

Physical ability and health in a non-steroid population of 77 adult patients with Duchenne muscular dystrophy B. Werge, J. Rahbek, A. Madsen, J. Marquardt, U. Werlauff, B. Steffensen.

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INTRODUCTION

Life expectancy for adults with Duchenne muscular dystrophy (DMD) has increased due to medical and respiratory treatment, and care givers who must address the care needs of this group of patients are faced with other types of demands. However, knowledge about physical ability and its impact on body function in adult patients is scarce. Denmark has a tradition for invasive respiratory intervention in DMD and – as a result of this – a major group of adult patients.

AIM

The aim of the study was to assess physical ability and living conditions in the steroid naïve Danish adult DMD population. This study presents the results from a section about the

METHODS

All Danish patients \geq 18 years (n=87) with a confirmed clinical diagnosis of DMD were invited to participate in the study. Each patient was interviewed in his home by two professionals from the National Danish Rehabilitation Centre for Neuromuscular Diseases (RCfM) about his physical ability, health, social relations and daily life using a comprehensive questionnaire. Motor function was assessed by means of the EK2 scale. Descriptive statistics (median, range, spearman's Rho) were used to illustrate distribution of data.

CONCLUSION

Respiratory problems in Danish adult patients with DMD are well managed, but cardiac assessment and medication were not initiated in all patients. Constipation is a major problem that impacts daily life and needs to be addressed in the management of adult patients with DMD. Despite the progressive loss of muscle strength, the adult patient endures sitting in his wheelchair all day, and is able to operate his electrical wheelchair and computer; however, pain due to sitting position is a common problem.

RESULTS

Eighty-six patients accepted the invitation. Nine were subsequently excluded, two due to treatment with steroids, seven due to cessation of ambulation ≥ 13 years. Median age of 77 patients was 27 years (18-46); cessation of ambulation was 9.0 years (5-12).



OROFACIAL

68/77 patients could eat cut up food but avoided hard and chewy food. 26 patients



VENTILATION

Seventy-two patients needed ventilator support: 58 patients used invasive ventilation (IV) on tracheostomy and 56 of those used the ventilator 24-h. 14 patients used non-invasive ventilation (NIV) by a nasal mask during the night; 44/58 patients on IV had used NIV in 30 months (1 -120) before they were tracheostomized.

Continuous Positive Airway Pressure (CPAP) was used intermittently to mobilize mucus by 17 patients; of those, 8 used NIV and 5 IV. Lung infections were rare; only 7 patients had been hospitalized due to pneumonia or lung infections during the previous year. All 77 patients were regularly assessed at a specialized pulmonary centre, 62 patients had received vaccination against infection with influenza, 46 patients against

experienced regular problems with swallowing food; 12 patients had a G-tube for nutritional support. Thirty-one patients spoke in a low voice; 28 patients spoke indistinctly; this was not related to age. Although speech problems were more common in tients on IV than in patients using NIV, the majority (31/58) of patients on IV did not have speech problems. All patients used an un-cuffed tracheostomy tube day and night.

SITTING ABILITY

All patients spend the day in their electrical wheelchair which has adjusted seat and electrical tilt and back; for additional body-support, 71 had an electrical belt and 54 had adjustable lateral body supports. 9/77 patients could balance when sitting outside the wheelchair. In their wheelchair, 29 patients could sit upright without support of the back rest, 48 patients needed permanent back and neck support to sit.



pa-



UPPER LIMB FUNCTION

Upper limb function was related to age. All but two patients operated their wheelchair by hand (n = 67) or by alternative steering (n = 8). 11 patients could feed themselves, 66 pa-



HEART

Sixty-five patients were assessed regularly by a heart specialist, 12 were not. Cardiac treatment is illustrated in the table.

MEDICAL DRUGS	Ace inhibitors	Beta blockers	Diuretic	Pacemaker
Number of patients	32/77	19/77	12/77	3/77

BOWEL AND BLADDER

Forty-five patients experienced regular stomach discomfort; most problems were caused by constipation, and 48 patients took drugs against this. Nine patients had bladder incontinence, 5 had bowel incontinence. One patients had a catheter, one patient had an ostomy. Three patients had been hospitalized due to bowel and bladder problems in the previous year.

PAIN AND FATIGUE

52/77 said they felt pain, 31 had problems/pain due to their sitting posture primarily from buttocks, back and lower limbs. 24 of these experienced pain on a daily basis, and 19 said that pain made daily activities more difficult. 13 used non-prescription analgesics.

Twenty-five patients said that fatigue was a problem every day or during the week; 17 of them said that it restricted their level of activity.

Fifty-six patients had a pelvic obliquity of more than 2 cm measured as the difference between the two anterior iliaca superior.

Obliquity was present in patients who had had scoliosis surgery as well as patients who had not been operated, but there was a tendency to a more pronounced obliquity in the non-operated group.



tients were fed by an assistant.



Upper limb function illustrated by EK2, item 5 (Ability to move arms)



SURGICAL INTERVENTIONS AND ORTHOSES

SURGERY	NUMBER OF PATIENTS
Spine	52/77
Hip flexors	23/77
Hamstrings	16/77
Achilles tendon	60/77

Ankle-foot7321Brace559 1 with spinal surgery/8 without	
Brace 55 9 1 with spinal surgery/ 8 without	
	t
Standing frame70	