Mental Retardation

Background

- Treatment for MR began in 1850 with facilities focused on skills training and fostered caring, warm environment
  - In the US, 20 opened by 1890
- Directors of these schools founded the AAMR, helping to promote research, intervention, and social policy
- Multidisciplinary research has always played a huge role in the development of the field

Background

- Many "dark" episodes in history of MR in the U.S., including
  - Transformation into "warehouses" from home-type environment
  - Eugenics movements of the early 20th century
  - Sterilization laws during the 1920s
- Shift since 1970's to reintegration and "mainstreaming" of people with MR into "normal" life

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MR Features

- Three main criteria shared by every classification system

1. Significant limitations in intellectual functioning
2. Significant limitations in adaptive behavior
3. Limitations present before 18 years old

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DSM-IV-TR Diagnostic Criteria

- Significantly sub-average intellectual functioning
  - An IQ of approximately 70 or below on an individually administered IQ test
  - With standard error, scores from 65-75 usually considered "about 70"
  - In infants, a clinical judgment of significantly sub-average intellectual functioning

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DSM-IV-TR Diagnostic Criteria

- Concurrent deficits or impairments in present adaptive functioning in at least two of the following areas
  - Communication
  - Self-care
  - Home living
  - Social/interpersonal skills
  - Use of community resources
  - Self-direction
  - Functional academic skills
  - Work
  - Leisure
  - Health
  - Safety

DSM-IV-TR Diagnostic Criteria

- Onset before age 18
- Levels of MR based on IQ scores
  - Mild (317; 50-55 to approximately 70)
  - Moderate (318.0; 35-40 to 50-55)
  - Severe (318.1; 20-25 to 35-40)
  - Profound (318.2; below 20-25)
  - Severity Unspecified (319)

Developmental Course

- For most people, IQs are not stable when measured at 1 year old, but begin to show high stability starting at age 4
- For MR people, however, IQs are highly stable from early in life to adulthood
  - Least stable at mild MR range, most stable with IQs below 50

Developmental Course

- Down Syndrome MRs show IQ decreases over time
  - Not getting dumber, just developing at a much slower rate than peers
  - Show adaptive plateaus at 7-11 years old
- Fragile-X MRs have steady IQ and adaptive functioning increases until puberty
- Intensive, early intervention programs have been shown to boost IQ scores by 10-15 points

Epidemiology

- Population prevalence estimates of approximately 1-2%
  - Lack of standardized definitions and testing conditions hampers results
- 20-40% more males diagnosed with MR
  - May be due to sex-linked disorders such as Fragile-X

Epidemiology

- Prevalence
  - Standard view is 3%, but this is based on several assumptions that seem unacceptable.
  - Jane Mercer, a proponent of a 1% prevalence rate suggests the four assumptions are as follows:
    - IQ as the sole criterion
    - IQ stability
    - System issues (diagnostic practice)
    - Life expectancy (death rates)
  - The result of these four factors is that prevalence rates of MR are generally below 3%

Epidemiology

- Sex differences
  - More males than females- reasons why are unclear:
    - Some argue susceptibility of the male CNS
    - Differential registration by parents
    - Sex-linked disorders

Epidemiology – SES & Ethnicity

- MR more common among low SES and minority groups
- Relationship between mild MR and parental SES and its highly correlated measure, parental IQ
  - Classic study by Reed & Reed (1965)
- Association of mild MR and race is more complicated
  - Test bias, de-emphasis by schools of the importance of IQ in diagnostic decisions - the Larry P. v. Riles case in CA and its effects

Epidemiology

- Mild MR more prevalent among lower SES groups
- No differences in levels of severe or profound MR across SES
  - Points to environmental influence and factors

Developmental Theories

- Similar-sequences approach said that retarded and non-retarded children pass through same sequence, but slower
  - Strong research support for this occurring
- Similar-structures said that retarded children have the same "organization of intelligence" as non-retarded children
  - Supported for familial retardation, but organic MR do show numerous deficits

