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MUSCULAR DYSTROPHY IN ADULTS: A FIVE-YEAR FOLLOW-UP

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ABSTRACT. The aim was to describe the natural history of adults with hereditary muscular dystrophies, including myotonic dystrophy, with respect to muscular function, ventilation and electrocardiogram. In a prospective study, 46 subjects were followed over a period of five years. In 1991 and 1996, their muscle function was assessed according to an observation scheme and their lung vital capacity was measured by spirometer. Electrocardiograms were obtained in 1991, 1993 and 1996. Deterioration of muscular function was seen with regard to both the functional muscle tests and the vital capacity. The proportion of pathological electrocardiograms increased from 38% in 1991 to 54% in 1996 in the 26 patients with myotonic dystrophy without an increase in clinically detected cardiac abnormalities. Timely examinations using standard methods can reveal medically important information on deterioration, which often passes clinically unnoticed because of the insidious progress of the diseases.

Key words: muscular dystrophy; myotonic dystrophy; myopathia distalis tarda hereditaria; chronic disease; Brooke observations; functional deterioration; respiratory function; electrocardiography; follow-up; rehabilitation.

INTRODUCTION

The muscular dystrophies (MD), including myotonic dystrophy (MyD), are inherited muscular diseases with varying degrees of severity of progressive weakness (27). Clinical features and medical problems have been studied mostly by means of cross-sectional studies. Longitudinal or prospective follow-up studies on the health status of adult patients with different types of MD contribute important knowledge for planning and evaluation of rehabilitation programmes. Considering the changed health picture in Sweden and other industrial countries and the higher proportion of chronic diseases in the population, there is a need for convincing studies which can guide rehabilitation, both in terms of need and expected effectiveness (17).

In a previous study, all adult individuals with neuromuscular diseases were identified in the county of Örebro, Sweden (1). In 1991, all eligible subjects with MD were examined regarding disability, respiratory function, electrocardiography and quality of life (2, 3). Another study concerned coping and quality of life (4). At that time, there were no comprehensive follow-up studies published on adults with MD. Therefore, we initiated a prospective longitudinal follow-up of this cohort separated into three groups: (i) myotonic dystrophy, a systemic disorder with stigmata from several organs, where distal muscle weakness is conspicuous; (ii) myopathia distalis tarda hereditaria, distinguished by a clinical picture with only distal muscle weakness; and (iii) other MD, where the disease always involves weakness in proximal muscles. Since then, a study from California has been published on adults with MD showing profiles of impairment and disability in patients followed for up to 10 years (13, 18–21).

The objective was to describe the natural history and loss of functionality by means of a five-year follow-up study on adults in Sweden with MD in terms of muscular functional tests, respiratory function and electrocardiographic changes.

MATERIALS AND METHODS

Study group

The patient selection was based on a population survey of neuromuscular diseases in the county of Örebro in Sweden (1). In 1991, the cohort included 57 individuals with muscular dystrophy (MD) in the age range of 16–64. The distribution by diagnosis has been reported earlier (2–4). Informed consent was received from all for a five-year follow-up study. At the final examination in 1996, 46 individuals participated. The two most frequently occurring diagnoses in the study group were MyD and myopathia distalis tarda hereditaria (MDTH). The third group of subjects represented the diagnoses limb girdle MD (including both clinically and genetically quite different

Table I. *Distribution of the study group in 1996 by number, diagnosis, gender, age and duration of disease*

Muscular dystrophies (MD)	*n*	M/F ratio	Age, mean	Disease duration, mean years (SD)
Myotonic dystrophy	26	0.53	44	24 (9.0)
Myopathia distalis tarda hereditaria	8	0.33	64	22 (11.7)
Other MD (total)	12	0.71	49	28 (9.7)
Limb girdle MD	7			
Facioscapulohumeral MD	3			
Becker MD	1			
Emery-Dreifuss MD	1			
All	46	0.53	49	25 (9.7)

Note: None of the individuals with MyD were congenitally affected.

entities), Fascioscapulohumeral MD, Becker MD and Emery-Dreifuss MD. Although this group was heterogeneous from both clinical and genetic standpoints, their MD caused considerable weakness in the proximal muscles, and because of the low numbers they were treated as a group, in the present study named 'Other MD'. The distribution of the study group by number, diagnosis, sex and age is shown in Table I.

