

Mental Retardation

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MENTAL RETARDATION is **global developmental delay** (due to some pathogenic process) that results in **significant limitation of intellect or cognition** → poor adaptation to demands of everyday life.

- affects **1-3% population** in developed countries.
 - improvements in obstetric and neonatal care, rubella vaccination, Rh₀(D) immune globulin use, etc have reduced *INCIDENCE* of MR; but increase in survival of very-low-birth-weight infants has kept *PREVALENCE* constant.

DSM-IV-TR defines mental retardation as follows:

1. **IQ ≤ 70** (i.e. 2 standard deviations below mean IQ of general population)
2. Concurrent **deficits / impairments in adaptive functioning** in at least 2 of following areas: communication, self-care, home living, social/interpersonal skills, use of community resources, self-direction, functional academic skills, work, leisure, health, safety.
3. Onset **before age 18 years**.

Inability to function independently (not low IQ) is most important feature of MR!

Four degrees of retardation (DSM-IV):

1. **Mild** (80-85%): IQ ≈ 50-70 (major deficits are in judgment) – especially common in lower socioeconomic class (vs. other degrees are distributed evenly).
 2. **Moderate** (10-15%): IQ ≈ 35-50
 3. **Severe** (3-7%): IQ 20-35
 4. **Profound** (1-2%): IQ < 20
- although mental retardation is classified as axis II disorder, it is not considered mental illness as such; it is system of identifying *people who need social support and special educational services* to carry out tasks of everyday living.
 - 40-70% patients have diagnosable **psychiatric disorders** (but examination is especially difficult in nonverbal persons).
 - *mood disorders* (esp. depressive) are quite common if patients are socially rejected by other children.
 - individuals are **vulnerable to abuse** - because of difficulties in reporting it and tendency to want to please others.
 - **self-injurious behavior** is common in certain mental retardation syndromes (esp. Lesch-Nyhan syndrome).
 - anorexia and bulimia nervosa are relatively rare, but mental retardation is predisposing for **pica** and **ruminantion**.
 - greater degree of mental retardation suggests greater amount of brain damage - incidence of concomitant **sensory** and **motor deficits** increases (motor or sensory impairments can mimic cognitive impairment but are not in themselves causes of it).
 - primary manifestations of MR:
 - 1) **delayed intellectual development** (in mild MR, may not be recognizable until preschool age); it is much more common to experience slow progress than developmental arrest
 - 2) **immature behavior**
 - 3) **limited self-care skills**.

ETIOLOGY

- various disorders leading to permanent brain damage.

No cause can be identified in 30% cases of *severe mental retardation* and in 50% cases of *mild mental retardation*.

- **physical causes** tend to cause moderate-to-profound retardation.
- **disadvantaged environment** is more likely in mild retardation.

Prenatal Causes

1. **Chromosomal aberrations** (e.g. **Down syndrome**)
2. **Disorders with autosomal-dominant inheritance** (e.g. tuberous sclerosis)
3. **Disorders with autosomal-recessive inheritance** (e.g. most metabolic disorders: phenylketonuria, homocystinuria, lysosomal storage diseases, mucopolysaccharidoses).
4. **X-linked mental retardation** (e.g. **fragile X syndrome** - most common inherited form of mental retardation and, after Down syndrome, its most common genetic form!).
5. **Brain malformations**
6. **Maternal infections** - viral infections (e.g. rubella, CMV, toxoplasmosis, HIV), syphilis.
7. **Toxic substances** (esp. **ethanol** - most important of teratogenic substances!)
8. **Toxemia of pregnancy**
9. **Placental insufficiency, poor nutrition** (e.g. iodine deficiency → cretinism)

Perinatal Causes

1. **Infections** (esp. **herpes simplex** type 2 encephalitis)
2. **Perinatal asphyxia**
3. **Prematurity, extremely low birth weight** → intracranial hemorrhage and hypoglycemia.
4. **Kernicterus**.

Postnatal Causes

1. **Infections** (meningitis, encephalitis).
2. **Toxic substances** (esp. lead poisoning)
3. **Brain tumors** and associated treatments.
4. **Near-drowning**.
5. **Psychosocial problems** (development is severely affected in depriving [nonstimulating] environment, even if adequate physical care is provided): poverty, parental mental illness.
6. **Psychotic illness in child**.

Malnutrition and **environmental deprivation** are most common causes of MR worldwide!

DIAGNOSIS

- can be made only with use of **standardized psychometric testing** administered by trained professional: N.B. impairments in sensorimotor, visual, auditory, language, and social skills may make valid assessment difficult (because there are few standardized tests for children with cerebral palsy, deafness, blindness, or autism).

