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Living with Muscular Dystrophy

Illness Experience, Activities of Daily Living, Coping, Quality of Life and Rehabilitation

BY

BIRGITTA NÄTTERLUND

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ABSTRACT
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The overall aim was to study and gain knowledge about what it means living with muscular dystrophy and to study rehabilitation out from the patients perspective, among adults with muscular dystrophy in three Swedish counties: Örebro, Östergötland and Norrbotten. The thesis comprises two qualitative and three quantitative studies. Thirty interviews about illness experience were subjected to content analysis and 37 interviews about perceived support in rehabilitation were analysed according to phenomenological guidelines. Data were also collected by the Assessment of Problem-focused Coping (APC), the ADL Staircase, the elf-report ADL, the Mental Adjustment to Cancer Scale, the Sickness Impact Profile, the Psychosocial well-being questionnaire and the Quality of Life Profile. The APC was developed for assessment of problem-focused coping and covers also questions concerning the extent to which activities are experienced as problems and satisfaction with activities. The result shows that the experience of illness is largely similar in the three diagnostic groups (proximal MD, Myotonic muscular dystrophy, Myopathia distalis tarda hereditaria). The persons reported many restrictions of everyday activities, most often in mobility and transportation. Over half were dependent on other people in activities of daily living, and the illness was experienced mainly as having negative consequences for everyday life. A lower quality of life may be partly explained by a reduced capacity for activities. Problem-focused coping was used only to a limited extent, and 'Fighting spirit' was the dominant coping strategy. Rehabilitation was experienced as very valuable, particularly the education about the muscle disease, technical aids, grants and physical training. Over a five-year period, disability and quality of life of the study participants deteriorated significantly, and the dependence on other people increased.

Key words: Muscular dystrophy, illness experiences, activities of daily living, coping, quality of life, rehabilitation.

Birgitta Nätterlund, Department of Public Health and Caring Sciences: Section for Caring Sciences, Uppsala University, Uppsala Science Park, SE-751 83 Uppsala, Sweden

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"Life is what happens to you
While you are making other plans"
(John Lennon)

ORIGINAL PUBLICATIONS

The present thesis is based on the following five studies, which will be referred to in the text by their Roman numerals:

I. Nätterlund, B., Sjödén, P-O. & Ahlström, G. The illness experience of adult persons with muscular dystrophy. Submitted.

II. Nätterlund, B. & Ahlström, G. (1999). Problem-focused coping and satisfaction with activities of daily living in individuals with muscular dystrophy and postpolio syndrome. Scandinavian Journal of Caring Sciences, 13, (6), 26-32.

III. Nätterlund, B. & Ahlström, G. (2001). Activities of daily living and quality of life in persons with muscular dystrophy. Journal of Rehabilitation Medicine, 33, In press.

IV. Nätterlund, B., Gunnarsson, L-G. & Ahlström, G. (2000). Disability, coping and quality of life in individuals with muscular dystrophy: a prospective study over five years. Disability and Rehabilitation, 22 (17), 776-785.

V. Nätterlund, B. & Ahlström, G. (1999). Experience of social support in rehabilitation: a phenomenological study. Journal of Advanced Nursing, 30 (6), 1332-1340.

Reprints of Studies II-V were made with permission from the publishers.

Abbreviations

ADL	Activities of daily living/daily activities
APC	Assessment of Problem-focused Coping
FSH	Facioscapulohumeral muscular dystrophy
I-ADL	Instrumental activities of daily living (cleaning, shopping, transportation, cooking)
ICIDH	International Classification of Impairment, Disability and Handicap
MAC	Mental Adjustment to Cancer (MAC) scale
MD	Muscular dystrophy
MDTH	Myopathia distalis tarda hereditaria/Morbus Welander
MyD	Myotonic muscular dystrophy/dystrophia myotonica
P-ADL	Personal activities of daily living (bathing, dressing, toileting, transfer, continence, feeding)
PPS	Postpolio syndrome
SIP	Sickness Impact Profile
The WHOQOL Group	The World Health Organization Quality of Life Group
WHO	World Health Organization

CONTENTS

INTRODUCTION

The present thesis focuses on adults with Muscular Dystrophy (MD), their experiences of illness, activities of daily living, coping, quality of life and perceived support in rehabilitation. A rehabilitation project "Programme for improved quality of life in muscular dystrophy" (Ahlström & Gunnarsson 1997) was carried out as a part of this thesis. The rehabilitation programme was based on previous research (Ahlström 1994), which had shown that adults with MD encounter a variety of illness-related problems in their daily life and that they find these problems hard to deal with by themselves. MD is a progressive neurological disease group including several types of hereditary, primary and incurable muscular diseases involving muscular weakness, which usually progresses relatively slowly (Brooke 1986, Edwards 1989, Harper 1988, Walton & Gardner-Medwin 1988). The development of a progressive disease may cause great stress in the afflicted individual (Boutaugh & Brady 1996). A progressive disease like MD involves successive transitions to new situations due to repeated losses of functional capacity (Ahlström 1994, Ahlström & Gunnarsson 1996, Walton & Gardner-Medwin 1988). The capacity to perform activities of daily living is essential for a person to maintain his or her roles in the family, at work and in society as a whole (Kielhofner 1992). Since there is no cure or treatment for MD, it is important to give recurrent support and care both to the persons´ with MD and to their families, assisting them to manage their daily life and to find meaningful occupations (Wilcock 1993).

Chronic illness

A chronic illness can mean both incapacity and retention of capacity, it may appear suddenly or develop slowly, be progressive or remain stationary, and have consequences for the rest of the person's life. Chronic illness can shorten life, but does not necessarily do so (Sidell 1997). An illness is chronic if it lasts more than

three months (Lapham & Ehrhart 1986, Pollin 1994). One of the earliest formal definitions of chronic illness was given in 1949. This definition is "all impairments or deviations from normal that have one or more of the following characteristics: are permanent; leave residual disability; are caused by non-reversible pathological alteration; require special training of the patient for rehabilitation; or may be expected to require a long period of supervision, observation or care" (Dimond 1984, p. 2). Since 1949, other definitions have added the time dimension (Pollin 1994), lifestyle (Hymovich & Hagopian 1992) and symptom management (Kinzel 1993).

During recent decades, there has been an increase in the number of people suffering from chronic illnesses in society. Such illnesses create difficulties of performance in different kinds of occupation. People with chronic illness have to adjust their lifestyle to the limitations that may follow (Hwu 1995). A chronic illness may create social and financial burdens, which affect the family in various ways, for instance by creating a need for various types of support in daily life (Kielhofner 1992, Sidell 1997). The literature often focuses on the general impact of chronic illness and has failed to address the afflicted persons' experience of occupation in daily life. Many studies fail to illuminate what people do and how they integrate illness into daily life (Michael 1996).

Muscular dystrophy

The term muscular dystrophy was introduced in the mid-19th century and has enjoyed international acceptance for 150 years (Kuhn 1990). More than 30 different types have been described in the literature, varying in their heredity and rate of progression, and at present there is no cure (Bartalos 1991). These diseases may have their onset in childhood, adolescence or adulthood (Eggers & Zatz 1998, Walton & Gardner-Medwin 1988). In an extensive Swedish population study, the total prevalence of MD was 39 persons (aged 5-79 years) per 100,000 head of

population, which gives an estimated total figure of 3,200 persons in Sweden (Ahlström et al. 1993). Prevalence varies between the types of MD, the most common for adults being myotonic muscular dystrophy (MyD) with 18.2 (age 5-67 years), followed by proximal MD 14.1 (age 9-70 years) (all diagnoses) and myopathia distalis tarda hereditaria (MDTH) 5.6 (age 49-79 years) per 100,000 (Ahlström & Gunnarsson 1997, Ahlström et al. 1993). The cardinal characteristic for all types of MD is a progressive degeneration of muscle cells and/or muscle fibres. Different muscle groups are involved in the different types of MD (Brooke 1986, Walton & Gardner-Medwin 1988).

Myotonic muscular dystrophy (MyD) is often considered separately from the other forms of MD. Onset may be at any time from birth to adulthood. There is often an effect on the endocrine system and the cardiac muscle, and there are often cataract, dysphagia, frontal baldness and mental changes. MyD is therefore regarded as a systemic disease. Symptoms vary in their degree of severity and comprise myotonia and muscular weakness, where the distal muscles are the first to be affected. Since proximal muscles are involved there can be great difficulty in walking. Facial muscles are also involved, giving rise to dysarthria and "the myotonic face" (Ahlström et al. 1993, Forsberg 1990, Griggs et al. 1995, Harper 1985, Sirotkin-Roses 1991, Walton & Gardner-Medwin 1988).

Some of the diagnoses included in the *Proximal muscular dystrophy group* are Becker MD, limb–girdle MD, facioscapulohumeral MD and Emery–Dreifuss MD. Becker MD affects the pelvic girdle in particular. Onset is usually between the ages of 5 and 15 years, but may be later, and the majority of patients are still able to walk when they are 20. The disease progresses more slowly and more gently than does Duchenne MD (the onset of which is in early childhood) (Griggs et al. 1995, Harper 1985, Kakulas 1999, McDonald et al. 1995, Walton & Gardner-Medwin 1988).

Limb–girdle MD is the least well-defined in this group. Onset is usually in late childhood or the teens. The first symptom is that the person finds it difficult to walk, because it is the pelvic girdle in particular which is affected. The muscle involvement resembles that of Becker MD, but is characterised by a great variability, ranging from severe to milder forms. There is no facial involvement and the upper extremities and the shoulder girdle are seldom affected (Griggs et al. 1995, Harper 1985, Stübgen & Stipp 1997).

Facioscapulohumeral MD is the most benign of the progressive muscular dystrophies. The symptoms usually emerge between the ages of 7 and 20. Characteristic of the disease is involvement of the shoulder girdle, in combination with weakness of the facial and neck muscles. The upper extremities are most often affected. Speech may be affected because of facial muscle weakness. The pelvic girdle and the thigh muscles are only moderately affected, which means that the ability to walk is retained until the advanced stage (Griggs et al. 1995, Harper 1985, Kilmer et al. 1995).

The onset of Emery–Dreifuss MD occurs in early childhood or in the teens. Muscular weakness in the upper extremities involves the biceps and triceps. In the legs, the peroneal muscles are more affected than the proximal muscles. Shortening of the Achilles tendon may occur, affecting the ability to walk. Marked contractures may develop at an early stage, most often affecting the joints of the elbow and neck. Some persons are only mildly affected throughout their lives. Cardiac involvement is an important and potentially life-threatening feature, even for patients with minimal limb involvement (Griggs et al. 1995).

Myopathia distalis tarda hereditaria (MDTH), also known as Welander distal myopathy, seldom afflicts persons under 40 and has a slow rate of progress. Although it is the commonest of the distal myopathies, it occurs hardly anywhere

except in Sweden and parts of Finland. The distal muscular weakness usually occurs first in the muscles of the thumb and/or index finger, giving rise to a clumsiness in precision movements which spreads to the other fingers and later the arms. Gradually, distal muscular weakness develops in the lower extremities, and eventually there is difficulty in walking (Borg et al. 1998, Harper 1985, Åhlberg et al. 1997). Except for MDTH, all types of MD can affect the respiratory musculature. Because of the different natural histories of these diseases it is of interest to compare their consequences for the afflicted persons..

Activities of daily living

Usually the term occupation includes several activities. Occupation refers to engagement in activities, tasks and roles for the purposes of meeting the requirements of living (Christiansen 1991, Levine & Brayley 1997) It is central in human experience, supplying a person with what is needed to meet the needs for survival (Wilcock 1998). Occupation is a part of everyday life, and demonstrates the person's cultural, intellectual, moral and physical attributes. Only by doing can a person tell what he/she is or what he/she hopes to be. When people have capacities to meet the challenges of their occupations, this results in an optimal state relating to health, well-being and quality of life (Christiansen et al. 1999). Activity and rest are important components of healthy living (Wilcock 1998) and the concept of activity is central in occupational therapy (Törnquist 1995). WHO (2000) defines activity as follows: "Activity is the execution of a task or action by an individual. It represents the individual perspective of functioning" (p. 99). Wilcock (1998) focuses on activity as the state of being active and by this she means that activity refers to actions. Activity is something that may be used as a therapeutic aim, and it may be passive or active (Levine & Brayley 1997). The term "activity limitations" in current International Classification of Impairment, Disability and Handicap (ICIDH) (2000), has replaced "disability" in the earlier ICIDH (1980). Disability was defined as

follows “difficulties an individual may have in executing activities” (World Health Organization 1980, p.99). Disability is not included in the latest version of the ICIDH-2 (World Health Organization 2000). An activity limitation may range from a slight to a severe deviation in terms of quality or quantity in performing the activity in a manner or to the extent that is expected of people without the condition of illhealth (World Health Organization 2000).

Activities of daily living may be taken to include personal care, home management, leisure, studies and work. These activities are usually described in two categories: Personal activities of daily living (P-ADL) and Instrumental activities of daily living (I-ADL). P-ADL may comprise toileting, dressing, eating, grooming, ambulation and bathing. I-ADL may comprise communication, shopping, cooking, housekeeping, laundry and transportation (Sonn & Hulter Åsberg 1991, Törnquist 1995). Disability is often assessed in terms of dependence and independence from other persons in activities of daily living (Sonn & Hulter Åsberg 1991). A person’s environment often plays a decisive part, and it can make activity easier as well as harder (Törnquist 1995).

