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DESCRIPTION

Mental retardation has been recognized through history as an abnormality. In 1799, French physician Marc Itard worked with a child, known as the "wild boy of Aveyron," with the goal of adapting him to society. Although Dr. Itard failed to cure the boy, who he named "Victor," it was one of the first attempts to treat an individual with mental retardation (Lane, 1976). During the summer of 1846, Dr. Buckminster Brown visited the Hospital for the Cure and Education of Cretins on the Abendberg, canton of Berne, Switzerland. According to Dr. Brown, cretins and idiots were isolated from the population of the institution, which was located on the Swiss Alps, with the hope that they could be treated using treatments such as gymnastic exercise. At that time, individuals with mental retardation were known as innocents, simpletons, cretins, or idiots (Brown, 1847). In fact, derogatory terms such as idiot, fool, moron, and imbecile were applied in diagnostic and legal terminology to refer to individuals with mental retardation (Volkmar & Dykens, 2002).

Today, individuals with mental retardation are still associated with their deficits such as lack of intelligence (IQ test scores of 70 or less) or lack of adaptive social skills. However, there has been a change in the focus on mental retardation from what a person cannot do to what a person can do. In addition, there is greater emphasis on inclusion rather than exclusion. With treatment and assistance, some individuals with mental retardation have overcome their condition in order to function in society with relative normality. For instance, consider the case of Christopher Burke (1995) who suffers from Down's syndrome (a form of mental retardation), yet he became an actor who starred in the TV show Life Goes On, despite his condition. The purpose of this chapter is to present a detailed coverage of the essential neurological, neuropsychological, and other aspects of mental retardation.

NEUROPATHOLOGY/PATHOPHYSIOLOGY

In many ways, it is futile to attempt to discuss the neuropathological underpinnings of mental retardation. In essence, it is best conceptualized as a symptom as opposed to a disorder in and of itself. Mental retardation has been seen as a consequence of genetic/chromosomal abnormalities, disrupted neurological development, and perinatal factors. Genetic/chromosomal abnormalities constitute the most common basis for mental retardation. Trisomy 21 (i.e., Down's syndrome), Trisomy 18 (Edward's syndrome), Trisomy 13 (Patau's syndrome), Trisomy 8, Trisomy 5 (Cri-du-chat syndrome), Fragile X syndrome, Klinefelter's syndrome, Turner's syndrome, and phenylketonuria are just a few of the genetic chromosomal disorders in which mental retardation commonly presents as a symptom.

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All cephalic disorders, when children survive, have shown a substantial risk of mental retardation in addition to an array of other neurological symptoms. These include colpocephaly, holoprosencephaly, hydranencephaly, lissencephaly, porencephaly, and schizencephaly. This list obviously does not include cephalic presentations such as anencephaly as death often occurs prior to or immediately following birth. For some of these more severe forms, infants may live for up to a year or slightly more, but this is quite rare.

Perinatal factors linked with increased risk of mental retardation, among other neurological defects, include infectious processes, drug or toxic substance exposure, and trauma. Infectious processes include cytomegalovirus, herpes simplex, and rubella. Drug and toxic substance exposure include alcohol, cocaine, and the array of other illicit substances. Finally, maternal trauma can include irradiation, maternal suffocation, or physical injury among other things.

By all means, the presentations reported are not all inclusive. Rather, the emphasis is on the broad correlates of genetic/chromosomal abnormalities, infectious processes, and prenatal trauma. The interested reader is encouraged to read the literature on these individual presentations in regard to their pathological link to mental retardation. Many of which are included in this text. Beyond these, there is literature demonstrating links between gestational time, birth weight, and neonatal and early childhood environment and intellectual capacities.

