Evidence for early impairment of verbal intelligence in Duchenne muscular dystrophy

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Marsh, G. G., and Munsat, T. L. (1974). *Archives of Disease in Childhood*, **49**, 118. **Evidence for early impairment of verbal intelligence in Duchenne muscular dystrophy.** Intelligence tests were carried out on 34 boys with muscular dystrophy. The 16 who were mildly physically disabled scored significantly lower on the verbal scale (mean 85.3) of the Wechsler Intelligence Scale for Children (WISC) than on the performance scale (mean 97.6). The 18 who were moderately or markedly physically disabled showed no significant difference between their mean verbal (87.6) and performance (89.7) scores. The results are interpreted as suggesting an early and nonprogressive impairment of verbal intelligence in Duchenne muscular dystrophy.

The occurrence of intellectual impairment above the expected frequency in children with Duchenne muscular dystrophy (DMD) has been amply documented in published reports (Dubowitz and Crome, 1969; Prosser, Murphy, and Thompson, 1969; Zellweger and Niedermeyer, 1965). There has been less agreement about the nature of this intellectual impairment, however. Though DMD, a progressive neuromuscular disease, would be expected to impair scores on tests requiring use of hands, Zellweger and Niedermeyer (1965) and Zellweger and Hanson (1967) have observed that dystrophic children often obtain performance IQ scores that are higher than their verbal IQ scores. The majority of boys in these two studies were relatively unimpaired in their motor functioning, with only a small number of them in wheelchairs. In studying dystrophic children confined to wheelchairs, Sherwin and McCully (1961) anticipated greatly lowered performance IQ scores but found very little discrepancy between the verbal and performance IQ scores. 4 of the boys, despite considerable motor impairment, obtained higher performance IQs than verbal IQs.

Prosser *et al.* (1969) analysed verbal and performance IQ scores for a group of 39 dystrophics, ranging in age from 5 to 18. No significant

difference was found between the verbal and performance IQ scores. The apparent contradiction between Prosser *et al.*'s findings and the observations mentioned above may be related to differences in the severity of motor disability in their respective samples. Severe motor disability would be expected to lower scores on performance subtests and, in the case of DMD, should be more noticeable in the older age range as the disease progresses and the child becomes less agile in using his hands. Some support for this appears in Zellweger and Niedermeyer's (1965) study of DMD children, aged 3 to 16, who were relatively free from motor impairments; 16 of their 23 patients scored higher on the performance subtests. In the Sherwin and McCully study (1961) in which all children, aged 10 to 15 years, were wheelchair dependent but able to attend regular public schools, the motor disability was greater and only 4 of the 15 dystrophic boys obtained performance IQ scores higher than verbal scores. Prosser *et al.* do not report the effect of disease severity on the verbal and performance IQ scores separately. However, because their patients ranged in age up to 18 years, there is a strong possibility that many of the older dystrophics were more severely impaired in the use of their hands, which may have suppressed scores on the performance subtests more than in the other studies.

In no published study have the dystrophic

children's verbal and performance intelligence test results been based exclusively on the complete Wechsler Intelligence Scale for Children (WISC) (Wechsler, 1949) and examined in light of their disability ratings. In most studies two and even three different intelligence tests have been administered in order to cover the range of ages in the samples (Allen and Rodgin, 1960; Kozicka, Prot, and Wasilewski, 1971; Prosser *et al.*, 1969; Sherwin and McCully, 1961; Zellweger and Hanson, 1967; and Zellweger and Niedermeyer, 1965). Contrary to the assumption followed by those researchers that intelligence tests such as the WISC, the Wechsler Adult Intelligence Scale (WAIS, Wechsler, 1955), and the Stanford-Binet Intelligence Scale (S-B, Terman and Merrill, 1937) are comparable in estimating IQ means and can be used interchangeably, Hannon and Kicklighter (1970) reported that the WAIS produces a significantly higher IQ score of approximately 7 IQ points over the WISC for 15- to 16-year-olds in the less-than-average intelligence range. The WISC was noted to obtain higher scores for the average and above-average adolescents than the WAIS. The S-B has been noted to give higher IQ scores to children of normal mental abilities at the younger age levels (Barclay and Carolan, 1966) than the WISC. Thus, by treating the scores from these tests as equivalent, real differences may have been obscured. For example, because the WISC age range is from 5 years to 15 years 11 months, in order to obtain IQ scores for boys older than 15 years 11 months, the WAIS has to be administered. In Prosser *et al.*'s study the age range extended to 18 years. The IQ results on their older dystrophic boys which were based on the WAIS possibly contain test-related inflated IQ values which could obscure actual lowering of IQ scores.