Coping

The concept of coping is frequently used in stress research to denote phenomena related to the management of stress and difficulties. This concept is used to give a theoretical explanation of how people think, feel and behave in stressful situations in attempts at managing these. Stress is a natural part of life, and the managing of stress is of decisive importance for the individual’s well-being. Coping may be seen as a process including cognitive appraisal and reappraisal of the dynamic person-environment relationship (Lazarus 1991, Lazarus & Folkman 1984). Lazarus (1984) defines coping as “constantly changing cognitive and behavioural efforts to manage specific external and/or internal demands that are appraised as taxing or exceeding

the resources of the person" (p. 141). Usually, two types of coping are distinguished: emotion-focused coping and problem-focused coping. Emotion-focused coping concerns the ways in which the individual handles emotions associated with stressful situations or activities. Emotion-focused coping aims to alleviate the stress reaction even if the actual circumstances remain unaltered. Problem-focused coping denotes steps taken to handle the source of the stress, i.e. to address the problem itself Research has shown that individuals usually employ more than one coping strategy in one and the same situation. Thus, emotion-focused and problem-focused coping may occur together (Ahlström 1994, Christiansen 1991).

Living with a progressive disease means living with progressive impairment and disability for a number of years. It is of great importance for the person's well-being how he or she manages the stresses occasioned by the disability (Benner & Wrubel 1989). This situation places great demands on the individual regarding coping. In order to attain a successful social adjustment, the individual must use more than one coping strategy (Christiansen 1991). A study by McColl et al (1995) reveals that social support may exert a direct effect on coping, whereas the reverse was not demonstrated. The person's capacity for coping partly depends on the support he or she receives from the family and the social network, and social support is regarded as a coping resource (Lazarus & Folkman 1984, McColl et al. 1995). Previous studies have revealed that persons who are suffering from severe progressive diseases are often able to manage difficulties in a fashion that is successful to themselves (Ahlström & Karlsson 2000, Ahlström & Sjödén 1996, Ramund & Stensman 1988). Altering life orientation and life values appears to be a coping strategy used by persons with chronic illness (Ahlström & Sjödén 1996). Coping includes all that the person does to handle stress regardless of whether the results are good or bad. The way in which a person manages a situation depends very much on the resources available within that person and in the social network. When coping and its results

are being measured, these two factors must be kept apart and assessed separately (Lazarus & Folkman 1984).

Quality of life

The present thesis employs the concept of quality of life to reflect satisfaction with activities of daily living, health-related quality of life and well-being. According to the WHOQOL Group (1995), quality of life is defined as "An individual's perception of their position in life in the context of the culture and value systems in which they live and in relation to their goals, expectations, standards and concerns" (p.1405). This is a broad concept, covering the person's physical and mental health and their relationship to salient features of the environment (Abresch et al. 1998, The WHOQOL Group 1995). Thus the term quality of life is closely related to 'subjective satisfaction' and 'a sense of well-being' (Mayers 1995). Quality of life is subjective and for this reason, self-report is considered to be the most valuable method for acquiring data (Bach 1995, Post et al. 1999). Quality of life is often used as an indicator of the extent to which interventions are beneficial for the person with chronic disease. When physical status and functional abilities are the focus of research this is often referred to as health-related quality of life. In this case, quality of life may be measured in terms of the relative absence of negative experiences (Ahlström 1994, Naess 1994, Spilker 1990).

The term satisfaction reflects an important dimension of a person's evaluation of specific areas of life. The concept of quality of life has a wider frame of reference than satisfaction, including more reactions, i.e. experience of self-esteem, meaning, anxiety and restlessness (Naess et al. 1987). Satisfaction with activities depends on individual performance components: physical, mental, socio-cultural and spiritual. These components give value and meaning to life in interaction with the environment (Post et al. 1999, Whalley Hammel 1995). A Swedish research group

has described eight domain-specific items concerned with life satisfaction (Self-care ADL, leisure, vocational situation, financial situation, sexual life, partnership, family life, and contact with friends and acquaintances) and their influence on satisfaction with life as a whole (happiness) (Brännholm & Erhardsson 1994, Fugl-Meyer et al. 1991). In the study by Fugl-Meyer et al (1991), these authors found that all eight domains were closely and positively associated with level of happiness in a group of 201 persons, aged 22-55 years.

Quality of life is often used as an indicator of the extent to which interventions are beneficial for the person with a chronic disease. When physical status and functional abilities are focus of the research, this being known as health-related quality of life. In this case quality of life is often assessed in terms of the relative absence of negative experiences (Ahlström 1994, Spilker 1990, Naess 1994).

The World Health Organization (2000) defines well-being as "a general term encompassing the total universe of human life domains including physical, mental and social aspects, that make up what can be called a good life. Health domains are a subset of domains that make up the total universe of human life" (p.163). Wilcock (1998) defines well-being as "a subjective assessment of health, which is less concerned with biological function than with feelings such as self-esteem and a sense of belonging through integration" (p. 98). Happiness, success and health are important parts of well-being, that are influenced by the person's environment, physical and mental strength, health and personality (Wilcock 1998). Well-being is related to social support, community cohesion, marital state, education and religious attitudes, beliefs and activities (Nordenfelt 1994, Wilcock 1998). These factors affect the person's well-being in either a positive or a negative direction (Nordenfelt 1994). Physical-, mental- and social well-being are parts of an integrated system, and cannot easily be separated. When people experience occupational well-being in a physical sense, it becomes possible for them to perform activities which they are in need of or

which they want to do, which makes effective use possible of both social and mental capacity (Wilcock 1998). Quality of life and well-being are sometimes considered to be synonymous concepts (Naess 1994, Post et al. 1999). Quality of life is regarded as a psychological concept, and Naess (1987) puts the main emphasis on the emotional experience of having a good life. The definition of Naess (1987) rests on four values in life, a person being considered to have a good quality of life when he or she is active, relates well to others, and possesses self-esteem and a basic mood of happiness.

The contents and definition of the concepts of quality of life vary with respect to its theoretical origin and the empirical field of application (Abresch et al. 1998, Calman 1987, Post et al. 1999). There is, however, a consensus that quality of life as a concept is subjective, individual and multi-dimensional (Abresch et al. 1998, Ahlström 1994, Post et al. 1999). Over the past ten years, quality of life has successively become the overriding goal of successful rehabilitation (Abresch et al. 1998). Only one study on quality of life in adults with muscular dystrophy has been identified in the literature (Ahlström 1994). In the present thesis, three dimensions of quality of life were employed in order to obtain a broader knowledge of quality of life as a consequence of living with MD. Satisfaction with ADL, well-being and health-related quality of life are complementary dimensions of quality of life.

Rehabilitation

The health panorama in Sweden has been changing in recent decades, and between 1960 and 1990 health care costs increased by 235% (Höök 1995). Parallel to improvements in medical technology, there has been a continuing increase in the number of patients surviving illnesses and accidents. This has brought about an increased need for rehabilitation with heavier demands (Baum 1991, Kielhofner

1992). Earlier discharge from hospital increases the need for rehabilitation resources (Baum 1991).

Rehabilitation has a long tradition, and it is an essential part of medical care today, but it is defined and interpreted in different ways. The term rehabilitation refers to "a process aimed at enabling persons with disabilities to reach and maintain their optimal physical, sensory, intellectual, psychiatric and/or social functional levels, thus providing them with the tools to change their lives towards a higher level of independence" (United Nations 1994, rule 3, paragraph 23). "Rehabilitation may include measures to provide and/or restore functions, or compensate for the loss or absence of a function or for an activity limitation. The rehabilitation process does not involve initial medical care. It includes a wide range of measures and activities from more basic and general rehabilitation to goal-oriented activities, for instance, vocational rehabilitation" (United Nations 1994, rule 3, paragraph 23).

The process of rehabilitation should include assessment, goal description, planning, intervention and evaluation. It is necessary that the goals are formulated clearly and that they are adapted to the individual's potentials and needs (Söderback 1995). Two essentials for attaining the goals of rehabilitation are for the right measure to be taken at the right time, and for the rehabilitation to be conducted in a way that lies as close as possible to the everyday life of the individual (Söderback 1995). Rehabilitation is affected both by the patient's own activities and by the various interventions of the rehabilitation team. The patient and the team members often have different perspectives on the aims of the rehabilitation, which may result in conflicts. For this to be avoided, it is important that staff and patient set up the goals together (Söderback 1995). Rehabilitation should include social support and a teaching process enabling the individual to learn coping strategies with the aim of improving well-being (Bach 1995, McColl 1995, Robert et al. 1995).

The provision of social support is an important task during rehabilitation and in the broader context of community integration (Choi & Wodarski 1996, McColl 1995, McColl & Skinner 1995, Robert et al. 1995). A number of studies show that social support has positive effects for persons with chronic illness (McColl 1995, McColl et al. 1995, McColl & Skinner 1995). Social support offers a way of understanding individual health (McColl & Skinner 1995, Sarason et al. 1990, Thoits 1986), and it has been shown to have a highly beneficial effect on life stress and chronic illness (Kaplan & Toshima 1990, Sarason et al. 1990). Sarason et al (1990) mean that measures of social support can be divided in three categories: 1) The network model focuses on the individual's social integration into a group and the interconnectedness of those within that group. 2) The received support model focuses on what the person actually receives or reported to have received, i.e. through the intervention in the rehabilitation program. 3) The perceived support model focused on support the person believes to be available if he or she should need it (Sarason et al. 1990). Social support consists of the actions that others perform to assist a particular person (Thoits 1986). House's (1981) three-factor model of social support was used as a guide to plan the rehabilitation programme in the present study: Instrumental support, Emotional support, and Information support. House's theory of social support is the basis for Thoits' (1986) conceptualisation of social support and coping, and Thoits (1986) considers social support to be a form of coping assistance. The limitations in performing daily activities also create life strains that require continuous coping (Boutaugh & Brady 1996).

The definition formulated by the research group in charge of the rehabilitation programme, carried out as a part of this thesis, is in some ways similar to that of WHO, but it places a clearer emphasis on quality of life: "Rehabilitation means that staff in an interactive process confirm the competence of the individual, creating increased resources for that individual. The aim is to improve quality of life for an individual who has difficulties in solving or managing experienced problems as a

result of illness, experienced illness and/or disabilities" (Ahlström & Gunnarsson 1997, p. 19). This means that rehabilitation activities should enable the individual to live a life as good as possible despite MD. The first reason for developing a rehabilitation programme was that a specific programme for adults with MD was lacking in the literature. The second reason was that earlier research has shown that persons with MD experience many illness-related problems in daily life (Ahlström & Sjödén 1996).

The ability to manage activities in everyday life varies greatly between persons, according to their disability. This means that persons with different types of MD require different types of treatment. Over and above the treatment of symptoms and improvement in physical function, it is necessary for rehabilitation programmes to deal with emotional issues such as stress, hopefulness and self-esteem (Pain et al. 1998), the physical and psychosocial environment and cognitive aspects (Johnston et al. 1992, Söderback 1995, Wressle et al. 1999).

Most of the existing research on muscular dystrophy is concerned with fundamental pathophysiological and genetic questions, or with children with MD. Studies on muscular dystrophy in adults are few and of recent date. Traditionally, in the nursing and medical services there has been a sceptical attitude towards the possible usefulness of rehabilitation for persons with muscular dystrophy, since the illness is both progressive and incurable. For this reason, it has been the aim of the present dissertation to develop increase our understanding both of what it means to live with muscular dystrophy and also of how an individually tailored rehabilitation programme is perceived by the patient.

AIMS

The overall aim of this thesis is to gain further knowledge about what it means to live with muscular dystrophy (MD) and about patient perceptions of rehabilitation. Specific aims are as follows:

- To study illness experiences and their possible variation between different types of MD (Study I)
- To study activities of daily living (Studies I, II, III, IV)
- To study coping in activities of daily living (Studies II, IV)
- To study experience of quality of life (Studies II, III, IV)
- To study changes over time and relationships between activities of daily living, coping and quality of life (Study IV)
- To study whether there are any gender differences and differences between various types of MD with respect to activities of daily living, coping and quality of life (Studies II, III, IV)
- To study perceived support in a rehabilitation programme designed for persons with muscular dystrophy (Study V).

MATERIAL AND METHODS

Designs

This thesis employs qualitative (Studies I and V) as well as quantitative methods (Studies II, III and IV) and includes a study on instrument development for problem-focused coping (Study II). The following phenomena were studied among adults with muscular dystrophy: Illness experience, Activities of daily living (ADL), Coping, Quality of life, and Perceived support in rehabilitation.

Studies I and V have a descriptive design and are based on qualitative interviews. Illness experience was assessed by two semi-structured interviews (Study I). The transcribed interviews were analysed by narrative content analysis (Lieblich et al. 1998). Perceived support in rehabilitation was assessed by a semi-structured interview (Study V) and analysed by a phenomenological approach inspired by Giorgi (1996).

Studies II and III have a mainly descriptive design and are based on questionnaires about activities of daily living (Studies II and III), coping (Study II), and quality of life (Studies II and III). In Study III the relations between activities of daily living, coping and quality of life were investigated. Study IV is a prospective five-year study (1991-1996), focusing on changes regarding disability, coping and quality of life in adults with MD. It also includes an investigation of the relations between these variables. Studies II-IV include investigations of differences in these respects depending on gender or diagnosis.

Subjects

The study group comprises adults with muscular dystrophy (MD) from three count counties in Sweden; the county of Örebro in the central part of Sweden (Studies I-V), the adjacent county of Östergötland (Study III), and the county of Norrbotten in the northern part of the country (Study II). In Study II, a comparison group was included consisting of persons with postpolio syndrome (PPS). The common characteristic of all these persons is that they have progressive muscular weakness. Because of the various natural histories of the different diagnoses of muscular dystrophies, they were compared in this thesis. The subjects in Östergötland were included mainly to evaluate the rehabilitation programme with qualitative parameters (Ahlström et al. unpublished). The number of persons and their characteristics are shown in Table I.

