
Challenges in Cognition and Learning

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Agenda

- Definitions
- Reflection & Research
- Intellectual Disabilities- Mental Retardation
- Down's Syndrome
- Reflection

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Definitions

- Spastic muscles
- Hypotonic muscles
- Adaptive skills
- Nondisjunction
- Translocation
- Trisomy
- Mosaic Trisomy
- hypothyroidism

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Research

- Why is the term “Mental Retardation” so controversial?
- Search the web and identify at least two other terms commonly used to identify this population.

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Intellectual Disabilities and Early development

- Ongoing process that begins with embryo and continues through the life span
- Closely linked to CNS development
- Most rapid intellectual growth in first few years of life
- Progresses in a cephalocaudal fashion
- Follows a fixed sequence
 - The fixed sequence allows for early assessment of problems by comparing rates of developmental milestones with norms.
- Four areas assessed:
 - Gross motor
 - Fine Motor
 - Language
 - Social Adaptive

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- **Gross Motor Skills**
 - Progress from reflexive movement to running and jumping by the age of 2
- **Fine Motor Development**
 - Progress from clenched fists to scribbling and drawing by 18 months
- **Language Skills**
 - Progress from crying at birth to two and three word phrases by the age of 2
- **Social-Adaptive Skills**
 - Progress from recognizing the mother to playing with other children and independent feeding skills.

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■ A Review of Piaget

- The Sensorimotor Stage: Birth to 18 months
 - Thinking linked to sensorimotor schemes involved with grasping, reaching and vocalizing. Child becomes interested in external world.
- The Preoperational Stage: 2-7 years
 - Can use symbol to represent objects not present, language skills improve. Child can pretend and make up a story.
- The State of Concrete Operations: 7-12 years
 - Can order and classify numbers, classes, relations and other aspects of the physical world. Can generalize to new learning situations. Understand right from wrong.
- The Stage of Formal Operations: 12 years of age
 - Thinking no longer limited to concrete events and is able to project into the future and make long term goals. Develop sensitivity to the feelings of others and become self conscious.

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Developmental Delay

- Term used to indicate atypical neurodevelopment in which there is a failure to achieve age-appropriate milestones
 - Early in life detected by inadequate sucking, floppy or spastic muscles, or lack of visual or auditory response
 - Later delays in sitting, walking and lack of language
 - When a significant delay is present, mental retardation is likely diagnosis
 - Not always indicative of a severe disability

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Developmental Delay

- Early Identification
 - Rationale is to intervene when the CNS is malleable and responsive to habilitation
 - Early identification more common in severe impairments
 - Mild impairments often not recognized until school age
 - Developmental screening should be done by pediatrician on a yearly basis
 - Early identification must be multifaceted as screening tools are not that accurate
 - At risk children should be provided early intervention even if delays do not present themselves at the current time
 - Evidence of a significant delay should trigger a comprehensive evaluation

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Developmental Delay

- Early Intervention Services
 - Traditionally begin in elementary school, but due to changes in laws, early identification programs now include children from birth to 21 years of age.
 - In Infant-Toddler Programs an IFSP is developed that includes
 - Statement of present levels of performance
 - Statement of family concerns and priorities
 - Statement of outcomes expected of the child and family and timelines that determine progress
 - Specific early intervention services provided
 - Name of service coordinator responsible for implementing plan
 - In Preschool and IEP is developed that will provide the child with an appropriate education delivered in the least restrictive environment.

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Mental Retardation

- Definition is controversial, there is general agreement that a person with this disability must have
 - An Significantly subaverage intellectual functioning
 - impairment resulting from an injury, disease or abnormality that existed before age 18
 - An impairment in adaptive abilities
- IQ scores
 - Mild (requiring intermittent support) 50-55 to approx. 70
 - Moderate (requiring limited support) 35-40 to 50-55
 - Severe (requiring extensive support) 20-25 to 30-35
 - Profound (requiring pervasive support) below 20-25

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Mental Retardation

- Age of onset
 - Onset must be during childhood when the child is in the developmental years.
 - Mental retardation must be present during developmental brain growth
- Adaptive Impairments
 - Must also be impaired in their ability to adapt or function in daily life
 - Must include at least two of the following impairments in adaptive behaviors: Communication, self-care, home living, social/interpersonal skills, use of community resources, self-direction, functional academic skills, work, leisure, health and safety.

