a variety of other worries and foreboding. This disorder is more common in women, and often related to chronic environmental stress. Its course is variable but tends to be fluctuating and chronic. The sufferer must have primary symptoms of anxiety most days for at least several weeks at a time, and usually for several months. These symptoms should usually involve elements of apprehension, motor tension, and autonomic overactivity (lightheadedness, sweating, tachycardia or tachypnoea, epigastric discomfort, dizziness, dry mouth etc).

### F41.2 Mixed anxiety and depressive disorder

### F41.9 Anxiety disorder, unspecified

## F42 Obsessive-compulsive disorder

The essential feature of this disorder is recurrent obsessional thoughts or compulsive acts. (For brevity, "obsessional" will be used subsequently in place of "obsessive-compulsive" when referring to symptoms). Obsessional thoughts are ideas, images or impulses that enter the individual's mind again and

again in a stereotyped form. They are almost invariably distressing (because they are violent or obscene, or simply because they are perceived as senseless) and the sufferer often tries, unsuccessfully, to resist them. They are, however, recognized as the individual's own thoughts, even though they are involuntary and often repugnant. Compulsive acts or rituals are stereotyped behaviours that are repeated again and again. They are not inherently enjoyable, nor do they result in the completion of inherently useful tasks. The individual often views them as preventing some objectively unlikely event, often involving harm to or caused by himself or herself. Usually, though not invariably, this behaviour is recognised by the individual as pointless or ineffectual and repeated attempts are made to resist it; in very long-standing cases, resistance may be minimal. Autonomic anxiety symptoms are often present, but distressing feelings of internal or psychic tension without obvious autonomic arousal are also common. There is a close relationship between obsessional symptoms, particularly obsessional thoughts, and depression.

Obsessive-compulsive disorder is equally common in men and women, and there are often prominent anankastic features in the underlying personality. Onset is usually in childhood or early adult life. For a definite diagnosis, obsessional symptoms or compulsive acts, or both, must be present on most days for at least 2 successive weeks and be a source of distress of interference with activities. The obsessional symptoms should have the following characteristics:

(a) they must be recognised as the individual's own thoughts or impulses;
(b) there must be at least one thought or act that is still resisted unsuccessfully, even though others may be present which the sufferer no longer resists;
(c) the thought of carrying out the act must not in itself be pleasurable (simple relief of tension or anxiety is not regarded as pleasure in this sense);
(d) the thoughts, images, or impulses must be unpleasantly repetitive.

## F43 Reaction to severe stress, and adjustment disorder

This category differs from others in that it includes disorders identifiable not only on ground s of symptomatology and course but also on the basis of one or other of two causative influences - an exceptionally stressful life event producing an acute stress reaction, or a significant life change leading to continued unpleasant circumstances that result in an adjustment disorder. In contrast, the disorders brought together in this category are thought to arise always as a direct consequence of the acute severe stress or continued trauma. The stressful event of the continuing unpleasantness of circumstances is the primary and overriding casual factor, and the disorder would not have occurred without its impact. Reactions to severe stress and adjustment disorders in all age groups, including children and adolescents, are included in this category.

## F43.0 Acute stress reaction

A transient disorder of significant severity which develops in an individual without any other apparent mental disorder in response to exceptional physical and/or mental stress and which usually subsides within hours or days. The stressor may be an overwhelming traumatic experience involving a serious threat to the security or physical integrity of the individual or of a loved person(s) (eg. natural catastrophe, accident, battle, criminal assault, rape), or an unusually sudden and threatening change in the social position and/or network of the individual, such as multiple bereavement or domestic fire. The risk of this disorder developing is increased if physical exhaustion or organic factors (eg. in the elderly) are also present. Individual vulnerability and coping capacity play a role in the occurrence and severity of acute stress reaction, as evidenced by the fact that not all people exposed to exceptional stress develop the disorder. There must be an immediate and clear temporal connection between the impact of an exceptional stressor and the onset of symptoms; onset is usually within a few minutes, if not immediate. In addition,

the symptoms:

(a) show a mixed and usually changing picture; in addition to the initial state of "daze", depression, anxiety, anger, despair, overactivity, and withdrawal may all be seen, but no one type of symptom predominates for long;

(b) resolve rapidly (within a few hours at the most) in those cases where removal from the stressful environment is possible; in cases where the stress continues or cannot by its nature be reversed, the symptoms usually begin to diminish after 24-48 hours and are usually minimal after about 3 days.

## F43.1 Post-traumatic stress disorder

This arises as a delayed and/or protracted response to a stressful event or situation (either short- or long-lasting) of an exceptionally threatening or catastrophic nature, which is likely to cause pervasive distress in almost anyone (eg. natural or manmade disaster, combat, serious accident, witnessing the violent death of others, or being the victim of trauma in intrusive memories ("flashbacks") or dreams, occurring against the persisting background of a sense of "numbness" and emotional blunting, detachment from other people, unresponsiveness to surroundings, anhedonia, and avoidance of activities and situations reminiscent of the trauma. Commonly there is fear and avoidance of cues that remind the sufferer of the original trauma. Rarely, there may be dramatic, acute burst of fear, panic or aggression, triggered by stimuli arousing a sudden recollection and/or re-enactment of the trauma or of the original reaction to it. There is usually a state of autonomic hyperarousal with hypervigilance, an enhanced startle reaction, and insomnia. The onset follows the trauma with a latency period which may range from a few weeks to months (but rarely exceeds 6 months). The course is fluctuating but recovery can be expected in the majority of cases. In a small proportion of patients the condition may show a chronic course over may years and a transition to an enduring personality change (see F62.0).

## F43.2 Adjustment disorder

States of subjective distress and emotional disturbance, usually interfering with social functioning and performance, and arising in the period of adaptation to a significant life change or to the consequences of a stressful life event (including the presence of possibility of serious physical illness). The stressor may have affected the integrity of an individual's social network (through bereavement or separation experiences) or the wider system of social supports and values (migration or refugee status). The stressor may involve only the individual or also his or her group or community. The onset is usually within 1 month of the occurrence of the stressful event or life change, and the duration of symptoms does not usually exceed 6 months, except in the case of prolonged depressive reaction (F43.21). If the symptoms persist beyond this period, the diagnosis should be changed according to the clinical picture present, and any continuing stress can be coded by means of one of the Z codes in Chapter XXI of ICD-10. Situations which may provoke adjustment disorders among people with mental retardation include resettlement from hospital, and other changes relating to staffing or residential services, as well as the stressful life events described above. People with autism and related disorders are more vulnerable, because of their intolerance of change.

Diagnosis depends on a careful evaluation of the relationship between:

(a) form, content, and severity of symptoms;

(b) previous history, and personality; and

(c) stressful event, situation, or life crisis.

## F44 Dissociative (conversion) disorders

For a definite diagnosis the following should be present:
(a) the clinical features as specified for the individual disorders in F44.-;
(b) no evidence of a physical disorder that might explain the symptoms;
(c) evidence for psychological causation, in the form of clear association in time with stressful events and problems or disturbed relationships (even if denied by the individual).

### F44.0 Dissociative amnesia

There is a profound loss of memory involving recent as well as remote events which cannot be explained by an organic disorder. Complete amnesia is rare; it is usually part of a fugue (F44.1) and, if so, should be classified as such.

### F44.1 Dissociative fugue

There is profound loss of memory for recent as well as distant events together with an apparently purposeful journey away from home or place of work, during which self-care is maintained. As with dissociative amnesia, differentiation from conscious simulation may be very difficult.

### F44.2 Dissociative stupor

The individual's behaviour fulfils criteria for stupor, but there is not evidence of a physical cause and there is positive evidence of psychogenic causation (such as recent stressful events or prominent interpersonal or social problems).

### F44.3 Trance and possession disorders

Only trance disorders that are involuntary and unwanted, and that intrude into ordinary activates by occurring outside (or being a prolongation of) religious or other culturally accepted situation should be included here.

Dissociative disorders of movement and sensation and mental retardation

In these disorders, the patient presents as having a physical disorder, although none can be found that would explain the symptoms. Assessment of the mental state and social situation suggests that the resulting disability is helping the patient escape a conflict, or express dependency or resentment indirectly. The disability may vary depending on the people present and the patient's mental state. Premorbid abnormalities or personality and social functioning are usually found. The symptoms may correspond to the patient's concept of physical disorder, rather than to anatomical or physiological principles, and someone with a similar illness may be known to the patient.

Dissociative convulsions and other disorders of movement and sensation are more common among people who also have non-dissociative impairments or disabilities. *The diagnosis should remain probable or provisional if there is any doubt about the contribution of actual or possible physical disorder, or if it is impossible to understand why the disorder has developed.* Isolated dissociative symptoms may be associated with affective, schizophrenic and other major mental disorder; these disorders are usually obvious and should take diagnostic precedence. Where a non-physical basis can be demonstrated, it may be difficult to distinguish between dissociative states and (conscious) simulation.

### F44.4 Dissociative motor disorder

The commonest varieties are loss of ability to move the whole or part of a limb or limbs.

### F44.5 Dissociative convulsions

Dissociative convulsions (pseudoseizures) may mimic the movements of epileptic seizures closely, but tongue-biting, serious bruising due to falling, and incontinence of urine are rare in dissociative convulsions, and loss of consciousness is absent or replaced by a state of stupor or trance.

### F44.6 Dissociative anaesthesia and sensory loss

Dissociative deafness and anosmia are far less common than loss of sensation or vision, the latter rarely being total and usually involving loss of acuity, blurring, or "tunnel" vision.

### F44.7 Mixed dissociative (conversion) disorders

### F44.8 Other dissociative (conversion) disorders

### F44.9 Dissociative (conversion) disorder, unspecified

## F45 Somatoform disorders

The main feature of somatoform disorders is repeated presentation of physical symptoms, together with persistent requests for medical investigations, in spite of repeated negative findings and reassurances by doctors that the symptoms have not physical basis. If any physical disorders are present, they do not explain the nature and extent of the symptoms or the distress and preoccupations of the patient. Even when the onset and continuation of the symptoms bear a close relationship with unpleasant life events or with difficulties or conflicts, the patient usually resists attempts to discuss the possibility of psychological causation; this may even be the case in the presence of obvious depressive and anxiety symptoms.

