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Neuropsychological profiles of persons with mental retardation and dementia

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Abstract

This study examined the use of neuropsychological tests to assist in the differential diagnosis of dementia among persons with mental retardation. The author compared performances of persons with mental retardation and dementia ($n = 10$) to persons with mental retardation without dementia ($n = 12$). Participants were matched by IQ (mild or moderate mental retardation), age, presence of Down syndrome, and gender. In addition, all participants in the dementia group had corroborative medical tests (i.e., imaging, EEG, or high tau low AB42 protein testing) consistent with diagnosis of dementia. Test performance was compared on measures of attention and executive functions, language, memory and learning, and a dementia screening. Results from MANOVAs and nonparametric tests revealed significantly lower performance for persons with mental retardation and dementia in all areas assessed. Cut-off scores were also developed for the sample in order to maximize sensitivity and specificity for the test battery. Despite the small sample size, these findings suggest that there are significant measurable differences in several neurocognitive domains between the two groups.

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1. Introduction

Due to recent medical advances and improved quality of life, the general life span of persons with mental retardation is increasing. As the life spans of persons with mental retardation increase, so will the number of individuals who will develop Alzheimer's disease or other forms of dementia. Within the last decade, publications have been developed to assist with diagnosis (Aylward, Burt, Thorpe, Lai, & Dalton, 1995), and treatment (Janicki, Heller, Seltzer, & Hogg, 1995) of dementia (particularly Alzheimer's disease) in persons with mental retardation. Burt and Aylward (2000) have proposed a test battery for purposes of assessing neuropsychological function in persons with intellectual disabilities. There is substantial literature in the field of neuropsychology addressing cognitive decline in persons with dementia. Yet, there is less research that examines neuropsychological differences between persons with intellectual disabilities who have dementia versus individuals without dementia in areas other than memory and adaptive functions.

Several screening measures have been developed over the years in order to address the need for identification of mental deterioration in persons with mental retardation. For example, Evenhuis (1992) developed the Dementia Questionnaire for Mentally Retarded Persons (DMR), which reportedly demonstrated good specificity. Gedy (1995) developed the Dementia Scale for Down Syndrome (DSMS), which reported good specificity and, depending on the study, moderate to good sensitivity. A recent study comparing these two instruments (Shultz et al., 2004), found that both scales were good in discriminating between groups. A number of other screening tools have been used to assist with identifying dementia including checklists (Visser et al., 1997) and mental status examinations (Folstein, Folstein, & McHugh, 1975; Haxby, 1989). Strydom and Hassiotis (2003) conducted a literature review pertaining to screening instruments for persons with dementia and intellectual disability. According to their review, the use of any of the instruments as single tools for screening of dementia should be interpreted with caution.

According to the *Diagnostic and Statistical Manual of Mental Disorders*, Fourth Edition, Text Revision (DSM-IV-TR; APA, 2000), dementia involves multiple cognitive deficits that include memory impairment. Cognitive disturbances including agnosia (failure to recognize or identify objects despite intact sensory function), apraxia (impaired ability to carry out motor activities despite intact motor function), aphasia (language disturbance), and executive functions (patient's ability to organize, plan, and carry out activities) may be present. A gradual onset and continuing cognitive decline characterize dementia of the Alzheimer's type. Vascular dementia is characterized by a stepwise regression. Focal signs/symptoms are usually reported, or presence of cerebrovascular disease is noted. Assessment of memory alone is not sufficient for evaluation of dementia. A battery of tests including free memory recall, learning, recognition memory, verbal fluency (i.e., generative-naming), confrontation naming, and praxis have been found helpful for staging dementia severity or tracking its progression (Bondi, Salmon, & Kaszniak, 1996; Kaszniak, Wilson, Fox, & Stebbins, 1986; Palmer, 2001; Welsh, Butters, Hughes, Mohs, & Heyman, 1992).

The literature is quite sparse as it relates to neuropsychological assessment for persons with mental retardation. Matthews (1974) was probably the first individual who attempted to standardize a neuropsychological test battery for persons with mental retardation. He

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