## NEUROPSYCHOLOGICAL/CLINICAL PRESENTATION

The clinical presentation of mental retardation remains consistent with its diagnostic criteria. Mental retardation is marked by significant limitations in both intellectual functioning and adaptive functioning, occurring prior to age 18. In regard to intellectual functioning, this corresponds with abilities two standard deviations or more below the population mean. Given mental retardation itself is best conceptualized as a symptom, it commonly coincides with other neuropsychological, academic, behavioral, emotional, and/or social deficits that may be specific to an underlying disease or disorder but not specific to mental retardation.

## DIAGNOSIS

There are multiple definitions of mental retardation, which share common points. The American Association on Mental Retardation (Luckasson et al., 2002) provides the following definition:

*Mental retardation is a disability characterized by significant limitations both in intellectual functioning and in adaptive behavior as expressed in conceptual, social, practical adaptive skills. This disability originates before age 18. (p. 8)*

The *Diagnostic and Statistical Manual of Mental Disorders*, fourth edition, text revision (*DSM-IV-TR*) (American Psychiatric Association [APA], 2000) defines mental retardation similarly. It states that the disorder is characterized by intellectual functioning that is significantly subaverage (that is, an IQ of approximately 70 or below) with onset before 18 years of age and concurrent deficits or impairments in adaptive functioning. The *DSM-IV-TR* provides separate codes for Mild, Moderate, Severe, and Profound Mental Retardation and for Mental Retardation, Severity Unspecified (APA, 2000).

Both definitions focus on deficits on intellectual and adaptive functioning and they agree that the mental retardation symptoms must appear before the age of 18. Through time, the primary tool to assess mental retardation, intellectual deficits, has been the use of IQ (intelligence quotient) tests such as the Wechsler Intelligence Scales for Children, 3rd Edition (WISC-III; Wechsler, 1991) Stanford-Binet Intelligence Scales, 4th Edition (Thorndike, Hagen, & Sattler, 1986a, 1986b), and Kaufman Assessment Battery for Children (K-ABC; Kaufman & Kaufman, 1983). Recently, adaptive skills have been incorporated in the definition of mental retardation; thus, adaptive tests are used in the assessment of mental retardation including the Vineland Adaptive Behavior Scales (Sparrow, Balla, & Cicchetti, 1984) and the Adaptive Behavior Assessment System (Harrison & Oakland, 2000). Today, the combination of intelligence and social adaptive measures of functioning are applied in the diagnosis of mental retardation.

Neuropsychological testing has been used to distinguish among individuals with mental retardation that might have another disorder. For instance, Palmer (2006) compared performances in neuropsychological testing among 10 individuals with comorbid mental retardation and dementia with 12 individuals with only mental retardation. The comparison was based on measures of attention, executive functions, language, dementia screening, as well as memory and learning. Palmer's results indicated that there are significant differences in neurocognitive measures between the two groups. Although participants from the dementia group showed more severe defects in memory and learning, agnosia, semantic verbal fluency as well as attention and executive functions, both groups did show significant neuropsychological deficits.

In another study, Shultz et al. (2004) evaluated screening tools for dementia in 38 older adults with mental retardation. The instruments used included the dementia scale for Down's syndrome (Gedy, 1995), the Dementia Questionnaire for Mentally Retarded Persons, and the Reiss Screen for Maladaptive Behavior. It was found that the dementia scale for Down's syndrome and the Dementia Questionnaire were accurate in assessing dementia. However, a slight difference in effectiveness from the Dementia Questionnaire might be because mentally retarded individuals show less cognitive symptoms of dementia, which puts the dementia scale for Down's syndrome at a disadvantage against the Dementia Questionnaire.

Neuropsychological tests have also been used in the assessment of cognitive deficits in individuals with mental retardation. Vicari, Albertini, and Caltagirone (1992) identified cognitive profiles of 32 adolescents with mental retardation using neuropsychological assessment, which included measures of verbal functions, memory, visuoconstructive and visuospatial skills. Results from Vicari et al. indicated that the neuropsychological test battery distinguished cognitive profiles of participants

GLOSSARY Mental retardation refers to substantial limitations in a person's level of present functioning. It is characterized by...