The present study was undertaken to investigate further the effect of severity of Duchenne muscular dystrophy on verbal and performance IQ, and to define in more detail the nature of the intellectual deficit in DMD.

Methods and materials

Subjects. 40 dystrophic boys with a firm diagnosis of DMD established by experienced clinicians were selected for the study. The diagnosis of DMD was established on each child on the basis of at least four of the following criteria: (1) typical clinical signs of Duchenne muscular dystrophy; (2) muscle biopsy consistent with DMD; (3) myopathic electromyogram; (4) raised serum enzymes, especially creatine phosphokinase; (5) consistent pedigree.

The first 40 available dystrophic boys who met the diagnostic, age, and commuting requirements were asked

to participate. Thus, the sample was not strictly random. 6 boys could not be included in the results; 4 were hyperactive and unmanageable in the testing situation; 1 child refused the tests; and one mother reported her child untestable because of mental retardation.

The 34 boys who completed the tests were categorized into two groups based on their physical independence ratings. These ratings were based on 15 daily activities, many of which describe personal grooming, feeding, dressing, writing, speaking, and ambulation. The 5 categories were (1) bedridden, (2) wheelchair with assistance, (3) wheelchair independent, (4) ambulatory limited, and (5) ambulatory unlimited. Categories 4 and 5 were combined to represent the mildly impaired group (group I). Categories 2 and 3 were combined to designate the moderately to markedly impaired group (group II). No patient was in category 1.

Test. The WISC (Wechsler, 1949) was selected as the test instrument because it offers a verbal IQ based on verbal answers and a performance IQ score based on subtests requiring use of the hands. Another advantage of the WISC is that it converts raw subtest data into mean scaled scores so that performance on the subtests can be compared. The scaled scores have been computed to provide for each subtest a mean scaled score of 10 and SD of 3.

Results

Descriptive data. Table I gives the characteristics of the 34 dystrophic boys. 26 of the 34 boys attended special schools for handicapped children; the remaining 8 were attending regular schools. The descriptive data in Table I show that the younger the child the less the physical disability; the older the child the greater the chance he will be wheelchair dependent. This is consistent with the uniform progress of the disease. In the sample of 34 boys, the crucial age appears to be age 9 when the children move into the more disabled category, group II.

In comparing the IQ test results of groups I and II with those of the WISC normative group of 1100 normal boys between the ages of 5 and 15 (Seashore, Wesman, and Doppelt, 1950), it is evident that the two dystrophic groups are considerably lower in IQ scores (Table II). Another way to look at the obtained IQ scores is to compare them with the WISC normative group by converting the obtained IQ score to the corresponding centile rank of the normative group. Group I's verbal IQ of 85.3 (dull normal range) is between the 10th and 20th centiles of the WISC normative group; their performance IQ of 97.6 (average range) is between the 40th and 50th centiles; and full scale IQ of 90.4 (average range) corresponds to the 25th centile.

