

Mental Retardation

Mental retardation is a congenital or early acquired (under 2 years of age) intellectual disability.

Classification

Classification according to ICD-10: Mental Retardation (<https://mkn10.uzis.cz/prohlizec/Ment%C3%A1ln%C3%AD%C2%A0retardace>) (F70-F79)

- **Mild mental retardation** – IQ 50–69
- **Moderately severe mental retardation** – IQ 35–49
- **Severe mental retardation** – 20–34
- **Profound mental retardation** – IQ less than 20

This *congenital or early acquired* (up to 2 years of age) intellectual disorder was previously referred to as oligophrenia or weak-mindedness. It is characterized by a serious impairment of mental abilities, which leads to a significant limitation of the adaptive functioning of the child or adult in their social environment. The basic diagnostic criterion is a low score on a comprehensive intelligence test (i.e. performance below 2 standard deviations from the mean). However, only the result of the test is not enough for the diagnosis, the individual in question must also fail to fulfill the demands expected of an individual of his age in his social environment (i.e. family). Impaired cognitive abilities tend to be unevenly distributed amongst people afflicted by this ailment. Delayed speech development and verbal intelligence are often observed along with behavioral disorders.

Mild mental retardation – IQ 50–69

If it is not a combined impairment, this diagnosis is usually made only in preschool age (sometimes also after starting school, which the child cannot handle).

- Achieving basic developmental milestones, especially of a motor nature, in the first year of life is normal or only slightly delayed.
- Speech development does not have to be delayed.
- **Retardation only becomes apparent when dealing with more complex tasks and situations.**
- **Children *do well in practical* (formerly special) schools. Only in exceptional cases, with a lot of help from the family, can they even master the elementary school curriculum.**
- Due to their increased trustworthiness and suggestibility, they are more susceptible to abuse from other people.
- The capacity for abstraction and logical reasoning is mainly affected.
- Thinking is stereotyped and not very flexible.
- Mechanical memory and visual motor skills may be at a good level.
- They may also suffer from specific developmental learning disabilities. These are even more common in them than in the general population, probably due to the common etiological basis of the CNS.

'Most of these people learn after primary school and are able to lead an independent life with occasional help from those around them in more complex new situations.'

Moderately severe mental retardation – IQ 35–49

The '*delay* in the development of these children is usually noticed already in the *infancy or toddler* age.

- Movement development is usually delayed.
- Speech development tends to be significantly retarded. During childhood, these people acquire at least a minimal vocabulary, which is enough for normal understanding.
- Some children master the curriculum of a practical school, but most of them attend a special school (in exceptional cases, children can be exempted from compulsory school attendance and the family takes responsibility for their education). In special schools, children are educated according to the program for practical schools, while they have more time to prepare. Emphasis is placed on mastering practical skills.
- Most children can handle self-care well and master simple work activities.
- As adults, they can work in "protected workshops" or in various programs of supported employment.

The environment cannot influence mental retardation as such, but it very significantly influences the emotional and social development of retarded children.

Severe mental retardation – IQ 20–34



Ceramics from a sheltered workshop

Severe retardation is evident from an early age.

- These children are mostly disabled in combination, i.e. they also have a physical deficit.
- They never learn to speak or learn only a few basic words.
- They are usually included in the rehabilitation classes of auxiliary practical schools. In these classes, they focus on mastering communication (in a normal way or with the use of special communication systems), the basics of self-service skills, and understanding common life situations.

With good management, they can be included in partial work activities as adults. Here, too, the great importance of caring for the family applies.

Profound mental retardation – IQ less than 20

Along with intellectual disabilities, these individuals also often have severe motor impairments. They are often capable of minimal movement or are completely immobile.

Most of the time, they do not acquire the basics of spoken language. Understanding tends to be limited to basic instructions. They react sensitively mainly to tactile stimuli and also to sounds, especially to the emotional timbre of the voice. Reaction to visual stimuli tends to be limited. They communicate with the environment through global reactions. In case of joy, they relax and smile, increasing tension is indicated by rising muscle tension and crying. Even these children tend to be placed in rehabilitation classes in special schools, where similar rules apply to them as for children with severe mental retardation. The goal of teaching here is not to master the curriculum but to make maximum use of each child's strengths and thus achieve the greatest possible self-sufficiency in everyday life.

Genetic causes

Common causes of mental retardation are **Down syndrome** and **Fragile X syndrome**^[1].

