

INTELLECTUAL DISABILITY

MBChB Level IV Tutorial

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INTRODUCTION

- Intelligence

- Sum of those aspects of mental life that relate to general cognitive abilities necessary for appraising and adapting to the environment
- Intelligence seems to be normally distributed within the population
- Is under both genetic and environmental control

INTRODUCTION...

- Neurodevelopmental disorders
 - Starts early in infancy and childhood
 - Impairment and delay in central nervous system
 - The course is fairly stable compared to other mental disorders which can remit and escalate

INTRODUCTION...

- Neurodevelopmental disorders include:
 - Intellectual disability
 - Autism spectrum disorders
 - Attention Deficit Hyperactivity disorder

ID DEFINITION

- Abnormal intellectual development may be due to slowness in development (retardation, delay) or distortions in development, or both
- Intellectual disability is a consequence of intellectual impairment
- Intellectual disability is a "disability characterized by significant limitations in both **intellectual functioning** and in **adaptive behaviour**, which covers many everyday social and practical skills"

ID DEFINITION...

Adaptive functioning/skills

- There are three sets of adaptive skills;
- *conceptual skills*- reading, numbers, money, time, and communication skills
- *practical life skills*- feeding, bathing, dressing, occupational skills, and navigational skills.
- *social skills*- understanding and following social rules and customs, obeying laws, and detecting the motivations of others in order to avoid victimization and deception

ID DEFINITION

- The impairment should be global, of early onset and long term
- The child's IQ should be less than 70
- The child should be functionally impaired in every day life skills
- * IQ tests are standardized tests, designed to have a mean of 100 and a standard deviation of 15.

CLASSIFICATION...

- Classification of intellectual disability by IQ
- Mild 50-55 to 70 (account for 85 % of ID)
- Moderate 35-40 to 50-55 (10% of ID)
- Severe 20-25 to 35-40 (3-4% of ID)
- Profound below 20 or 25 (1-2% of ID)

PREVALENCE

- 2-3% of the general population score in the mild ID (IQ 50-70)
- Moderate to profound (IQ < 50) occurs in 3-4 per 1000 children
- More boys than girls in special schools for learning disabilities

AETIOLOGY

- Genetic

- Chromosomal abnormalities

- Trisomy 21: Down Syndrome

- Accounts for about a third of all instances of intellectual disability with IQ less than 50 (mean IQ of about 45)
 - There is a relationship between the proportion of cells showing trisomy 21 and the severity of ID

AETIOLOGY

- Chromosomal abnormalities
 - Trisomy 13 (Patau's)
 - Trisomy 18 (Edwards)
 - Not compatible with life, although some survive several months
 - Cri-du chat syndrome: partial deletion of short arm of chromosome 5

AETIOLOGY

- Metabolic disorders
 - Phenylketonuria
 - Galactosemia
- Prenatal
 - Maternal illnesses
 - Rubella, syphilis, toxoplasmosis
 - Endocrine disorders
 - malnutrition
 - Toxins
 - Lead, alcohol
 - Placental dysfunction

AETIOLOGY

- Perinatal causes
 - Birth asphyxia
 - Prematurity complications
 - Kernicterus
- Post natal
 - Head Injury (accidental / non accidental)
 - Toxins
 - CNS infections
 - Seizures
 - Environmental deprivation
- idiopathic

CLINICAL FEATURES

- First identified or suspected antenatally if there is known family history of a genetic disorder associated with ID, or both parents have ID
- May be identified soon after delivery when a condition known to be associated with ID is identified e.g. Down Syndrome
- Biochemical screening identification soon after birth can identify children with hypothyroidism, phenylketonuria

CLINICAL FEATURES

- Diagnosis may be delayed at times for several years until the consequences of generalised development become obvious. E.g. where physical features are subtle and degree of impairment is mild or moderate (autism, fragile X syndrome)
- Severity of ID, social circumstances and sometimes underlying aetiology influence mode of presentation

CLINICAL FEATURES

- Features include

- Delayed gross motor milestones in the first year of life (* can be unreliable influenced by familial and cultural factors)
- Delayed social milestones such as smiling, attachment behaviour
- Speech and language delay
- Deafness may be suspected due to lack of response to sound, lack of appropriate words
- Failure to make educational progress

CLINICAL FEATURES

- Clinical features will depend more especially on:
 - Severity of the impairment
 - Associated physical and psychiatric conditions
 - Quality of care and education received
 - Aetiology of the intellectual disability

CLINICAL FEATURES

- Mild: appearance unremarkable, normal language, can live independently as adults, need help coping with family responsibilities
- Moderate: Language is affected with simple speech. Activities of daily living acquired over time e.g. dressing, feeding, attention to hygiene. Problems with extended activities like handling money and directions

CLINICAL FEATURES

- Severe: development greatly slowed. Look after themselves eventually after close supervision. Engage in simple social activities
- Profound: dependent even for simple activities of daily living.

ASSESSMENT

- Four components
- Assessment is not a-once-and-for-all exercise.
 1. Investigation of the cause
 - Frequently not possible
 - Basic right to know
 - Provide relief
 - Assists in focusing towards the future
 - A treatable cause may be found
 - Amniocentesis, routine biochemical tests, physical examination may reveal stigmata consistent with an obvious chromosomal abnormality

ASSESSMENT

2. Identification of associated physical and psychiatric problems

- Sensory disorders are common in children with ID
- General physical and neurological examination. Cerebral palsy can occur in 25% of children with moderate to severe ID
- Behavioural and emotional problems: ADHD (10% of mod-severe ID), Autism spectrum disorders,
- Self injurious behaviour (10-15% of severe ID): Conditions associated with self injurious behaviour include: Lesch-Nyhan, fragile X, Prader Willi, Smith-Magnesis

ASSESSMENT

3. Determination of level of functioning

- General intelligence
- Speech and language
- Gross and fine motor development
- Sensory issues
- Social and personal development

ASSESSMENT

4. Assessment of family functioning, care, expectations and coping capacity
 - A major source of difficulty is mismatch between the child's abilities and potential, and parental perceptions of what the child is and is capable of.

MANAGEMENT

Important components

- Breaking the news
 - Nature
 - Likely causes
 - Likely further developments
 - Break the news as soon as possible to both parents
 - Arrange for subsequent follow-up meetings
 - Psycho-educational: genetic counselling where available, likely implications on the child in the future, future parental pregnancies
 - Psychotherapeutic: sympathetic and supportive listening, moving family towards practical solutions

MANAGEMENT

Important components...

- Counselling on promotion of development
 - Child will more help in the acquisition and retention of new skills from parents, teachers
 - Help needs to be provided at an appropriate level for the child
 - Early intervention programs
 - Link up parents with various professionals as early as possible

MANAGEMENT

Important components...

- Dealing with associated disabilities and behavioural problems
 - Common physical deficits: impairment of vision, hearing, epilepsy, cerebral palsy
 - Common psychiatry deficits: ADHD, Autism spectrum disorders
 - Other psychological problems: aggression, self injury, obsessive compulsive behaviour

MANAGEMENT

Important components...

- Advising on appropriate education
 - Provide information on medical matters (meds, physiotherapy, hearing aids, spectacles)
 - Child's attention span, persistence, level of activity, capacity to form relationships, language and communication skills

MANAGEMENT

Important components...

- Genetic counselling where applicable
- Providing social and emotional support
 - Parents get support from various sources- relatives, friends, neighbours, friends, social workers, teachers
 - Regular examinations and health checks

Questions and clarifications...