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Hand function, activity limitation, and health-related quality of life in patients with polymyositis and dermatomyositis

THESIS FOR DOCTORAL DEGREE (Ph.D.)

by

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ABSTRACT

Background: Polymyositis (PM) and dermatomyositis (DM) are rare idiopathic inflammatory myopathies. Common clinical features are proximal muscle weakness and reduced muscle endurance, which can lead to activity limitation and reduced health-related quality of life (HRQoL). The current body of knowledge about hand function, activity limitation, and HRQoL in patients with PM and DM is limited.
Aim: The overall aim of this thesis was to describe and explore hand function, activity limitation, work ability, and HRQoL in patients with PM and DM.
Method: Four papers with cross-sectional, over-time, or intervention pilot study designs have been applied in this thesis. Descriptive, comparable, over time, and correlational statistics have been used. In all, 143 patients with PM and DM participated in the studies.
Results: The results in this thesis showed that both women and men with PM and DM have reduced grip force and HRQoL compared to gender- and age-matched values from the literature. The reduced grip force and HRQoL were measured at the time of diagnosis in both women and men. Women had a reduced grip force at years 1-4 and at 6 years after diagnosis, while the men were affected up to 2 years after diagnosis. The HRQoL was rated lower than the normative values up to 6 years after diagnosis in women and 2 years following diagnosis in men. The grip force had a moderate to high correlation to the HRQoL dimensions of Role Physical, General Health, Vitality, and Mental Health. A hand exercise intervention seemed to be feasible to perform with good adherence but generated few individual improvements in hand function and activity performance why the protocol needs to be adjusted. Patients with reduced hand grip strength also demonstrated activity limitation (according to the Disability of the Arm, Shoulder, and Hand questionnaire) and reduced dexterity. In patients with PM and DM, 44% worked full-time (40 h/week), 31% worked part-time, and 25% were on full-time sick leave. More than 50% of patients with PM and DM self-rated their work ability as “poor” or “less good”. Physically strenuous work components were present “quite often” to “very often” in up to 79% of the patients and were more prevalent in patients on sick leave ≥2 years. For those working, interfering factors in the work environment concerned task and time demands. Supporting factors were the meaning of their work, interactions with co-workers, and others. A low self-rated work ability was correlated moderate-high with a low percentage of full-time employment, the presence of work-related risk factors, and constraints in the work environment.
Conclusion: Patients with PM and DM have reduced hand grip strength, lower ratings on HRQoL, and poor to less good self-rated work ability and the low grip strength may influence HRQoL whereas the proximal weakness seems to affect the ability to work. Measures of hand function and work ability should be included in care of patients with PM and DM to guide interventions that could minimize impairment and as measures in the evaluation of treatment.

SAMMANFATTNING

Bakgrund: Polymyosit (PM) och dermatomyosit (DM) är ovanliga inflammatoriska sjukdomar. Vanliga symtom vid PM och DM är nedsatt muskelstyrka och uthållighet i de proximala musklerna, dvs. de som ligger nära bålen. Den nedsatta muskelfunktionen påverkar de dagliga aktiviteterna och livskvaliteten. Kunskapen är begränsad om hur handfunktion, aktivitetsförmåga och livskvalitet är påverkad hos personer med PM och DM.
Syfte: Målet med denna avhandling var att undersöka handfunktionen, aktivitetsförmågan, arbetsförmågan och livskvaliteten hos personer med PM och DM.
Metod: Avhandlingen innehåller fyra delarbeten med sammanlagt 143 patienter med PM och DM. Tvärsnitt, över tid- och intervention pilot design har använts i denna avhandling. Vid dataanalys har beskrivande-, jämförande-, över tid- och sambandsstatistik använts.
Resultat: Resultatet i avhandlingen påvisar att både kvinnor och män med PM och DM har nedsatt gripkraft och livskvalitet. Både hos kvinnor och män var gripkraften och livskvaliteten nedsatt vid sjukdomsdebut. Hos kvinnorna var gripkraften och livskvaliteten även nedsatt upp till 6 år efter sjukdomsdebut medan hos männen upp till 2 år efter sjukdomsdebut.
Handträningsintervention tycks vara möjlig att genomföra med god följsamhet men endast få individuella förbättringar i handfunktion och aktivitetsförmåga kunde ses, varför träningsprogrammet behöver justeras.
Av patienter med PM och DM i arbetsför ålder arbetade 44 % heltid (40 timmar/vecka), 31 % arbetade deltid och 25 % var heltidssjukskrivna. Mer än hälften självskattade sin arbetsförmåga som mycket låg eller låg.
Fysiskt ansträngande moment i arbetet (arbetsrelaterade riskfaktorer) förekom ganska till mycket ofta hos upp till 79% och oftare hos de patienter som hade varit sjukskrivna mer än 2 år. För de som arbetade ansågs tidskrav och arbetsuppgiftens krav vara ett hinder för att bevara arbetsförmågan. I arbetsmiljön ansågs arbetets värde och betydelse, interaktion med arbetskamrater och andra som ett stöd för att bevara arbetsförmågan. Den självskattade arbetsförmågan hade måttliga till höga samband till arbetsgrad (procent i arbete), arbetsrelaterade riskfaktorer samt hinder och möjligheter i arbetsmiljön.
Konklusion: Patienter med PM och DM har nedsatt gripkraft, livskvalitet och arbetsförmåga. De symtom som är sjukdomsspecifika tycks påverka förmågan att fortsätta arbeta. Det finns ett behov av att undersöka handfunktion och arbetsförmåga hos personer med PM och DM i vården för att minimera funktionsnedsättning och för att utvärdera behandling.