Eleven individuals could not be followed up. Five were deceased, four from pneumonia (3 MyD and 1 'Other MD') and one from mammary cancer ('Other MD'). Two persons had moved (1 MyD and 1 'Other MD') and two persons ('Other MD') were unwilling to participate because they needed continuous ventilator support. Two persons were excluded because they had the non-progressive disease myotonia congenita.

Muscular function test

The functional test was based on an observation scheme proposed by Brooke (6) and modified for computerized analyses in the examinations made in 1991 (3). The test includes 18 different observations of the subject, including gait, stepping onto a footstool/chair, rising from the floor and from a chair. Each observation was graded from 0 (normal) to 4 (fail). Scores of relative disability were calculated for every test in addition to a compiled score, i.e. the mobility index (3). Thus, 0 = normal function and 100 = the individual fails in all tests.

Motor ability was also assessed according to a modified grading scale (3, 6, 29) with regard to hips/legs (0–12), arms/shoulders (0–6) and bulbar function (0–4), where 0 = normal function.

All participants were examined by a neurologist, in 1991 by LGG (3) and in 1996 by KD. The examinations in 1996 were performed blinded for the result from 1991. The same manual with standardized instructions (3) was used on both occasions. In order to achieve a reliable examination, the two neurologists co-operated in the first 10 cases and a consensus of almost 100% was obtained in their judgements.

Body Mass Index (BMI)

All individuals were measured for body weight and height. The BMI was calculated in 1991 and 1996.

Respiratory function test

Vital capacity (VC) and the forced expiratory volume in 1 second (FEV_1) were measured in the sitting position. The FEV_1, expressed as the percentage of VC (FEV%), was calculated, and all values were expressed as a percentage of the predicted, age-adjusted, normal values. A bellows spirometer (Vitalograph Ltd., Buckingham, UK) was used for all individuals at the beginning of the study in 1991 (2). At the follow-up in 1996, an electronic spirometer (Vitalograph Alpha) was used instead in most cases. Five individuals were tested with both pieces of equipment at the follow-up with high concordance; the electronic spirometer registered 2% lower values for VC. Three individuals were not examined with spirometry at the follow-up because of severe weakness in their respiratory muscles, difficulty in obtaining reliable values (2 cases) and refusal (1 case).

Electrocardiography

Standard 12-lead electrocardiograms (ECGs) were recorded in 1991, 1993 and 1996. All the ECGs in 1991 and most of those in 1993 were recorded on a Hewlett-Packard 4750 instrument. Some of the ECGs in 1993 and all in 1996 were recorded on a Siemens-Elema Megacart. All the ECGs from 1991–1996 were re-evaluated and interpreted by the same clinical physiologist (MB). Standard interpretation criteria were used (7, 24). Recognized ECG abnormality/pathology was classified as follows: (i) rhythm disturbances, including sinus bradycardia and sinus tachycardia; (ii) conduction disturbances, including AV-block I, left anterior hemiblock (LAH), left bundle branch block (LBBB) and right bundle branch block (RBBB); (iii) hypertrophy of atria and ventricles; and (iv) Q-waves and STT-changes.

Each individual ECG was assessed as normal, abnormal or pathological. Abnormal ECGs showed at least one of the following signs: sinus bradycardia, sinus tachycardia, LAH without other conduction disturbances, isolated high R/S quotient in V1 or unspecified STT changes. Pathological ECGs showed at least one of the following signs: AV block I, LBBB, RBBB, right atrial hypertrophy, ventricular hypertrophy, pathological Q-waves or pathological STT changes.

Statistical analysis

Beyond descriptive statistics, a two-tailed *t*-test for dependent observations was used for data on interval scale (respiratory function). The Wilcoxon signed-ranks test was used for the other data on ordinal scales. Spearman's rank correlation (*rho*) was applied. In all analyses, a probability of <0.05 was accepted as indicating statistical significance.

