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Sexual Dysfunction in Male Spinal cord Injuries

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男性脊髓損傷病人性功能異常之探討

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134 例罹患脊髓損傷半年以上並曾在台大醫院復健科住過院之病患被選爲本研究的對象。由於國人性觀念保守，本研究以郵寄問卷方式進行。總共有55份回覆，回收率爲 41.4 %。其中 2 例男性病患已去世，另外有 8 例爲女性病患，但她們的回答都不完整，因此這10 例均被排除於本研究之外。剩下45 例男性病患的年齡由20 至57 歲不等，平均年齡爲 33.9 歲。他們的罹病期爲半年至14 年，平均爲 5.3年。

本研究結果顯示，73.3% 之男性脊髓損傷患者能保有勃起功能，37.8% 具有射精功能，22.2% 有性交經驗，有 3 例（6.7 % ）還使其配偶受孕。損傷位置越高者越能保有勃起功能。反之，位置越低者較能保有射精功能。

由於男性脊髓損傷患者常併有不等程度的性功能異常，在接受適當的肢體復健後，著者認爲仍需給予適當的有關性生活之諮詢和必要的醫療處置。

INTELLECTUAL FUNCTIONING IN DUCHENNE MUSCULAR DYSTROPHY —A PRELIMINARY REPORT

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Intelligence tests with Wechsler Intelligence Scale for Children (WISC) were carried out on 10 boys with Duchenne muscular dystrophy (DMD) and 10 age-matched controls. The verbal IQ scales came close to be significantly lower in DMD group (mean 77.2) than in control (mean 90.5). Verbal ability was found to be reduced prominantly in comparison with the performance test in DMD patients.

Four patients with mildly physically disabled scored significantly lower verbal, performance and full IQ scales than that in the 6 markedly physically disabled. The results are interpreted as a preliminary finding of the intellectual impairment and the significant difference of verbal disability compared to performance test in the patients with DMD. The impairment of intellect also tend to be comparable with increasing physical disability.

Key words: intellectual function, Duchenne muscular dystrophy

Introduction

Duchenne muscular dystrophy (DMD) is an X chromosome linked recessive genetic disorder usually affecting males. It has been known for a long time that the disease can be associated with different degrees of mental deficiency. This was noted initially by Duchenne himself[1,2] and has been documented by many authors since then.[2,3]

The cause of mental impairment in DMD is, however, poorly understood. The early studies were attributed this finding to psychosocial, emotional or psychoeducational factors. Most reports since 1960 had concluded that the decreased intellectual functioning was related to the influence of some physiological parameters of the disease.[4-8]

Marsh and Munsat[5] firstly reported an early impairment of verbal intelligence in DMD patient. Karagan, et al[6] also demonstrated more significant differences in verbal IQ than on performance tests, with low scores on subtests requiring memory for patterns, numbers and verbal labeling. They supposed the involvement of central nervous system with cognition dysfunction in this disease.

The present study was undertaken to evaluate the intellectual functioning, comparing to the findings of verbal and performance intellect in the DMD patients.

Materials and Methods

Our study was made on 10 boys with DMD, ranging from age of 8 to 14 years, in the Department of Physical Medicine and Rehabilitation of National Taiwan University Hospital. The diagnosis was based on the typical clinical pictures and on such ancillary examinations as electromyography, muscle enzyme activity and muscle biopsy. Patient's physical ability was classified according to the Functional Classification by Vignos, et al[9] as Table 1.

The Wechsler Intelligence Scale for Children (WISC) was administered to all of these DMD patients and 10 age-matched boys without physical or mental disabilities in schooling.

Student's t-test was used in the comparison of IQ scales between the DMD and control groups. Paired t-test was used in the comparison of verbal and performance IQ sacles in the DMD patients.

Results

The verbal, performance, full IQ scales and functional classification of these 10 patients are listed in Table 2. Mean verbal and performance IQ scales for the DMD and control groups are shown on Table 3.

Verbal IQ of DMD group comes close to being significantly lower than of the control group (t-test, $0.05 < P < 0.1$). This finding suggests the impendence and full IQ scales are not significantly different in both disease and control groups ($P > 0.1$).

In control group, mean verbal IQ of 90.5 and performance IQ of 91.2 are not different ($P > 0.1$). But in the DMD group, mean verbal IQ of 77.2 is significantly lower than the mean performance IQ of 80.9 (paired t-test, $P < 0.01$). This presents the fact that verbal disability is prominant in the DMD patients.

Table 4 reveals the IQ scales of DMD patients divided by 2 groups under the Vignos's functional classification of 1 to 3 and 4 to 9. The latter is more physically disabled that they can not climb stairs or walk without devices. In the comparison of IQ scales

between group I and II, verbal IQ was decreased in group II (t-test, $P < 0.01$) and performance IQ was also decreased in group II ($P < 0.02$). These meaned that the intellectual impairment were significantly related to the increasing physical disability.

In the comparison of verbal and performance IQ scales between the 2 groups of DMD, there was no significant difference in group I (paired t-test, $P > 0.1$), but with significantly lower mean verbal IQ of 64.3 than mean performance IQ of 65.7 ($P < 0.05$). This appeared the deterioration of verbal intelligence was prominantly related to the increasing physical disability.

DISCUSSION

Our preliminary study of the 10 boys with an established diagnosis of DMD comes close to being significant impairment of the intellectual function. Verbal ability was found to be reduced prominantly in comparison with the performance test. These findings are correlated to the recognition of most authors' studies.[4-8]

The cause of mental retardation in DMD patients is, however, poorly understood. The fact of association between the mental defect and the muscular disease is by no means constant. Some authors[8,10] attributed mental retardation in DMD patients to reduced motor efficiency beginning in childhood with early immobilization and the associated difficulties in learning and emotional disturbances.