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Prevalence of Mental Retardation

- Prevalence depends on definition used, method of ascertainment, and the population studied.
 - According to classic definition 2.5% of population is affected
 - Cultural and socially disadvantaged removed, prevalence is between 0.8% and 1.2%
 - Prevalence not increased since the 1940's
 - Natural balance between improved health care and the emergence of new diseases.
- Associated Impairments
 - Mental retardation is frequently accompanied by associated impairments
 - Cerebral palsy, visual impairments, seizure disorders, communication impairments, feeding difficulties, psychiatric disorders, and attention deficit/hyperactivity disorder
 - Associated impairments may make it difficult to distinguish mental retardation from other development delays

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Causes of Mental Retardation

- There are two overlapping populations
 - Mild retardation is associated with lower socioeconomic status
 - May have a genetic component compounded by environmental variables
 - More severe retardation linked to biological causes in two thirds of cases
 - Most common diagnosis: Fragile X syndrome, Down syndrome, and FAS.
 - The earlier the problem exists, the more severe its consequence
 - Chromosomal disorders, hereditary syndromes, insults in the first and second trimester and other pregnancy problems cause the most severe problems.

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Mental Retardation

- Medical Diagnostic Testing
 - No single method exists for all causes of mental retardation
 - Diagnostic testing must include historical information and medical examination
 - Might include: chromosomal study, metabolic investigation, EEG, or MRI depending on the concerns and outward appearance of child
 - How extensively a cause is pursued depends on:
 - What is the degree of mental retardation
 - Is there a specific diagnostic plan to follow
 - Are the parents planning to have children
 - Do the parents want a diagnosis?

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- Psychological Testing
 - Infants is the Bayley Scales of Infant Development
 - Over the age of 2 are: Stanford Binet Intelligence Scale and the Wechsler Intelligence Scales
- Infant Development Tests
 - The Bayley Scales assess
 - Fine motor, gross motor, language, visual problem solving skills and behavior from 1 month to 3 ½ years of age.
- Intelligence Tests on Children
 - Stanford Binet used most often with children 18 months or older
 - Asses four areas of intelligence
 - Visual abilities, abstract/visual thinking, quantitative reasoning, and short-term memory
 - Wechsler Preschool and Primary Scales of Intelligence most often used with children with a mental age of 3-7
 - Wechsler Intelligence Scale for Children most often used with children who function above a 6 year mental age

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■ Tests of Adaptive Functioning

- In addition to intelligence testing, adaptive skills must be measured
 - Most commonly used is the Vineland Adaptive Behavior Scales
 - Involves parent, caregiver/teacher with questions about adaptive behaviors that include
 - Communication, daily living skills, socialization, and motor skills
 - Other scales used are:
 - Woodcock-Johnson Scales of Independent Behavior,
 - American Association of Mental Deficiency Adaptive Behavior Scale
 - There is usually a good correlation between scores on the intelligence and adaptive scales

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Intervention Approaches for Mental Retardation

- Most useful approach is multidisciplinary directed at many aspects of the individual's life including education, social, and recreational activities, behavior problems and associated impairments
- Educational Services
 - The single most important focus
 - Must be relevant to child's needs and address areas of strength and weakness
 - To develop education plan, developmental level, level of support and goals for learning and independence must be established
- Leisure and Recreational Needs
 - Social and recreational needs must be addressed
 - Adolescents frequently do not have opportunities for appropriate social interactions
 - Need to participate in dances, trips, dating and other typical social events

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Prognosis

- Depends on the underlying cause
- The presence of associated medical and developmental disabilities
- The capabilities of the family
 - Many adults with **mild to moderate** mental retardation gain economic and social independence
 - Most marry and live successfully independently or in supervised settings.
 - Life expectancy is not adversely affected
 - Many adults with more **severe** mental retardation live at home or in a supervised setting.
 - They may have successful work in the community
 - Adults with **profound** mental retardation may perform simple tasks in a closely supervised workshop
 - These adults usually live with parents, or in group homes

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Down Syndrome

- Was one of the first symptom complexes associated with mental retardation to be identified as a syndrome.
 - Three types of chromosomal abnormalities lead to Down syndrome
 - Trisomy 21
 - Results from nondisjunction, mostly commonly during meiosis I of the egg
 - Translocation
 - Involves the attachment of the long arm of an extra chromosome #21 to chromosome #14, #21, or #22.
 - Mosaic trisomy
 - Some but not all cells have the defect
 - Resulting from the nondisjunction during mitosis of the fertilized egg
 - IQ is 10-30 points higher and have fewer medical complications than trisomy 21 or translocation

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■ Prevalence

- Has decreased since the 1970s due to prenatal diagnosis
 - Linked with maternal age
 - No age effect on translocation, but one third inherit the gene from parents who are carriers
 - This genetic risk can be diagnosed prior to pregnancy
 - Trisomic Down syndrome occurs in more males (59%) than females (49%)
 - Translocation Down syndrome occurs more often in females(74%)

■ Etiology

- If an individual has three copies of “critical region” on chromosome #21, he or she will have the clinical features of Down syndrome
 - Trisomy causes malformation as a result of incomplete rather than deviant development of the embryo
 - Normally formed heart with incompletely closed chambers
 - Multiple brain abnormalities include delayed myelination, fewer neurons, decreased synaptic density and decreased acetylcholine neurotransmitter receptors