Numerical chromosomal aberrations

 For more information see *Down Syndrome*, *Fragile X Syndrome*, *Klinefelter Syndrome*.

The most common cause of congenital mental retardation is **Down syndrome** (*trisomy 21*). **The average IQ at 5 years is around 50, but intelligence shows significant inter-individual differences**^[1]. **In addition to this, the typical facies is manifested: epicanthus, low-set ears, macroglossia, hypertelorism, wide root of the nose, strong everted lips, white spots in the iris (Brushfield spots), short and wide neck. In addition, hypotonia, deformities of the hands and feet' (brachymesophalangial, clinodactyly of the 5th finger, four-finger groove, sandal groove), skeletal anomalies' (short stature, hyperextension of joints, coxa valga, splayed iliac blades), developmental defects of organs (stenosis of the duodenum, pancreas annulare, atresia of the anus, megacolon, prolapse rectum, hypogonadism).** The increased incidence of "congenital heart defects" (atrial septal defect, ventricular septal defect, atrioventricular canal, tetralogy of Fallot, patent ductus Botall) and "leukemia" (up to 10-30x higher than the population) is very serious. Complications from congenital heart defects and an increased incidence of leukemia reduce the average survival time of patients with Down syndrome to just over 40 years.



Child with Down Syndrome

- Klinefelter syndrome (karyotype 47 XXY)^[1]

Structural chromosomal aberrations

Fragile X syndrome (amplification of the CGG sequence in the FMR₁ gene) is the cause of approximately 7% of severe and 4% of mild mental retardation^[1]. The average IQ is around 50. Other clinical signs include large earlobes, an elongated face, hyperactivity and, in boys, macroorchidism.

- Syndrome Cri du chat syndrome (5p deletion)
- Wolf-Hirschhorn syndrome (4p deletion)

Microdeletion syndromes

In microdeletion syndromes, mental retardation is a common symptom. It occurs, for example, in the following syndromes^[1]:

- Angelman syndrome (maternal 15q11.2 microdeletion),
- Prader-Willi syndrome (microdeletion of paternal 15q11.2),
- Miller-Dieker syndrome (microdeletion 16p13.3),
- Smith-Magenius syndrome (microdeletion 17p11.2),
- DiGeorge syndrome,

- Sphrintzen syndrome.

Congenital metabolic disorders

- branched-chain amino acid metabolism disorders (e.g. Leucinosi)

Secondary causes

Secondary causes include endocrinopathy, congenital infections and exogenous embryofetopathy.

Endocrinopathy

- Congenital hypothyroidism

Congenital infections

- Congenital cytomegalovirus infection^[2]
- Syphilis congenita recens^[2]
- Congenital toxoplasmosis^[2]
- African trypanosomiasis^[2]

Exogenous Embryofetopathy

- Alcohol embryopathy^[1]
- Hydantoin embryopathy^[1]

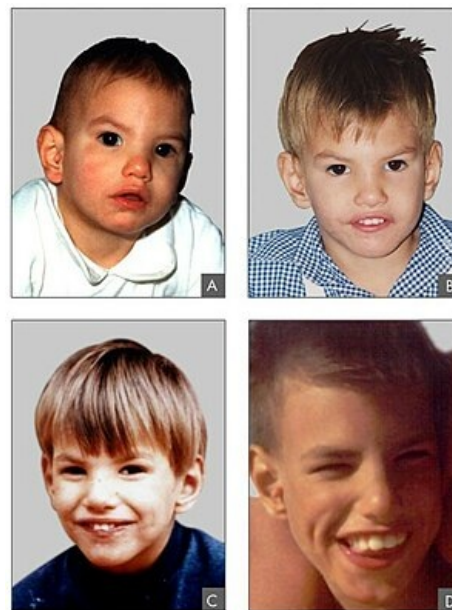
Links

Related Pages

- Intelligence and its disorders
- Down Syndrome
- Hypotonia (pediatrics)

Reference

1. MUNTAU, Ania Carolina. *Pediatrics*. 4. edition. Prague : Grada, 2009. 581 pp. pp. 37-49. ISBN 978-80-247-2525-3.
2. SEDLÁČEK, Dalibor – ŠUBRT, Ivan – DORT, Jiří. Kongenitální infekce – současný stav. *Pediatric pro praxi* [online]. 2007, y. 8, vol. 2, p. 72–76, Available from <<http://www.pediatricpropraxi.cz/artkey/ped-200702-0002.php>>. ISSN 1803-5264.



Cri du chat Syndrome