Persons with muscular dystrophy in the county of Örebro. This subgroup was identified in a comprehensive survey in the county of Örebro (Ahlström et al. 1993) and has been included in previous research (Ahlström 1994). It comprises persons with different types of diagnosis of MD: myotonic muscular dystrophy (MyD), myopathia distalis tarda hereditaria (MDTH) and various types of proximal muscular dystrophy: Becker MD, limb-girdle MD, facioscapulohumeral MD and Emery-Dreifuss MD. The subgroup in the county of Örebro is included in all five studies, although with varying numbers of persons as shown in Table I. These persons have the following levels of education: 38 persons have compulsory school or earlier equivalent, 15 upper secondary school, and 5 a university or college education.

Persons with muscular dystrophy in Östergötland. This subgroup was selected from the patient file at the Department of Neurology, Linköping University Hospital (Table I). The same selection criteria regarding diagnosis and age, as were used in the county of Örebro, were employed. The subgroup comprises 39 persons (Study III). The educational levels in this subgroup are unknown.

Table I. Demographic data on persons with MD and the PPS (Studies I-V)

	Study I	Study II	Study III	Study IV	Study V
Offered participation	67	124	79	57	52
EXCLUDED					
Not progressive MD	2	-	-	2	-
DROPOUTS					
Poor health	-	4	2	2	1
Deceased				5	
Declined	7	-	-	-	14
Long visit abroad	-	1	-	-	-
Change of domicile	-	-	-	2	-
Language difficulties	-	-	-	1	-
Number of subjects	**58**	**119**	**77**	**45**	**37**
COUNTIES/ DIAGNOSES					
Örebro	58	33	38	45	37
Östergötland	-	-	39	-	-
Norrbotten	-	46	-	-	-
PPS Comparison group	-	40	-	-	-
GENDER					
Men	24 (41%)	57 (48%)	30 (39%)	16 (36%)	16 (57%)
Women	34 (59%)	62 (52%)	47 (61%)	29 (64%)	21 (43%)
AGE mean (range)	47 (21-64)	52 (20-81)	49 (24-74)	44 (25-64)	50 (23-69)
DISEASE DURATION mean (range)	22 (3-43)	Unknown	23 (4-69)	24 (8-46)	24 (6-45)

Persons with muscular dystrophy in Norrbotten. This subgroup comprises 46 adults with myotonic muscular dystrophy (MyD) (Study II). These persons were identified in a survey of the population of Norrbotten (Lexell et al. 1999). This study included the first 46 patients of a total of 102 identified. The educational level in this subgroup is unknown.

Comparison group: Persons with postpolio syndrome (PPS). This subgroup comprises 40 persons with symptoms characteristic of PPS. These persons were

identified in the earlier population study in the county of Örebro (Ahlström et al. 1993) and have been included as a comparison group for testing the APC (Study II). The educational level in this subgroup is unknown.

Procedure and data collection

An overview of the data collection methods used in the studies is shown in Table II.

Illness experiences

Illness experience was described through interviews on two occasions in patients' homes at an interval of 3 months in 1990-1991 (Study I). The interviews were conducted in the form of conversations and with the assistance of two interview guides. The main questions concerned five topics: Onset of disease and learning of diagnosis, Experiencing the disease, Managing problems in everyday life, Changes experienced in everyday life due to the disease, and Thoughts of the future. The time dimension of past, present and future formed the structure for the interview. Follow-up questions were asked in order to elicit answers in narrative form. The number of such questions varied according to the comprehensiveness of the responses and the range of problems brought up. The interviews lasted an average of one and a half hours. They were taped and then transcribed verbatim.

Activities of daily living

Data on activities of daily living (ADL) were based on three self-report ADL instruments: Assessment of Problem-focused Coping (APC) which has been recently developed as a self-report assessment instrument for problems, problem-focused coping and satisfaction with ADL (Study II), the ADL Staircase (Hulter Åsberg & Sonn 1989, Sonn & Hulter Åsberg 1991), and the Self-report ADL (Ahlström 1994, Ahlström & Gunnarsson 1996) (Studies III, IV).

Table II. Overview of data collection in Studies I-V

Instruments	Study I n=58	Study II n=119	Study III n=77	Study IV n=45	Study V n=37
ILLNESS EXPERIENCE					
Semi-structured interview	X	-	-	-	-
ACTIVITIES OF DAILY LIVING					
Assessment of Problem-focused Coping (APC)	-	X	-	-	-
ADL Staircase	-	-	X	X	-
Self-report ADL	-	-	X	X	-
COPING					
Assessment of Problem-focused coping (APC)	-	X	-	-	-
Mental Adjustment to Cancer (MAC) scale	-	-	-	X	-
QUALITY OF LIFE					
Satisfaction with ADL					
Assessment of Problem-focused Coping (APC)	-	X	-	-	-
Well-being					
Psychosocial well-being questionnaire	-	-	-	X	-
Health-related quality of life					
Sickness Impact Profile (SIP)	-	-	-	X	-
Quality of Life Profile	-	-	X	-	-
PERCEIVED SUPPORT IN REHABILITATION					
Semi-structured interview	-	-	-	-	X

Data were collected by the APC on one occasion during the period from September 1995 to May 1996 for the three study groups. The data collection for the study persons with MD in the counties of Örebro and Norrbotten was performed in conjunction with a hospital visit. Persons in the PPS completed the APC during a home visit (Study II).

The data collection using the ADL Staircase was conducted in the form of structured interview questions at the hospital for the persons in Örebro. For the persons in Östergötland, the data collection was done in their homes (Study III). The data collection at the five-year follow up in Study IV with the ADL Staircase was made in 1991 at a home interview, and in 1996 as an interview in conjunction with a hospital visit. The self-report ADL was completed by both groups in a postal questionnaire in Studies III and IV.

Coping

Data regarding coping were gathered with two coping questionnaires, the APC (Study II), and the Mental Adjustment to Cancer (MAC) scale (Berglund et al. 1994, Greer et al. 1992) (Study IV). The data collection using the APC has been described above for activities of daily living. Persons in the PPS group completed the APC during a home visit. The MAC was completed in a postal questionnaire in 1991 and 1996.

Quality of life

Data on quality of life were collected with four self-report instruments. These instruments are APC (Study II), the Psychosocial well-being questionnaire (Kaasa et al. 1988a, Kaasa et al. 1988b) (Study IV), the Sickness Impact Profile (SIP) (Bergner et al. 1976a, Sullivan et al. 1986) (Study IV), and the Quality of Life Profile (Ahlström & Karlsson 2000) (Study III).

Data regarding satisfaction with ADL were collected using the APC. The data collection with the APC has been described above for activities of daily living (Study II). The interviewees completed the Quality of Life Profile independently, but in the presence of interviewers (Study III). The Psychosocial well-being questionnaire and the SIP were completed in a postal questionnaire in 1991 and 1996 in Study IV.

Perceived support in rehabilitation

Perceived support in rehabilitation was investigated through interviews intended as a qualitative evaluation (Patton 1990) individually adapted and tailored rehabilitation programme for adults with MD. The rehabilitation programme was performed by a multi-professional team (Study V). The programme was administered in six groups that were formed according to diagnosis, age and degree of disability so that they were as homogeneous as possible. This programme was designed on the basis of previous research (Ahlström 1994) and included of 4 sessions from the spring of 1995 to the autumn of 1996, including individual (75%) as well as group measures (25%).

Session 1: One day. *Contact day*. The participants met the rehabilitation team for individual examination. Individual rehabilitation plans were drawn up. Session 2: Four days (two weeks later). *Programme days*. Individual measures in accordance with rehabilitation plans, involving e.g. adaptation of home, trying out of technical aids, examination, information concerning self-care programmes and (if necessary) consultation with the orthopaedic shoemaker. Session 3: Two-three days (six months later). *Follow-up programme*. Completion of earlier individual measures in accordance with the rehabilitation plans, and group activities. The number of days was decided by the size of the groups. Session 4: Two days (18 months later). *Concluding measures*. Individual measures, individualised discussion for the purpose of evaluation and group discussion. Group activities in sessions 2-4 included discussions about the participants' life situation and the situation regarding relatives, education, water gymnastics, and evening activities. Planning of the design of further rehabilitation was also performed.

An interview was conducted at the hospital at the end of the programme. An interview guide was used, based on the sub-goals of the rehabilitation programme (Ahlström & Gunnarsson 1997) and three types of social support (House 1981):

1. Instrumental support (e.g. 'Please describe any problem that you have had help in solving'), 2. Informative support (e.g. 'Please describe your experiences of the training'), and 3. Emotional support (e.g. 'Please describe how you felt about the discussion of questions dealing with your everyday life or that of your relatives'). Follow-up questions were asked within all three areas for purposes of clarification and to obtain more detailed accounts. The interviewees were encouraged to start by describing how they had perceived the rehabilitation. The interview lasted 45-90 minutes and was taped and then transcribed verbatim.

Instruments

The instruments about ADL, coping and quality of life used in Studies II, III, and IV are presented below and in Table II.

Assessment of Problem-focused Coping (APC) (Study II)

This is a self-report instrument consisting of two parts. It has been developed to highlight the patient's competence in solving his or her own problems. Only Part 1 is employed in this study. It has been developed primarily for persons with muscular dystrophy and other types of muscular weakness. A manual has been developed for use of the APC (Nätterlund & Ahlström 1997). The APC items were derived from 810 interview statements on "difficulties experienced as a direct result of disease" in a study by Ahlström and Sjödén (1996). This yielded a total of 32 basic activities that were refined, broadened and classified into five general Occupational Forms: Personal Care, Home Management, Leisure, Mobility and transportation, and Work (Table III). Basic activities are wide-ranging activities that occur in people's everyday life. There are between 2 and 11 actions for each basic activity. An Action is a more specific and targeted, for instance, "wash yourself", which is one of the actions within the Personal care of Occupational Form (Table III). There is no ranking of the actions by levels of difficulty.

Table III. Occupational Forms, basic activities and actions in each of the basic activities of the APC (Study II)

Occupational Forms	Basic activities	Number of actions
PERSONAL CARE	Personal hygiene	9
	Dressing and undressing	6
	Eating and drinking	3
	Toilet	5
HOME MANAGEMENT	Shopping	9
	Cooking	11
	Washing up	5
	Baking	4
	Washing	5
	Cleaning	6
	Mending clothes	5
	Child care	4
LEISURE	Reading	4
	Writing	3
	Music	7
	Handicraft	6
	Cinema, theatre, concerts etc	2
	Gardening	7
	Keeping fit	3
	Swimming	3
	Skiing	5
	Ballsports	8
	Skating	3
MOBILITY AND TRANSPORTATION	Indoor mobility	8
	Outdoor mobility	7
	Drive car	3
	Cycle	2
	Public transportation	3
WORK	Physically easy work	10
	Physically demanding work	11
	Work under stress	7
	Verbal communication	3

Problems. The questions on problems concern performance of the basic activities, using the following response alternatives: Not doing the activity, Doing it without problems, and Doing it with problems. The individual is asked to mark one of these alternatives for each of the 32 basic activities.

Problem-focused coping. The coping theory of Lazarus and Folkman (1984) and categories in a previous study (Ahlström & Sjödén 1996) formed the basis for five problem-focused coping categories and the response alternatives for each of the 32 basic activities were: "Devices and tricks": I usually perform the activities in a new/different way, e.g. with new muscle movements or by using devices and tricks; "Aids": I usually perform the activities by employing technical aids; "Own technical solutions": I usually perform the activities using my own technical solutions; "Accept help": I usually accept help or ask someone else to do it; and "Avoid": I usually avoid doing things that cause problems (Ahlström 1994, Ahlström & Sjödén 1996). The individual is asked to mark those alternatives that apply to his or her current situation. Thus, an individual could mark more than one alternative of coping for each of the 32 basic activities.

Satisfaction. The last section concerns satisfaction with each of the 32 actions. Satisfaction involves four response alternatives, coded in the following way: 3 = functions well, 2 = does not function very well but I accept it, 1 = does not function very well but I consider that it cannot be changed, and 0 = does not function very well and I want to find a better solution. The individual is asked to mark one of these four alternatives for each of the 32 activities even if he or she does not at present perform the activity.

Part 1 of the APC comprises 12 response alternatives for each of the 32 basic activities, 3 of them dealing with *Problems*, 5 with *Problem-focused coping* and 4 with *Satisfaction*. The numbers of responses (Problem and Problem-focused coping) and values (Satisfaction) for all basic activities within each Occupational Form were added and expressed as a percentage of the maximum value obtainable for each particular Occupational Form. Nine percentage scores were calculated for each of the five Occupational Forms: three for problems, five for problem-focused coping, and one for satisfaction (total score). Moreover, a total score was calculated

for Problem-focused coping (total index). High values mean more problems with activities of daily living, more use of problem-focused coping and high satisfaction with activities of daily living.

The APC was tested in two pilot studies. The persons with MD in the county of Örebro completed this instrument on two occasions, both in the pilot study in conjunction with the start of the rehabilitation programme and six months later. This demonstrating that the APC questions were relevant and comprehensible to the study persons (Study II). The result of a psychometric test demonstrated reasonable internal consistency (alpha, mean 0.70, range 0.41-0.90), confirming that the instrument possesses acceptable homogeneity. Internal consistency was calculated for problems, coping, and satisfaction scores in each of the Occupation Forms: Personal Care, Home management, Leisure, Mobility and transportation (Work was excluded because of only eight study persons were in employment). Test-retest reliability over a period of six weeks varied from low (rho 0.33) to high (rho 1.0) for different basic activities (Tollén & Ahlström 1998, 2000).

