RCFM

# Living conditions and quality of life in adults with Duchenne muscular dystrophy – a Danish survey

A. Madsen, J. Rahbek, B. Werge, J. Marquardt, O. Gredal, B. Steffensen.

The National Danish Rehabilitation Centre for Neuromuscular Diseases

mail: anma@rcfm.dk

## INTRODUCTION

Improvements in treatment methods have increased life expectancy for people with Duchenne muscular dystrophy (DMD), and the population of adults with DMD in Denmark has increased with 19 % during the past 10 years. During their childhood, boys with DMD are seen with frequent intervals at hospitals and at the Danish Rehabilitation Centre for Neuromuscular Diseases (RCFM), whereas evaluations tend to be less frequent in adulthood.

## AIM

The aim of the study was to assess physical abilities, living conditions, social relations and quality of life in our adult population of men with DMD. This study presents the results from the part of a questionnaire survey that concerned living conditions and quality of life.

## METHODS

All Danish men with DMD ≥ 18 years (n=87) with a confirmed clinical diagnosis of DMD according to the diagnostic criteria (ENMC 1997) were invited to participate in the study. Each individual was interviewed in his home by two professionals from RCFM about his physical ability, health, social relations and daily life using a comprehensive semi-structured questionnaire with both fixed and open-ended questions. A VAS scale was used to indicate the level of agreement to a specific statement. Descriptive statistics (median and range) were used to illustrate distribution of data.

## CONCLUSION

Danish adult men with DMD live an independent life by means of assistive devices and personal assistance, but few of them complete an educational program, and very few get a job or a family. Activities of daily living are time consuming, and as they become older, most days are spent at home with a helper as the only company. Still, the adult man is capable of operating his wheelchair independently and stay in touch with family and friends by means of a computer or other electronic devices. Despite worries about health issues, they consider life to be satisfactory and felt they were able to participate in desired activities.

# RESULTS

Eighty-six men with DMD responded to the invitation and were interviewed. Nine were subsequently excluded; two due to treatment with steroids, seven due to cessation of ambulation ≥ 13 years. A total of 77 respondents were included in the data analysis. Median age of the 77 respondents was 27 Years (18-46).

At their own request, fifteen respondents received help to answer the questions from a parent or a helper.

All respondents spent their day out of bed in a powered wheelchair. All but two were able to operate their wheelchair and/or use a computer by hand (n = 67) or by alternative steering (n = 8). 72/77 respondents used mechanical ventilation; 58 were on invasive ventilation by tracheostomy and 14 used non-invasive night time ventilation (NIV) by a nasal mask.

## TYPES OF ACCOMMODATIONS

Forty-nine respondents lived away from home; 45 had their own place, four lived in a public assisted facility. Mean age for leaving home was 23.5 y.

Among the youngest men aged 18-20, 11/13 lived with their parents. In the age group ≥ 26, 7/45 lived with their parents.

Four respondents lived with a partner; two of them had fathered a child.

## PERSONAL ASSISTANCE

All but 4 respondents had qualified for 24-h personal assistance. Median age for establishing a 24-h personal assistance scheme was 21.5 years (12-27). The 4 respondents who did not have personal assistance around the clock were all ≤ 20 years.

The majority received help to manage their personal assistance scheme from their parents, a personal assistant, an agency or the municipality. Some only needed help with salary payments.

Forty respondents qualified for extra hours which they could use for spending a night away from home, etc. Eleven respondents found they did not have sufficient help when away from home on weekends or vacations.

A personal assistant team usually consisted of 5-6 helpers – both men and women were employed as helpers; 30/77 respondents employed one or more family members as helpers.

## EDUCATION

All 77 respondents had completed elementary school and all but 5 respondents felt that they had had sufficient help in elementary school. Sixteen of these also had completed Upper secondary education. Twenty-two respondents had supplemented elementary school with a stay at a lower secondary-level boarding school and 25 had attended a folk high school. Nine of the respondents had completed a business degree and three has completed a Bachelor's degree. Thirty-one regretted not having prioritized education more highly.

## PROFESSIONAL EXPERIENCE AND MAINTENANCE

Thirteen respondents had work experience, and at the time of the study, three were employed in paid part-time jobs; 70 respondents received a pension and 3 received money from the State Educational Grant and Loan Scheme. In addition, 73/77 received money from the municipality to cover excess expenses derived from their functional impairment and personal assistance scheme. All but four respondents said they were able to make ends meet financially.

