

Mental Retardation - Current Definition

- Since 1992, focus is adaptive
- Definition revised by AAMR in 2002
- Two aspects: deficits in intellectual functioning and in adaptive behavior
- Subaverage intelligence
 - IQ 70-75 or below, 2 SDs or 2nd percentile
- Two or more areas of adaptive deficit
 - communication
 - self-care/home living/social skills
 - community use/health and safety
 - functional academics/leisure/work
- Manifested in early development, before age 18 yrs.

Mental Retardation - Classification System (p. 110 and Table 4-1)

Mild/ Educable	55-69 IQ	Often achieve early secondary level skills; some independence
Moderate/ Trainable	40-54 IQ	Attain early elementary level; some support for living skills
Severe	25-39 IQ	Develop some self-help skills; require supportive living
Profound	0-24 IQ	Limited achievements for self-care; not independent as adults

Mental Retardation and Cognition

- Typically developing research groups
 - Chronological age peers = CA matches
 - Mental age (younger) peers = MA matches
- Cognitive development is generally same as in non-MR population but slower
 - MR = MA matches < CA matches
- Attention and focus is relative strength
 - MR = CA matches
- Attention to salient features, symbolism, organization and recall relatively poorer than predicted by IQ
 - MR < MA matches < CA matches

Language Deficits Associated with MR

- 50% of MR have lang skills = MA
- 50% of MR have lang skills < MA
 - 25% poorer expressive only
 - 25% both rec/exp relatively poorer
- **Syntax and Morphology**
 - MLU<3 MR=MA; MLU>3 MR<MA
 - still acquire in developmental order
- **Phonology** - increased incidence of speech problems in MR pop
- **Semantics** - MR=MA, yet overall tend to be more literal and concrete than peers
- **Pragmatics** - acquisition similar but difficulty with pronouns and requesting clarification

Terms Associated with Chromosomal and Genetic Syndromes

- **Autosomal disorders** – affects one of the 1-22 # pairs of chromosomes
- **Sex chromosome disorders** – affects sex chromosomes, X or Y
- **Gene** – chromosomes consist of genes, genes direct production of proteins
- **Dominant vs. Recessive Inheritance** – 1 vs. 2 affected genes lead to traits
- **X-linked Inheritance** – traits inherited from genes on the X chromosome
- **Trisomy** – 3 chromosomes instead of 2

Down Syndrome

- **Most common and best known chromosomal abnormality**
 - 1 in every 700 newborns
 - Note that it is genetic, but not inherited
- **Trisomy 21**
 - extra chromosome, position 21
- **Associated with increased maternal age**
- **National Down Syndrome Society web site**

Physical Features of Down Syndrome

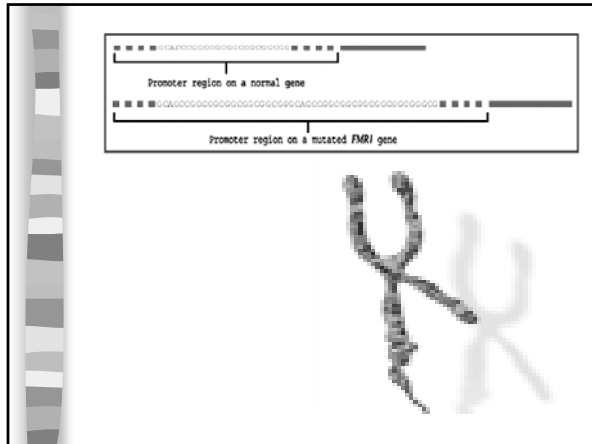
- round, flat face with midface hypoplasia
- epicanthic folds (vertical fold over inner corner of eye)
- eyes slant up (palpebral slant)
- small mouth w/large, protruding tongue
- short broad neck
- Brachycephaly – distance from front to back of head is shortened
- broad hands, short fingers, and transverse palmar crease
- overall hypotonic or low muscle tone