Table II. *Mean of disability with regard to muscular function in percent assessed by Brooke's functional tests in all 46 subjects in 1991 and 1996*

	MyD (n = 26)		MDTH (n = 8)		Other MD (n = 12)		All (n = 46)	
Tests	1991	1996	1991	1996	1991	1996	1991	1996
Brooke's functional tests:								
Mobility index	23	48**	26	45*	56	69**	32	52**
Gait	12	35**	16	27*	29	56**	17	39**
Stepping onto footstool	16	39**	21	32	52	67*	26	45**
Stepping onto chair	38	68**	34	50*	77	79	47	68**
Rising from chair	21	29*	13	25	56	67	29	38**
Rising from floor	33	60**	50	66	69	81*	45	66**
Rising from chair/floor	27	44**	31	45	63	74**	37	52**

* $p = < 0.05$, ** $p = < 0.01$.
MyD = myotonic dystrophy, MDTH = myopathia distalis tarda hereditaria, MD = muscular dystrophy.

RESULTS

Evident deterioration of the muscular functions was seen in all the functional tests (Table II). Among the diagnostic subgroups, the decline was most obvious for subjects with MyD. In the compiled mobility index, a statistically significant deterioration was found for all subgroups. With regard to the graded function in arms and shoulders, the same pattern was found, but the bulbar dysfunction had deteriorated only for MyD (Table III).

The base level in the grading scale, i.e. the ability to walk and climb stairs without assistance, was achieved by 23 out of 26 patients with MyD, 6 out of 8 with MDTH and 3 out of 12 with 'Other MD'. Another two subjects with MyD and five with 'Other MD' managed the stairs with the aid of a railing. During the period 1991–1996, a slightly higher degree of disability was

Table III. *Distribution of subjects with regard to diagnostic group and disability graded in fixed levels with respect to arm/shoulder and bulbar function (6)*

	MyD (n = 26)		MDTH (n = 8)		Other MD (n = 12)		All (n = 46)	
Function	1991	1996	1991	1996	1991	1996	1991	1996
Arms/shoulders (0–6)								
0 Can abduct arms in full circle and lift 2 kg with one hand above eye level	23	23	6	6	4	3	33	32
1 As above, but cannot lift 2 kg with one hand above eye level			2	1	3		5	1
2 As above, but with elbows flexed	1					1	1	1
3 Cannot raise hands above head, but can raise a glass of water to mouth	1	1		1	3	5	4	7
4 Can raise hands to mouth, but not a glass		1			2	1	2	2
5 Cannot raise hands to mouth, but can use hands to pick up pennies from the table	1	1				2	1	3
6 Cannot raise hands to mouth and has no useful function of hands								
Bulbar (0–4)								
0 Normal speech and swallowing	9	5	8	8	9	9	26	22
1 Speech and/or swallowing is abnormal, but presents no practical difficulty	14	16			3	3	17	19
2 Speech is occasionally difficult to understand and swallowing or the swallowing difficulty causes occasional choking (on a daily basis)	3	5					3	5

MyD = myotonic dystrophy, MDTH = myopathia distalis tarda hereditaria, MD = muscular dystrophy.

Table IV. *Mean of the vital capacities assessed by spirometry*

Expressed as:	MyD ($n = 26$)		MDTH ($n = 8$)		Other MD ($n = 9$)		All ($n = 43$)	
	1991	1996	1991	1996	1991	1996	1991	1996
Litre, l (SD)	2.9(1.0)	2.7*(1.0)	3.5(0.9)	3.3(0.8)	3.4(1.2)	3.1(1.3)	3.1(1.0)	2.9**(1.0)
Range, l(SD)	1.4–4.7(1.0)	1.1–4.2(0.9)	2.1–4.7(0.9)	2.1–4.5(0.8)	1.2–5.1(1.2)	0.8–5.1(1.3)	1.2–5.1(1.0)	0.8–5.1(1.0)
% predicted	69.9	68.0	93.5	91.8	75.4	68.3	75.5	72.5

* $p = < 0.05$, ** $p = < 0.01$.
MyD = myotonic dystrophy, MDTH = myopathia distalis tarda hereditaria, MD = muscular dystrophy.

found in three subjects with MyD and in four with 'Other MD'. In 1991, 7 individuals (MyD 3, 'Other MD' 4) were confined to wheelchairs, as compared to 10 in 1996 (MyD 3, MDTH 1, 'Other MD' 6).