TABLE I
Descriptive characteristics of 34 dystrophic boys

Case no.	Age (yr) (mth)		Age at diagnosis (yr)	Independence rating*	Appliance†	School/class‡	WISC		
							Verbal IQ	Performance IQ	Full scale IQ
1	5	8	5	5	0	R/K	80	87	82
2	6	7	5	5	0	R/K	65	80	70
3	6	10	5	5	0	R/K	75	79	75
4	7	1	2	4	0	Sp/1	85	113	98
5	7	4	3	5	0	Sp/2	92	108	100
6	7	10	2	4	0	Sp/2	96	110	103
7	7	11	5	4	0	Sp/2 & 3	87	107	96
8	8		4	5	0	R/2	104	128	117
9	8	2	4	4	0	Sp/2	76	83	77
10	8	2	5	4	0	R/2	86	110	97
11	8	3	2	4	0	Sp/2	67	67	64
12	8	5	4	5	0	Sp/3	66	80	70
13	8	7	4½	4	0	Sp/Ungraded	82	87	83
14	8	9	8	5	0	Sp/3	87	101	93
15	8	9	4	5	0	R/4	111	93	103
16	9		4	2	1	Sp/3	65	78	68
17	9		5	2	1	Sp/Ungraded	79	86	80
18	9	2	4½	2	1	Sp/3	90	100	94
19	9	5	3	3	1	Sp/2	69	58	60
20	9	6	5	2	1 & 2	Sp/4	80	89	83
21	9	9	5	3	1	Sp/2	100	100	100
22	10		5	3	1	Sp/4	100	94	97
23	10	1	3	3	1	Sp/4	109	108	109
24	10	5	3	4	0	Sp/4	106	128	118
25	10	6	5	2	1	Sp/5	92	96	93
26	10	11	4	2	1	Sp/5	91	96	93
27	11		2½	2	1	R/5	84	89	85
28	11	2	7	3	1	Sp/5	87	99	92
29	11	3	4	3	1	Sp/5	99	76	87
30	11	5	7	3	1	Sp/6	99	104	101
31	11	11	3	2	1	R/7	101	115	109
32	12	7	2½	2	1	Sp/6	85	86	84
33	13	2	5	2	1	Sp/Ungraded	65	65	63
34	15	6	5	2	1	Sp/9	81	75	76

*See categories 1-5 under Methods and Materials.

†0, none; 1, wheelchair; 2, leg braces.

‡R, regular school; Sp, special school for the handicapped; K, kindergarten; 1-9, 1st-9th grades.

WISC, Wechsler Intelligence Scale for Children.

TABLE II
Mean age, independence ratings, and mean WISC scores for groups I and II and WISC normative group

	Group I (no. = 16 boys) (mildly impaired)			Group II (no. = 18 boys) (moderately to markedly impaired)			WISC normative group (no. = 1100 boys, age 5 to 15; Seashore <i>et al.</i> , 1950)	
	Mean	SD	t	Mean	SD	t	Mean	SD
Age	7.92	1.08		10.85	1.67			
Independence rating*	4.50	0.51		2.38	0.50			
Verbal IQ	85.31	14.03		87.55	12.84		101.2	15.3
Performance IQ	97.56	18.18		89.66	14.87		100.3	15.6
Verbal-Performance Score†	-12.25	11.40	4.29‡	-2.11	9.31	0.96		
Full scale IQ	90.37	16.51		87.44	14.22		100.8	15.6

*See Methods and Materials.

†Refers to verbal score minus the performance score.

‡P < 0.001 level.

Group II's verbal IQ of 87.5 (dull normal range) corresponds to the 20th centile of the WISC normative group; their performance IQ of 89.7 (upper range of dull normal) corresponds to the 25th centile; and their full scale IQ of 87.4 corresponds to the 20th centile.

Evidence for an early and nonprogressive verbal deficit. The dystrophic boys with mild physical disabilities (group I) scored significantly lower on their verbal IQ (85.3) than on their performance IQ (97.6) ($t = 4.29$, $P < 0.001$) (Table II). Since our patients were all males, it is possible that this finding is due to a sex difference based on the assumption that boys generally obtain lower verbal IQs than performance IQs. However, Seashore *et al.* (1950) have shown that boys in the normative sample obtained comparable means for both verbal and performance IQs (101.2 for verbal and 100.3 for performance IQs). No significant difference was observed between verbal (87.6) and performance (89.7) for the group II patients ($P = 0.96$, $P > 0.2$).

Nature of intellectual deficit. Table III presents the mean WISC subtest scores for groups I and II. Though none of the subtest comparisons between the two groups reached statistical significance, the coding task, a performance subtest that requires the patient to write symbols for numbers, came close to being significant ($t = 1.85$, $0.1 < P < 0.05$).