Developmental Theories

- Family studies have shifted from pathology-focused to how families deal with the stress and coping styles
- Double ABCX model
  - Crisis of raising child with MR (X) is a function of child's characteristics (A), family's resources (B), and family's perceptions (C)

Other Theories

- Eco-Cultural perspectives
  - What role does MR child fulfill in family system?
- Social role theory
  - MR child is merely fulfilling a societal role
- Behavior theories
  - MR people can be changed using behavior modification for adaptive behaviors, helping parents, and gaining vocational success

Types of MR

- Two-groups approach divides MR into:
  - Familial retardation is when MR is present without known organic causes
  - Organic MR is caused by genetic problems
    - e.g., Down Syndrome, Fragile-X, Williams Syndrome, Prader-Willi Syndrome

Two-Group Approach

	Organic	Cultural–familal
Definition	Individual shows a clear organic cause of mental retardation	Individual shows no obvious cause of retardation; sometimes another family member is also retarded
Characteristics	More prevalent at moderate, severe, and profound levels of retardation	More prevalent in mild mental retardation
	Equal or near-equal rates across all ethnic and SES levels	Higher rates within minority groups and low-SES groups
	More often associated with other physical disabilities	Few associated physical or medical disabilities
Causes[a]	Prenatal (genetic disorders, accidents *in utero*)	Polygenic (i.e., parents of low IQ)
	Perinatal (prematurity, anoxia)	Environmentally deprived
	Postnatal (head trauma, meningitis)	Undetected organic conditions

[a]Causes are suspected for cultural-familial mental retardation.

Types of Organic MR

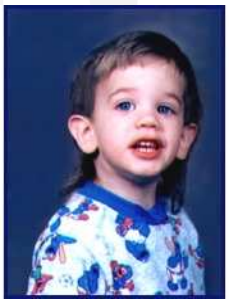
Down Syndrome

- Moderate MR; slowing rate of development; social strengths; weaknesses in grammar and speech

Types of Organic MR

Fragile-X

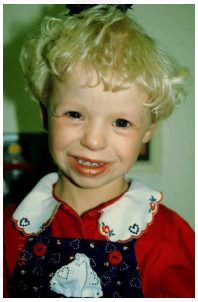
- Long narrow face, prominent ears, jaw, and forehead, high arched palate, flat feet, hyperextensible joints, enlarged testicles
- Moderate MR; more males than females; strength in Gestalt reasoning, weakness in sequential processing; autistic and ADHD like behaviors



Types of Organic MR

Williams Syndrome

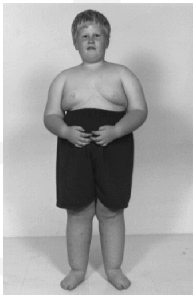
- Small upturned nose, long philtrum (upper lip length), wide mouth, full lips, small chin, and puffiness around the eyes
- Heart and blood vessel problems, feeding problems
- Usually have good verbal and language skills



Types of Organic MR

Prader-Willi Syndrome

- Typically causes low muscle tone, short stature, incomplete sexual development, mild MR, problem behaviors, and a chronic feeling of hunger that can lead to excessive eating and life-threatening obesity
- Note the inverted V-shaped upper lip, small hands, and truncal obesity



Co-morbidity

- Dual diagnosis rates vary from 10-50%
  - Much more common than in non-retarded peers
- MR people have been shown to have the majority of other DSM-IV disorders
  - Problems with applying criteria to persons with MR due to mental age appropriate behaviors
  - Some proposals for alternative checklists for problems like anxiety and depression

Co-morbidity

- Fragile-X's show high rates of hyperactive, attention, speech, and autistic-type problems
- Prader-Willi show failure to thrive initially, followed by hyperphagia and OCD-like food behaviors
  - Also show mood and psychotic disorders