LIST OF SCIENTIFIC PAPERS

I. **Regardt M, Welin Henriksson E, Alexanderson H, Lundberg IE. Patients with polymyositis or dermatomyositis have reduced grip force and health-related quality of life in comparison with reference values: an observational study. Rheumatology (Oxford). 2011;50(3):578-85. Epub 2010/11/26.**

II. **Regardt M, Schult M-L, Alexanderson H, Alemo Munters L, Dastmalchi M, Dani L, Lidén M, Lundsten K, Lundberg IE, Welin Henriksson E. Grip force and health-related quslity of life in patients with polymyositis and dermatomyositis at different time points. (Submitted)**

III. **Regardt M, Schult ML, Axelsson Y, Aldehag A, Alexanderson H, Lundberg IE, Elisabet Welin Henriksson. Hand Exercise Intervention in Patients with Polymyositis and Dermatomyositis: A Pilot Study. Musculoskeletal care. 2014. Epub 2014/03/14.**

IV. **Regardt M, Welin Henriksson E, Sandqvist J, Lundberg IE, Schult M-L. Work ability in patients with polymyositis and dermatomyositis: An explorative and descriptive study. (Submitted)**

LIST OF ABBREVIATIONS

ACSM	American College of Sports Medicine
AFS	Swedish Work Environment Authority's statute book
AWC	Assessment of Work Characteristics
AWP	Assessment of Work Performance
BP	Bodily Pain
CK	Creatine Kinase
DASH	Disability of the Arm Shoulder and Hand
DM	Dermatomyositis
EPM-ROM scale	Escola Paulista de Medicina-Range of Motion-scale
GAT	Grip Ability Test
GH	General Health
HAQ	Health Assessment Questionnaire
HRQoL	Health-related quality of life
ICD-10	International Statistical Classification of Disease and Related Health Problems
ICF	International Classification of Functioning, Disability and Health
ICIDH	International Classification of Impairments, Disabilities and Handicaps
IMACS	International Myositis Assessment and Clinical Studies
MAP	Myositis Activities Profile
MCP	Metacarpophalangeal
MH	Mental Health
MMT-8	Manual Muscle Test in 8 muscle groups
MoHO	Model of Human Occupation
N	Newton
OMERACT	Outcome Measures in Rheumatology
PF	Physical Functioning
PGA	Physician Global Assessment of Disease Activity
PIP	Proximal interphalangeal
PM	Polymyositis
RA	Rheumatoid Arthritis

RE	Role Emotional
ROM	Range Of Motion
RP	Role Physical
RPE Borg CR	Rating of Perceived Exertion Borg Category Ratio
SF	Social Functioning
SF-36	Short Form-36
SweMyoNet	Swedish Myositis Network
VAS	Visual Analogue Scale
VT	Vitality
WAI	Work Ability Index
WEIS	Work Environment Impact Scale
WHO	World Health Organization

1 Background

The background will describe and explore hand function, activity limitation, work ability, and health-related quality of life (HRQoL) in patients with polymyositis (PM) and dermatomyositis (DM) using the framework of the International Classification of Functioning, Disability, and Health (ICF) (1).

1.1 Polymyositis and dermatomyositis

Polymyositis and DM are idiopathic, chronic inflammatory diseases leading to muscle weakness and low muscle endurance that result in restrictions in activity performance and a low HRQoL (2-4). Polymyositis and DM are rare diseases with an annual incidence of between approximately 6-8 cases per 100,000 person-years and an annual prevalence of 14-17 persons per 100,000 (5). Data from Sweden in 1993 show approximately the same prevalence: between 3.3-7.4 cases per 100,000 (6). These disorders often affect persons who are still of a working age, as the majority of the patients are 45-64 years old at disease onset (5). There are approximately twice as many women than men with PM and DM (5).