ADL Staircase (Studies III and IV)

The ADL Staircase is a further development of the Katz ADL Index (Katz et al. 1970). It measures dependence/independence with regard to ten activities of ADL. The ability to perform each activity is assessed on a three-grade scale: independent, partly independent, or dependent. For each item, each step of the scale carries specific definitions. Six items concern personal care (P–ADL), i.e. bathing, dressing, toileting, transfer, continence and feeding (Katz et al. 1963, Katz et al. 1970, Katz et al. 1983, Sonn & Hulter Åsberg 1991) and four concern instrumental activities of daily living (I–ADL), i.e. cleaning, shopping, transportation and cooking (Hulter Åsberg & Sonn 1989, Sonn & Hulter Åsberg 1991).

The ADL Staircase is graded from 0 to 10, and the grades are expressed as follows: *Independent, grade 0,* = Independent in all ten activities; *Dependent in I-ADL, grades 1-4*, 1= Dependent in one activity, 2= Dependent in cleaning and one more activity, 3= Dependent in cleaning, shopping and one more activity, 4= Dependent in cleaning, shopping, transportation and one more activity; and *Dependent in I- and P-ADL, grades 5-10,* 5= Dependent in all I-ADL and one P-ADL, 6= Dependent in all I-ADL, bathing and one more P-ADL, 7= Dependent in all I-ADL, bathing, dressing, and one more P-ADL, 8= Dependent in all I-ADL, bathing, dressing, toileting and one more P-ADL, 9= Dependent in all I-ADL, bathing, dressing, toileting, transfer and one more P-ADL, and 10= Dependent in all activities. The category *Other* is employed for persons dependent in two or more activities but not classifiable as above. A prerequisite for a reliable result is that this category does not exceed 5%. The instrument has a high degree of reliability and validity for several diagnoses (Hulter Åsberg & Sonn 1989, Katz et al. 1963, Sonn 1995, Sonn & Hulter Åsberg 1991). The higher the value, the greater the degree of dependence in ADL.

Self-report ADL (Studies III and IV)

This self-report instrument has been developed inductively from interviews with persons with MD (Ahlström 1994, Ahlström & Gunnarsson 1996). It comprises 29 items concerning difficulties with activities of daily living. In the revised version, the subject marks one of the following answers: “No difficulty” or “Can do it but don’t” (0.0), “Sometimes difficulty” (0.33), “Always difficulty” (0.66) and “Fails” (1.0). The results are given in the form of a percentage of the maximum number of points with regard to each of the following indices, as developed from a factor analysis (Ahlström & Gunnarsson 1996): Ambulation (10 items), Arm strength (9 items, Finger strength (5 items), Finger subtle function (4 items) and Total index (29 items). The instrument has been tested and has been found to show good construct validity and to have empirical relevance (Ahlström 1994, Ahlström &

Gunnarsson 1996, Ahlström & Karlsson 2000). The higher the percentage score, the greater the difficulty.

Mental adjustment to cancer (MAC) scale (Study IV)

The MAC scale is a self-report instrument, developed for patients with cancer, for assessment of emotion-focused/cognitive coping. The MAC scale consists of 40 items, each with four alternative answers according to a 1-4 Likert scale, 'Definitely does not apply to me', 'Applies partly to me', 'Applies quite well to me', 'Definitely applies to me' (Berglund et al. 1994). The MAC scale covers five categories of coping: 'Fighting spirit' (16 items), 'Helplessness/ hopelessness' (6 items), 'Anxious preoccupation' (9 items), 'Fatalism' (8 items), and 'Avoidance' (1 item). Responses are totalled within each category. Standardised scores were computed by subtracting the sample mean from the individual scores and dividing by the sample standard deviation to provide a Z-score. The category producing the highest Z-score was selected as the dominant coping category for the patient (Greer et al. 1992, Lampic et al. 1994, Nordin 1998). The instrument has been judged to be acceptably reliable and valid (Greer et al. 1989, Watson et al. 1989, Watson et al. 1988). The MAC has been translated to Swedish (Berglund et al. 1994) and adapted for patients with MD (Ahlström & Sjödén 1993, 1996) by replacing the word 'cancer' with 'muscular dystrophy', and by modifying three items as indicated by the italicised parts as follows: 'I am firmly convinced that I am *not going to get worse*' (originally: 'going to get better'), 'I have made up my mind that *the worst is over now and I want to forget that I have the disease*' (originally: 'it is over now and I am leaving the disease behind me'), 'I am worried that the *muscular dystrophy is going to get worse*' (originally: 'cancer is going to come back or get worse'). This adapted version of the questionnaire has shown acceptable reliability and validity as compared with results based on interview data (Ahlström & Sjödén 1996).

Psychosocial well-being questionnaire (Study IV)

This instrument comprises twelve questions concerning psychosocial well-being during the past seven days. The *'Psychosocial well-being Index'* consists of ten questions (five positive and five negative), each pair covering a particular psychosocial issue. The answers are given in accordance with a five-point scale of possible answers ranging from 'never' to 'all the time' (giving values from 1 to 5). One question is designed to assess *Satisfaction* with life and one question *Happiness*, and these responses are given on a seven-grade scale. A low score represents a high quality of life. The instrument has been found to possess acceptable reliability and validity in the original studies of cancer patients (Kaasa et al. 1988a, Kaasa et al. 1988b). It is based on Naess´s definition of quality of life (Naess et al. 1987).

Sickness Impact Profile (SIP) (Study IV)

This instrument measures health-related quality of life in terms of dysfunction. It is a self-evaluation instrument comprising 136 items divided into the following twelve categories: Sleep and rest, Eating, Emotional behaviour, Body care and movement, Home management, Mobility, Social interaction, Ambulation, Alertness behaviour, Communication, Work, and Recreation and pastimes. These categories are in turn categorised in three indices: 'Physical Index' (Ambulation, Mobility, Body care and movement), 'Psychosocial Index' (Social interaction, Alertness behaviour, Emotional behaviour, Communication), and 'Independent Categories' (Sleep and rest, Eating, Work, Home management, Recreation and pastimes). Together, all categories form the 'Total Index'. Affirmative answers are given weighted points in accordance with the manual and the sum of each index and categories are presented as percentages of the maximum scores (Bergner et al. 1976a). The higher the percentage, the poorer the health-related quality of life. Developed in the USA in the 70s, the SIP was translated into Swedish in 1981, and both versions have proved

to have good psychometric qualities (Bergner et al. 1976a, Bergner et al. 1976b, Sullivan 1988, Sullivan et al. 1986, Sullivan et al. 1990).

Quality of Life Profile (Study III).

This instrument is a self-assessment questionnaire and has been developed inductively on the basis of 120 interviews with people with MD (Ahlström & Karlsson 2000). It is designed for persons with progressive diseases, and can be regarded as a health-related quality of life instrument. The instrument has 44 items, grouped as follows: *Life-picture* (I item), *Life-areas* (19 items), *Problems* (15 items) and *Acceptance* (6 items).

Life-picture comprises a global assessment of the consequences of the disease. The respondent marks one of the four alternative, as best agrees with his or her situation. *Life-areas* comprises 19 items concerning the following: occupation (1 item), education (1 item), employment (1 item), family life (5 items), social life (3 items), dwelling (2 items), independence (5 items), and leisure (1 item). There are four alternative answers about the consequences of the muscular disease for each of the 19 items“ in a negative direction”, “don’t know in what direction” and “has not been important”. *Problems* comprises six items concerning mobility, eight concerning fatigue, and one concerning pain, sleep, interests and leisure. *Acceptance* comprises six items concerning how the person and those closest to the person accept the limitations caused by the disease. In the case of both *Problems* and *Acceptance*, the person can mark one or more items if applicable (Ahlström & Karlsson 2000).

Life-areas, *Problems* and *Acceptance.* Each of these can be used to form a positive and a negative index. These indices represent the sum of marked answers “in a positive direction” direction” (higher quality of life) and “in a negative direction”

(lower quality of life) The instrument has been tested in a study of persons with postpolio syndrome, and has been found to have acceptable discriminant validity (Ahlström & Karlsson 2000). The different indices give a picture of the persons perceived situation. The score level is mainly nominal.

Analysis of interviews

Narrative analysis (Study I)

Inductive content analysis (Lieblich et al. 1998) was used to analyse participants' reports about illness experiences. In order to obtain an overall picture of the interview answers, all 116 interviews were first read (n=58). Thereafter the interviews with 15 persons (30 interviews) (proximal MD 5, MyD 5, MDTH 5) were chosen for an analysis, which was performed by the author. The chosen interviews were judged to be particularly information-laden with respect to the purpose of the study; the persons spoke freely and in detail of their experience of the disease, telling how it had affected their lives, family, work and activities of daily living. The 15 persons were men and women and of different ages. Brief interviews with persons with MyD who suffered markedly from dysarthria or mild cognitive effects were excluded because these conditions prevented them from reflecting upon their situation.

The two interviews for each person were analysed as a coherent interview text. The interview text was split into meaning units. A meaning unit is a coherent expression of meaning comprising usually more than one sentence. The meaning units were sorted under the theme headings. The themes were derived from the interview text. A detailed categorisation was made of the meaning units within each theme. These categories were compared and refined in a smaller number of categories for each diagnosis group. Three core narratives were then formulated on the basis of the identified themes and categories, one for each diagnosis group. The intention of a

core narrative is to communicate the core of the content and the expressions which best agree with the interview material (Viney & Bousfield 1991).

Validation of the three core narratives was performed by the researcher who had not performed the analysis but who had conducted the interviews, using deductive testing categories to classify the responses from the analysis in each diagnosis group. This was done for the two interviews with the 43 persons who were not selected for the core narratives.

Phenomenological analysis (Study V)

A procedure inspired by a phenomenological method (Giorgi 1996) was employed for analysis of the verbatim transcripts of the interviews about perceived support in rehabilitation. *Reading of interviews*: The 37 interviews were read so as to gain a "sense of the whole" of the material. Each interview was then analysed separately. *Discrimination of meaning units:* After a further reading, the interview text was split into meaning units. *Transformation:* Each meaning unit was transformed systematically in two stages, each raising the level of abstraction. This analysis was subjected to bracketing, which is to say that the researchers' previous knowledge and understanding was minimised.

A validation was performed, the aim of which was to guarantee that the interviewer had faithfully followed the interview text in the analysis. The validation of the 37 transformed meaning units was performed by an independent judge, as described in Study V. The interviewer and the independent judge compared all the transformed meaning units from the 37 interviews. If there were any differences the original text was read again and discussed. The transformation was altered so as to make it as true to the original as possible.

Synthesis of transformed meaning units and validation of the synthesis. A synthesis was formulated with the aim of obtaining a phenomenological description of the participants' perceptions of support in the rehabilitation process. One synthesis was formulated for each interviewee by the interviewer and an independent judge. The syntheses were compared and refined so as to provide as reliable a description as possible on the basis of the interview text. The 37 final syntheses were formulated in a *General description.* This included nine themes, constituting the expression of the results.

Statistical analysis

The statistical analyses that have been used in the II, III and IV are shown in Table IV, and a significance level of $p<0.05$ was chosen.

Table IV. Statistical methods used in the quantitative studies (Studies II - IV)

	APC	Self report ADL	ADL Staircase	MAC	SIP	Quality of Life Profile	Psychosocial well-being questionnaire
Gender differences							
Unpaired t-test	II	III, IV	-	IV	IV	III	IV
Mann–Whitney	-	-	III, IV	-	-	-	-
Differences, county group							
Unpaired t-test	-	III	-	-	-	III	-
Mann–Whitney	-	-	III, IV	-	-	-	-
Differences, MD group							
ANOVA	II	III	-	-	-	III	-
Kruskal–Wallis	-	-	III	-	-	-	-
Changes over time							
Paired t-test	-	IV	-	IV	IV	-	IV
Wilcoxon´s test	-	-	IV	-	-	-	-
Correlations							
Spearman's (rho)	-	-	III	-	-	III	-
Pearson's (r)	-	III	-	III	III	-	-

Ethical considerations

The thesis is based on 5 studies including 4 study groups in 3 counties. The studies have been examined by the Research Ethics Committee at the Örebro Medical Centre Hospital, Sweden. [Study I, journal numbers 1086/87 and 643/90, Studies II - V, journal numbers 991/95 and 112/95 (multi-centre study) and Study IV, journal number 701/93]. For all the study groups, both written and oral information was given concerning the aim and design of the studies. Informed consent, confidential treatment and presentation of data have been routine procedures, as has the emphasis on voluntary participation that can be terminated at any time during the course of the studies. Interview questions which were felt likely to arouse strong feelings, were avoided, i.e. questions about the future for persons with very great disabilities and symptoms of MD.

RESULTS

Summaries of Studies I-V

Illness experiences

From the analysis of the interviews, there emerged six main themes, and a total of 45 categories were identified in the interview text in the proximal MD group and the MyD group, Table V, and 41 categories in the MDTH group. The themes constituted the expression of the results: Experience of first symptoms, Learning of the specific diagnosis, Experience after being told the diagnosis, Experience of the present, Experience of managing daily living and the Future. These themes and categories were used in the formulation of three core narratives, one for each type of MD. The three core narratives are presented separately in Study I.