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■ Early Identification

- Women over 35 are offered prenatal screening identification prior to birth to allow the physician to provide genetic counseling and appropriate medical evaluation of the newborn
- Due to distinctive pattern of physical features, identified easily at birth
- Diagnosed based on eight phenotypic characteristics including
 - Three palm print pattern, Brushfield spots, ear length, internipple distance, neck skinfold, and widely spaced first toes

■ Medical complications of Down Syndrome

- Increased risk of abnormalities in almost every organ system
- Congenital Heart Disease
 - Two thirds have endocardial cushion defect (connection between the atria and ventricles), ventricular septal defect, and atrial septal defect
 - Major complications are congestive heart failure due to pulmonary vascular obstructive disease

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- **Medical Complications in Down Syndrome**
- **Sensory Impairments**
 - Both hearing and vision problems are prevalent
 - Ophthalmic disorders include refractive errors, strabismus, nystagmus, blepharitis, tear duct obstruction, cataracts and ptosis
 - Hearing loss can be conductive, sensorineural or both
 - Conductive problems due to narrow throat and immune deficiency that causes recurrent ear infections
- **Endocrine Abnormalities**
 - Hypothyroidism 28 more time prevalent
 - Diabetes more than twice the usual prevalence
 - Half are overweight by the time they are in early childhood
 - All have short stature with men averaging 5 feet and women averaging 4 ½ feet in height.

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- **Medical Complications in Down Syndrome**
- **Orthopedic Problems**
 - Increased prevalence due to ligament abnormalities
 - Include atlanto-axial subluxation, hip dislocation, patella instability, and flat feet.
 - Develop juvenile rheumatoid arthritis-like disorder
- **Dental Problems**
 - Most serious is periodontal disease
 - Involves gingivitis and loss of alveolar bone
 - Due to general low resistance to infections
- **Gastrointestinal Disorders**
 - Found in 5% of children with problems in feeding, vomiting and aspiration pneumonia in the newborn period.
 - Malformations include stenosis or atresia of the duodenum, imperforate anus, enlarged colon, tracheo-esophageal fistula, or esophageal atresia, and pyloric stenosis.

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■ Medical Complications in Down Syndrome

- Seizure Disorders
 - Epilepsy occurs in 6% of children with Down syndrome
 - Wide variety of types of seizures
- Hematologic Disorders
 - Almost every cellular element of the blood system has been found to be at risk for abnormality
 - Rarely lead to severe problems
 - Have a 1 in 150 chance of developing leukemia compared to 1 in 2,800 in the general population
- Skin Conditions
 - Most common are eczema, inflammation of the lips and dry and scaly skin.

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■ Neurodevelopmental and Behavior Impairments

- Typically have gross motor skill delay due to central hypotonia
 - Gross motor milestones are delayed
 - Sit up at 1 year of age and walking at age 2
- Cognitive delays often not noticed until after the age of 2
 - Significant delays become evident due to the lack of language development with the first word usually at 24 months
 - Poor verbal short term memory, but stronger visual-motor skills
- Stereotyped as being happy and amiable
 - Actual profiles the same as typically developing children
 - Do have behavioral and psychiatric disorders including ADHD, ODD, aggressive behavior, phobias, eating disorders, elimination disorders, Tourette syndrome, autism and self-injurious behaviors
 - May experience a deterioration of cognitive or psychological functioning in adolescence
 - May be attributed to unrecognized hypothyroidism
 - Increased prevalence of Alzheimers

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Evaluation and Treatment

- All children with Down Syndrome need to be periodically evaluated for many of their medical complications
 - Echocardiogram in the newborn period
 - Within 6 months complete ophthalmic evaluation
 - Auditory Brainstem ABR test for hearing
 - Polysomnogram for sleep apnea
 - Screening for congenital hypothyroidism
 - X-rays for atlantoaxial subluxation
- Early Intervention
 - Parents need to be aware of variety of services available to them
 - Early intervention education programs
 - Supplemental Security Income SSI
 - Parent support groups both locally and nationally
 - Respite care options

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Down's Syndrome

- Prognosis
 - Since the 1970 prognosis has improved due to parent advocacy groups
 - Children are raised in their own homes and are the pioneers of mainstreaming and inclusion
 - Life expectancy and quality of life have improved
 - Supported employment opportunities have allowed adults to hold real jobs in the community

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Summary

- Development is a step-by step process linked to the maturation of the CNS
- Mental retardation impairs development of intellectual functioning and adaptive skills.
- In mild to moderate mental retardation, the underlying cause can be unclear
- In the severe and profound population, there is usually a definable cause
- Vast majority have only mild to moderate mental retardation
- Early intervention is important to enable the child to develop and use all capabilities
- Down Syndrome of one major cause of mental retardation
- Children with Down Syndrome may have multiple health issues but current prognosis is much better due to medical intervention
- The social and personal needs of individuals with intellectual disabilities are similar to the needs of the general population

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Reflection

- How does your new knowledge of individuals with intellectual disabilities change the way you may provide early intervention to these children and their families?

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