Assessment of INTELLIGENCE (i.e. IQ):

- A. **Revised Wechsler scales** (Wechsler Preschool and Primary Scale of Intelligence [WPPSI], Wechsler Intelligence Scale for Children [WISC], and Wechsler Adult Intelligence Scale [WAIS]).

B. Revised Stanford-Binet intelligence test.

Assessment of ADAPTATION (i.e. ability to function in real life) - **Revised Vineland Adaptive Behavior Scales** (data are derived from interview with parents or caretaker and do not require participation of person being assessed).

- diagnosis rarely is made in young children because of lack of stability of test scores for children younger < 5 years (**children < 5 years** with cognitive difficulties are considered to be **DEVELOPMENTALLY DELAYED**).
DEVELOPMENTAL DELAY is less specific term than mental retardation, because it implies only that child's **performance at present time is delayed** compared to peers of same age. This label does not predict future performance!

TREATMENT

Mental retardation is irreversible!

- referral to early intervention program (in center with multidisciplinary team that evaluates and serves children with MR) during infancy may decrease severity of disability.
- provide child with **stimulating environment**.
- training of adequate **social skills** and **self-help skills** are often more important than **academic skills**.
- recent trend is to move mentally disabled individuals out of institutional settings and into more community-based living arrangements (such as group homes) - **DEINSTITUTIONALIZATION** - this trend underscores idea that mental retardation cannot be categorized as mental illness in strictest sense of term - these individuals should be viewed as merely population requiring more-than-average help in daily chores of living.

Every effort should be made to have child live at home or in community-based residence!

- BEHAVIORAL DISORDERS** are reason for most psychiatric referrals and out-of-home placements for people with MR (behavior problems are often situational, and precipitating factors can usually be found!!!).
 - in many cases, **BEHAVIORAL PROBLEMS** respond to psychosocial (behavioral) interventions (e.g. managing environment, behavior-decelerating / accelerating techniques).
- many people with mild ÷ moderate MR can support themselves, can live independently, and can be successful at jobs that require basic intellectual skills.

BEHAVIORAL PROBLEMS

- use of psychotropic medications to treat **BEHAVIORAL PROBLEMS** of patients who are developmentally disabled often has been controversial; even clinicians who are convinced of beneficial effects of certain medications prefer to use such agents only when specific **DSM-IV-TR** diagnosis is present.
- psychoactive drugs without behavioral therapy and environmental changes is rarely helpful.

Selection of Medications for Target Symptoms:

Condition	Preferred Medications
Self-injurious behavior	Newer atypical antipsychotic Anticonvulsant/mood stabilizer Also consider SSRI
Physical aggression to people or property	Newer atypical antipsychotic Anticonvulsant/mood stabilizer Also consider SSRI
Nonaggressive agitation	Anticonvulsant/mood stabilizer Also consider newer atypical antipsychotic, SSRI
Suicidal ideation/behavior	SSRI Also consider anticonvulsant/mood stabilizer
Anxiety	SSRI Buspirone Also consider benzodiazepines
Hyperactivity	Psychostimulant Also consider α 2-agonist, anticonvulsant/mood stabilizer
Insomnia	Trazodone Also consider zolpidem
Psychiatric or behavioral problems with comorbid epilepsy	Divalproex/valproic acid Carbamazepine Also consider gabapentin

When all else fails → consider **CLOZAPINE**

DROOLING

- one option is **Salivary Duct Clips** - control drooling → improved caregiving, reduced aspirations.

PROGNOSIS

- it is **unrealistic to expect** catch-up and achievement of normalcy.
- life expectancy** may be shortened (more severe retardation and greater immobility, higher is mortality risk).

IQ	Abilities				
	Mental	Language	Education	Work	Daily Living
> 115	Superior		unrestricted		
85-115	Average		unrestricted		
70-85	Borderline	Usually normal	Remediation needed	Employable	Independent ADL, may need some living help
55-70	Mild mental deficiency	Normal or impaired	Limited ability	Employable at selected tasks	Need variable amounts of living help
40-55	Moderate mental deficiency	Normal or impaired	Very Limited	May be capable of simple tasks	Dependent for living, may need ADL help
25-40	Severe mental deficiency	Limited or absent	Minimal functional	None or minimal	Need ADL help
< 25	Profound mental deficiency	Absent	--	--	Totally dependent, often nonambulatory and incontinent

ADL, Activities of daily living

- when no cause for mental retardation can be found, multifactorial inheritance is most likely (3-5% recurrence risk).

Viktor's NotesSM for the Neurosurgery Resident
Please visit website at www.NeurosurgeryResident.net