Somatoform disorders and mental retardation

Repeated presentation of physical symptoms and persistent requests for medical or nursing

attention or investigation are not uncommon among some people with mental retardation. The distinction between categories within F45 may be difficult, as may the assessment of the degree of conscious motivation for the behaviour.

## F45.0 Somatization disorder

The main features are multiple, recurrent, and frequently changing physical symptoms, which have usually been present for several years before the patient is referred to a psychiatrist. A definite diagnosis requires the presence of all of the following.

(a) at least 2 years of multiple and variable physical symptoms for which no adequate physical explanation has been found;
(b) persistent refusal to accept the advice or reassurance of several doctors that there is no physical explanation for the symptoms;
(c) some degree of impairment of social and family functioning attributable to the nature of the symptoms and resulting behaviour.

## F45.2 Hypochondriacal disorder

For a definite diagnosis, both of the following should be present:

(a) persistent belief in the presence of at least one serious physical illness underlying the presenting symptom or symptoms, even though repeated investigations and examinations have identified no adequate physical explanation, or a persistent preoccupation with a presumed deformity or disfigurement.
(b) persistent refusal to accept the advice and reassurance of several different doctors that there is not physical illness or abnormality underlying the symptoms.

Hypochondriacal complaints may be used by some people with mental retardation as a means of seeking attention, especially from medical or nursing personnel. If the behaviour is clearly a deliberate attempt to influence events, a psychiatric diagnosis should not be made.

## F45.3 Somatic autonomic dysfunction

## F45.4 Persistent somatoform pain disorder

The complaint is of severe, persisting and distressing pain which cannot be explained by a physiological process or physical disorder. The pain occurs in association with emotional conflict or psychosocial problems that are sufficient to justify the conclusion that they are the main causative influences. The result is usually an increase in support and attention.

## F45.8 Other somatoform disorders

## F45.9 Somatoform disorders, unspecified

## F48 Other neurotic disorders

## F48.0 Neurasthenia

Considerable cultural variations occur in the presentation of this disorder; two main types occur, with substantial overlap. In one type, the main feature is a complaint of increased fatigue after mental effort, often associated with some decrease in occupational performance or coping efficiency in daily tasks. In the other type, the emphasis in on feelings or bodily or physical weakness and exhaustion after only minimal effort, accompanied by a feeling of muscular aches and pains and inability to relax. In both types, a variety of other unpleasant physical feelings, such as dizziness, tension headaches, and a sense of general instability, is common. Definite diagnosis requires the following:

(a) either persistent and distressing complaints of increased fatigue after mental effort, or persistent and distressing complaints of bodily weakness and exhaustion after minimal effort;
(b) at least two of the following: feelings of muscular aches and pains; dizziness; tension headaches; sleep disturbance; inability to relax; irritability; dyspepsia;
(c) any autonomic or depressive symptoms present are not sufficiently persistent and severe to fulfil the criteria for any of the more specific disorders in this classification.

## F48.1 Depersonalisation-derealisation syndrome

## F48.9 Neurotic disorder, unspecified

## F50-F59 Behavioural syndromes associated with physiological disturbances and physical factors

## F50 Eating disorders

Eating disorders and mental retardation

Overeating and unusual dietary preferences are relatively common among people with mental retardation, but eating disorders such as anorexia nervosa and bulimia nervosa are less frequent. The latter disorders have been described in association with mental retardation generally, and specifically with Down's syndrome and Turner syndrome. The factors which are thought to contribute to anorexia and related disorders include cultural expectations and societal pressures to be thin. May people with mild and moderate mental retardation were protected against such pressures in the past through policies or segregation and institutionalisation, and such disorders may become more prevalent in societies where policies of deinstitutionalisation and community care are implemented.

Regurgitation, rumination and psychogenic vomiting are seen among people with mental retardation, and care should be taken to identify and treat associated (often secondary) physical disorders such as hiatus hernia and reflux oesophagitis.

## F50.0 Anorexia nervosa

Anorexia nervosa is a disorder characterised by deliberate weight loss, induced and/or sustained by the patient. For a definite diagnosis, all the following are required:

(a) Body weight is maintained at least 15% below that expected (either lost or never achieved), or Quelet's body-mass index is 17.5 or less. Prepubertal patients may show failure to make the

expected weight gain during the period of growth.

(b) The weight loss is self-induced by avoidance of "fattening foods" and one or more of the following: self-induced vomiting; self-induced purging; excessive exercise; use of appetite suppressants and/or diuretics.

(c) There is a body-image distortion in the form of a specific psychopathology whereby a dread of fatness persists as an intrusive, overvalued idea and the patient imposed a low weight threshold on himself or herself.

(d) A widespread endocrine disorder involving the hypothalamic-pituitary gonadal axis in manifest in women as amenorrhoea and in men as a loss of sexual interest and potency. There may also be elevated levels of growth hormone,raised levels of cortisol, changes in the peripheral metabolism of the thyroid hormone, and abnormalities of insulin secretion.

(c) If the onset is prepubertal, the sequence of pubertal events is delayed or even arrested (growth ceases; in girls the breasts do not develop and there is a primary amenorrhoea; in boys the genitals remain juvenile). With recovery, puberty is often completed normally, but the menarche is late.

## F50.1 Atypical anorexia nervosa

This term should be used for those individuals in whom one or more of the key features of anorexia nervosa (F50.0), such as amenorrhoea or significant weight loss, is absent, but who otherwise present a fairly typical clinical picture.

## F50.2 Bulimia nervosa

Bulimia nervosa is a syndrome characterised by repeated bouts of overeating and an excessive preoccupation with the control of body weight, leading the patient to adopt extreme measure so as to mitigate the "fattening" effects of ingested food. The term should be restricted to the form of the disorder that is related to anorexia nervosa by virtue of sharing the same psychopathology. The age and sex distribution is similar to that of anorexia nervosa, but the age of presentation tends to be slightly later. Repeated vomiting is likely to give rise to disturbances of body electrolytes, physical compilation (tetany, epileptic seizures, cardiac arrhythmias, muscular weakness), and further severe loss of weight. For a definite diagnosis all the following are required:

(a) There is a persistent preoccupation with eating, and an irresistible craving for food; the patient succumbs to episodes of overeating in which large amounts of food are consumed in short periods of time.

(b) The patient attempts to counteract the "fattening" effects of food by one or more of the following; self-induced vomiting; purgative abuse, alternating periods of starvation; use of drugs such as appetite suppressants, thyroid preparations or diuretics. When bulimia occurs in diabetic patients they may choose to neglect their insulin treatment.

(c) The psychopathology consists of a morbid dread of fatness and the patient sets herself or himself a sharply defined weight threshold, well below the Premorbid weight that constitutes the optimum or healthy weight in the opinion of the physician. There is often, but not always, as history of an earlier episode of anorexia nervosa. Bulimia nervosa must be differentiated from gastrointestinal disorder, personality disorder and depressive syndromes.

## F50.3 Atypical bulimia nervosa

## F50.4 Overeating associated with other psychological

## disturbances

## F50.5 Vomiting associated with other psychological disturbances

Apart from the self-induced vomiting of bulimia nervosa, repeated vomiting may occur in dissociative disorders (F44.-), in hypochondriacal disorder (F45.2) when vomiting may be one of several bodily symptoms and in pregnancy when emotional factors may contribute to recurrent nausea and vomiting.

Psychogenic vomiting may be seen in association with mental retardation, and may have a function (such as anxiety reduction) which maintains the behaviour. It must be distinguished from disorders such as regurgitation and rumination, which may be coded using F50.8

## F50.8 Other eating disorders

This category includes disorders such as rumination and regurgitation, which are not uncommon among adults and children with mental retardation. Feeding disorder of infancy and childhood (F98.2) and pica of infancy and childhood (F98.3) are more specific categories which may be applicable for some children with mental retardation.

## F50.9 Eating disorder, unspecified

## F51 Non-organic sleep disorders

This section includes only those sleep disorders in which emotional causes are considered to be a primary factor. Sleep disorders of organic origin such as Kleine-Levin syndrome (G47.8) are coded in Chapter VI (G47.-) of ICD-10. Nonpsychogenic disorders including narcolepsy and cataplexy (G47.4) and disorders of the sleep-wake schedule (G47.2) are also listed in Chapter VI, as are sleep apnoea (G47.3) and episodic movement disorders which include nocturnal myoclonus (G25.3). Finally, enuresis (F98.0) is listed with other emotional and behavioural disorders with onset specific to childhood and adolescence, while primary nocturnal enuresis (R33.8), which is considered to be due to a maturational delay of bladder control during sleep, is listed in Chapter XVIII of ICD-10 among the symptoms involving the urinary system.

Sleep disorders of organic origin, or where an organic factor seems to play a major factor in causation, should be coded using G47.-. Such disorders are not uncommon in association with some specific developmental disorders such as Prader-Willi syndrome.

F51.0 Nonorganic insomnia
F51.2 Nonorganic hypersomnia
F51.2 Nonorganic disorder of the sleep-wake schedule
F51.3 Sleepwalking (somnambulism)
F51.4 Sleep terrors (night terrors)

F51.5 Nightmares
F51.9 Nonorganic sleep disorder, unspecified

## F54 Psychological and behavioural factors associated with disorders or diseases classified elsewhere

## F60-69 Disorders of adult personality and behaviour

### F60-F62 Specific personality disorders, mixed and other personality disorder, and enduring personality changes

These types of condition comprise deeply ingrained and enduring behaviour patterns, manifesting themselves as inflexible responses to a broad range of personal and social situations. The represent either extreme or significant deviations from the way the average individual in a given culture perceives, thinks, feels, and particularly relates to others. Such behaviour patterns tend to be stable and to encompass multiple domains of behaviour and psychological functioning. They are frequently, but not always, associated with various degrees of subjective distress and problems in social functioning and performance. Personality disorders differ from personality change in their timing and the mode of their emergence: they are developmental conditions, which appear in childhood or adolescence and continue into adulthood. They are not secondary to another mental disorder or brain disease, although they may precede and coexist with other disorders. In contrast, personality change is acquired, usually during adult life, following severe or prolonged stress, extreme environmental deprivation, serious psychiatric disorder or brain disease or injury (See F07.-_).

