Mental Retardation

Last updated: April 24, 2019

ETIOLOGY

DIAGNOSIS

TREATMENT

PROGNOSIS

Mental retardation is a global developmental delay (due to some pathogenic process) that results in significant limitation of intellect or cognition → poor adaptation to demands of everyday life. It affects ~3% of population in developed countries.

- Improvements in obstetric and neonatal care, rubella vaccination, Rh(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 in judgment) – especially common in lower socioeconomic class (vs. other degrees are distributed evenly).
2. Moderate (10-15%): IQ = 50-35
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.

- Mood disorders (exp. 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 (exp. Lesch-Nyhan syndrome).
- Anorexia and bulimia nervosa are relatively rare, but mental retardation is predisposing for pica and rumination.

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 impairments but are not characteristics of MR).

- Primary manifestations of MR:
  1) delayed intellectual development (in mild MR, not recognizable until preschool age); it is much more common to experience slow progress than developmental arrest
  2) immature behavior
  3) limited self-care skills.

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- 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 (exp. ethanol: most important of teratogenic substances!)
8. Teratogenesis of pregnancy
9. Placental insufficiency. poor nutrition (e.g. iodine deficiency → cretinism).

Perinatal Causes

1. Infections (exp. 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 (exp. lead poisoning).
3. Brain tumors and associated treatments.
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]).
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 psycho-social (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:

<table>
<thead>
<tr>
<th>Condition</th>
<th>Preferred Medications</th>
</tr>
</thead>
<tbody>
<tr>
<td>Self-injurious behavior</td>
<td>Newer atypical antipsychotic, Anticonvulsant/mood stabilizer</td>
</tr>
<tr>
<td>Physical aggression to people or property</td>
<td>Newer atypical antipsychotic, Anticonvulsant/mood stabilizer</td>
</tr>
<tr>
<td>Nonaggressive agitation</td>
<td>Anticonvulsant/mood stabilizer</td>
</tr>
<tr>
<td>Suicidal ideation/behavior</td>
<td>SSRI, Also consider anticonvulsant/mood stabilizer</td>
</tr>
<tr>
<td>Anxiety</td>
<td>SSRI, Buspirone, Also consider benzodiazepines</td>
</tr>
<tr>
<td>Hyperactivity</td>
<td>Psychostimulant, Also consider R-2 agonist, anticonvulsant/mood stabilizer</td>
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<tr>
<td>Incontinence</td>
<td>Trazodone, Also consider spasmolytics</td>
</tr>
<tr>
<td>Psychiatric or behavioral problems with comorbid epilepsy</td>
<td>Newer anticonvulsant/mood stabilizer, Carbamazepine</td>
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When all else fails – consider CLAMAPINE

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 |
--- | --- | --- | --- | --- | --- | --- |
< 75 | Superior | unrestricted | unrestricted | | | |
75-85 | Average | | | | | |
55-70 | Mild mental deficiency | Usually normal | Remediation needed | Employable | Independent ADL, may need some living help |
40-55 | Moderate mental deficiency | Normal or impaired | Limited ability | Employable at selected 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).