TABLE III
Mean WISC subtest scores for groups I and II

	Group I (no. = 16)	Group II (no. = 18)	Difference in mean scores
<i>Verbal subtests</i>			
Information	7.81	8.11	+0.30
Comprehension	7.68	6.88	-0.80
Arithmetic	7.06	6.88	-0.18
Similarities	7.87	8.66	+0.79
Vocabulary	7.87	9.50	+1.63
(Digit span)	(7.31)	(7.05)	-0.26
<i>Performance subtests</i>			
Picture completion	9.37	7.88	-1.49
Picture arrangement	9.06	9.16	+0.10
Block design	10.87	9.55	-1.32
Object assembly	10.31	9.27	-1.04
Coding	8.80	6.72	-2.08
Verbal IQ	85.31	87.55	+2.24
Performance IQ	97.56	89.66	-7.90
Full Scale IQ	90.37	87.44	-2.93

Note: Digit span scores were not included in computing individual WISC IQs in the present study.

Group I's lower scores on all the verbal subtests in comparison with their performance subtests appear to be related to a mild impairment in verbally mediated cognitive functioning which is apparent in the early stages of the disease when the patient shows little disability in the use of his hands. A low verbal IQ score with a higher performance IQ score has been associated with reading disability (Clements and Peters, 1962; Ackerman, Peters, and Dykman, 1971) and with impaired reading, spelling, and arithmetic scores (Rourke, Young, and Flewelling, 1971). The verbal deficit in DMD appears to be a relatively generalized one that interferes with verbal subtests measuring fund of general information, practical knowledge and judgement, verbal abstract thinking, expressive vocabulary, mentally calculated arithmetic problems, and rote learning. Partial corroborative evidence of a generalized intellectual deficit may be inferred from the fact that 23 of the 34 dystrophics were placed in school classes below those of their age-related peers (Table I).

Group II, the older and more physically disabled dystrophics, showed a similar but not as clear-cut a pattern, possibly because their physical disability is selectively interfering with the performance subtests.

Discussion

The results suggest a resolution of the conflicting findings in published reports regarding discrepancies between verbal and performance IQs in Duchenne muscular dystrophy. A greater verbal impairment in relation to performance IQ was clearly observed in the younger dystrophics who were still physically active and in relatively good motor control. They scored on average 12 points higher on performance IQ than verbal IQ. In the older, more disabled dystrophics who were less facile with their hands, this difference was not observed.

In analysing the effect of disease severity on verbal and performance IQs by comparing the younger, less physically impaired dystrophics with the older, more disabled dystrophics, it becomes apparent that the impairment in verbal IQ is early and nonprogressive, whereas performance IQ shows no corresponding impairment early in the disease process but lower scores tend to emerge as the disease advances and are related to the severity of the disease process. The finding of an early, nonprogressive impairment of verbal IQ supports the contention of Rosman (1970) and Prosser *et al.* (1969) that there is a cerebral defect that is a genetically determined developmental abnormality appearing early in DMD, but contradicts Prosser *et al.*'s conclusion that the intellectual impairment is

uniform, with both verbal and performance IQs equally impaired.

The main conclusion from this study is that the organic brain damage appears to be genetically determined, is present early in intellectual development, and tends to be verbal in nature.

The main implication of these findings is that when the dystrophic boy becomes wheelchair dependent he is probably penalized on the WISC Performance subtests because he is less agile with his hands. In estimating the intellectual functioning of the more severely physically handicapped dystrophics, it is well to keep in mind that their test results probably underestimate their abilities to some extent on the performance subtests. It is not suggested that the use of the performance subtests be discontinued in estimating intellectual functioning for either the mildly or more severely disabled DMD patient. Relying on the verbal IQ alone to estimate a dystrophic boy's intellectual functioning also tends to underestimate his actual intelligence, because it is in the area of cognitive functioning measured by the verbal subtests that the early cognitive impairment is noted. The recognition that the intellectual impairment is nonprogressive in nature leads to some important therapeutic considerations in that teachers should not assume that the intellectual capabilities will deteriorate as the child gets weaker and eventually becomes confined to a wheelchair.

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