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REHABILITATION OF NEUROMUSCULAR DISEASE 1047-9651/98 $8.00 + .00

COGNITIVE, PSYCHOSOCIAL, AND EDUCATIONAL ISSUES IN NEUROMUSCULAR DISEASE

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The neuromuscular diseases (NMDs) represent a diverse group of disorders with onsets ranging from birth to adulthood and physical deficits from mild to severe. Individuals with NMD also vary significantly in terms of cognitive abilities, ranging from no appreciable deficits to severe impairment in functioning. This article will review the available research on cognitive function of individuals with Duchenne muscular dystrophy (DMD), myotonic dystrophy (MD), spinal muscular atrophy (SMA), Becker's muscular dystrophy (BMD), limb-girdle syndrome (LGS), facioscapulohumeral dystrophy (FSHD), hereditary motor sensory neuropathy (HMSN), and hereditary spinal cerebellar ataxia (HSCA) and the implications for psychosocial and educational function. There is substantial research on DMD and MD, but the other disease groups have received much less attention. The article will reflect this differential emphasis in the research literature.

METHODOLOGIC ISSUES

Before discussing reported findings concerning cognitive functioning and the various neuromuscular diseases, some general comments on research methodology—specifically, problems in assessment, statistical analysis, and design—are in order. Such a discussion may help readers understand methodologic flaws that result in conflicting results. Regard-

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ing assessment, most studies have relied largely on one of the three age-appropriate Wechsler intelligence scales (WIS) to assess cognitive function. These test batteries have well established reliability and validity when used appropriately. However, a review of the NMD literature raises several concerns regarding the use and interpretation of data produced by these batteries. First, several investigators have used the Full Scale Verbal and Performance scores obtained from the different age-appropriate versions of the WIS as if they were equivalent and have simply added them together for their analyses.[44, 55, 78] This practice ignores the differences that exist between the child and adult batteries. Furthermore, the average IQ scores of the revised versions of both the Wechsler Intelligence Scales for Children and the Wechsler Adult Intelligence Scales are approximately 5 to 6 and 7 to 8 points lower, respectively, than their original counterparts,[40, 45] making comparison of early and later studies questionable unless these differences are taken into account.

The second assessment problem noted with the use of the Wechsler Intelligence Scales concerns the emphasis placed on analyzing verbal-performance (VIQ-PIQ) differences. Both scales comprise subtests that entail many different cognitive abilities.[45] The two factor VIQ-PIQ interpretive approach to the Wechsler batteries often does not render the best representation of the data. This is particularly true for children and individuals with impaired cognition, where data tend to be more accurately summarized by a three-factor model composed of a verbal comprehension factor, a perceptual organizational factor, and a third factor often called the freedom from distractibility factor, typically reflecting attention/concentration abilities.[40, 45] Further, much emphasis has been placed on VIQ-PIQ differences that, while statistically significant, may have limited clinical meaning. Added to this is the question of the appropriateness of using the performance subtests when assessing individuals with orthopedic handicaps. This is especially true for children, because the contribution of speed to the final score greatly increases with age. In general, the performance scale renders an underestimate of nonverbal ability for orthopedically handicapped children beyond the age of 9.[40]

Regarding statistical matters, much of the research in NMD has relied on multiple independent tests of significance of mean differences (e.g., t-tests). Many studies have performed up to 50 or more comparisons, utilizing the conventional probability level of $p < .05$ for each comparison. Such a procedure may result in finding statistically significant differences by chance alone. The practice of conducting multiple independent comparisons ignores the fact that the actual alpha level of the entire experiment rapidly increases beyond $p = .05$. For example, a study containing ten independent t-tests, each evaluated at the .05 level, would result in an approximate experimentwise {alpha} $\leq .40$. Sheerly by chance, up to 4 of these 10 comparisons would be expected to meet the $p \leq .05$ level and be declared significant when, in fact, this may not be the case. Conducting multiple independent tests of significance also

ignores the multifactorial relationships that may exist among the measures, possibly obscuring important aspects of the phenomena under investigation. Clearly, these statistical issues can seriously affect a study's findings and, ultimately, the literature base as a whole.

Small sample size, a characteristic of NMD research resulting from the low base rates, makes obtaining a representative sample of the disease population of interest quite difficult and often can lead to study findings that are highly sample-specific. Complicating the picture even further is the emerging awareness of the tremendous phenotypic variability that exists within these diseases and the possible presence of subtypes. The large heterogeneity of variance, combined with the small sample sizes and the associated restriction in both the statistical power and generalizability of the results, has resulted in a research literature that is highly variable and not infrequently contradictory.

Regarding research design, a review of the literature indicates the need for studies utilizing samples of sufficient size, with clinical controls, assessing a broad range of cognitive, psychologic and other variables, to enable investigators to better identify the multifactorial influences on the cognitive performance of individuals with various NMDs. Planned longitudinal studies, as opposed to the use of cross-sectional data, also are needed to address the question of possible deterioration or improvement in cognitive functioning. Although some longitudinal studies have been reported, few cite data obtained from objective measures subjected to formal statistical analysis. Further, much of the objective data of these studies are derived from smaller subsets of larger samples, introducing the possibility of significant sampling bias. The foregoing discussion should be kept in mind when reading subsequent sections of this article, because methodologic and statistical problems may contribute to the frequently conflicting results.

REVIEW OF THE LITERATURE: COGNITIVE FUNCTIONING

Duchenne Muscular Dystrophy

Since 1872, when Duchenne first described the disorder, it has been commonly accepted that DMD is associated with diminished intellectual capacity.[23] Although early research focused essentially on establishing the presence and extent of intellectual impairment, recent research has sought to provide a more detailed picture of the nature, course, and cause of the cognitive impairments and their relationship to psychosocial and educational concerns.

Extent of Cognitive Impairment

The earliest writers in the field, Duchenne, Gowers, and Erb, based upon clinical observations, recognized a relationship between intellec-