Each of the conditions in this group can be classified according to its predominant behavioural manifestations. However, *classification in this area is currently limited to the description of a series of types and subtypes, which are not mutually exclusive and which overlap in some of their characteristics.* Personality disorders are therefore subdivided according to clusters of traits that correspond to the most frequent or conspicuous behavioural manifestations.

The assessment should be based on as many sources of information as possible. Although it is sometimes possible to evaluate a personality condition in a single interview with the patient, it is often necessary to have more than one interview and to collect history data from informants.

Cultural or regional variations in the manifestations of personality conditions are important, but little is known about them. Personality conditions that appear to be frequently recognised in a given part of the world but do not correspond to any one of the specified subtypes below may be classified as "other" personality disorders and identified through a five-character code provided in an adaptation of this classification for that particular country or region. Local variations in the manifestations of a personality disorder may also be reflected in the wording of the diagnostic guidelines set for such conditions.

Personality disorder and mental retardation

It may be difficult to distinguish between specific personality disorders and other behaviour disorders occurring in the setting of mental retardation. It is important to formulate the possible factors which predispose to, precipitate and maintain maladaptive behaviours, and to make a diagnosis of personality disorder only when it is clear that the behaviour does not result from a specific psychiatric disorder (such as an affective disorder) or a physical disorder, and where individual personality

characteristics appear to be the most important single factor underlying the behaviour. Maladaptive behaviour such as aggression and persistent disregard for social norms may be associated with many underlying factors or disorders (autism, communication problems, disinhibition, etc). The function of the maladaptive behaviours shown by an individual may vary over time, necessitating a longitudinal assessment before a diagnosis of personality disorder can be confirmed. The context or contexts in which behaviour occur may help to differentiate between personality and other behavioural disorders.

Where there is evidence that behaviours which would otherwise fulfil criteria for a personality disorder are associated with an underlying physical disorder, categories within F06 and F07 should be considered (eg. F06.8, specified mental disorders due to brain damage and dysfunction and to physical disease, F07.0 organic personality disorder).

## F60 Specific personality disorders

A specific personality disorder is a severe disturbance in the characterological constitution and behavioural tendencies of the individual, usually involving several areas of the personality, and nearly always associated with considerable personal and social disruption. Personality disorder tends to appear in late childhood or adolescence and continues to be manifest into adulthood. It is therefore unlikely that the diagnosis of personality disorder will be appropriate before the age of 16 or 17 years.

Specific personality disorders are conditions not directly attributable to gross brain damage or disease, or to another psychiatric disorder, meeting the following criteria:

(a) markedly disharmonious attitudes and behaviour, involving usually several areas of functioning, eg. affectivity, arousal, impulse control, ways of perceiving and thinking, and style of relating to others;
(b) the abnormal behaviour pattern is enduring, of long standing, and not limited to episodes of mental illness;
(c) the abnormal behaviour pattern is pervasive and clearly maladaptive to a broad range of personal and social situations;
(d) the above manifestations always appear during childhood or adolescence and continue into adulthood;
(e) the disorder leads to considerable personal distress but this may only become apparent late in its course;
(f) the disorder is usually, but not invariably, associated with significant problems in occupational and social performance.

For different cultures it may be necessary to develop specific sets of criteria with regard to social norms, rules and obligations. For diagnosing most of the subtypes listed below, clear evidence is usually required of the presence of at least three of the traits or behaviours given in the *Clinical Descriptions and Diagnostic Guidelines.* As these specific disorders can seldom be diagnosed in mentally retarded people, only the heading are given.

F60.0 Paranoid personality disorder
F60.1 Schizoid personality disorder
F60.2 Dissocial personality disorder
F60.3 Emotionally unstable personality disorder
  F60.30 Impulsive type
  F60.31 Borderline type

F60.4 Histrionic personality disorder
F60.5 Anankastic personality disorder
F60.6 Anxious (avoidant) personality disorder
F60.7 Dependent personality disorder
F60.8 Other specific personality disorder
F60.9 Personality disorder, unspecified

## F61 Mixed and other personality disorders

## F62 Enduring personality changes, not attributable to brain damage and disease

## F63 Habit and impulse disorders

By convention, the habitual excessive use of alcohol or drugs (F10-F19) and impulse and habit disorders involving sexual (F65.-) or eating (F52.-) behaviour are excluded.

### F63.1 Pathological fire-setting (pyromania)

The disorder is characterised by multiple acts of, or attempts at, setting fire to property or other objects, without apparent motive, and by a persistent preoccupation with subjects related to fire and burning. There may also be an abnormal interest in fire-engines, and other fire-fighting equipment, in other associations of fires, and in calling out the fire service.

The essential features are repeated fire-setting without any obvious motive such as monetary gain, revenge, or political extremism; an intense interest in watching fires burn; and reported feelings of increasing tension before the act, and intense excitement immediately after it has been carried out. Pathological fire-setting should be distinguished from deliberate fire-setting (where an obvious motive is present), fire-setting by a young person with conduct disorder and fire-setting by an adult with sociopathic personality disorder, organic personality disorder or schizophrenia. Dementia or acute organic states may also lead to inadvertent fire-setting, acute drunkenness, chronic alcoholism or other drug intoxication (F10.F19) are other causes.

It may be difficult to establish the main motivation underlying the setting of a fire or fires by someone with mental retardation. For example, there maybe a combination of excitement, tension-reduction, lack of awareness of the seriousness of the behaviour and a desire for change (eg. a change of residence).

### F63.2 Pathological stealing (Kleptomania)

The disorder is characterised by repeated failure to resist impulses to steal objects that are not acquired for personal use or monetary gain. The objects may instead be discarded, given away, or hoarded. There is an increasing sense of tension before, and a sense of gratification during and immediately after, the act. Although some effort at concealment is usually made, not all the opportunities for this are taken. The theft is a solitary act, not carried out with an accomplice. The individual may express anxiety,

despondency, and guilt between episodes of stealing from shops (or other premises) but this does not prevent repetition. Cases meeting this description alone, and not secondary to one of the disorders listed below, are uncommon. Pathological stealing should be distinguished from recurrent shoplifting, organic mental disorders (with memory impairment) and depressive disorders.

### F63.3 Trichotillomania

A disorder characterised by noticeable hair loss due to a recurrent failure to resist impulses to pull out hairs. The hair-pulling is usually preceded by mounting tension and is followed by a sense of relief or gratification. This diagnosis should not be made if there is a pre-existing inflammation of the skin, or if the hair-pulling is in response to a delusion or a hallucination.

## F64 Gender identity disorder

## F65 Disorders of sexual preference

### F65.2 Exhibitionism

A recurrent of persistent tendency to expose the genitalia to strangers (usually of the opposite sex) or to people in public places, without inviting or intending closer contact. There is usually, but not invariably, sexual excitement at the time of the exposure and the act is commonly followed by masturbation. This tendency may be manifest only at times of emotional stress or crises, interspersed with long periods without such overt behaviour. Exhibitionism is almost entirely limited to heterosexual males who expose to females, adult or adolescent, usually confronting them from a safe distance in some public place. Most exhibitionists find their urges difficult to control and ego-alien. If the witness appears shocked, frightened, or impressed, the exhibitionist's excitement is often heightened.

### F65.3 Voyeurism

### F65.4 Paedophilia

A sexual preference for children, usually of prepubertal or early pubertal age. Some paedophile are attracted only to girls, others only to boys, and others again are interested in both sexes. Paedophilia is rarely identified in women. Contacts between adults and sexually mature adolescents are socially disapproved, especially if the participants are of the same sex, but are not necessarily associated with paedophilia. An isolated incident, especially if the perpetrator is himself an adolescent, does not establish the presence of the persistent or predominant tendency required for the diagnosis. Included among paedophilia, however, are men who retain a preference for adult sex partners but, because they are chronically frustrated in achieving appropriate contacts, habitually turn to children as substitutes. Men who sexually moles their own prepubertal children occasionally approach other children as well, but in either case their behaviour is indicative of paedophilia.

## F66 Psychological and behavioural disorders associated with sexual development and orientation

## F68 Other disorders of adult personality and behaviour

### F68.0 Elaboration of physical symptoms for psychological reasons

Physical symptoms compatible with and originally due to a confirmed physical disorder, disease, or disability become exaggerated or prolonged due to the psychological state of the patient. An attention-seeking (histrionic) behavioural syndrome develops, which may also contain additional (and usually nonspecific) complaints that are not of physical origin. The patient is commonly distressed by this pain or disability and is often preoccupied with worries, which may be justified, of the possibility of prolonged or progressive disability or pain.

### F68.1 Intentional production or feigning of symptoms or disabilities, either physical or psychological (factitious disorder)

In the absence of a confirmed physical or mental disorder, disease, or disability, the individual feigns symptoms repeatedly and consistently. For physical symptoms this may even extend to self-infliction of cuts or abrasions to produced bleeding, or to self-injection of toxic substances. The imitation of pain and the insistence upon the presence of bleeding may be so convincing and persistent that repeated investigations and operations are performed at several different hospitals or clinics, in spite of repeatedly negative findings. *Malingering,* defined as the intentional production or feigning of either physical or psychological symptoms or disabilities, motivated by external stresses or incentives, should be coded as Z76.5 or ICD-10 and not by one of the codes in this book.

### F68.8 Other specified disorder of adult personality and behaviour

## F69 Unspecified disorder of adult personality and behaviour

## F80-89 Disorders of Psychological Development

Disorders included in F80-89 have three characteristics in common:

(a) an onset during infancy or childhood;

(b) delayed development of functions that are strongly related to biological maturation of the nervous system;

(c) a steady course, unlike the remissions and relapses that are generally characteristic of psychiatric disorders

As in the general population, the functions most affected by developmental disorders in mentally retarded people include language, motor coordination and/or visuo-spatial skills. Impairments in these functions are common among mentally retarded children and adults: specific developmental disorders are therefore difficult to detect among this population. Also, whereas these conditions are far more common in males than females in the general population, this effect is less marked in the mentally retarded.