The results of the three core narratives showed that the learning of the diagnosis stood out as something traumatic for all the interview persons, regardless of the specific diagnosis. Several interview persons felt that they had not been given sufficient information about the disease at an early stage of its progress. They stated that they were given information on the hereditary aspect when given the diagnosis but that they did not fully understand the meaning of this information. Most of the study persons had a long duration of illness, the average being 22 years. Despite this, only two clear transitions appeared in their narratives. The clearest transition was that from a healthy person to a person with a disease, which appeared at the time when they received the diagnosis. The second transition, from independence to dependence on help in daily life, was described most clearly by those who had great disabilities.

Table V. Themes and categories, concerned with the past, present and future

Time dimension	THEMES Categories
PAST	EXPERIENCES OF FIRST SYMPTOMS Symptoms during schooldays or other training period LEARNING OF THE DIAGNOSIS Reaction to learning of the diagnosis Contact with doctor Being asked to come and see the doctor Examinations Seeing the doctor EXPERIENCE AFTER BEING TOLD THE DIAGNOSIS Emotional experiences Physical experiences: Walking, Rising from a sitting position Lifting, Going up and down stairs Being able to speak Leisure activities
PRESENT	EXPERIENCE OF THE PRESENT Emotional experiences, Personality now, Evaluation of life Being able to speak of the disease Attitudes of others and social support, Information from medical staff Support from family, Relationships, Information from family and friends Knowledge of disease and heritability, Having a hereditary disease Information on the disease from health care staff Other problems: different symptoms, Medicines and forms of treatment Attitudes of colleagues at work, Adaptation of work Friends and social relationships, Making new acquaintances Leisure activities Physical training Financial considerations EXPERIENCE OF MANAGING DAILY LIVING Dwelling; Dressing; Personal care; Home management Transfer/Transport; Aids; Planning; Managing problems/tricks
FUTURE	FUTURE Feelings experienced on facing the future with regard to disease Daily living, Work, Leisure activities

Narratives concerning the present time showed that these persons are fighting and are not willing to regard themselves as either sick or disabled. Several of the interview persons mentioned it as an advantage that the deterioration of their activities is gradual, giving them time to reflect on what is happening. They found

it painful to think of the future. That reminds them that the disease takes a progressive course and that they will get worse. The core narratives for Proximal MD and MDTH were those that could most easily be generalised to others with the same diagnosis on the basis of the deductive validation procedure. A great variety of life and illness experiences appeared in the interviews that were not selected for inductive content analysis (Study I).

The results of the three core narratives showed that progressive muscular weakness causes many problems in daily life for persons with MD. These persons struggle to manage by themselves to avoid becoming dependent on others. The interview persons have clearly suffered losses regarding their independence in everyday living, in leisure activities and at work. It became clear from the narratives that their activity limitations had increased the proportion of sedentary occupations. For the most part, the illness experience was similar irrespective of the particular diagnosis. There were similar reactions to learning of the diagnosis, a similar feeling of uncertainty about the future, and similar accompanying psychosocial consequences of having a hereditary disease.

Activities of daily living

The results of Study II (n=119) with regard to activities of daily living assessed by the APC showed that most problems were experienced within Mobility and transportation (indoor and outdoor) and Work. There were only a few significant differences between the MD group in Örebro and the MyD group in Norrbotten. More activities with problems appeared in Personal care for the MD group in Örebro than for the MyD group in Norrbotten ($p<0.01$). The MD group in Örebro also reported more basic activities with problems within Leisure than did the MyD group in Norrbotten ($p<0.05$) (Table VI).

Table VI. Responses regarding activities of daily living within the Occupational Forms (Study II)

Occupational Forms	MD (n=33) mean%	MyD (n=46) mean%	PPS (n=40) mean%	Total (n=119) mean%
PERSONAL CARE				
Not doing the activity	5	0	2	2
Doing it without problems	59	83	75	74
Doing it with problems	36	17	23	24
HOME MANAGEMENT				
Not doing the activity	30	32	37	33
Doing it without problems	37	42	38	39
Doing it with problems	33	26	25	28
LEISURE				
Not doing the activity	51	54	54	53
Doing it without problems	29	37	28	32
Doing it with problems	20	9	18	15
MOBILITY AND TRANSPORTATION				
Not doing the activity	23	26	31	27
Doing it without problems	36	45	30	37
Doing it with problems	41	29	39	36
WORK	n=29	n=15	n=9	n=53
Not doing the activity	27	20	35	26
Doing it without problems	39	42	48	41
Doing it with problems	34	38	17	33

Few differences appeared between the two MD groups (in Örebro and in Norrbotten) and the PPS group (Study II). The PPS group reported more basic activities with problems in Leisure than did those with MyD in Norrbotten ($p<0.05$) (Table VI). There were gender differences within the total study group of 119 persons within Personal care. Men reported more basic activities without problems than did women (mean% 80 vs 68, $t=2.0$, $df=117$, $p<0.05$). Differences also appeared for Home management, where women reported more basic activities with problems (mean% 37 vs 17, $t=5.1$, df 117, $p<0.001$). The men avoided more basic activities within Leisure compared to the women (mean% 16 vs 8, $t=2.0$, $df=117$, $p<0.05$).

Within the MD group (n=33) in Örebro (Study II), the persons with MDTH and proximal MD had problems with more basic activities within Personal care than did the persons with MyD (mean%: MDTH vs MyD, 57 vs 18, $p<0.01$; proximal MD vs MyD, 48 vs 18, $p<0.05$). The persons with MDTH reported more activities with problems within Work (mean%: MDTH vs MyD, 57 vs 21, $p<0.01$: MDTH vs proximal MD 57, vs 35, $p<0.05$) and within Leisure than did persons with MyD (mean%: MDTH vs MyD, 31 vs 12, $p<0.05$). The women in the MD group in Örebro reported more basic activities with problems than did men within Home management (unpublished data).

The ADL Staircase (n=77) showed that 52% were dependent in I-ADL (grades 1-4) and 18% in both P-ADL and I-ADL (grades 5-10). The persons with MyD showed significantly more dependence than the persons with MDTH ($z=2.34$, $p<0.05$). The MD group in Östergötland showed significantly more dependence (85 vs 58%) than the MD group in Örebro ($z=2.7$ $p<0.01$) (Study III).

The result for the total group in Study III (n=77) showed that persons with MDTH had the greatest disability for activities requiring distal muscular strength according to the Self-report ADL. Persons with proximal MD had the greatest disability for Ambulation. Persons with MyD showed both proximal and distal muscular weakness. The persons with MyD and MDTH showed significantly more difficulties regarding 'Finger strength' than did the persons with proximal MD (MyD vs proximal MD $t=3.13$, $df=63$, $p<0.01$; MDTH vs proximal MD $t=2.91$, $df=41$, $p<0.01$). The greatest disability of the three diagnosis types concerning 'Finger subtle function' was experienced by the persons with MDTH (MDTH vs proximal MD $t=8.03$, $df=41$, $p<0.001$; MDTH vs MyD $t=2.98$, $df=44$, $p<0.01$), followed by MyD, and the least disability was shown by persons with proximal MD (Study III).

Over a five-year period, the total group in Study IV (n=45) showed a significant increase in dependence in I-ADL and P-ADL (grades 5-10) (1991: 9%; 1996: 11%, $p<0.05$), assessed by the ADL Staircase. Persons with MyD showed the greatest increase of dependence in I-ADL and P-ADL over the five years ($z=2.31$, $p<0.05$). There were no gender differences in 1991 or 1996 regarding the dependence in ADL assessed by the ADL Staircase (Study IV).

The group in Study IV (n=45) also showed a significant deterioration over the five-year period according to all indices of the Self-report ADL (Ambulation, Arm strength, Finger strength and Total index) ($p<0.001$) apart from 'Finger subtle function'. The women demonstrated more disability than did men with respect to finger strength among persons with MyD in 1991 ($t=2.13$, $df=24$, $p<0.05$).

To sum up, most of the problems reported concerned mobility indoors as well as outdoors, and work. Over half of the group were dependent on others, chiefly in activities requiring mobility. Nearly half of the group were employed, and heavy work was experienced as problematic by half of these. There was a significant deterioration over five years for the total study group regarding activities of daily living accompanied by an increase in dependence in P-ADL and I-ADL. Difficulties in both proximal and distal muscles varied with the type of MD. Few significant diagnosis-related differences emerged regarding activities of daily living.

Coping

The APC data for coping (n=119, Study II) showed little use of problem-focused coping (Devises and tricks, Aids, Own technical solutions, Accept help, Avoid). Problem-focused coping was used least for Leisure. For Home management, the MD group in Örebro used 'Aids' in significantly more activities ($p<0.01$), and

‘Accept help’ in fewer activities ($p<0.05$) than did the MyD group in Norrbotten. The MD group in Örebro also used their ‘Own technical solutions’ for more activities within Leisure than did the MyD group in Norrbotten ($p<0.05$) (Table VII) (Study II). Within the MD group in Örebro, there were no significant differences between diagnoses with respect to coping assessed by the APC.

The PPS group in Study II used the coping strategies ‘Aids‘ and ‘Avoid’ in more activities within Mobility and transportation than did the MD group in Örebro (‘Aids’ $p<0.01$, ‘Avoid’ $p<0.05$) (Table VII). Of the total number of persons in the study (n=119), 53 were working, and of these the men reported using ‘Aids’ in more activities within Work than did the women (mean% 10 vs 0, $t=2.4$, $df=117$, $p<0.05$). The women in the PPS group used ‘Aids’ in more activities within Personal care (mean% 20 vs 11, $t=2.2$, $df=117$, $p<0.05$), and Home management (mean% 3 vs 10, $t=3.2$, $df=117$, $p<0.01$), as compared to the men. More men than women (mean% 16 vs 8, $t=2.0$, $df=117$, $p<0.05$) used the coping strategy ‘Avoid’ when they had problems in performing activities within Leisure. More men used ‘Aids’ in more activities within Work as compared to women (mean% 10 vs 0, $t=2.4$, $df=117$, $p<0.05$) (unpublished data).

The results of Study IV (n=45) showed no significant differences over the five-year period with regard to the coping strategies assessed by the MAC (Fighting spirit, Helplessness/hopelessness, Anxious preoccupation, Fatalism, Avoidance). ‘Fighting spirit’ was the predominant type of coping strategy in most persons in both 1991 and 1996. The persons with MDTH showed a reduction in ‘Fatalism’ during the five-year period (mean 17 vs 14, $p<0.05$). No significant differences appeared between the three diagnosis types or between genders with regard to coping strategies assessed by the MAC during the five-year period (Study IV).

Table VII. Responses with regard to problem-focused coping within the five Occupational Forms (Study II)

Problem-focused coping	MD (n=33) mean%	MyD (n=46) mean%	PPS (n=40) mean%	Total (n=119) mean%
PERSONAL CARE				
Devices and tricks	24	26	31	27
Aids	20	13	16	16
Own technical solutions	12	3	4	6
Accept help	10	14	11	12
Avoid	0	3	6	3
PF coping Total	13	12	13	12
HOME MANAGEMENT				
Devices and tricks	16	10	9	11
Aids	11	4	7	7
Own technical solutions	7	5	4	6
Accept help	20	34	28	28
Avoid	8	9	11	9
PF coping Total	12	12	12	12
LEISURE				
Devices and tricks	7	8	6	7
Aids	4	2	4	3
Own technical solutions	4	1	2	2
Accept help	6	5	7	6
Avoid	11	11	14	12
PF coping Total	6	8	6	7
MOBILITY AND TRANSPORTATION				
Devices and tricks	19	14	16	16
Aids	8	10	21	13
Own technical solutions	4	5	3	4
Accept help	9	15	11	12
Avoid	6	14	18	13
PF coping Total	9	12	13	11
WORK	n=29	n=15	n=9	n=53
Devices and tricks	13	32	17	21
Aids	5	3	8	5
Own technical solutions	5	3	6	5
Accept help	13	5	19	12
Avoid	13	12	11	13
PF coping Total	10	11	11	10

PF-coping Total = Total of all five problem-focused coping strategies

In summary, the subjects used problem-focused coping relatively infrequently in spite of the fact that the predominant coping was 'Fighting Spirit'. The most used problem-focused coping strategy was 'Devices and tricks'. Few differences in problem-focused coping were found between the different MD groups or between the sexes. The emotive/cognitive coping strategies were stable during the five-year period.

Quality of life

The highest satisfaction with ADL was reported for Personal care and the lowest for Mobility and transportation as assessed by the APC (Table VIII). Three of eleven basic activities within Leisure for which the study group most commonly desired a better solution were keeping fit, swimming, and writing. Gender differences appeared only for Home management, where the women wished for changes in more basic activities than did the men ($t=2.1, df=117, p<0.05$) (Study II).

Within the MD group in Örebro (Study II), persons with MyD reported a higher satisfaction with their performance of ADL than did persons with MDTH the performance of Personal care (mean%: MyD vs MDTH, 95 vs 73, $p<0.05$), Mobility and transportation (mean%: MyD vs MDTH, 84 vs 54, $p<0.01$) and Work (mean%: MyD vs MDTH, 83 vs 62, $p<0.05$). Persons with proximal MD showed a higher satisfaction with activities within Home management than did persons with MDTH (mean%: proximal MD vs MDTH, 88 vs 61, $p<0.05$) (unpublished data).

The total study group including the PPS group (n=119) reported a high score for satisfaction, the lowest for Mobility and transportation and the highest for Personal care. The PPS group showed a lower satisfaction with activities within Leisure than did the MD group in Örebro and the MyD group in Norrbotten ($p<0.01$) (Study II) (Table VIII).