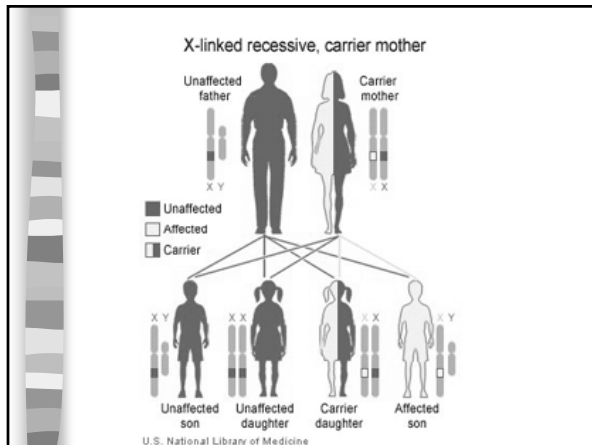
Speech/Language Characteristics of Down Syndrome

- **Speech problems severe**
 - associated with oral/facial anomalies and low muscle tone
- **Hearing problems prevalent**
 - conductive (83% of pop)
 - sensorineural (10-15%)
 - mixed loss (8%)
- **Cognitive impairment has impact on speech/language development**
 - 50-60% pop has IQ range from 30 to 50

Fragile X Syndrome

- **Most common hereditary cause of MR**
- **Identification and diagnosis of Fragile X only since the 1980s, FMR1 gene**
- **“fragile” site on X, actually repeats of proteins**
- **Birth incidence**
 - 1/4000 males and 1/8000 females
- **Pre-mutation versus a full mutation**
- **Also, carrier versus expression**





Physical Features of Fragile X

- long, narrow face
- large, cupped ears
- hyperextensible joints/double-jointed thumbs
- flat feet, part of the connective tissue differences like the double-jointed aspect
- large testicles or macroorchidism (after age 8 years or in puberty)
- National Fragile X Foundation Photo Gallery

Speech/Language Characteristics of Fragile X

- **Language impairment varies with cognition**
 - some children are learning disabled, others have mild to severe mental retardation
- **rapid speech rate, “cluttering”**
- **mumbling and jargon, also part of cluttering**
- **echolalia**
- **topic maintenance and content problems**
- **perseveration**

Behavioral Characteristics of Fragile X

- **Behavior**
 - aggression
 - hyperactivity
 - impulsivity
 - short attention span
- **Sensorimotor Integration Differences**
 - low muscle tone
 - resistance to change
 - toe walking
 - hand flapping
- **2-6% of individuals with autism also have Fragile X syndrome**

Maternal Substance Abuse and Language Impairment

- **teratogen** - substance that crosses placental barrier, known to cause abnormal structural development of embryo
- **Types of Teratogens:**
 - alcohol, illegal drugs, cigarettes
 - radiation
 - legal drugs (retinol)
 - maternal infections
- **Factors that affect teratogen exposure:**
 - inherent susceptibility
 - quantity
 - critical periods of development
- **Also caregiver environment is at risk**

Fetal Alcohol Spectrum Disorder (FASD)

- **Fetal Alcohol Syndrome (FAS)**
 - 3 diagnostic criteria
 - Pre- and/or post-natal growth deficiency
 - Facial dysmorphology
 - Neurobehavioral effects
- [National Organization on FAS web site](#)
- [Fact Sheet from NOFAS click here](#)
- **Partial Fetal Alcohol Syndrome (PFAS)**
 - milder version, less dramatic physical effects
 - Used to be termed Fetal Alcohol Effects or FAE
- Difficult to diagnose without knowledge of maternal alcohol use



Facial Dysmorphology Associated with FAS

Discriminating Features

- Short palpebral fissures
- Flat midface
- Short nose
- Indistinct philtrum
- Thin upper lip



Associated Features

- Epicanthal folds
- Low nasal bridge
- Minor ear anomalies
- Micrognathia

FAS and Language Impairment

- Tied to cognitive ability (Avg. IQ=70)
- Learning problems with poor math skills
- Poor abstract reasoning, poor judgment
- Better expressive than receptive language
- Poor vocabulary and comprehension (oral and written)
- Other behavior problems
 - hyperactive/distractible
 - social isolation
 - poor impulse control/lack of inhibition