In some cases - notably of autism - a period of normal development precedes the disorder. However, the symptoms and natural history of these cases have much in common with other developmental disorders, with which they are classified.

Diagnosis of Specific Developmental Disorders

It is important to distinguish specific developmental disorders from general intellectual retardation. To make the diagnosis of specific developmental disorder in addition to mental retardation, the following guidelines should be followed.

## F80 Specific developmental disorders of speech and language

These are disorders of language development, not due to an identifiable cause (neurological or speech mechanism abnormalities, sensory impairments, environmental factors, etc). In diagnosing such disorders in mentally retarded individuals, two kinds of difficulty are commonly encountered.

(i) The differentiation from language delay which is simply part of global delay or mental retardation. (Where this occurs, the language delay should not be assigned a coding in this group.) Many individuals with mental retardation do have specific deficits in language development. Where language developmental delay is more severe than the general level of retardation - and this is apparent in everyday life - a specific developmental disorder of speech and language may be coded *in addition* to the F70-79 code. A deficit in the score on a measure of languages or speech development of at least two standard deviations more severe that the global delay, indicates the presence of a specific developmental disorder of language or speech.

(ii) The diagnosis of language delay in the presence of deafness, neurological or structural abnormality: all of which are common in mentally retarded people. Severe deafness and abnormalities such as uncorrected cleft lip-palate disrupt language development. Where language delay occurs in the presence of a severe abnormality of this king, it should not be coded separately. However, a developmental language disorder may be diagnosed where associated with mild deafness, neurological or structural abnormality, where these are deemed insufficient to cause a language delay.

*Includes;*

F80.0 Specific speech articulation disorder
F80.1 Expressive language disorder
F80.2 Receptive language disorder
F80.3 Acquired aphasia with epilepsy (Landau-Kleffner syndrome)
F80.8 Other developmental disorders of speech and language
F80.9 Developmental disorder of speech and language, unspecified

## F81 Specific developmental disorders of scholastic skills (SDSS)

These disorders are distinct from delayed scholastic development caused by poor education, acquired brain trauma, uncorrected visual or hearing problems, or disease. The main difficulty in diagnosing them among mentally retarded individuals lies in the distinction from scholastic delay which is a result of the general level of intellectual functioning.

The diagnosis of SDSS is reserved for a deficit two standard deviations below that to be expected of a general intellectual level, where this has been present from early in life. However, where such an impairment has been acquired later in life, and not present from early in development, the performance on testing will generally not indicate this.

*Includes;*

F81.0 Specific reading disorder
F81.1 Specific spelling disorder
F81.2 Specific disorder of arithmetical skills
F81.3 Mixed disorder of scholastic skills
F81.8 Other developmental disorders of scholastic skills
F81.9 Developmental disorder of scholastic skills, unspecified

## F82 Specific developmental disorder of motor function

In this disorder, there is a severe delay of motor coordination that is not simply a reflection of general mental retardation or any neurological disorder, and is not the direct result of any visual or hearing deficit. As with all developmental disorders, an appropriate standardised test aids diagnosis. The diagnosis should be confined to coordination deficits which have been present from early in life.

## F83 Mixed specific developmental disorders

This category is confined to those cases where a combination of different specific developmental disorders occurs, but none predominates sufficiently to qualify as a main diagnosis. The category should be used where dysfunctions meet the criteria for two or more of F80, F81 and F82.

## F84 Pervasive developmental disorders

The pervasive developmental disorders (pdd) feature abnormal social behaviour and communication, and a narrow range of interests and activities which are both unique to the individual and carried out repetitively. Unlike the specific developmental disorders, the pdds affect a wide range of functioning, particularly social behaviour and language. Also, unlike general mental retardation, the pdds do not have the same direct impact upon other aspects of learning and functioning, such as scholastic learning - where areas or "islets" of normal or high ability commonly occur in persons affected by pdds - or the potential to acquire self-care skills. However, due to their inherent language and social disabilities, the pdds do have some general, or "pervasive", effect upon learning other skills. In most cases, pdds have their onset in infancy.

These behaviours are very common in mentally retarded individuals, and especially the more severely retarded. However, as with other developmental disorders, the category F84 should only be used if the developmental delay is clearly out of keeping with the general level of retardation.

Certain identifiable causes of mental retardation - such as Fragile X Syndrome, congenital Rubella

and tuberous sclerosis - are frequently associated with a pdd. However, pdd should be diagnosed in a mentally retarded individual only on the basis of behavioural features. Where a pdd forms part of the *behavioural phenotype* of a specific cause of mental retardation, the pervasive developmental disorder, the cause of the mental retardation, the pervasive developmental disorder, the cause of the mental retardation, and the degree of the mental retardation itself (F70-F79) should be are separately coded on the appropriate axes.

The majority of pdds belong to the group of **autistic spectrum disorders** (childhood autism F84.0, atypical autism F84.5, and Aspergers syndrome F84.5). There remains some controversy regarding the status and classification of these developmental disorders, particularly Asperger's syndrome.

## F84.0 Childhood autism

Childhood autism is defined by:

(i) onset before the age of three years
(ii) social interaction disability
(iii) communication disability
(iv) restricted repetitive behaviour

### Social interaction disability

The basic disability is in the capacity to develop reciprocal social interaction, that is, a manner of behaviour which takes heed of, and responds to, the needs and behaviour of others. In mentally retarded subjects, this impairment commonly presents with:

(i) lack of response to other people's emotions
(ii) deficient of inappropriate use of social signals

### Communication disability

The disability is in the area of the capacity to use and understand the significance of language. In mentally retarded cases, this may appear as:

(i) lack of *social* usage of whatever language skills are present
(ii) impaired imagination
(iii) a "mechanical" style of expression, with little flexibility or variation and
(iv) lack of accompanying gesture to add meaning to communication

The detection and assessment of communication disability depends upon appraisal of the functions, particularly receptive and expressive language and neuromuscular coordination, plus general level of mental retardation.

### Restricted repetitive behaviour

An apparent preference for rigidity and routine in a wide range of aspects of daily living. There are many examples:

(i) insistence to perform routines in nonfunctional rituals
(ii) motor stereotypies, especially in more severely retarded subjects
(iii) stereo typed interests, such as preoccupation with particular routes when travelling
(iv) resistance to change in personal environment (including moving of personal effects of furniture
(v) specific attachments to unusual, typically non-soft objects or interest in particular aspects of objects (such as smell or feel)

For any of these to be taken as an indicator of the presence of autism, it must be out of keeping with the general level of retardation. In more severely retarded individuals, only obvious and severe impairments of this type should be considered evidence of childhood autism.

All of these deficits may, in some individuals, occur as a result of general mental retardation. In assessing more severely retarded individuals, social interaction disability may be of more value in reaching the diagnosis of autism. As in the diagnosis of all developmental disorders in mentally retarded individuals, the coding should only be applied where the deficit is not simply due to the level of mental retardation.

In addition to these specific diagnostic features, mentally retarded people with autism frequently show a variety of other non-specific problems such as self-injury (eg by headbanging or wrist biting), sleep disturbance, disturbances of eating behaviour (eg pica), temper tantrums and aggression. Most mentally retarded people who have autism lack spontaneity and initiative, and have difficulty applying themselves to any creative tasks (even when these are well within their general intellectual capacity). The manifestations of autism in any one individual change from childhood through to adulthood and into later adulthood, but a broadly persistent pattern is seen with continuity of deficits and socialisation, at any age, providing there is clear evidence of onset of the disorder within the first three years of life.

## F84.1 Atypical autism

This pervasive developmental disorder differs from autism in terms of *either* age of onset *or* failure to fulfil all three sets of diagnostic criteria. The diagnosis is applicable where one of these conditions applies, in a case which would otherwise qualify for the diagnosis of Childhood Autism:

- features of childhood autism (see above F84.0) begin after the age of three years;
- there are insufficient demonstrable abnormalities in one or two of the three areas of psychopathology required for the diagnosis of autism (namely, reciprocal social interactions, communications, and restrictive, stereotyped, repetitive behaviour) in spite of the characteristic abnormalities being present in the other area (s).

## F84.5 Asperger's syndrome

This disorder is characterised by:

- social interaction disability of autistic type (see F84.0)
- restricted repetitive interests and activities (described in F84.0)

Asperger's syndrome is said by some authorities to differ from autism primarily in that there is no general delay or retardation in language or in cognitive development. However, the diagnosis is applicable to mentally retarded individuals who, while having no general retardation of language, do have highly deviant idiosyncratic or repetitive language.

Most affected individuals are markedly clumsy. The condition occurs predominantly in males (in a ration of about 8 males to 1 female). The abnormalities persist into adolescence and adult life, and this is particularly characteristic of mentally retarded individuals affected by Asperger's syndrome. In some cases, often in individuals of borderline intelligence or with very mild degrees of mental retardation, psychotic episodes occasionally occur in early adult life.

## F84.2 Rett's Syndrome

This is a condition of unknown aetiology, so far reported only in girls, which has been included in this Axis because of the occurrence of an autistic-like picture at an early stage in the course of the disorder.

This condition is characterised by:

- midline handwringing stereotypies
- hyperventilation (often with a periodic episodic character)
- loss of purposive hand movements

The disorder has a characteristic onset, course, and pattern of symptomatology. This comprises:

1. apparently normal or near normal early development
2. at age 7 to 24 months, partial or complete loss of acquired hand skills and of speech, together with deceleration in head growth
3. at age 2 to 3 years, social and play development is arrested, but social interest tends to be maintained, or recovered later
4. in mid-childhood, truncal ataxia and apraxia associated with scoliosis or kypho-scoliosis tend to develop, and sometimes there are choreoathetoid movements
5. severe mental retardation invariably results
6. by age 8 years fits frequently develop

In making the diagnosis of Rett's Syndrome, it should be borne in mind that the most characteristic feature is the loss of purposive hand movements and acquired fine motor manipulative skills, accompanied by the loss or partial loss or lack of development of language, and the occurrence of the distinctive stereotyped "handwashing" movements, with the arms flexed in front of the chest or chin. Failure to gain bowel and bladder control, excessive drooling and protrusion of the tongue are also very common. In about half the cases, spinal atrophies with severe motor disability develop in adolescence or adulthood. Rigid spasticity may become manifest later, and is usually more pronounced in the lower than in the upper limbs.