Table VIII. Degree of satisfaction within the Occupational Forms (Study II)

	MD (n=33) mean%	MyD (n=46) mean%	PPS (n=40) mean%	Total (n=119) mean%
PERSONAL CARE	88	90	88	89
HOME MANAGEMENT	78	85	75	80
LEISURE	81	82	68	77
MOBILITY AND TRANSPORTATION	73	75	65	71
Number of persons working	n=29	n=15	n=9	n=53
WORK	77	77	70	76

The result (n=77) showed that the consequences of the disease were for the most part negative for the whole group with regard to Life-areas and Problems assessed by the Quality of Life Profile (Study III). There were more negative than positive consequences within Life-areas and Problems (Life-areas positive: mean 1.2, Life-areas negative: mean 4.6; Problems positive: mean 0.8, Problems negative: mean 3.8). With regard to Acceptance, almost half (44%) agreed with the statement "I have entirely accepted the restrictions caused by the disease" and (41%) with "I have to some extent accepted the restrictions caused by the disease". Somewhat less than half, i.e. 44% agreed "Life would have had more to offer if the disease hadn't got in the way, but I don't go around thinking about it". Thirty-five percent agreed with the statement "Life hasn't become worse because of the disease, I have a good life", 11% with the statement "Of course I'm disappointed, since the disease hampers me", and 10% with "I have developed as a person: I probably wouldn't have had the personal strength I do have if I had been in perfect health". The MD group in Östergötland reported more negative consequences of the disease, regarding Problems than did the MD group in Örebro ($t=3.37$, $df=69$, $p<0.01$). Otherwise, there were no significant differences between counties, diagnoses or genders.

There was a deterioration over five years for the whole study group (n=45) for the Physical index ($p<0.001$) as assessed by the SIP (Table IX). Persons with MyD and proximal MD showed a deterioration in 'Body care and movement' within the Physical index ($p<0.01$). There was also deterioration for persons with MyD in 'Social interaction' within the Psychosocial index ($p<0.01$).

Table IX. Mean values (SD) of Health-related quality of life, according to the Sickness Impact Profile in 1991 and 1996 in persons with MD (Study IV) (paired t-test)

	MyD (n=26)		MDTH (n=8)		Proximal MD (n=11)		Total (N=45)	
	1991	1996	1991	1996	1991	1996	1991	1996
	mean (SD)		mean (SD)		mean (SD)		mean (SD)	
PHYSICAL INDEX	10.0 (15.8)	14.6* (18.1)	12.2 (12.2)	13.0 (13.0)	13.1 (13.7)	20.4* (17.6)	11.2 (14.5)	15.7*** (17.1)
PSYCHOSOCIAL INDEX	6.8 (10.3)	9.27* (12.7)	7.0 (6.5)	7.1 (4.9)	3.3 (3.3)	5.7 (8.8)	6.0 (8.5)	8.0* (10.8)
Independent Categories								
Sleep and rest	14.0 (14.7)	15.2 (18.0)	8.5 (13.1)	7.0 (8.3)	13.3 (9.84	10.4 (9.6)	12.8 (13.3)	12.6 (15.1)
Eating	0.6 (2.3)	2.5 (6.3)	2.0 (4.0)	4.0 (7.5)	0.5 (1.6)	3.0 (7.2)	0.8 (2.6)	2.9* (6.5)
Home management	21.6 (29.0)	26.3 (27.8)	21.2 (23.7)	15.1 (24.1)	21.9 (30.9)	27.6 (29.4)	21.6 (28.0)	24.6 (27.3)
Recreation and pastimes	13.4 (19.2)	17.1 (20.2)	15.4 (20.4)	18.3 (16.9)	22.7 (20.0)	20.1 (21.3)	16.0 (19.5)	18.1 (19.5)
Employment	31.6 (30.7)	25.8 (30.7)	47.8 (31.5)	0** [(1)]	25.7 (29.1)	24.3 (30.8)	33.0 (30.7)	20.9 (29.1)
TOTAL INDEX	10.4 (12.4)	13.3* (14.1)	12.0 (10.1	9.5 (8.1)	10.0 (9.0)	14.0* (12.4)	10.7 (11.1)	12.9* (12.7)

[(1)] =Six of eight persons with MDTH were on retirement pension, *$p<0.05$, **$p<0.01$, ***$p<0.001$

The persons with proximal MD showed deterioration in 'Ambulation' ($p<0.05$) and 'Body care and movement' ($p<0.05$) within the Physical Index over the five-year period. The persons with MDTH showed deterioration in 'Communication' within the Psychosocial Index ($p<0.05$). Two items were particularly evident here: 'I find it difficult to write by hand or type' and 'I write only when I have to sign something' (Study IV).

The whole study group in Study IV (n=45) showed a deterioration over five years for the index of 'Satisfaction' assessed by the Psychosocial well-being questionnaire (1991: mean 2.47; 1996: mean 2.91, $p<0.05$). This change was significant also for the persons with MyD (1991: mean 2.42; 1996: mean 2.88, $p<0.05$). By contrast, there were no significant changes over the five-year period for 'Psychosocial well-being' or 'Happiness'.

In summary, the highest degree of satisfaction with activities of daily living was noted for Personal care and the lowest for Mobility and transportation. Muscular dystrophy had mostly negative consequences, and nearly half stated that life would have offered more without it. There was a significant deterioration over five years with regard to health-related quality of life (SIP) and to satisfaction with ADL.

Relationships between activities of daily living, coping and quality of life

Activities of daily living vs Coping.

There were only weak correlations (r=0.40 or less) both in 1991 and 1996, between ADL (ADL Staircase, the Self-report ADL) and coping (MAC). Thus, activities of daily living appear to be unrelated to the MAC coping strategies (Study IV).

Activities of daily living vs Quality of life.

In Study IV, there were strong correlations between ADL (ADL Staircase, Self-report ADL) and quality of life (SIP) in most of the indices except Employment, in 1991 and 1996 (n=45). The total index correlated significantly with all ADL variables (Table X). The strongest correlations demonstrated were found between 'Ambulation' in the Self-report ADL and the 'Physical index' in the SIP, and between the ADL Staircase, and 'Physical index'. These results were shown for measurements performed in 1991 and 1996 (Table X).

Table X. Correlations (r) between the Self-report ADL, the ADL Staircase and the Sickness Impact Profile (n=45), (Study IV)

SIP	The Self-report ADL										ADL staircase	
	Ambulation		Arm strength		Finger strength		Finger subtle function		Total index		Total	
	1991	1996	1991	1996	1991	1996	1991	1996	1991	1996	1991	1996
PHYSICAL INDEX	0.87***	0.90***	0.69***	0.72***	0.45**	0.67***	0.47**	0.64***	0.83***	0.71***	0.83***	0.80***
PSYCHOSOCIAL INDEX	0.56***	0.62***	0.53***	0.44**	0.43**	0.52***	0.52***	0.31***	0.63***	0.60***	0.61***	0.64***
Independent Categories												
Sleep and rest	0.45**	0.50***	0.31*	0.49***	0.40**	0.42**	0.18	0.45**	0.43**	0.52***	0.45**	0.45**
Eating	0.43**	0.57***	0.48***	0.59***	0.43**	0.49**	0.38**	0.56***	0.52***	0.62**	0.52**	0.41**
Home management	0.78***	0.76***	0.67***	0.70***	0.51***	0.60***	0.46**	0.46**	0.79***	0.73***	0.79***	0.74***
Recreation and pastime	0.50***	0.69***	0.51***	0.50***	0.48***	0.43**	0.44**	0.40***	0.59***	0.62***	0.42**	0.67***
Employment	0.32*	0.23	0.38*	0.18	0.24	0.13	0.27	0.14	0.37*	0.15	0.32*	0.2
TOTAL INDEX	0.78***	0.85***	0.70***	0.69***	0.52***	0.66***	0.53***	0.37***	0.82***	0.81***	0.80***	0.81***

*p<0.05, **p<0.01, ***p<0.001

There were no significant correlations between the ADL indices (ADL Staircase, Self-report ADL) and the three indices in the Psychosocial well-being questionnaire (Psychosocial well-being, Happiness, Satisfaction) apart from a weak correlation (r=0.30) between 'Finger strength' and 'Psychosocial well-being' (Study IV). These data suggest that 'Psychosocial well-being' is not strongly related to the Self-report ADL indices.

Study III showed strong correlations between on the one hand the ADL Staircase, all indices in the Self-report ADL and on the other hand the total of negative answers in the Quality of life Profile ($p<0.001$). There was a weak correlation between the ADL Staircase and Life-areas negative (Quality of life Profile) (rho=0.33), and a moderate correlation with Problems negative (rho=0.51). There was also relatively weak correlation between the four ADL indices in Self-report ADL and Life-areas negative (r=0.33-0.42) and a moderate correlation with Problems negative (rho=0.50-0.54). These findings imply that higher scores of disability correlates with lower health-related quality of life (Study III).

Coping vs Quality of life.

The correlations in Study IV between coping (MAC) and quality of life (SIP, Psychosocial well-being questionnaire) were weak to moderate, and unsystematically distributed among the different indices in the data from 1991 and 1996. There were a few significant correlations between three indices in the SIP (Psychosocial index, Sleep and rest, and Eating) and 'Helplessness/ hopelessness' (MAC) in 1991 (r=0.30-0.45) and for two indices in 1996 (Psychosocial index, r=0.44 and Total index, r=0.34). There were also weak correlations between four indices in the SIP (Physical index, Psychosocial index, Home management, Total index) and 'Fatalism' in 1996 (r=0.32-0.43) and between one index in SIP (Home management) and 'Avoidance' 1996 (r=0.34).

Correlations between 'Fighting Spirit' (MAC) and 'Psychosocial well-being' (Psychosocial well-being questionnaire) were higher in 1996 than in 1991 (r=0.36 vs 0.64). There were also significant correlations between 'Fighting Spirit' and 'Happiness' (r=0.53) and between 'Fighting Spirit' and 'Satisfaction' (r=0.52) in 1996 (Study IV). Also for Helplessness/hopelessness' and the three indices in the Psychosocial well-being questionnaire, a higher correlation appeared after five years (Psychosocial well-being: r=0.43 vs 0.53, Happiness: r=0.32 vs 0.54, and Satisfaction: r=0.41 vs 0.57).

In summary, there was strong correlations between ADL and health-related quality of life, but there were only weak to moderate correlations between ADL and coping. This indicates that it is necessary to assess ADL besides coping and psychosocial well-being because they were relatively independent of each other. An increase in 'Helplessness/hopelessness' and 'Fighting Spirit' was associated with low quality of life in terms of psychosocial well-being.

Perceived support in rehabilitation

The 37 syntheses, one for each interview person, formed nine themes: Psychosocial support, Meeting other people with MD, Knowledge and learning, Adjustment to daily life, Coping with illness-related problems, Adjustment at work, Management of physical disability, Medical examination and treatment, and Involvement of relatives (Study V).

The experience of the participants was that they received more *Psychosocial support* from the staff than they had expected. The staff were knowledgeable about muscular dystrophy, took time, listened, and understood the problems of the participants. Most of the participants felt that it was valuable to engage in *Meeting other people with MD*. Their experience was that they gave each other advice and support in the conversation groups. Only a few felt no need to meet the other

participants with MD. The participants felt that they had gained *Knowledge and learning* regarding disease, heredity, technical aids, grants and physical training. Only a few had previous knowledge of these matters.

The participants experienced an *Adjustment to daily life*, which involves making everyday life easier by adapting the home, using technical aids and practical advice. Some of the participants were in need of further adaptation of their homes after the end of the rehabilitation (i.e. managing the stairs in their homes or using small shower cubicles). Several had been afraid of starting to use technical aids, in particular the wheelchair, but they had understood with time that using aids gives a greater feeling of freedom and independence. The participants were also given help in applying for various financial grants to facilitate everyday life.

Those participants who were still employed felt that they could not manage to work full-time, since it was too strenuous, physically and mentally, to be both working and looking after the family. Some of the participants were allowed *Adjustment at work* through adaptation of their place of work or help in reducing their working hours. More than half felt that they were receiving support and advice with *Management of physical disability*. They were more daring in taking exercise after gaining knowledge of the value of keeping fit and understanding better the reasons for doing so. Most were too tired, however, when they came home from work, to do exercises, or they were in pain, which prevented them.

Some participants felt that they had become more cheerful, and felt that they had gained more energy after the rehabilitation. They felt that the *Medical examination and treatment* during the rehabilitation period was highly beneficial. The *Involvement of relatives* in the rehabilitation was very valuable, since it gave the relatives a better understanding of their disease and situation. Many of the study persons experienced good support from their relatives, but some of them felt that it

was hard to talk about their disease, particularly with their children. In addition they were also worried that their children would get their muscle disease.

In summary, the participants felt that the rehabilitation had been valuable and that the staff had listened to their problems and given them advice. They had gained valuable knowledge of their muscle disease and its hereditary nature. They found it was very valuable to meet others with the same disease. The support was given both by the staff and the other participants with MD.

DISCUSSION

Illness experiences

The present study shows that the most common reactions to being told the diagnosis were disappointment, aggression, anxiety or despair (Study I), which confirms previous studies on persons with MD (Ahlström & Sjödén 1996, Brunnback 1987, Faulkner & Kingston 1998). Unpleasant events, which affect the individual emotionally, may be recalled decades later (Christianson 1992). When the persons were told that they had a hereditary, progressive disease they would not at first believe this. An earlier study of women with MD by Faulkner and Kingston (1998) showed that one person in four found it hard to accept the disease. When an event takes place that is experienced by the individual as very traumatic, denial is a common defence reaction (Christianson 1992). The interviews showed that the illness experiences were similar irrespective of the specific diagnosis. With this knowledge in mind, the diagnosis of an incurable disease with a progressive course should be announced with great care and respect for the individual. Most of those who are given the diagnosis of MD are young, and careful attention should be paid to the age and life situation of the person.