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with mental retardation. Furthermore, findings suggest that (a) since cognitive deficits of mental retardation vary depending on the skills impaired and the severity of the deficits, then mental retardation impairment is heterogeneous across all skills; and (b) a set of neuropsychological assessments exploring single cognitive functions is necessary to accurately identify and understand cognitive profiles from individuals with mental retardation.

Assessment of mental retardation has been a complicated issue for educators, psychologists, and mental health professionals particularly because the cognitive and language skills of individuals with mental retardation make the assessment difficult (Smith, 2005). For instance, Flynn (2000) suggested a change to adaptive behavior tests instead of intelligence tests because of the lack of justification of an IQ criterion as related with impaired adaptive behavior. Similarly, Graue et al. (2007) compared Wechsler Adult Intelligence Scale, 3rd Edition (WAIS III) scores among 26 participants with mild mental retardation and 25 community volunteers who feigned mental retardation. Consistent with criticisms toward intelligence measures, Graue et al. found no significant difference between IQ scores, thus the scores from WAIS III did not distinguish individuals who feigned from individuals with genuine mental retardation.

Despite the social and professional criticisms, intelligence measures are supported as the primary tool in the diagnosis of mental retardation. Umphress (2008) compared IQ test scores from the Reynolds Intellectual Assessment Scales (RIAS; Reynolds & Kamphaus, 1998) and the WAIS III to investigate whether results are comparable when measures are given by a same tester on the same day. Umphress found similar IQ scores in general, but significant difference between RIAS and WAIS III scores less than 80 was present. Similarly, Watkins and Campbell (1992) found the WAIS-R to be stable and reliable in a sample of 50 adults with mental retardation during 2–5 years, which was consistent with results from Rosen, Stallings, Floor, and Nowakowska (1968).

Neuropsychological testing has also been applied to identify individuals who might be feigning mental retardation. For example, Marshall and Happe (2007) studied which neuropsychological tests of effort and motivation would be appropriate if feigning of cognitive deficits might be present. They administered a comprehensive neuropsychological battery to 100 mentally retarded participants including the WAIS III (Wechsler, 1997), the Wechsler Memory Scale (WMS) III (Wechsler, 1997), the forced choice recognition portion of the California Verbal Learning Test II (CVLT-II; Delis, Kramer, Kaplan, & Ober, 2000), and Vocabulary Digit Span Test from the WAIS III Digit Span Test (Mittenberg, Theroux-Fichera, Zilinski, & Heibronner, 1995). It was found by Marshall and Happe that the scores from the forced portion of the CVLT-II, WMS III, and V-DS difference score are appropriate to distinguish individuals who might be feigning mental retardation.

## TREATMENT

John F. Kennedy's administration proposed a new approach toward mental retardation in the 1960s with a focus on prevention, treatment, and rehabilitation (Kennedy, 1963). Such a policy marked a shift in terms of the importance of mental retardation as a relevant issue in society. Thus, an emphasis has been placed not only on the treatment of mental retardation as a disorder but also efforts have been directed toward finding ways to promote increased social engagement of individuals with mental retardation. Dykens (2006) supported the application of positive psychology on mental retardation with a focus on positive mental states such as happiness, contentment, hope, engagement, and strengths. Similarly, Favell, Realon, and Sutton (1996) examined individuals with severe and profound mental retardation from two intermediate care facilities. According to Favell et al. facial expressions could be applied as a practical method of measuring happiness in individuals with mental retardation.

Other treatments of mental retardation have the objective of integrating individuals with mental retardation to relatively normal life. For instance, LeBlanc, Hagopian, and Maglieri (2000) found that token economy with response cost procedure is effective in eliminating inappropriate social interaction, verbal aggression, and inappropriate sexual behavior in 26-year-old mentally retarded males. In another study, it was found that interactions between peer buddies improved communication behaviors, reciprocity of interactions, and enhanced range of communication behaviors in five high school students with mental retardation (Hughes et al., 2002).

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