In contrast to autism, both severe self-injury and complex stereotyped preoccupations or routines are uncommon.

## F84.3 Other childhood disintegrative disorder

Any pervasive developmental disorder (other than Rett's syndrome) that is defined by a period of normal development prior to onset, and by definite loss, over the course of a few months, of previously acquired skills in at least several areas of developmental, together with the onset of characteristic abnormalities of social, communicative and behavioural function. Such disorders often begin with a period of a vague illness in which the child becomes restive, irritable, anxious and overactive. This is generally followed by first impoverishment and then loss of speech and/or language, and accompanied by disintegration of behaviour, both in terms of a loss of behavioural skills and an increase in behavioural disturbance. 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 the period of some months is followed by a plateau and then some degree of limited improvement.

Prognosis for these disorders is very poor, most resulting in severe mental retardation. There is

some controversy and uncertainty about the extent to which such conditions are separate from autism. In some cases the disorder can be shown to be due to some associated encephalopathy, but the diagnosis of a childhood disintegrative disorder should be made on the behavioural features and the longitudinal changes in these across time. As with other developmental disorders, any associated neurological condition (which can generally be presumed to be the cause) should be separately coded, as should the degree of mental retardation at the time of the assessment.

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

This is an ill-defined disorder of uncertain nosological validity. The diagnosis is applicable where there is:

- moderate or severe mental retardation (IQ below 50)
- in childhood, hyperactivity and inattention
- stereotyped behaviours

In adolescence, the overactivity tends to be replaced by underactivity, a pattern that is *not* usual in hyperkinetic children of normal intelligence. It is also common for the syndrome to be associated with the variety of developmental delays either specific or global. These children tend not to benefit from stimulant drugs (in which respect they are unlike those with an IQ in the normal range) and indeed many such mentally retarded children exhibit a severe dysphoric reaction (sometimes with psychomotor retardation) when given stimulants.

It is unclear 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-.

### F84.8 Other pervasive developmental disorders

### F84.9 Pervasive developmental disorder, unspecified

This residual diagnostic category 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 of any of the other F84 codes can not be met.

## F88 Other disorders of psychological development

## F89 Unspecified disorder of psychological development

## F90-F98 Behavioural and emotional disorders with onset usually occurring in childhood and adolescence

Many of the disorders described in this section are common among children with mental retardation. Where the disorders have an onset in childhood, but continue into adult life, these categories may be used to describe the affected adults. The extent and nature of the general developmental delay in a child with mental retardation will influence the expression of specific psychiatric disorder (for example: a child with mental retardation and hyperkinetic disorder may be at greater risk because of a diminished awareness of danger). Psychiatric disorders may last longer in mentally retarded children. They are more difficult to diagnose because of the impairments in communication, associated with mental retardation.

## F90 Hyperkinetic disorders

Clinical description

This group of disorders is characterised by: early onset, a combination of overactive, poorly modulated behaviour with marked inattention and lack of persistent task involvement, and pervasiveness over situations and persistence over time of these behavioural characteristics.

Hyperkinetic disorders always arise early in development (usually in first 5 years of life). Their chief characteristics are lack of persistence in activities that require cognitive involvement, and a tendency to move from one activity to another without completing any one, together with disorganised, ill-regulated, and excessive activity. These problems usually persist through school years and even into adult life, but many affected individuals show a gradual improvement in activity and attention. Hyperkinetic children are often reckless and impulsive, prone to accidents, and find themselves in disciplinary trouble because of unthinking (rather than deliberately defiant) breaches of rules. Their relationships with adults are often socially disinhibited, with a lack of normal caution and reserve, they are unpopular with other children and may become isolated. Cognitive impairment is common, and specific delays in motor and language development are disproportionately frequent. Hyperkinetic disorders are several times more frequent in boys that in girls. Associated reading difficulties (and/or other scholastic problems) are common.

Diagnostic guidelines

The deficits in persistence and attention should be diagnosed *only if they are excessive for the child's age and IQ.* Learning disorders and motor clumsiness occur with undue frequency, and should be noted separately (under F80-F89) when present.

The characteristic behaviour problems should be of early onset (before age 6 years) and long duration. The diagnosis of hyperkinetic disorder can be made in adult life but attention and activity must be judged with reference to developmentally appropriate norms. When hyperkinesia was present in childhood, but has disappeared and been succeeded by another condition, such as dissocial personality disorder or substance abuse, the current condition rather than the earlier one is coded.

The cardinal features are impaired attention and overactivity; both are necessary for diagnosis and should be evident in *more than one situation.* The associated features not sufficient for diagnosis, or even necessary, though they may support it. Social disinhibition, recklessness, and impulsive flouting of social rules may all occur. Hyperkinetic is divided as follows in children of normal intelligence; with mentally retarded patients the differentiation may be difficult.

### F90.0 Disturbance of activity and attention
### F90.1 Hyperkinetic conduct disorder

This coding should be used when the overall criteria for both F90 and for F91 are met.

## F90.8 Other hyperkinetic disorders

## F90.9 Hyperkinetic disorders, unspecified

This category should only be used where criteria for F90 are fulfilled, but differentiation cannot be made between F90.0 and F90.1

## F91 Conduct disorders

Clinical description

Conduct disorders are characterised by a repetitive and persistent pattern of dissocial, aggressive, or defiant conduct. Such behaviour, when at its most extreme for the individual, should amount to major violations of age-appropriate social expectations, and is therefore more severe than ordinary childish mischief or adolescent rebelliousness. Isolated dissocial or criminal acts are not in themselves ground for the diagnosis, which implies an enduring pattern of behaviour. Features of conduct disorder can also be symptomatic of other psychiatric conditions, in which case the underlying diagnosis should be coded. Disorders of conduct may in some cases proceed to dissocial personality disorder (F60.2). Conduct disorder is frequently associated with adverse psychosocial environments, including unsatisfactory family relationships and failure at school, and is more commonly noted in boys.

Diagnostic guidelines

Judgments concerning the presence of conduct disorder should take into account the child's developmental level. Examples of the behaviours on which the diagnosis is based include the following excessive levels of fighting or bullying; cruelty to animals or other people; severe destructiveness to property; fire-setting, stealing, repeated lying, truancy from school and running away from home, unusually frequent and severe temper tantrums, defiant provocative behaviour, and persistent severe disobedience. Any one of these categories, if marked, is sufficient for the diagnosis, but isolated dissocial acts are not.

Exclusion criteria include uncommon but serious underlying conditions such as schizophrenia, mania, pervasive developmental disorder, hyperkinetic disorder, and depression. The diagnosis is not recommended unless the duration of the behaviour described above has been 6 months or longer.

Conduct disorder may be further subdivided as follows though these distinctions may be difficult with mentally retarded patients.

## F91.0 Conduct disorder confined to the family context

## F91.8 Other conduct disorders

## F91.9 Conduct disorder, unspecified

## F92 Mixed disorders of conduct and emotion

## F92.9 Mixed disorder of conduct and emotions, unspecified

## F93 Emotional disorders with onset specific to childhood

Anxiety disorders may be overlooked in mentally retarded children who cannot describe their mood clearly. As with mentally retarded adults, it is important to identify the conditions which may result from changes in routine or environment.

## F93.0 Separation anxiety disorder of childhood

The key diagnostic feature is a focused excessive anxiety concerning separation from those individuals to whom the child is attached (usually parents or other family members), that is not merely part of a generalised anxiety about multiple situations. The condition may present an inappropriate fear of being alone; refusal to go to sleep apart from the attachment figure, or repeated physical symptoms on occasions of separation.

## F93.1 Phobic anxiety disorder of childhood

This category should be used only for developmental phase-specific fears when they meet the additional criteria that apply to all disorders in F93, namely that the onset is during the developmentally appropriate age period, the degree of anxiety is clinically abnormal, and the anxiety does not form part of a more generalised disorder.

## F93.2 Social anxiety disorder of childhood

This category should be used only for disorders that arise before that age of 6 years, that are both unusual in degree and accompanied by problems in social functioning, and that are not part of some more generalised emotional disturbance. Children with this disorder show a persistent or recurrent fear and/or avoidance of strangers; such fear may occur mainly with adults, mainly with peers, or with both. The fear is associated with a normal degree of selective attachment to parents or to other familiar persons. The avoidance or fear of social encounters is of a degree that is outside the normal limits for the child's age and is associated with clinically significant problems in social functioning.

## F93.3 Sibling rivalry disorder

The disorder is characterised by the combination of: evidence of sibling rivalry and/or jealousy, onset during the months following the birth of the younger (usually immediately younger) sibling and emotional disturbance that is abnormal in degree and/or persistence and associated with psychosocial problems. The emotional disturbance may take any of several forms, often including some regression with loss of previously acquired skills (such as bowel or bladder control) and a tendency to babyish behaviour. Frequently, too, the child wants to copy the baby in activities that provide for parental attention, such as feeding. There is usually an increase in confrontational or oppositional behaviour with the parents, temper tantrums, and dysphoria exhibited in the form of anxiety, misery or social withdrawal. Sleep may become disturbed and there is frequently increased pressure for parent attention, such as at bedtime.

## F94 Disorders of social functioning with onset specific to childhood and adolescence

## F94.0 Elective mutism

The condition is characterised by a marked, emotionally determined selectivity in speaking, such that the child demonstrates his or her language competence in some situations but fails to speak in other (definable) situations. Most frequently, the disorder is first manifest in early childhood; it occurs with approximately the same frequency in the two sexes, and it is usual for the mutism to be associated with marked personality features involving social anxiety, withdrawal, sensitivity, or resistance. Typically, the child speaks at home or with close friends and is mute at school or with strangers, but other patterns (including the converse) can occur. The diagnosis presupposes:

(a) a normal, or near-normal, level of language comprehension.
(b) a level of competence in language expression that is sufficient for social communication.
(c) demonstrable evidence that the individual can and does speak normally or almost normally in some situations.