It became clear that there were two clear transitions in the study persons' narratives, i.e. phases requiring a change in behaviour due to the illness. One was the transition from the identity of being healthy to being a person with a progressive illness, a process that started when the diagnosis was announced. The transition from a state of independence in daily living to the state of needing help and support (Study V) has also been documented in two previous studies (Faulkner & Kingston 1998, Sidell 1997). A transition is a health status change with consequences for roles, relationships, expectations or capacities, and it also indicates changes in the needs of a person (Meleis 1991). A transition during the course of an illness requires that the individual integrate new knowledge in order

both to modify behaviour and to adapt to a new social context (Meleis 1991, Parkes 1971, Viney & Bousfield 1991). Earlier studies show that psychosocial problems may be partly explained by major functional impairments in MD, which can result in a lowered quality of life (Ahlström 1994, Eggers & Zatz 1998, Fowler et al. 1997). Important changes that characterise a transition in the life of an individual may cause that individual to re-evaluate requirements and life values (Meleis 1991, Parkes 1971).

Despite difficulties that cause great strain in everyday life, most persons had accepted their situation and had tried to make the best of it. This is reflected by the fact that these persons, when talking about their life, describe themselves as "living in the present". They want to regard themselves as neither ill nor disabled (Study I). The interview persons found it painful to think of the future as they knew that their illness will gradually grow worse. This has also been shown in an earlier study of MD (Faulkner & Kingston 1998). These persons may withdraw from others and from activities, which may lead to a social isolation and deterioration of the quality of life (Eggers & Zatz 1998, Hall et al. 1991). It is therefore important to identify social isolation, pay attention to it, and to consider it in rehabilitation efforts (Michael 1996). The way in which the individual copes with a transition and adjusts to change, and the way in which the environment affects the individual, are fundamental questions for medical care staff (Meleis 1991).

Activities of daily living

Study II shows that the muscular weakness affects many activities in everyday life, but also that some persons have succeeded in adapting their everyday life, reporting comparatively few activities as experienced problems. One explanation of this is that the time period during which the persons have been living with their illness has made possible adjustment to the slow, insidious deterioration of their activities. On average, the study group had had their illness for somewhat more

than two decades. The most common problems for the entire study group were experienced within Mobility and transportation, particularly indoors (Study II). This has also been found in studies of persons with the postpolio syndrome (Grimby & Thorén Jönsson 1994, Westbrook & McDowell 1991). Participants were least satisfied with the activities within this field. One explanation for the preponderance of indoor over outdoor problems may be that there was considerable avoidance of outdoor activities (Study II). The persons with MD (Study III) experienced problems with limited freedom of movement in daily activities and in engaging in leisure activities. The interviews revealed that the proportion of sedentary activities increases when the muscular weakness progresses. Previous research has shown that the progressive course of the illness brings about significant deterioration of functioning capacity over a five-year period (Dahlbom et al. 1999). This results in repeated losses of important functions, which subject the person to considerable stress in managing new situations (Hainsworth 1994).

The persons with MD in Östergötland were more dependent on others in ADL than were the persons in Örebro. This was true also with regard to the number of negative problems (Study III). This may be due to the fact that the persons with MD in Örebro had been identified in a previous population study within Örebro (Ahlström et al. 1993), which may be expected to have included more persons with milder forms of MD. A further explanation is that the responses from the persons in Örebro may have been affected by measures taken in conjunction with the rehabilitation programme. Only a few persons in Östergötland had been given traditional rehabilitation during the corresponding period (Ahlström & Gunnarsson 1997). Therefore it cannot be excluded that the difference in ADL dependence may to some extent be due to the persons with MD in Örebro having been given technical aids adapted to suit them. They may also have gained a better insight into

how they could make best use of their remaining capacity for activity owing to the rehabilitation.

The persons with MyD showed the greatest dependence on others in activities of daily living, although those with proximal MD reported greater functional limitations in walking. One explanation of this may be that those with MyD experience a higher degree of distal muscular weakness (Study IV). Impairments of cognitive function may also contribute to the finding that persons with MyD are more dependent on others in ADL. The persons with MDTH showed the greatest limitations in activities calling for distal muscular strength, which is in accordance with previous studies and the nature of the disease (Ahlström & Gunnarsson 1996, Åhlberg et al. 1997).

Persons with MyD showed a significant deterioration, both in the capacity to walk and as an increased dependence in ADL (Study IV). In addition to the initial distal muscular weakness in MyD, the proximal muscles are affected as time goes on, and the difficulties in walking may lead to the persons becoming confined to a wheelchair (Harper 1988, Sirotkin-Roses 1991). In an earlier study (Dahlbom et al. 1999), the same persons with MyD showed the most marked deterioration in walking capacity as assessed by an observation scheme, Brooke's function test (Brooke 1986). In contrast to the present five-year follow-up study, the result from Dahlbom et al. (1999) also showed deterioration for proximal MD and MDTH. This suggests that self-report shows an underestimation compared with results based on observation. In the literature, underestimation in self-reporting has been explained by idealisation (Naess 1994). Another explanation may be that the activity limitations grow to be natural for the persons with MD since the slowly progressive nature of the deterioration gives them time to adapt to their muscular weakness. Thus they need not think about it all the time, but live more by what they can do than by what they cannot (Study IV). The persons with proximal MD

showed deterioration in activities with regard to both arm strength and finger strength (Study IV). This may suggest that the illness has advanced distally (Brooke 1986).

Among the persons with MD who were employed, almost half experienced problems with heavy work (Study II). This is not surprising, since heavy work demands muscular strength in both upper and lower extremities. Despite this, only a few left heavy work to colleagues or avoided it. Only a small number of persons used technical aids, which may be due to lack of knowledge about aids that can make work easier (Study II). One person in three in the study group stated that the illness had reduced their chances of employment on the labour market (Study IV). Several types of work demand muscular strength, and accurate movements in both fine and gross motor patterns. This makes it difficult to find a suitable job, which may be one explanation of the lower employment level or lack of work found earlier (Velozo 1993). The impairment of cognitive functions for persons with MyD may also be one explanation of reduced working capacity and joblessness (Kakulas 1999, Meyer & Boake 1993). Moreover, a low level of education may reinforce the difficulties in finding a job not requiring physical strength.

In summary, the studies on ADL show that persons with MD have a great need for rehabilitation in this respect. With improved knowledge of the situation of people with MD, the rehabilitation team as well as the occupational therapists can target measures involving technical aids and adaptations of home and workplace to make the life situation of these persons easier

Coping

Problem-focused coping was used only to a small extent in daily activities. The strategy that was used most was “Devices and tricks” (Study II). The persons with MD have gradually learnt to use such “Devices and tricks” to manage their daily

activities. A previous study shows that emotion-focused coping tends to dominate (Ahlström & Sjödén 1996). In the prospective longitudinal study, it became apparent that the coping strategies were comparatively stable over the five-year period (Study IV), which agrees with previous studies of cancer patients (Nordin 1998). An explanation of this may be that the MAC-scale only measures emotion-focused or cognitive coping (Ahlström & Sjödén 1996). Another explanation may be that the MAC-scale assesses stable coping strategies. The finding of mainly unchanged coping suggests that the MAC-scale measures the t*rait* rather than the *state* aspect of coping, for which reason instrument is not suitable for assessment of changes over time (Nordin & Glimelius 1997). 'Fighting Spirit' turned out to be the predominant coping strategy (Study IV), which confirms findings in earlier studies on cancer patients (Lampic et al. 1994, Nordin & Glimelius 1997). The persons with MDTH had the highest mean age compared with those with MyD and proximal MD. The former group also showed the highest scores on 'Fatalism' at the first assessment, but 'Fatalism' decreased until the second assessment. All eight persons with MDTH had taken part in a rehabilitation programme (Study V), which may be one explanation of the reduction of 'Fatalism'. It may be assumed that increased knowledge about the disease and the support which the medical services and society can offer will help to reduce fatalistic beliefs and their importance for events in daily life and the way that life is led.

The persons with MyD gave only meagre accounts of "ways of solving problems" and experiences as they face the future. This finding may be related to effects of the illness on the cognitive faculties. Persons with cognitive functional disabilities (Walton & Gardner-Medwin 1988) are in particular need of coping support for making things easier in their daily lives, something that needs to be paid considerably more attention in the work of caring and rehabilitation.

In summary, the present study shows that the persons themselves do not use problem-focused coping to solve their many everyday problems. However, they do try to find solutions, which is shown in the 'Fighting spirit' attitude to problems. This suggests that the problems of ADL make great demands on the individual. The prevalence of this form of coping must not be taken to suggest that persons with MD can manage their life situation in such a way that they experience a high quality of life. Previous research in persons with MD has shown that 'Fighting spirit' may even be related to a poorer quality of life (Ahlström 1994). In this respect, the rehabilitation team has an important advisory and supportive task to perform for persons with MD relating to problem-focused, and emotion-focused coping.

Quality of life

One-third of the persons stated that life had not deteriorated due to their disease even though they did report few positive consequences (Study III). This way of experiencing things may be a result of their having had the disease for a long period, during which they have adjusted to the changed conditions of life (White et al. 1992). At the same time, the result showed few favourable consequences of MD, the clearest being emotional support and choice of dwelling (Study III). An experience of having a good life, in other words, a high quality of life, has been demonstrated in other studies irrespective of the degree of physical ill-health (Montgomery et al. 1996, Ramund & Stensman 1988, Spilker 1990). Another explanation that may be partly applicable here is idealisation. Previous research shows that when interpreting self-reports it is important to bear in mind that a certain degree of overestimation of the person's own quality of life may be present (Ramund & Stensman 1988). This may find expression in the way in which the study persons tend to give varying degrees of positive reports when they are interviewed or responding to surveys (Kajandi 1994). Positive re-evaluation may to

some extent consist of idealisation (Lazarus 1993). In this sense idealisation may be seen as a coping strategy aiming at the attainment of well-being.

The persons with MD showed a significant deterioration over a five-year period regarding physical and psychosocial aspects of health-related quality of life, and satisfaction with ADL. This was most noticeable in persons with MyD (Study IV). This deterioration of well-being and satisfaction agrees with earlier results from persons with MD, which have shown a lack of psychosocial adjustment, depression, anxiety and passivity (Fowler et al. 1995). The results of two prospective longitudinal studies (Dahlbom et al. 1999, Johnson et al. 1995) show that persons with MyD have the greatest psychosocial problems and the poorest psychosocial well-being by comparison with remaining MD groups.

The increased dependence on other persons showed strong correlations with a poorer quality of life (Study IV). This suggests that a lower quality of life can partly be related to disability, which has been demonstrated in previous studies on muscular dystrophy and postpolio syndrome (Ahlström 1994, Ahlström & Gunnarsson 1996, Ahlström & Karlsson 2000). Of the three types of diagnosis, persons with MDTH reported the greatest number of positive disease consequences. One explanation may be that they are less dependent in ADL and were not restricted in their activities due to their disease to such a degree as the other two groups (MyD, proximal MD).

There were weak to moderate correlations between coping and quality of life (Study IV). Higher levels of ‘Helplessness/hopelessness’ and ‘Fighting Spirit’ correlated with a poorer quality of life (Study IV), which agrees with results in a previous study (Ahlström 1994). This contrasts with results from cancer research, which have shown that ‘Fighting Spirit’ is related to improved psychosocial well-being (Greer & Watson 1987, Lampic et al. 1994, Watson et al. 1991). This

discrepancy may be due to differences in the conditions imposed by the disease, since patients with cancer can probably fight and obtain palliative treatment for tiredness, nausea and pain, while patients with muscular weakness cannot overcome their paralysis. If this assumption is correct, 'Fighting Spirit' may be a reflection of an unrealistic notion of what can be controlled in a progressive disease like MD.

In summary, the more problems persons experience in everyday life, the less satisfaction they experience according to the psychosocial well-being questionnaire. Somewhat less than half had accepted their disease entirely. Most of the persons had had their disease for more than two decades, which suggests that the temporal factor alone cannot explain the extent to which a person accepts his or her situation.

Perceived support in rehabilitation

The experience of the study persons was that the goals of rehabilitation were adapted to the individual according to his or her needs (Study V). On the basis of the interview, it is possible to suggest that participants had increased their confidence in being able to structure, predict and understand the problems caused by their disease. The multi-professional team was perceived to provide a humane way of encountering the patients, and as a result the patients felt that they were seen and confirmed. For several of the study persons, the rehabilitation programme was the first time they had been able to compare themselves with others on the same terms, without needing to feel that they were outsiders or in some way different from other people. This result suggests the value of rehabilitation in small groups and together with persons having the same disease (Study IV). Both formal groups (i.e. groups led by rehabilitation staff) and informal groups (such as self-help groups) are valuable for persons with chronic diseases (Hildingh et al. 1995, McColl 1995). The group constitutes a cornerstone in a network support. The

pattern of social support in a group may be described as a relation between caring, belonging, sharing and confidence. Social support may slow down the deterioration of people's health and help them remain in the community (Choi & Wodarski 1996). Persons who had taken part in the rehabilitation had found it very valuable to be able to talk to rehabilitation staff about their illness experiences. They experienced support in working on their emotions and verbalising their apprehension and anxiety at the thought of living with an incurable, progressive disease (Study V). Thus they had gained a chance to prepare themselves with alternative coping strategies to deal with future stress as demonstrated in an earlier study (Edwards 1989).