The body mass index increased in the group of MyD from 23.3 to 25.4 ($p < 0.01$), in MDTH from 25.6 to 26.0 (non-significant), and in the 'Other MD' group from 22.0 to 23.5 ($p < 0.05$).

At the beginning of the study in 1991, two individuals had ventilator support at home ('Other MD' 2). Five years later, another five individuals needed nocturnal assisted ventilation (MyD 1, 'Other MD' 4). Smoking habits remained fairly unchanged over the five years: two out of seven stopped smoking and one started. Low average VC in relation to predicted values was seen in 1991 in MyD and 'Other MD' (Table IV). Over the five-year period, the VC decreased significantly in MyD.

All ECGs from MDTH patients were normal except for that of one patient who had developed an AV-block I in 1996 (Table V). The percentage of pathological ECGs in the whole study group increased from 30 (14/46) in 1991 to 39 (18/46) in 1993, and finally to 41 (19/46) in 1996. Progression of ECG abnormalities occurred almost exclusively in patients with MyD. Twelve out of 26 individuals in this group had advancing pathology. The percentage of patients with pathological ECGs increased from 38 (10/26) in 1991 to 54 (14/26) in 1996. During

Table V. *The ECG abnormalities in subjects with MD (*n = *46)*

ECG abnormalities	MyD ($n = 26$)			MDTH ($n = 8$)			Other MD ($n = 12$)		
	1991	1993	1996	1991	1993	1996	1991	1993	1996
Rhythm									
Sinus bradycardia	1	0	1	0	0	0	0	0	0
Sinus tachycardia	0	2	0	0	0	0	0	0	0
Total	1	2	1	0	0	0	0	0	0
Conduction disturbances									
AV-block 1	5	6	8	0	0	1	0	0	0
LAH	5	6	4	0	0	0	1	1	1
LBBB	3	4	7	0	0	0	0	0	0
RBBB	0	1	1	0	0	0	0	0	0
Total	13	17	20	0	0	1	1	1	1
Hypertrophy									
Right atrial	0	0	0	0	0	0	2	2	1
Ventricular	5	6	6	0	0	0	3	4	4
Total	5	6	6	0	0	0	5	6	5
QRS & STT-changes									
Q-waves	6	7	9	0	0	0	2	2	2
STT-abnormalities	6	9	14	0	0	0	1	2	3
Total	12	16	23	0	0	0	3	4	5
ECG assessment									
Abnormal	6	6	7	0	0	0	2	2	3
Pathological	10	13	14	0	0	1	4	5	4

Note: In many cases the subject has more than one ECG abnormality.
MyD = myotonic dystrophy, MDTH = myopathia distalis tarda hereditaria, MD = muscular dystrophy.

the five-year period, one male with MyD had a malignant arrhythmia, but he recovered fully.

DISCUSSION

The goals and objectives of a rehabilitation program depend on the natural history of the disease (11). Research on MD has focused mostly on children with Duchenne's and Becker's MD. In the present five-year follow-up study on adults with different types of MD, identified in a previous population survey (1), we found evident deterioration of muscular function with regard to both functional muscle tests and vital capacity, and in most groups of patients, the decline was statistically significant. In addition, an increase in pathological ECG findings was found, mainly for patients with MyD, in contrast with no increase of clinical symptoms from the heart, except for in one patient with MyD.

Treatment of MD should be prospective in order to inhibit deformity, prolong independent ambulation and maximize functional capabilities. It should not only be limited to palliation (9, 11). Research-based knowledge is a precondition for effective rehabilitation. There has been an opinion for many years that exercise, especially strengthening exercise, accelerates weakness in MD. This conception has not been verified by research studies, and today exercise-training programmes are recommended early in the course of the disease (when the muscle fibre degeneration and weakness are minimal). The exercise should be performed at submaximal levels (12). The results of the present study emphasize that lung and heart impairment should also be considered when planning rehabilitation.