Autism Spectrum Disorders

Historical Context

- Bleuler first used "autism" to describe schizophrenics who had lost touch with reality
- Kanner and Asperger described different types of autistic children in the early 1940's
  - Kanner saw language deficits, echolalia, pronoun reversal, social interaction problems
  - Asperger saw many of same problems, but without language deficits

Historical Context

- "Refrigerator mothers" and "freezer fathers" seen as responsible for autism
- Giving children MMR vaccinations was also thought to cause autism
- Both have been thoroughly discounted, but persist in the public imagination

Early Signs

- Group of deficits affecting social, affective, linguistic, behavioral, and cognitive development

TABLE 9.1. Early Symptoms of Autism

Social behavior	Typically develops	Behavior in children with autism[a]
Looking at faces	Birth	Less[b] at 12 months
Following person's gaze	6–9 months	Less at 18 months
Turning when name called	6–9 months	Less at 9 and 12 months
Showing objects to others	9–12 months	Less at 12 months
Pointing at interesting objects	9–12 months	Less at 12 and 18 months
Pointing to request	9–12 months	Not delayed at 18 months
Symbolic play	14 months	Absent at 18 months

[a]Data compiled from Baranek (1999), Baron-Cohen et al. (1996), and Osterling and Dawson (1994)—three studies comparing children with autism and children with typical development.
[b]"Less" indicates that this behavior was observed significantly less often in children with autism than in children with typical development at this chronological age.

DSM-IV-TR Criteria

- Delays in one of the following areas prior to age 3
  - Social interaction
  - Language as used in social communication
  - Symbolic or imaginative play
- Not better accounted for by Rett's Disorder or Childhood Disintegrative Disorder
- At least six items from (1), (2), and (3)

DSM-IV-TR Criteria

1) Qualitative impairment in social interaction, as manifested by at least two of the following
   - Marked impairment in the use of multiple nonverbal behaviors such as eye-to-eye gaze, facial expression, body postures, and gestures to regulate social interaction
   - Failure to develop peer relationships appropriate to developmental level
   - A lack of spontaneous seeking to share enjoyment, interests, or achievements with other people (e.g., by a lack of showing, bringing, or pointing out objects of interest)
   - A lack of spontaneous seeking to share enjoyment, interests, or achievements with other people (e.g., by a lack of showing, bringing, or pointing out objects of interest)

DSM-IV-TR Criteria

2) Qualitative impairments in communication as manifested by at least one of the following
   - Delay in, or total lack of, the development of spoken language (not accompanied by an attempt to compensate through alternative modes of communication such as gesture or mime)
   - In individuals with adequate speech, marked impairment in the ability to initiate or sustain a conversation with others
   - Stereotyped and repetitive use of language or idiosyncratic language
   - Lack of varied, spontaneous make-believe play or social imitative play appropriate to developmental level

DSM-IV-TR Criteria

3) Restricted repetitive and stereotyped patterns of behavior, interests, and activities, as manifested by at least one of the following
   - Encompassing preoccupation with one or more stereotyped and restricted patterns of interest that is abnormal either in intensity or focus
   - Apparently inflexible adherence to specific, nonfunctional routines or rituals
   - Stereotyped and repetitive motor mannerisms (e.g., hand or finger flapping or twisting, or complex whole-body movements)
   - Persistent preoccupation with parts of objects

Core Symptoms

- Deficits in social abilities seem to be due to impairments in understanding and responding to social information
- Secure attachment patterns are seen in 40-50% of autism children (65% in normal population)
- Impairments in imitative abilities, both immediate and deferred, that have been linked to expressive language deficits later

Core Symptoms

- Joint attention (referential looking) skills are impaired, compared to others of same intellectual level
- Decreased orientation to stimuli, especially social stimuli (speech, faces)
  - May be partially responsible for joint attention and imitation deficits
- Facial perception is also impaired
  - May focus on different area of face than normal (mouth instead of eyes)

