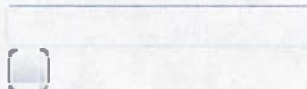


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Mental Retardation

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Background

Mental retardation (MR) or intellectual disability (ID) is a descriptive term for subaverage intelligence and impaired adaptive functioning arising in the developmental period (< 18 y). MR/ID and other neurodevelopmental disabilities are seen often in a general pediatric practice.

Terminology for MR/ID has been particularly challenging as the term *mentally retarded* carries significant social and emotional stigma. The American Association for Intellectual and Developmental Disability (AAIDD) has been particularly influential in terminology changes such that most professionals working in the field now refer to mental retardation as intellectual disability. The *DSM-V* is expected to adopt this new terminology.^[1]

Developmental delay is often used inappropriately as synonymous with MR/ID. Developmental delay is an overly inclusive term and should generally be used for infants and young children (< 5 y) in which the diagnosis is unclear, such as those too young for formal testing.^[1]

Approximately 10% of children have some learning impairment, while as many as 3% manifest some degree of MR/ID. The population prevalence of these combined disorders of learning rivals that of the common childhood disorder asthma.

MR/ID originates during the developmental period (ie, conception through age 18 years) and results in significantly subaverage general intellectual function with concurrent deficits in functional life skills. The diagnosis of MR/ID requires an intelligence deficit of at least 2 standard deviations (SDs) below the mean IQ. This generally translates into an intelligence quotient (IQ) score of 70-75, given a population mean of 100. Equivalent deficits in at least 2 areas of functional life skills or adaptive skills also must be present to meet the diagnostic criteria for MR/ID. Adaptive skills encompass functional life skills within the domains of communication, self-care, home living, social and interpersonal skills, use of community resources, self-direction, functional academic skills, work, leisure, health, and safety.

MR/ID is currently categorized broadly as follows.^[2]

Table 1. Intellectual disability categorization (Open Table in a new window)

Category	IQ score* (SD below mean)	Proportion of MR/ID	Educational level/adaptive skills	Intensity of supports required	Prevalence in total population
Mild		85%			0.9-2.7%

	50-55 to 70		Up to about 6th grade; vocational	Intermittent, especially under stress	
	(2-3)				
Moderate	35-40 to 50- 55	10%	up to about 2nd grade; unskilled or semi-skilled, supervised	Limited; usually supervised	0.3-0.4%
	(3-4)				
Severe	20-25 to 35- 40	4%	May learn words; elementary self-care skills	Extensive; closely supervised group or family home	
	(4-5)				
Profound	< 20-25 (>5)	1%	Little to no self-care skills	Constant aid and supervision	
*IQ scores are considered +/-5 points due to measurement error.					

The *DSM-V* is widely expected to change the definition of MR/ID, relying less on specific IQ levels.^[3] IQ scores can vary based on age, instrument, and practitioner. Furthermore, testing prior to school age does not correlate well with future performance. IQ scores can be measured using assessment instruments, such as the Stanford-Binet, Wechsler Adult Intelligence Scale (WAIS), or Wechsler Intelligence Scale for Children (WISC-IV). Nonverbal children can be tested with the Leiter International Performance Scale (Leiter-R).

The second component of diagnosis, adaptive skills, is usually measured with a self-reported or parent/caregiver-reported inventory, such as the Vineland Adaptive Behavior Scales, Second Edition (VABS-II). The *DSM-V* diagnosis is expected to require adaptive measurements of less than 2 SDs as compared to the population mean, with standard scores of 70 or less, in at least 2 of the following domains:^[3]

- Conceptual skills (communication, language, time, money, academic)
- Social skills (interpersonal skills, social responsibility, recreation, friendships)
- Practical skills (daily living skills, work, travel)

MR/ID also can be categorized as syndromic, if associated with dysmorphic features, or nonsyndromic, if not associated with dysmorphism or malformations. The understanding of specific MR/ID syndromes is expanding with recent molecular genetic advances. More than 800 recognized syndromes listed in the Online Mendelian Inheritance in Man (OMIM) database are associated with MR/ID, reflecting clinical diagnostic advances in the field. The most common associated chromosomal abnormality is trisomy 21, or Down syndrome. The most common X-linked abnormality associated with MR/ID is fragile X syndrome. However, for most cases of MR/ID, no specific genetic abnormalities are found.

Some forms of MR/ID are due to nongenetic factors and may be identifiable by their associated dysmorphism and clinical presentation. Examples include prenatal exposure to teratogens (eg, anticonvulsants, warfarin, alcohol) or prenatal thyroid dysfunction. Prenatal and postnatal exposure to lead and the associated decrement in IQ may increase an individual's chance of functioning in the MR/ID range.

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