Brooke functional tests appeared to give a reliable estimate of increasing muscle weakness over time. The functional muscle tests constituted 18 different observations. In the present study, improved function was noted in fully 3% of all observations, mainly concerning the tests of stepping onto a chair and rising from the floor without hand support. These tests might be somewhat hazardous to judge, and can be influenced by the degree of encouragement given. However, in 97% of all the 828 observations done over the five-year period, the estimation was either unchanged or an increased disability was seen, in agreement with the patients' reports. The Brooke tests were easy to perform according to the observation scheme and the strict manual used. Since more than one observer was involved in 1991 and 1996, and there was a time lapse of five years between examinations, the reliability may have been weakened. In addition, the deterioration rates were not adjusted for normal age-related decline in muscle strength. However, the Brooke examinations revealed and graded the functional disability and served to complement the neurological examination.

In the Californian study, the muscular function was assessed for 51 participants, 70% of whom were able to walk and climb stairs without assistance (18). Calculated for all 75 participants (before drop-outs), this means that 20% were not able to climb stairs, as compared to 10% in the present study. However, in the present study, the Brooke functional tests (Table II) proved to be much more discriminative and sensitive over time with regard to the lower extremities.

The mobility index in MyD increased from 23 to 48, indicating a marked increase of disability over the five years. Although this score is compiled from several tests approximately in accordance with an interval scale, it would not be appropriate to interpret this increase as indicating that the disability was twice as severe after the five-year follow-up period. In subjects suffering from MDTH affecting mainly distal muscles, the decline in mobility index was nearly as marked as in the other groups, and the result was not explained by the progress in single subjects. This indicates that a seemingly benign MD also causes evident disability (28). In the heterogeneous group of 'Other MD', the disability was most pronounced according to the mobility index, and there was a decline in function in both the upper and lower extremities.

Respiratory function declined in MyD and 'Other MD', in most cases mildly, but in some individuals severely, as illustrated by the fact that seven individuals in the study group required assisted ventilation at the end of this study. Respiratory abnormalities, including overt respiratory failure in MyD, are well known from previous studies (5), as is the decline in respiratory function and possible need for ventilator support in late stages of other types of MD (26). For the patients with MyD, the mean VC was 70% of that predicted. This was in accordance with the previously mentioned study from California where the mean was 76% ($\pm$19%) (18). Patients with 'Other MD' may also suffer from respiratory impairment (2, 14, 19, 20) not always obvious from clinical examination alone. Regular measurements of lung function should therefore be part of the evaluation of all individuals with MD to facilitate detection of persons at risk of developing deficient ventilation (26).

In the present study, the progression of ECG abnormalities occurred mainly in patients with MyD.

Conduction disturbances and pathological Q-waves were especially rapidly progressive. The ECGs were abnormal/pathological in a total of 80% (21/26 in 1996) of the cases, as compared to 75% in the Californian study (18), and the profile of pathology in that study was also similar to the results in the present study. It seems likely that progressive fibrosis and fatty infiltration of the conductive tissue in the heart are the main causes of increasing conduction disturbances in MyD (8, 10, 23). As for the pathological Q-waves, the underlying pathophysiology appears more complex. In addition to the fibrosis and fatty infiltration of both ventricles, severe coronary heart disease has been demonstrated in patients with MyD (10, 23). A third possible cause of pathological Q-waves is left ventricular hypertrophy (15). Left ventricular hypertrophy is a common finding in patients with MyD (8, 10, 23). A natural history of progressive cardiac conduction disturbances has been well documented previously (16, 22, 25). The results in the present study suggest a similar development of pathological Q-waves.

The present study focuses on clinically relevant information on muscular weakness, ventilatory dysfunction and cardiac involvement provided by standard tools and methods used in adults afflicted with insidious progress of muscular dystrophy. The results may have implications for rehabilitation interventions. In a forthcoming study, we investigate what it means to live with MD in terms of Activities of Daily Living (ADL), coping and quality of life over a five-year period.

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