Core Symptoms

- Show little preference for familiar versus non-familiar faces
  - Seem to use part of brain used by normals to process non-human objects
- Emotional expression and perception may be impaired
  - Mixed research supporting both
  - May be less impaired in older subjects due to compensatory skills

Core Symptoms

- Lack of and then long-term delay in symbolic play is one of the earliest signs of autism
- Delays in language precursors (joint attention, symbolic play) cause significant problems with language
  - About 50% have historically not developed language

Core Symptoms

- Deviant language is seen if it develops
  - Echolalia, abnormal prosody, pronoun reversal (or only using names later in life)
- Social / pragmatic language is most impaired
  - Use of irrelevant details, perseveration, inappropriate shifts in topic, ignoring social cues and intentions of other person in conversation
  - May be in part due to "mind-blindness"

Core Symptoms

- Semantic language also impaired
  - Acquisition of grammar and syntax, use of skills they acquire
  - Language comprehension is very concrete and literal
- Ritualistic, repetitive behaviors or interests seen very frequently
  - Lower-level (motor movements) and higher-level (routines and interests)

Related Symptoms

- Expression of frustration through self-injury (head banging, slapping, hair pulling)
  - May be more linked to MR
- Lack of need for sleep or sleep disturbances
  - Seems to improves over time, but more research needed
- Eating disturbances, mostly unusual food preferences and rituals
  - Continue into adulthood

Related Symptoms

- Abnormal fears of everyday objects
  - May be related to abnormal sensory responses
- Over and under reaction to sounds
- Hypersensitive to certain textures, but high pain tolerances

Lifetime & Development

- Symptoms can generally be seen before 12 months, but by 24 months at least
  - Important to distinguish between "current" and "lifetime" symptoms
- Have to consider the developmental appropriateness of behavior
  - Given high rates of comorbidity with MR, assess based on mental age, not chronological

Asperger's Disorder

- Relatively intact intellectual and language functioning accompanied by social impairments seen in autism
- Appropriate but unusually intense interests
- Increased motor clumsiness
- More "object-focused" than "people-focused"
- Little research to differentiate from high-functioning autism

Differential Diagnosis

- Difficult to differentiate between autism, Asperger's, and PDD-NOS
- Considered distinct diagnoses in DSM-IV-TR, but no good instruments to differentiate them
- Instead, have to rely on diagnostic criteria and clinical judgment

Differential Diagnosis

- Developmental Language Disorders and childhood-onset schizophrenia can resemble autism
- Rates of comorbidity vary greatly by disorder
  - 40-69% have MR
  - Widely varied rates of depression and anxiety
  - Tic behaviors more common than in population
  - Higher rates of seizure disorders

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Course & Prognosis

- Two typically seen courses
  - Symptom onset before 12 months
  - Regular development followed by loss of skills or regression before 3 years (primarily language skills)
- Very poor prognosis with no intervention
  - 75% will not live independently, even with early interventions
  - Higher IQs and earlier development of social communication skills are related to better prognosis

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Prevalence

- Population estimates range from 16-62 per 10,000 across all PDDs
- 3-4:1 male to female ratio
  - Females tend to have lower intellectual functioning and more severe symptoms
  - Higher functioning females, however, show less severe symptoms than matched males

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Early Detection & Intervention

- Both are crucial for long-term prognosis
  - Most parents report symptoms before 12 months, but average diagnosis is at 4 years
- Instruments for early screening are available, but have some limitations
- Lack of translation from university-based intervention programs to school-based has hampered early intervention programs

Early Intervention

- Effective programs have these in common:
  - Curriculum focusing on attention / compliance, motor imitation, communication, appropriate toy use, and social skills
  - High structured environments with low student-to-staff ratio
  - Strategies for generalizing learned skills
  - A functional approach to problem behaviors
  - Focus on skills used in transitioning from intervention program to preschool
  - High levels of family involvement