Cocaine and Birth/Development

- Possible outcomes of cocaine exposure:
 - premature detachment of the placenta
 - prematurity
 - congenital malformations, microcephalia
 - <50% have negative birth histories
- Newborn characteristics:
 - irritable, poor feeding, high-pitched cry, abnormal sleep patterns, tremors, stiffness
 - recover in first year
- Developmental problems are similar to those seen in children in chaotic homes

Cigarette Smoking and Birth/Development

- Possible outcomes:
 - low birth weight
 - prematurity
 - growth retardation
 - SIDS
 - cognitive achievement limitations

Neglect/Abuse and Language Impairment

- 5 types (Nelson)
 - physical abuse
 - sexual abuse
 - emotional abuse
 - neglect (physical and emotional)
- "Mutual Influence"
 - chicken and egg dilemma
 - Extreme example = "Genie"
- Children adopted from Russian orphanages
 - More overall behavior problems than for other international adoptees
 - Impairment of attachment
 - Hyperactivity and conduct disorders
 - Cognitive impairments in problem-solving abilities and cause-and-effect thinking



Language Skills and Abuse/Neglect

- Severely neglected children and abused children consistently earn lower scores on language tests than control groups
- Possible that the abuse causes the child to withdraw from the needed stimulation, also brain injury is possible
- Possible language characteristics:
 - unable to use language to express feelings
 - difficulty sustaining narratives and conversations
 - poor problem solving skills using language

Selective Mutism Defined

- Aka elective mutism
- (def) consistent failure to speak in social situations despite the ability to speak in other situations
- Selective Mutism Foundation
- Social anxiety is a prominent feature
- Criteria:
 - duration more than one month
 - not English as Second Language
 - interferes with educational, occupational, and social functioning
 - no developmental or psychiatric disorder (autism, schizophrenia, MR, PDD)

Selective Mutism Characteristics

- Usually speak with family at home, but won't speak at school or in public
- Onset in preschool years
- Generally normal language skills
- Rare disorder, Mostly girls
- Possible personality traits:
 - shy, anxious, negative, oppositional, controlling, manipulative, withdrawn, clingy
- Cause is unknown, possible genetic link
 - family history of extreme shyness, social phobia, or anxiety disorder
- No evidence to suggest link between selective mutism and child abuse

Treating Selective Mutism

- Behavioral
 - SLP or psychologist uses behavioral approach to reinforce increasing talking
 - desensitization
 - family-involvement
 - audio feedforward - tape child speaking and then play tape in situations where child does not typically speak
- Pharmacological treatment
 - Prozac (fluoxetine)
- Educational
 - modified curriculum

Autism Spectrum Disorders

(Box 4-10, pp 135-136 Paul)

- Autism
- PDD-NOS - Pervasive Developmental Disorder - Not otherwise specified
- Rett's syndrome - deceleration in head growth and related retardation, stereotypic hand movements, typically girls
- Childhood disintegrative disorder - onset of autism or PDD after apparently normal development for at least 2 years after birth
- Asperger's syndrome - like autism without the cognitive and language impairments

Autism

- **Criteria for diagnosing autism (DSM-IV, 1994) see Paul, p. 135**
 - 2 characteristics of social interaction impairment
 - Poor eye contact, doesn't engage or enjoy interactions
 - 1 characteristic of communication impairment
 - Delayed or absent speech, stereotypic or repetitive
 - 1 behavioral/motor characteristic
 - Sameness, hand flapping, rituals and preoccupations
 - Delays before age 3 in social, language or play, not Rett's or CDD.

Two patterns of Autism

- **Early onset**
 - 2/3 parents of children with autism report that their children were different from birth, significant developmental delays in walking, talking
- **Late onset (aka Regressive type)**
 - 1/3 parents of children with autism report normal development with regression around 12-24 months

Autism Facts and Figures

- **Cause is neurological, not psychiatric**
- **Increased incidence in family members, so genetic factor**
 - Genes on 4 chromosomes associated with autism in siblings
- **Environmental triggers are suspected**
 - "no such thing as a genetic epidemic"
- **Great range in functioning - "low, middle, and high functioning"**
 - 60% have IQ <50
- **Current estimates of autism are 1 in 150 births**
 - 1992-93: 68 children with autism in AL, 2003-04: 1319
- **4X more males**