The persons felt that they had been given valuable information on technical aids and on adaptations in their home, but they were striving to manage on their own without technical aids as long as possible. The greatest resistance described was that to using a wheelchair, which was felt by several of the participants to be an irrevocable development (Study V). For persons with a progressive neurological disease, emphasis is placed on programmes for adjusting and maintaining functions (Driessen et al. 1997) and the rehabilitation team can play an important role in providing social support (Hays & Kowaske 1995, Keeling et al. 1996, McColl 1995). Patients with a progressive disease live in a dynamic process of change and often have to re-evaluate their own capacities and goals, which is essential for their health (Nordeson et al. 1998). The rehabilitation team should focus on the active participation of the individual, and on that individual's own capacity to identify needs, solve problems and adjust to life changes (Choi & Wodarski 1996).

Methodological discussion

Validation (credibility) was tested in the two qualitative studies (I, V). In Study I the attempts to inductively identify the categories in the interview text were made to assess the credibility of the finding. Credibility is defined as "A criterion for

evaluating the data quality of qualitative data referring to confidence in the truth of the data" (Polit & Hungler 1999, p.699). The validation was done by a researcher who had not participated in the analysis but who had conducted the interviews. The previous knowledge gained by the researcher when conducting the interviews and gained through previous research experience was assumed not to have influenced the results. The result of the deductive analysis gives no information about the content of the categories but only about whether the specific categories could be identified or not. The life situation of the study persons was decisive for whether the categories could be identified or not. Thus, exceptions were persons who had very mild or severe types of the MyD. With a person at work, for example no conversation was initiated about early retirement pension, which is the reason why this category is lacking. When the disease state was mild, the question concerning use of a ventilator was never posed. For ethical reasons, interview questions on the future were not asked in interviews with persons with very severe disease states. The result of the test of the credibility showed good transferability. The validation indicates that the transferability of the core narratives to the respective groups varies from acceptable to good.

The results of Study I are judged to be valid for illness experiences in adults with MD, since the experiences were largely similar in the three core narratives, despite different diagnoses. It should be pointed out that cognitive effects are not reflected in the core narrative regarding MyD, since the selection included only the persons with the most narrative responses, this being an example of purposive sampling (Polit & Hungler 1999). However, persons with milder cognitive deficits and dysarthria were among those who were covered by the validation work. Those with severe cognitive deficits (n=4) or severe mental illness (n=1) were excluded earlier from the research project (Ahlström & Sjödén 1996). The purposive sampling brought about the limitation that persons with difficulties in expressing themselves due to dysarthria or cognitive effects were not included in the inductive analysis.

Credibility was assessed also for the analyses of the interviews about rehabilitation (Study V). The phenomenological method implies that faithfulness to the interview text is guaranteed by the exactitude in transformation and the formulation of syntheses (Giorgi 1996). In the present study, however, validation was performed by independent judges for both transformation and the formulation of synthesis for the 37 interviews. The validation demonstrated that the interviewer (an occupational therapist) and the independent judge (a nurse) had arrived at the same content in the transformations apart from differences in the choice of words. The interviewer included both large and small efforts performed in everyday life in the term 'making things easier in everyday life', while the independent judge interpreted this as only referring to large efforts. It was decided to use the term 'making things easier in everyday life' for both small and large efforts. Thus, there was good interview judge agreement. This was judged to favour credibility since the interviewer and the independent judge had different experiences and occupational backgrounds.

In the next stage of the validation procedure, the researchers formulated a synthesis for each study person (n=37) on the basis of the second transformation. The syntheses were compared. Where differences were found there was a discussion of changes based on the criterion that the syntheses should reproduce the second transformation and the basic text as faithfully as possible. These 37 syntheses then formed the basis for the formulation of a general structure. In the literature, the question of whether validation is warranted in phenomenological research is a subject of debate. Those who argue against credibility testing state that the exactness of the method itself guarantees credibility (Beck 1994, Giorgi 1997). Usually, few interview persons are included in phenomenological studies (Giorgi 1996). In the present study, validation was performed since the author had limited experience of qualitative analysis. The large number of interviews promotes credibility through a broader data base for the general synthesis.

The statistical methods were chosen on the basis of the scale level of the data and instructions in existing manuals for the instruments used. Most of the correlations based on ordinal data were computed using non-parametric methods. Some parametric correlations were performed on data from the ADL Staircase (Study IV). Overall results computed with different correlational methods were similar.

Few instruments have been developed specifically for adults with MD. There is a need not only for general instruments for making comparisons but also for specific instruments to describe the unique aspects of this patient group (Ahlström 1994). These dual rationales guided the selection of instruments in this study. Some aspects of the instruments are presented below.

The APC is a recently developed self-report instrument concerned with the way the persons sees his or her own situation in activities of daily living and the coping strategies that are used. This assumes that the person has the capacity to evaluate his or her own situation. Part 1 of the APC aims at making a general survey of the person's situation, and it may be seen as a screening instrument. A limitation is that this part gives no information on how often the person uses coping strategies, which is assessed in Part 2. The instrument provides no information on the person's degree of activity restriction, which must be made up for by the occupational therapist using other forms of assessment. One possible merit of the APC is to make use of the person's competence to decide what is a problem and in what areas the person feels he or she needs help. A further advantage of the APC is that it covers many areas of activities.

Further research is needed to assess the reliability and validity of the APC for patient groups with other diagnoses. The instrument showed acceptable internal consistency (alpha 0.70), while test-retest values varied greatly for different parts. Activities such as walking outside varied in their degree of difficulty between

seasons. Test-retest reliability should be assessed in a larger group in a future study in order to further test the reliability of the instrument (Tollén & Ahlström 1998). The construct validity was judged to be acceptable on the basis of the inductive analysis on which the instrument is based. The two pilot studies show that the APC questions were relevant and comprehensible to the study persons (Study II).

The persons completed the APC in conjunction with the start of the rehabilitation and six months later. This is judged not to have affected the result, since the persons cannot be supposed to be able to remember their answers on the first occasion after such a long time period. After the pilot study, some of the questions were refined and the response format was modified. The few differences that emerged between MD in Örebro and MyD in Norrbotten and in relation to the PPS group suggest that the rehabilitation had not affected the APC data in any appreciable extent (Study II).

The Self-report ADL was developed inductively for persons with MD (Ahlström 1994) and has been tested for construct validity in a previous study (Ahlström & Gunnarsson 1996) and in a study of the postpolio syndrome (Ahlström & Karlsson 2000). Previous research has shown acceptable reliability and validity for adults with these types of disorders (Ahlström & Gunnarsson 1996, Ahlström & Karlsson 2000). The present study is the first in which the instrument was used in repeated assessments over a period of time (Study II). The results show an increase in disability over time, which attests to the validity of the Self-report ADL for persons with MD.

The MAC-scale has been tested psychometrically and has been used in several Swedish studies in the field of cancer research (Berglund et al. 1994, Nordin et al. 1999). This instrument has shown satisfactory values for reliability and validity (Nordin et al. 1999). The sensitivity of the instrument for measuring changes over

time may be questioned on the basis of results of the present study. The need for further testing in longitudinal studies has been pointed out also in previous research (Lampic et al. 1994, Nordin & Glimelius 1997).

The Quality of Life Profile has been developed for persons with muscular weakness, and it is intended as a health-related quality of life instrument. It provides data on positive as well as negative consequences of the disease, and it is based on statements of persons with MD at interviews and on questionnaire data from studies using the Sickness Impact Profile (Ahlström & Karlsson 2000). The Quality of Life Profile has mainly a nominal scale level apart from the indices, which constitute a total of the number of answers. The discriminatory validity has proved to be acceptable in a previous study (Ahlström & Karlsson 2000). The usefulness of this instrument is, however, limited in that it assumes that the persons have lived with the disease for a number of years, having fallen ill while still young. The instrument is at the same time unique in this respect.

Study IV shows the strongest correlation between on the one hand Ambulation (Self-report ADL) and the ADL Staircase and on the other the Physical index (SIP). These instruments contain items regarding capacity for activity. For this reason it was not unexpected to find strong correlations between these indices. This has also been shown in previous research (Ambulation/ADL Staircase and Physical index in SIP, r=0.82), (Ahlström 1994).

The results of the five-year follow-up (Study IV) cannot be directly compared with those of other published longitudinal research (Dahlbom et al. 1999, Fowler et al. 1995). These studies differ with respect to selection, namely consecutive patients (Fowler et al. 1995) compared with selection from the population. The three studies also differ in their theoretical basis, and they use different assessment instruments.

The study concerning perceived support from the rehabilitation programme has a weak design, because it was limited to a qualitative assessment performed only after the intervention. A quantitative study using a before and after assessment of disability, coping and quality of life was also carried out. The results show very few changes after the intervention (Ahlström et al. unpublished data). The present qualitative evaluation gives no indications of the effect of rehabilitation, but only a view from the patient perspective. The risk of these persons giving socially desirable answers concerning the rehabilitation programme must be kept in mind.

Another limitation in the qualitative evaluation of the rehabilitation programme is that the persons had participated in previous research (Ahlström 1994). For this reason they may be expected to have a positive attitude since they have previously been the subject of attention. This is likely especially because these patients have been shown to have a low consumption of health care (Ahlström 1994, Ahlström & Gunnarsson 1997). Some of the staff had chosen to participate in the project, which may be expected to have enhanced their sense of involvement in the programme. It is hard to tell how far this has affected the result. One measure that was taken to reduce such an effect is that those responsible for the project, or staff on the rehabilitation team did not conduct the interviews in the qualitative evaluation of the rehabilitation.

The data concerned with activities of daily living, coping and quality of life were collected with the help of rehabilitation staff in the counties of Örebro, Östergötland and Norrbotten. This is judged not to have affected the results since the instruments were of a self-reported character. The ADL Staircase, which was used for a structured interview, was already well-known by all who were engaged in the data acquisition. Staff engaged in the collection of data were also given detailed information on procedures and this information was given to all by those responsible for the project.

The extent to which the results can be generalised to other groups with MD is judged to be fairly good for the quantitative assessment. The main reason is that the study included three MD groups from different counties, between which there were very few differences. Also, the instruments used were either specifically developed for MD or had been tested in previous studies of this group and were judged to be valid.

The MD group in Örebro county was identified in a total survey. The MD group in Östergötland was selected from the patient file at the Department of Neurology, Linköping University Hospital. These subjects comprise the same diagnoses and were in similar age to those in the county of Örebro. The MD group in Norrbotten was identified in a population survey. These samples were also comparable with respect to gender, mean age and disease duration.

The results of the two qualitative studies may be assumed to be representative of other persons with MD. The reasons for this are the validation procedures that have been conducted to guarantee credible results, and the fact that the interviews were performed with persons representing the population (Ahlström et al. 1993). There is some doubt regarding the qualitative evaluation of the rehabilitation programme. This result may have been affected by the fact that the persons had previously been paid attention to and may therefore have given "socially desirable" answers. If this is the case, it suggests that the repeated follow-ups themselves constitute a form of psychosocial support, making things easier in the lives of these persons who are living with a hereditary, incurable disease that takes a progressive course, and that underlines the importance of recurrent rehabilitation measures for persons with MD.

Summary

There is little previous research on the ways in which adults with MD experience their life situation and their activities of daily living. Therefore, the aim of this thesis

was to get knowledge about what it means to live with MD in terms of illness experiences, coping, and quality of life and about patient perceptions of rehabilitation. The results may be summarised as follows.

- When the diagnosis of an incurable, progressive, hereditary disease was announced, this was experienced in a similar way irrespective of the diagnosis, and the results were anxiety, fear, aggressiveness, and worry about the future. It was a painful experience to think of the future and to be reminded that the disease will progress continually, although the very slowness of the progression was felt to be a favourable factor. An initial transition, from a healthy person to a person with a disease, became evident when the diagnosis was announced. A second transition, from independence to dependence on help in daily life, was described most clearly by those whose activities suffered the greatest restrictions.

- More than half of the study persons were dependent on other persons in activities of daily living, and this dependence increased significantly over a five-year period. Mobility indoors and outdoors was reported as the most common problem in activities of daily living. The persons with proximal MD showed most problems with regard to walking, and those with MyD showed problems in activities demanding either distal or proximal muscular strength. Regarding activities calling for distal muscular strength, it became clear, not unexpectedly, that persons with distal MD experienced the greatest disability. Nearly half of the study persons were in gainful employment, and those who were, found that they had problems at work. Their muscular weakness had brought about an increasing amount of sedentary work over the period.

- The persons strive to manage by themselves, and this is reflected in the fact that ‘Fighting spirit’ was the dominant coping strategy. They used problem-focused coping relatively infrequently and the most used widely was ‘Devices and

tricks'. There were no differences between diagnoses for problem-focused coping and few differences between genders. Emotional and cognitive coping strategies showed no changes over a five-year period.

- The lowest degree of satisfaction in activities of daily life was shown for Mobility and transportation and there were three times as many negative as positive consequences of the disease, irrespective of the degree of restriction in activities. One person in six stated that they had not accepted the restrictions brought about by their disease, and fewer than half had partially accepted restrictions in activities. Physical and psychosocial aspects of health-related quality of life, and satisfaction deteriorated over the five-year period.

- There were moderate correlations between activities in daily life, but a weak correlations between coping and quality of life. However, The more use of 'Helplessness/ hopelessness' and 'Fighting spirit', was accompanied by a lower quality of life. The strongest correlation appeared between activities of daily living and health-related quality of life.

- The participants experienced the rehabilitation as highly valuable, receiving support from capable, committed rehabilitation staff and also adaptation of home and workplace, technical aids and practical advice. The participants exchanged advice and support, which was experienced as something very positive. According to the participants, the interest shown by relatives in the rehabilitation brought about increased understanding since the relatives acquired knowledge and were able to meet other people with MD.

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