The diagnosis requires that the failure to speak is persistent over time and that there is a consistency and predictability with respect to the situations in which speech does and does not occur. Care should be taken to ensure criterion (c.) is reached. *Some children with mental retardation may have speech abnormalities or be reluctant to speak for a variety of reasons.*

## F94.1 Reactive attachment disorder of childhood

Reactive attachment disorder nearly always arises in relation to grossly inadequate child care. It occurs in younger children who show contradictory or ambivalent social responses, especially at times of parting or re-union. The child averts their gaze when approached, appears miserable, resists comforting, and lacks emotional responsiveness. Unlike children with pervasive developmental disorder, the abnormal patterns of behaviour in reactive attachment disorder resolve when the child is given normal child care.

## F94.2 Disinhibited attachment disorder of childhood

This disorder is seen mainly in children raised in institutions from an early age. The child shows indiscriminately friendly and attention seeking behaviour with adults and associated behavioural disturbance.

## F94.8 Other childhood disorders of social functioning

## F94.9 Childhood disorders of social functioning, unspecified

## F95 Tic disorders

A tic is an involuntary, rapid, recurrent, non-rhythmic motor movement (usually involving circumscribed muscle groups), or vocal production, that is of sudden onset and serves no apparent purpose. Tics tend to be experienced as irresistible but they can usually be suppressed for varying periods of time.

Both motor and vocal tics may be classified as either simple or complex, although the boundaries are not well defined. Common simple motor tics include eye-blinking, neck-jerking, shoulder-shrugging, and facial grimacing. Common simple vocal tics include throat-clearing, barking, sniffing, and hissing. Common complex tics include hitting one's self, jumping and hopping. Common complex vocal tics include the repetition of particular works, and sometimes the use of socially unacceptable (often obscene) words (coprolalia), and the repetition of one's own sounds or words (palilalia). Tic disorders are substantially more frequent in boys than in girls and a family history of tics is common.

The major features distinguishing tics from other motor disorders are the sudden, rapid, transient, and circumscribed nature of the movements, together with the lack of evidence of underlying neurological disorder; their repetitiveness (usually) their disappearance during sleep; and the ease with which they may be voluntarily reproduced or suppressed. *The lack of rhythmicity differentiates tics from the stereotyped repetitive movements seen in some cases of mental retardation, or autism.*

### F95.0 Transient tic disorder

### F95.1 Chronic motor or vocal tic disorder

### F95.2 Combined vocal and multiple motor tic disorder (de la Tourette's syndrome)

A form of tic disorder in which there are, or have been, multiple motor tics and one or more vocal tics, although these need not have occurred concurrently. Onset is almost always in childhood or adolescence. A history of motor tics before development of vocal tics is common; the symptoms frequently worsen during adolescence, and it is common for the disorder to persist into adult life. The vocal tics are often multiple with explosive repetitive vocalisations, throat-clearing, and grunting and there may be use of obscene words or phrases. Sometimes there is associated gestural echopraxia which also may be of an obscene nature (copropraxia). As with motor tics, the vocal tics may be voluntarily suppressed for short periods, be exacerbated by stress, and disappear during sleep.

### F95.8 Other tic disorders

### F95.9 Tic disorder, unspecified

## F98 Other behavioural and emotional disorders with onset usually occurring in childhood and adolescence

### F98.1 Nonorganic enuresis

A disorder characterised by involuntary voiding of urine, by day and/or night, which is abnormal in relation to the individual's mental age and which is not a consequence of a lack of bladder control due to any neurological disorder, or epileptic attacks, or to any structural abnormality of the urinary tract.

The enuresis may have been present from birth (ie. an abnormal extension of the normal infantile incontinence) or it may have arisen following a period acquired bladder control. The later onset (or secondary) variety usually begins about the age of 5 to 7 years. The enuresis may constitute a monosymptomatic condition or it may be associated with a more widespread emotional or behavioural disorder.

## F98.1 Nonorganic encopresis

Repeated voluntary or involuntary passage of faeces, usually of normal or near-normal consistency, in places not appropriate for that purpose in the individual's own sociocultural setting. The condition may represent an abnormal continuation of normal infantile incontinence, it may involve a loss of continence following the acquisition of bowel control, or it may involve the deliberate deposition of faeces in inappropriate places in spite of normal physiological bowel control. The condition may occur as monosymptomatic disorder, or it may form part of a wider disorder, especially an emotional disorder (F93.-) or a conduct disorder (F91.-).

## F98.2 Feeding disorder of infancy and childhood

A feeding disorder of varying manifestations, usually specific to infancy and early childhood. It generally involves refusal of food and extreme faddiness in the presence of an adequate food supply and a reasonably competent care-giver, and the absence of organic disease. There may or may not be associated rumination (repeated regurgitation without nausea or gastrointestinal illness). It is important to differentiate this disorder from organic disease, anorexia nervosa, pica and conditions where the child readily takes food from adults other than the usual care-giver.

## F98.3 Pica of infancy and childhood

Persistent eating of non-nutritive substance (soil, paint chippings, etc.). Pica may occur as one of many symptoms of a more widespread psychiatric disorder (such as autism), or as a relatively isolated psychopathological behaviour; *only* in the latter case should this code be used. The phenomenon is most common in mentally retarded children, if mental retardation is also present, it should be coded (F70-F79). However, pica may also occur in children (usually young children) of normal intelligence.

## F98.4 Stereotyped movement disorder

Voluntary, repetitive, stereotyped, nonfunctional (and often rhythmic) movements that do not form part of any recognised psychiatric or neurological condition.

## F98.5 Stuttering (Stammering)

Speech that is characterised by frequent repetition or prolongation of sounds or syllables or words, or by frequent hesitations or pauses that disrupt the rhythmic flow of speech. Minor dyrhythmias of this type are quite common as a transient phase in early childhood, or as a minor but persistent speech feature in later childhood and adult life. They should be classified as a disorder only if their severity is such as markedly to disturb the fluency of speech.

### F98.6 Cluttering

### F98.8 Other specified behavioural and emotional disorders with onset usually occurring in childhood and adolescence

### F98.9 Unspecified behavioural and emotional disorders with onset usually occurring in childhood and adolescence

### F99 Mental disorder, not otherwise specified

This category is included in the classification for cases in which it is not possible to make a specific diagnosis. It should be necessary to use it very rarely.

# Axis IV
# Global Assessment of Psychosocial Disability
# WHO Short Disability Assessment Schedule (WHO DAS-S)

## I. Instructions

This instrument serves to rate disabilities in relation to the tasks and roles expected from the individual in his/her socio-cultural setting. It has been derived from the WHO Psychiatric Disability Assessment Schedule (WHO DAS) and produced in accordance with the principles embedded in the International Classification of Impairments, Disabilities and Handicaps (ICIDH). According to ICIDH: In the context of health experience, an *impairment* is any loss or abnormality of psychological, physiological or anatomical structure or function; In the context of health experience, a *disability** is any restriction or lack (resulting from an *impairment*) of ability to perform an activity in the manner or within the range considered normal for an individual in his socio-cultural setting; In the context of health experience, a *handicap* is a disadvantage for a given individual, resulting from an *impairment* or a *disability*, that limits or prevents the fulfilment of a role that is normal (depending on age, sex, and social and cultural factors) for that individual.

The rater is expected to rate disability (in relation to the individual's functioning without necessarily making distinctions in the above sense) by identifying the appropriate figure on the given scale of 0 to 5 for each of the specific areas of functioning. The anchor points and their definitions are as follows:

0 = No disability at any time i.e. the patient's functioning conforms to the norms of his/her reference group or socio-cultural context.

1 = Deviation from the norms in the performance of tasks or roles expected to be carried out by the patient in his/her socio-cultural setting.

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* The word disability, as it is used elsewhere in this Axis description, means any diminution of function. However within the WHO, ICIDH the term *disability* (italicized in this paragraph) has a more specific meaning, which is given here. It should be noted however, that the ICIDH is currently under revision, with a new version (and probably new definitions) in 1998).

2 = Deviation from the norms is conspicuous and dysfunctions interfere with social adjustment i.e. slightly disabled most of the time or moderately disabled some of the time.

3 = Deviation form the norms in most of the expected tasks and roles.

4 = Deviation from the norms in all the expected tasks and roles.

5 = Deviation from the norms has reached a crisis point i.e. the patient is severely disabled all the time.

When an individual has performed because s/he was supported by someone else (e.g., family, health worker, etc.), the present level of disability should be rated and a note should be made that this was "functioning with assistance".

In rating, all available information about the individual should be taken into account (e.g., interview, case notes, observation, reports from relatives, etc.). In rating each of the areas, please consider how strongly the individual is affected in performing expected tasks and roles, how many tasks and roles have been affected and how much of the time covered by the rating was characterised by disability.

# II. Rating

## 1. PERIOD COVERED BY RATING (tick appropriate box)

☐ current

☐ last month

☐ last year

☐ other (specify):.....

## 2. SPECIFIC AREAS OF FUNCTIONING (circle appropriate figure)

A. Personal care
Refers to personal hygiene, dressing, feeding, etc.

0 1 2 3 4 5

no disability gross disability

☐ functioning with assistance

B. Occupation
Refers to expected functioning in paid activities, studying, home-making, etc.

0 1 2 3 4 5

no disability gross disability

☐ functioning with assistance

C. Family and household
Refers to expected interaction with spouse, parents, children and other relatives. In rating, pay particular attention to performance in the context in which the individual lives.

0 1 2 3 4 5

no disability gross disability

☐ functioning with assistance

D. Functioning in broader social context
Refers to expected performance in relation to community members, participation in leisure and other social activities.