Environmental triggers

- **Some believe that over-vaccination is a factor in increase of autism**
 - 1940 3 vaccine doses before age 2, autism incidence was 1 in 10,000 births
 - 1980 8 doses before 2, incidence was 5 in 10,000
 - 2004 22-30 doses, incidence is 1 in 150
- **Thimerosal (form of mercury, which is a known neurotoxin)**
- **Birth hair mercury levels for autistic children is significantly lower than for nonautistic individuals – it may be an excretion problem**

Language/Communication Characteristics of Autism

- Failure to acquire language or sudden stop in development at 18-30 months
- 50% of population are nonverbal
- Severe impairments of comprehension
- Language learned “holistically” or unanalyzed whole forms
- Echolalia
- Possible apraxia of speech
- Pronoun reversals
- Unusual and inappropriate pitch, rate, rhythm of speech or voice
- Poor understanding of nonverbal communication
- Hand leading

Behavioral Characteristics of Autism

- Insistence on sameness, routine, perseveration
- Hypersensitivity to sounds
- May prefer to play alone
- Tantrums
- Biting, hitting, head banging, self-injurious
- Hand flapping, spinning, rocking
- Apparent insensitivity to pain
- Poor eye contact
- Fixation on certain objects

Possible Treatments

- **Traditional**
 - Speech-language therapy
 - Occupational therapy
 - Intensive educational/behavioral approaches, ABA
- **Nutritional/Dietary**
 - Gluten free and casein free diets
 - Vitamins, supplements, enzymes
 - Glutathione cream
- **Biomedical**
 - Chelation for heavy metal removal
 - Hyperbaric oxygen therapy

Defining ADHD

- Attention Deficit Hyperactivity Disorder (ADHD)
- Persistent pattern of inattention and/or hyperactivity-impulsivity, more frequent and severe than peers
- Present before age 7 years
- Symptoms in 2 or more settings
- Impairment of social, academic or occupational functioning
- NOT PDD, schizophrenia, psychotic disorder or other mental impairment

Criteria for ADHA

- Inattentive Criteria
 - little attn to details, difficulty with organization
 - short attn span, fails to finish tasks
 - easily distracted, forgetful
- Hyperactive Criteria
 - fidgets, squirms, leaves seat, runs or climbs
 - difficulty playing quietly
 - talks excessively
- Impulsive Criteria
 - blurts out answer before question completed
 - interrupts, intrudes, unable to wait turn

Identifying ADHD

- Fast growing disability category in children
- Prevalence estimates vary widely
- 9 X more frequent in boys
- Identified by parents with psychologists and physicians
 - Behavior rating scales (Connors')
 - Intelligence testing
- ADHD co-occurs with other disorders
 - Co-morbidity with other developmental disorders is very high
 - Tourettes or Tic disorders, LD, ODD

Intervention with ADHD

- **Pharmaceutical Intervention**
 - use of psychostimulants
 - Ritalin, Adderall, Dexedrine
- **Behavioral Intervention**
 - psychotherapy, cognitive-behavioral training, support groups, parent training programs
- **Educational Intervention**
 - preferential seating, modified curriculum

Specific Language Impairment (SLI)

- **Children who show significant deficits in language functioning ...in the absence of...**
- **a set of exclusionary criteria:**
 - no hearing impairment
 - no significant social-emotional impairment
 - no significant cognitive (nonverbal) deficits
 - no frank neurological differences

Problems with Defining SLI

- **Inclusionary criteria**
 - What is a significant deficit?
- **Exclusionary criteria**
 - ongoing middle ear problems
 - some, subtle social and emotional differences
 - intelligence testing limitations
 - neurological likely exist for all
- **Consider SLI when children have language deficits that are more deficient than cognitive skills or out of proportion with their IQ**

SLI Language Characteristics

- Pattern of development is “asynchronous”
- First sign is delay in first words
- Vocabulary or lexical development is delayed but similar to younger, typically developing children
 - SLI = LA peers < CA peers
- Syntax, morphology and phonology
 - SLI < LA peers < CA peers
- Pragmatics
 - LA peers < SLI = or < CA peers