However, Konicka, et al[11] found that mental development was not abnormal in the group of children with spinal muscular atrophy with comparable or more severe physical and educational difficulties than DMD children. They supposed that the physical disability was not the major factor in relation to the mental impairment.

Rosman's opinion[12] was that although mental retardation did not depend on the enviroment and the degree of mental retardation. He also found that the severity of histopathological changes in the muscle biopsy specimens were parallel to the patient's intellectual dysfunction. In our study of 2 groups of DMD patients divided by Vignos's functional classification, we found the intellectual impairment was significantly related to the increasing physical disability. This finding was correlated to the Rosman's Pathological study of myopathy in retarded DMD patients.

The most likely explanation for the intellectual impairment in DMD patients was a biochemical abnormality affecting the metabolism of brain as well as muscle, whereas traditionally it had been thought that DMD was a primary disease of muscle. It has recently been suggested that the disease may be secondary to a neuronal defect[13] or a generalized disorder of protein systesis and membrane[14]. So it has further been suggested that the intellectual impairment of DMD patients may be related to a central nervous system deficit.[5,15] and genetic implication.[7]

In our study of the DMD patients, verbal IQ is significantly impaired rather than performance IQ. This may be interpreted as the effect of some physical parameters of the disease upon central nervous system functioning. Karagan, et al[6] found more significant differences in verbal IQ than on performance test, with low scores on subjects requiring memory for patterns, numbers and verbal labeling. It has been suggested that the cognition deficit is more obviously in DMD patients and may be regarding to the concomitants of the central nervous dysfunction of the disease.

Other investigations demonstrated the possibility of central nervous dysfunction in DMD patients were, Kozicka[11] and Florek[16] used electroencephalograms in tests and showed 56% to 82% of patients with abnormalities. Their findings were together with the results of IQ tests and suggested the possibility of organic brain lesions of the DMD patients. Some authors' reports[4,12] also demonstrated the pathological changes of poorly developed cortical laminations, decreased brain weight, pachygyria and heterotopias within the cerebral white matters in retarded DMD patients.

Karagan and Sorensen[17] also found the verbal disability was not limited to DMD but was also present in Becker and Facioscapulohumeral types of muscular dystrophy. This appeared that various muscular dystrophy might involve cognitive deficits that could not be attributed to enviromental deprivation or psychosocial reactions to a chronic or fatal disease, but rather appeared to reflect involvement of higher central nervous system functions.

In attempting to identify the possible physical factors which relate to decreased intellectual functioning in DMD patients, it seems important to study brain-behavior relationships including intelligence, using age and stage grouppings which are more homogenous than used in the past. Also, longitudinal rather than cross-sectional designs are to be preferred.

Our preliminary report only shows a limited significance of the verbal intellectual dysfunction in the DMD patients. It requires more cases in broad age and stage ranges, and precise neuropsychological evaluations to the study in intellectual functioning of DMD patients.

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Table 1. Classification of Functional Ability in Patients with Duchenne Muscular Dystrophy by Vignos,et al.(9)

Class	Physical activity capacity
1	Walks and climbs stairs without assistance
2	Walks and climbs stairs with aid of railing
3	Walks and climbs stairs slowly with aid of railing(25 seconds for eight standard steps)
4	Walks unassisted and rises from chair but can not climb stairs
5	Walks unassisted but can not rise from chair or climb stairs
6	Walks only with assistance or walks independently with long leg braces
7	Walks in long leg braces but requires assistance for balance
8	Stands in long leg braces but unable to walk even with assistance
9	In wheelchair or bed

Table 2. WISC IQ Scores and Functional Classification of Boys with Duchenne Muscular Dystrophy.

Case	Age(year)	WISC*IQ Scales			Functional Class by Vignos, et al.
		Verbal IQ	Performance IQ	Full IQ	
1	12	94	109	101	1
2	8	93	110	101	2
3	13	102	109	105	2
4	8	97	86	91	3
5	11	79	78	77	4
6	13	77	83	78	4
7	8	49	35	37	4
8	14	57	48	48	4
9	10	59	61	56	9
10	9	65	89	74	9

*:Wechsler Intelligence Scale for Children

Table 3. WISC IQ Scales(Mean ± Standard Deviation) in Duchenne Muscular Dystrophy Patients and Control.

	DMD*(N=10)	Control(N=10)	Student's t-test
Verbal IQ	77.2 ± 18.0	90.5 ± 9.1	$t=1.98,\ 0.05<p<0.1$
Performance IQ	80.9 ± 24.7	91.2 ± 13.8	$t=0.55,\ p>0.1$
Full IQ	76.8 ± 22.4	90.1 ± 10.8	$t=1.6,\ p>0.1$

*:Duchenne Muscular Dystrophy

Table 4. WISC IQ Scales(Mean ± Standard Deviation) in Divided Groups of Duchenne Muscular Dystrophy.

	Group I (Class*1 to 3) N=4	Group II (Class*4 to 9) N=6	Student's t-test
Verbal IQ	96.5 ± 3.5	64.3 ± 10.7	$t=5.18,\ p<0.01$
Performance IQ	103.5 ± 10.1	65.7 ± 19.5	$t=3.2,\ p<0.02$
Full IQ	99.5 ± 5.2	61.7 ± 15.7	$t=4.15,\ p<0.01$

*:Class:Classification of Functional Ability of Duchenne Muscular Dystrophy by Vignos, et al.[9]