0 1 2 3 4 5

no disability gross disability

☐ functioning with assistance

## 3. TOTAL DURATION OF DISABILITY (tick appropriate box)

☐ less than one year

☐ 1 year or more

☐ unknown

## 4. SPECIFIC ABILITIES

Some patients may get a high rating of disability in one or more of the above areas, but may nevertheless have certain specific abilities that are an important asset with regard to the management and functioning of the individual in the community or family. Such an asset may be the skilful handling of a musical instrument, particularly good looks, physical strength or ease in social situations. Please describe the asset and given an illustration or example of its value. If this is the case, tick the box and describe briefly the specific abilities of the patient

☐ specific abilities are present

# Axis V
# Associated Abnormal Psychosocial Situations

## Introduction

Chapter XXI of ICD-10 comprises categories Z00-Z99 which are used to code 'factors influencing health status and contact with health services'. These should be used to record abnormal and psychosocial factors which are of relevance to the case but which are not classified as a disease, injury or external cause in categories A00-Y89 of ICD-10. For the purposes of Axis 5, social and psychological factors are grouped as follows:

- Factors of etiological significance
- Factors which affect the course of disorder
- Concurrent problems which affect treatment
- Consequences of the disorder
- Reasons for referral/contact

## Factors of etiological significance

These include factors which may affect early psychological development, including a personal or family history of mental and behavioural disorders or substance abuse. They exclude factors of etiological significance for mental retardation.

**Z55 Problems related to education and literacy.** *Excludes* disorders of psychological development (F80-F89).

Z55.1 Schooling unavailable and unattainable.

Z55.3 Underachievement in school.

Z55.4 Educational maladjustment and discord with teachers and classmates.

Z55.8 Other problems related to education and literacy (eg inadequate teaching).

Z55.9 Problem related to education and literacy, unspecified.

**Z61 Problems related to negative life events in childhood.** *Excludes* maltreatment syndromes (T74.-).

Z61.0 Loss of love relationship in childhood. *Includes* the loss of an emotionally close relationship, such as of a parent, a sibling, a very special friend or a loved pet, by death or permanent departure or rejection.

Z61.1 Removal from home in childhood. *Includes* admission to a foster home, hospital or other institution causing psychosocial stress, or forced conscription into an activity away from home for a prolonged period.

Z61.2 Altered pattern of family relationships in childhood. *Includes* the arrival of a new person into a family resulting in adverse change in child's relationships. It may include new marriage by a parent or birth of a sibling.

Z61.3 Events resulting in loss of self-esteem in childhood. *Includes* events resulting in a negative self-reappraisal by the child such as failure in tasks with high personal investment; disclosure or discovery of a shameful or stigmatizing personal or family event, or other humiliating experiences.

Z61.4 Problems related to alleged sexual abuse of child by person within primary support group. *Includes* problems related to any form of physical contact or exposure between an adult member of the child's household and the child that has led to sexual arousal, whether or not the child has willingly engaged in the sexual acts (eg any genital contact or manipulation or deliberate exposure of breasts or genitals).

Z61.5 Problems related to alleged sexual abuse of child by person outside primary support group. *Includes* problems related to contact or attempted contact with the child's or the other person's breasts or genitals, sexual exposure in close confrontation or attempt to undress or seduce the child, by a substantially older person outside the child's family, either on the basis of this person's position or status or against the will of the child.

Z61.6 Problems related to alleged physical abuse of child. *Includes* problems related to incidents in which the child has been injured in the past by any adult in the household to a medically significant extent (eg. fractures, marked bruising) or that involved abnormal forms of violence (eg. hitting the child with hard or sharp implements, burning or tying up the child).

Z61.7 Personal frightening experience in childhood. Includes an experience carrying a threat for the child's future, such as a kidnapping, natural disaster with a threat to life, injury with a threat to self-image or security, or witnessing a severe trauma to a loved one.

Z61.8 Other negative life events in childhood.

Z61.9 Negative life event in childhood, unspecified.

**Z62 Other problems related to upbringing.** *Excludes* maltreatment syndromes (T74.-).

Z62.0 Inadequate parental supervision and control. *Includes* lack of parental knowledge of what the child is doing or where the child is, poor control, lack of concern, or lack of attempted intervention when the child is in risky situations.

Z62.1 Parental overprotection. *Includes* pattern of upbringing resulting in infantilization and prevention of independent behaviour.

Z62.2 Institutional upbringing. *Includes* group foster care in which parenting responsibilities are largely taken over by some form of institution (such as a residential nursery, orphanage, or children's home), or therapeutic care over a prolonged period in which the child is in a hospital, convalescent home or the like, without at least one parent living with the child.

Z62.3 Hostility towards and scapegoating of child. *Includes* negative parental behaviour specifically focused on the child as an individual, persistent over time and pervasive over several child behaviours (eg. automatically blaming the child for any problems in the household or attributing negative characteristics to the child).

Z62.4 Emotional neglect of child. *Includes* the parent talking to the child in a dismissive or insensitive way, lack of interest in the child, of sympathy for the child's difficulties and of praise and encouragement, an irritated reaction to anxious behaviour and the absence of sufficient physical comforting and emotional warmth.

Z62.5 Other problems related to neglect in upbringing. *Includes* lack of learning and play experience.

Z62.6 Inappropriate parental pressure and other abnormal qualities of upbringing. *Includes* parents forcing the child to be different from the local norm, either sex-inappropriate (eg. dressing a boy in girl's clothes), age-inappropriate (eg. forcing a child to take on responsibilities above her or his own age), or otherwise inappropriate (eg. pressing the child to engage in unwanted or too difficult activities).

Z62.8 Other specified problems related to upbringing.

Z62.9 Problem related to upbringing, unspecified.

## Z81 Family history of mental and behavioral disorders

Z81.0 Family history of mental retardation (conditions classifiable to F70-F79).

Z81.1 Family history of alcohol abuse (conditions classifiable to F10.-).

Z81.2 Family history of tobacco abuse (conditions classifiable to F17.-).

Z81.3 Family history of other psychoactive substance abuse (conditions classifiable to F11-F16, F18-F19).

Z81.4 Family history of other substance abuse (conditions classifiable to F55).

Z81.8 Family history of other mental and behavioural disorders (conditions classifiable to F00-F99).

### Z82 Family history of certain disabilities and chronic diseases leading to disablement

Z82.0 Family history of epilepsy and other diseases of the nervous system (conditions classifiable to G00-G99).

Z82.7 Family history of congenital malformations, deformations and chromosomal abnormalities (conditions classifiable to Q00-Q99).

Z82.8 Family history of other disabilities and chronic diseases leading to disablement, not elsewhere classified.

### Z83 Family history of other specific disorders.

*Excludes* contact with or exposure to communicable disease in the family (Z20.-)

Z83.0 Family history of human immunodeficiency virus (HIV) disease (condition classifiable to B20-B24).

### Z86 Personal history of certain other diseases.

*Excludes* follow-up medical care and convalescence (Z42-Z51, Z54.-).

Z86.4 Personal history of psychoactive substance abuse (conditions classifiable to F10-F19). *Excludes* current dependence (F10-F19 with common fourth character.2), and problems related to use of alcohol (Z72.1), drugs (Z72.2), and tobacco (Z72.0).

Z86.5 Personal history of other mental and behavioral disorders (conditions classifiable to F00-F09, F20-F99).

Z86.6 Personal history of diseases of the nervous system and sense organs (conditions classifiable to G00-G99, H00-H95).

### Z91 Personal history of risk-factors, not elsewhere classified.

*Excludes* exposure to pollution and other problems related to physical environment (Z58.-), occupational exposure to risk-factors (Z57.-), personal history of psychoactive substance abuse (Z86.4).

Z91.8 Personal history of other specified risk-factors, not elsewhere classified, abuse NOS, or maltreatment NOS.

## Factors which affect the course of the disorder

These are significant life events which may be associated with the onset of an acute episode of a disorder, or the prolongation of a chronic disorder.

## Z56 Problems related to employment and unemployment

Z56.0 Unemployment, unspecified

Z56.1 Change of job

Z56.2 Threat of job loss

Z56.3 Stressful work schedule

Z56.4 Discord with boss and workmates

Z56.5 Uncongenial work, or difficult conditions at work.

Z56.6 Other physical and mental strain related to work

Z56.7 Other and unspecified problems related to employment

## Z59 Problems related to housing and economic circumstances.

*Excludes* inadequate drinking-water supply (Z58.6).

Z59.0 Homelessness

Z59.1 Inadequate housing (eg lack of heating, restriction of space, technical defects in home preventing adequate care, unsatisfactory surroundings). *Excludes* problems related to physical environment (Z58.-).

Z59.2 Discord with neighbours, lodgers and landlord

Z59.3 Problems related to living in residential institution (including as resident in a boarding-school). *Excludes* institutional upbringing.

Z59.4 Lack of adequate food. *Excludes* effects of hunger (T73.0), inappropriate diet or eating habits (Z72.4), and malnutrition (E40-E46).

Z59.5 Extreme poverty.

Z59.6 Low income.

Z59.7 Insufficient social insurance and welfare support.

Z59.8 Other problems related to housing and economic circumstances (eg foreclosure of loan, isolated dwelling, or problems with creditors).

Z59.9 Problem related to housing and economic circumstances, unspecified.

## Z60 Problems related to social environment

Z60.0 Problems of adjustment to life-cycle transitions.
Z60.1 Atypical parenting situation.

Z60.2 Living alone.

Z60.3 Acculturation difficulty, because of migration or social transplantation.

Z60.4 Social exclusion and rejection, including exclusion and rejection on the basis of person characteristics, such as unusual physical appearance, illness or behaviour. *Excludes* target of adverse discrimination such as for racial or religious reasons (Z60.5)

Z60.5 Target of perceived adverse discrimination and persecution. *Includes* persecution or discrimination, perceived or real, on the basis of membership of some group (as defined by skin colour, religion, ethnic origin, etc.) rather than personal characteristics. *Excludes* target of adverse discrimination such as for racial or religious reasons (Z60.5).