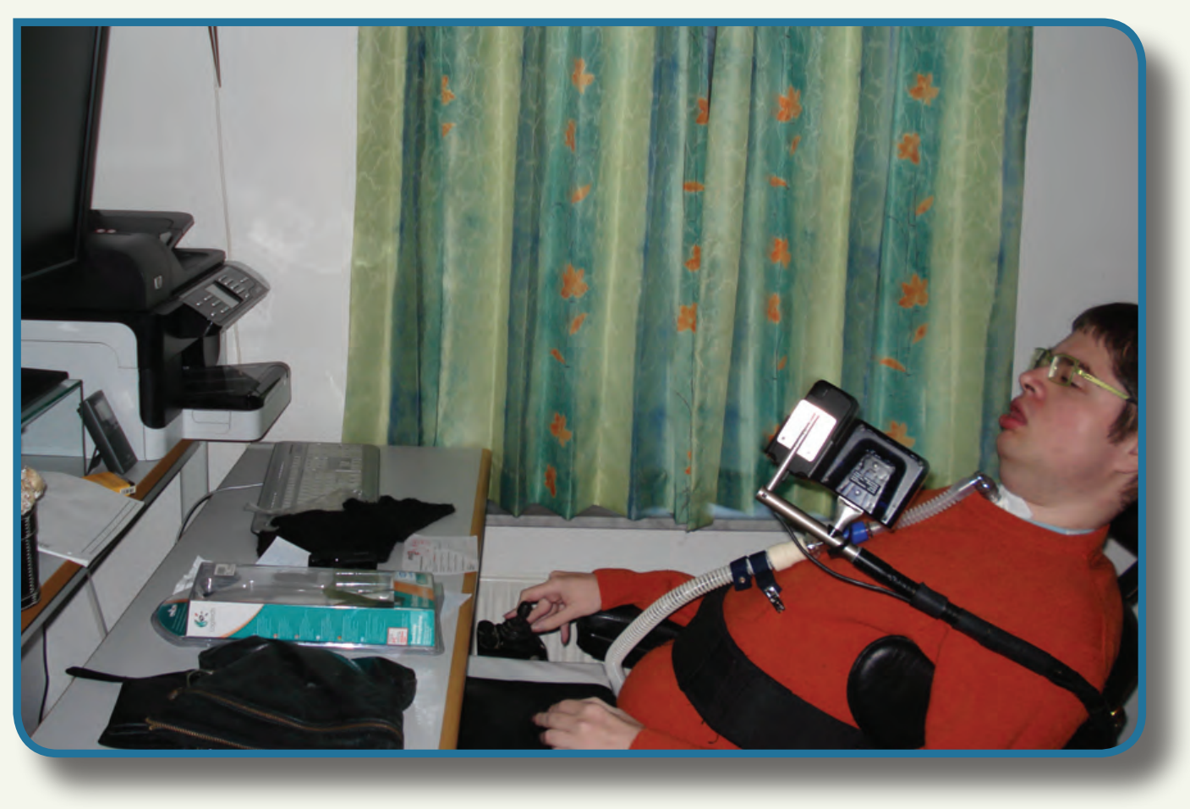
## A TYPICAL DAY FOR AN ADULT MAN WITH DMD

The adult man with DMD starts his day between 9 and 10 a.m. with variations ranging from 5.30 a.m. to 2.30 p.m. It takes between one and two hours to get out of bed, have his tracheostomy tended to, get mucous mobilization and cough assistance, go to the bathroom, get dressed, eat breakfast and have his teeth brushed. Before he is ready to begin his daily activities, it is already close to noon. Baths and/or bowel movements increase time spent on personal care and the older he gets, the more time he will tend to spend on personal care.

For adults not attending education or work, days are usually spent at home with his helper as his only company. On average, he spends 6 hours (0-14 hours) at his computer (gaming, reading the news, browsing the Internet, social media), and 4 hours (0-14 hours) watching TV, but usually both computer and TV are turned on all day. He communicates with family and friends over his computer or phone, but once a week he receives visits or goes to visit family and/or friends. His day doesn't feel long and boredom is not an option.

He usually participates in grocery shopping and planning of meals while daily routines such as cleaning and laundry are scheduled and carried out by his helpers. He eats two-three meals a day. Trips outside the home are made by car which is driven by his helper.

Median bedtime is 12 pm, but may vary from 7pm to 5 am. The older he is, the more time he spends in bed. He needs help to turn and position himself at least four times during the night.



## LEISURE TIME

77 respondents owned a car and drove 12.000 km (median) with a maximum of 40.000 km per year; 18 respondents had travelled abroad in their car the previous year. Thirteen respondents were active in sports – mainly wheelchair football. Other leisure interests were watching TV (e.g. sports and films), PC games, listening to music, activity on social media and visiting family and friends. 59/77 respondents felt they were able to participate in desired activities; if not, physical barriers were often cited as the reason.

## FAMILY AND FRIENDS

All respondents had good relationships with their family. Eight respondents were an only child, eight had a brother with DMD, and three respondents had lost a brother with DMD.

Almost everyone (n=74) had a least one friend. Respondents between 20 and 30 years of age had most friends, and the number of friends tended to drop with age. They had met their friends through the Danish patient organization (n=51), their hobby (n=44), or school (n=40), and some were former helpers (n=25).

## PROBLEMS AND WORRIES

The majority (n=66) said they generally felt safe in their daily lives but asked about certain issues concerning their neuromuscular disease, 46 said they were worried. Worries were mainly about loss of hand function, cardiac problems, and choking on food or water; 6 respondents worried about dying. 35 respondents thought they would live to become old-age pensioners. Nine respondents had not expected to reach their present age.

Other types of worries concerned their personal assistant scheme and cuts in social welfare.

Four respondents did not have anybody to share their worries with while the rest shared their worries with their parents (n=35), helpers (n=25) or friends (n=17).

## QUALITY OF LIFE

The majority (n=50) did not consider their disability to be a part of their identity, and they 'always' or 'mostly' felt they were respected by society in general like other people. They felt appreciated and loved and the majority (n= 62) considered their QoL to be 'good'. Being with family and friends greatly was important and highly valued.

Although many of them didn't feel lonely, 51 respondents missed having a partner, and eleven respondents had dreamt of fathering a child. The lack of a girlfriend/partner in their life was not what they had expected, and they wished they had given this higher priority. Thirty-three respondents had experienced sex with a partner or a prostitute but the majority (n=50) said their sexual needs were not satisfied. 29 respondents had found a method with which they could satisfy themselves sexually.