Z60.8 Other problems related to social environment

Z60.9 Problems related to social environment, unspecified

## Z63 Other problems related to primary support group, including family circumstances.

*Excludes* maltreatment syndromes (T74.-), and problems related to negative life events in childhood (Z61.-), or upbringing (Z62.-).

Z63.0 Problems in relationship with spouse or partner. *Includes* discord between partners resulting in severe or prolonged loss of control, in generalization of hostile or critical feelings or in a persisting atmosphere of severe interpersonal violence (hitting or striking).

Z63.1 Problems in relationship with parents and in-laws.

Z63.2 Inadequate family support

Z63.3 Absence of family member

Z63.4 Disappearance and death of family member, or assumed death of family member.

Z63.5 Disruption of family by separation and divorce, or estrangement.

Z63.6 Dependent relative needing care at home

Z63.7 Other stressful life events affecting family and household (eg normal anxiety about a sick person in the family, health problems within family, and ill or disturbed family member, or an isolated family).

Z63.8 Other specified problems related to primary support group (eg family discord NOS, high expressed emotional level within family, or inadequate or distorted communication within

family).

Z63.9 Problem related to primary support group, unspecified.

### Z64 Problems related to certain psychosocial circumstances

Z64.0 Problems related to unwanted pregnancy. *Excludes* supervision of high-risk pregnancy due to social problems. (Z53.7)

Z64.1 Problems related to multiparity. *Excludes* supervision of pregnancy with grand multiparity (Z35.4).

### Z65 Problems related to other psychosocial circumstances.

*Excludes* current injury.

Z65.0 Conviction in civil and criminal proceedings without imprisonment

Z65.1 Imprisonment another incarceration

Z65.2 Problems related to release from prison

Z65.3 Problems related to other legal circumstances (eg arrest, child custody or support proceedings, litigation, or prosecution).

Z65.4 Victim of crime and terrorism or of torture.

Z65.5 Exposure to disaster, war and other hostilities. *Excludes* target of perceived discrimination or persecution (Z60.5)

Z65.8 Other specified problems related to psychosocial circumstances

Z65.9 Problem related to unspecified psychosocial circumstances

## Concurrent problems which affect treatment

These are concurrent problems with a person's way of life which may require counselling and may otherwise affect the treatment programme.

### Z72 Problems related to lifestyle.

*Excludes* problems related to life-management difficulty (Z73.-), and socioeconomic and psychosocial circumstances (Z55-Z65).

Z72.0 Tobacco use. *Excludes* tobacco dependence (Z17.2)

Z72.1 Alcohol use. *Excludes* alcohol dependence (Z10.2)

Z72.2 Drug use. *Excludes* abuse of non-dependence-producing substances (F55) drug dependence (F11-F16, F19 with common fourth character .2)

Z72.3 Lack of physical exercise

Z72.4 Inappropriate diet and eating habits. Excludes behavioural eating disorders of infancy or childhood (F98.2-F98.3), eating disorders (F50.-), lack of adequate food (Z59.4), malnutrition and other nutritional deficiencies (E40-E64).

Z72.5 High-risk sexual behaviour

Z72.6 Gambling and betting. *Excludes* compulsive or pathological gambling (F63.0).

Z72.8 Other problems related to lifestyle, or self-damaging behaviour.

Z72.9 Problems related to lifestyle, unspecified.

**Z73 Problems related to life-management difficulty.** *Excludes* problems related to socioeconomic and psychosocial circumstances (Z55-Z65).

Z73.0 Burn-out, or state of vital exhaustion.

Z73.1 Accentuation of personality traits. *Includes* Type A behaviour pattern (characterized by unbridled ambition, a need for high achievement, impatience, competitiveness, and a sense of urgency)

Z73.2 Lack of relaxation and leisure

Z73.3 Stress, not elsewhere classified, or physical and mental strain NOS

Z73.4 Inadequate social skills, not elsewhere classified

Z73.5 Social role conflict, not elsewhere classified

Z73.6 Limitation of activities due to disability. *Excludes* care-provider dependency (Z74.-).

Z73.8 Other problems related to life-management difficulty

Z73.9 Problem related to life-management difficulty, unspecified

**Z76 Persons encountering health services in other circumstances**

Z76.5 Malingerer (conscious simulation), i.e. person feigning illness (with obvious motivation). *Excludes* factitious disorder (F68.1), and peregrinating patient (F68.1).

**Z91 Personal history of risk-factors, not elsewhere classified.** *Excludes* exposure to pollution and other problems related to physical environment (Z58.-), occupational exposure of risk-factors (Z57.-), and personal history of psychoactive substance

abuse (Z86.4).

Z91.1 Personal history of noncompliance with medical treatment and regimen.

Z91.2 Personal history of poor personal hygiene

Z91.3 Personal history of unhealthy sleep-wake schedule. *Excludes* sleep disorders (G47.-).

Z91.4 Personal history of psychological trauma, not elsewhere classified

Z91.5 Personal history of self-harm, parasuicide, self-poisoning, or suicide attempt.

## Consequences of the disorder

These are impairments, disabilities and handicaps which are consequences of mental and behavioural disorders and associated physical disorders. Although these are included in Axis 5, it is preferable to use the World Health Organisation *Classification of Impairments, Disabilities, and Handicaps* for this purpose.

### Z74 Problems related to care-provider dependency.

*Excludes* dependence on enabling machines or devices NEC (Z99.-).

Z74.0 Reduced mobility. *Includes* bedfast, chairfast.

Z74.1 Need for assistance with personal care

Z74.2 Need for assistance at home and no other household member able to render care.

Z74.3 Need for continuous supervision.

Z74.8 Other problems related to care-provider dependency

Z74.9 Problem related to care-provider dependency, unspecified

### Z99 Dependence on enabling machines and devices, not elsewhere classified

Z99.3 Dependence on wheelchair

# Appendix

## Standard assessment and measurement instruments for disorders of psychological development

Eight standardised assessment instruments are recommended for use in screening, diagnosing or assessing the severity of disorders of psychological development. The instruments listed have been chosen on the basis of their widespread (in many cases international) use, their general familiarity, availability of research concerning their reliability and validity. Several do not require particular specialist training or expertise in their administration.

Of the eight schedules, four (1 a-d below) provide an assessment over a range of developmental domains. These assessment measures are particularly applicable where more than one developmental disorder is suspected or being screened for. The other four assessment tools 2, a-d) focus, either entirely or principally, on disorders of one domain or aspect of development.

### Part 1

### (a) Vineland Social Maturity Scale (Sparrow et al 1984)

This well-established scale assesses a broad range of developmental domains including communication, motor skills and socialisation, in addition to general functioning in areas such as dressing, feeding, and ability to use public transport. The Vineland has proven to be useful in the assessment of children who, for whatever reason, do not cooperate in testing. The scale may be completed by interview with a reliable informant. An overall social age can be derived from the scale, which may be usefully compared to mental and chronological age.

### (b) The British Ability Scales (Elliot et al 1983) and Differential Ability Scales (Elliot 1990)

This recently introduced test measures a range of functions and educational attainments which include Reasoning, Speed, Spatial Imagery, Short Term Memory, Perceptual Matching and Retrieval and Application of Knowledge, and can be used to calculate an overall IQ.

### (c) The Adaptive Behaviour Scales (Nihira et al 1974)

Probably the most widely used tool for the assessment of social functioning in mentally retarded adults, and of proven use internationally. In addition to measurement of adaptive behaviour (Part 1 of ABS: independent functioning; physical development; economic activity; language development; numbers and time; domestic activity; vocational activity; self-direction; responsibility and socialisation), also contains section on the assessment of behaviour disturbance (Part 2).

### (d) The Portage Guide to Early Education (Bluma et al 1975)

This approach to the assessment and education of young children has now been adopted for widespread use internationally. It's strengths include: broad-based developmental assessment, including infant stimulation, socialisation, language, self-help, cognitive and motor; only brief period of training

required; active involvement of parent or carer.

## Part 2

### (a) Autism Behaviour Check List (Krug et al 1979)

Originally developed as a screening tool for use in education, this parent/carer postal questionnaire is of established value as a reliable indicator of problems in the area of autism and social developmental delay, and has been used in a number of countries.

### (b) Disability Assessment Scale (Holmes et al 1982)

This scale focuses on autistic and related social developmental and language disorders, and many of the behavioural problems which occur in affected individuals, including overt and antisocial behaviours.

### (c) British (Peabody) Picture Vocabulary Test (Dunn et al 1982)

This is a test of language comprehension, and is suitable for non-speaking children. The test booklet is largely pictorial, and has been found to be of use cross-culturally, occasionally by some modification of format. The age range covered is 3 to 19 years. The test can be employed without professional training, but is most often used by psychologists and speech therapists.

### (d) Reynell Scales of Language Development (Reynell 1977)

Two forms: comprehension and expressive language, over age range 1 month to 6 years. Of particular use in non-verbal individuals.

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ICD-9 code: 317 ICD-10 â€“ Mild mental retardation F70. An individual is diagnosed with mild mental retardation if they have an IQ score of 50-69,6 and the majority of cases fall within this category. Individuals with mild mental retardation typically develop social and communication skills adequate for self support, but may need assistance during times of unusual stress. Academic skills can be acquired up to the 6th grade level. Given appropriate supports, individuals with mild mental retardation can usually live successfully in the community, either in independent or supervised settings, and 8 Aliases & Classifications for Mental Retardation, X-Linked, with Cerebellar Hypoplasia and MalaCards integrated aliases for Mental Retardation, X-Linked, with Cerebellar Hypoplasia and Distinctive Facial Appearance: Name: Mental Retardation, X-Linked, with Cerebellar Hypoplasia and Distinctive Facial Appearance 57 13. Mental Retardation X-Linked with Cerebellar Hypoplasia and Distinctive Facial Appearance 75 29 6. X-Linked Mental Retardation with Cerebellar Hypoplasia and Distinctive Facial Appearance 12.Â  Diseases related to Mental Retardation, X-Linked, with Cerebellar Hypoplasia and Distinctive Facial Appearance via text searches within MalaCards or GeneCards Suite gene sharing: # Related Disease.