The disease course varies from a severe, progressive systemic disease with multi-organ involvement to a slowly progressive onset mainly affecting muscle performance (7). The prognosis varies, and the overall improvement of muscle performance is slow, taking place over a period of months. Approximately 20% of the patients go into remission and recover muscle strength, whereas the majority of the patients develop persisting muscle weakness and low muscle endurance that has a negative effect on muscle and activity performance (2, 3).

1.1.1 Definition and diagnosis

Polymyositis and DM affect both body function and body structure in the skeletal muscle, including the skin and other organs such as the oesophagus, heart, and lungs (7-11). These symptoms all contribute to the morbidity and mortality rate (<10%) that exists in patients with PM and DM (9, 11). The impairments in patients with PM and DM include reduced muscle strength and muscle endurance more commonly existing in the proximal muscles (7, 10), which can lead to difficulties in activities, participation and negatively

affects HRQoL (2-4). What distinguishes PM from DM is that DM also has rash, most commonly on the face, knuckles, and upper eyelids (7).

Bohan and Peter defined the criteria to diagnose PM and DM in the 1970s (12). In their definition, there are five major disease criteria to diagnose PM and DM (12) (Figure 1):

1. Symmetrical proximal muscle weakness
2. Elevation of serum muscle enzymes, such as creatine kinase (CK) and aldolase
3. Abnormal electromyographic findings, such as the following:
    - Short, small, polyphasic motor units
    - Fibrillations, positive sharp waves
    - Insertional irritability
    - Bizarre high-frequency repetitive discharges
4. Abnormal muscle biopsy findings, such as the following:
    - Mononuclear infiltration
    - Regeneration, degeneration
    - Necrosis
5. Dermatological features of DM, such as Gottron´s sign, papules, or heliotrope rash

Figure 1. Bohan and Peter's five major diagnostic criteria (12).

Polymyositis and DM can be further classified as definite, probable, or possible disease (12). For *definite DM,* the patient should have three or four of the diagnostic criteria (including rash), while for *definite PM,* the patient needs four criteria (without rash) (12). For *probable DM,* the patient should have three criteria (including rash), while at least three criteria (without rash) are required for *probable PM* (12). For *possible DM*, two criteria are required (including rash), and *possible PM* requires two criteria (without rash) (12). This thesis included patients with definite and probable PM or DM.

1.1.2 Treatment

Treatment in PM and DM is largely pharmacological, and physical therapy is also recommended as exercise can be used as intervention (7, 13-18). However, no studies have evaluated the effectiveness of different occupational therapy interventions in PM and DM.

Pharmacological treatment of PM and DM is based on high doses of glucocorticoids and immunosuppressive drugs, such as methotrexate and azathioprine. These drugs are used to minimize the usage of steroids (7, 13-15). A majority of the patients respond to the pharmacological treatment, but their effectiveness is inconclusive (7).

Despite the fact that most patients have a favourable response to pharmacological treatment, a majority still develops sustained disability (2, 3). Therefore, the importance of endurance exercise in addition to pharmacological treatment is increasingly being recognised (16).

Earlier patients with PM and DM were advised to refrain from exercise and physical activity since it was believed that exercise increased the inflammation in myositis (17). During the last two decades, several studies have shown that aerobic, resistance, and endurance exercise have been well tolerated without signs of increased disease activity or inflammation (14, 15, 17, 18). New evidence even suggests that exercise may reduce inflammation (17). The exercise interventions performed in PM and DM are intended to increase muscle strength in the proximal muscles and to improve aerobic capacity (13-15).

1.2 Theoretical framework

The ICF are used as the theoretical framework in this thesis for classification of functioning, disability, and health. The ICF describes the patients and the measures used as well as the results for the following ICF components: body functions, body structures, activities, and participation.

1.2.1 The International Classification of Functioning, Disability and Health (ICF)

There is a need for a common language regarding health since the World Health Organization (WHO) has the task to work for public health (19). The WHO requires statistical and epidemiological reports about public health from its members (19, 20). For several years, the International Statistical Classification of Disease and Related Health Problems (ICD-10, 1994) was used to determine these reports on death or mortality (20). However, the ICD-10 did not give a comprehensive picture about public health, and there was also a need to report health outcomes that were non-fatal (20). For instance, some conditions affect health and limit function in everyday life even though they are not considered fatal, e.g. depression (20).

Health can be described as follows: '[...] health is both a matter of how long one lives and how well one lives […]' (20) (p. 566). Therefore, a need developed for a classification system with a common language that would enable comparisons between countries and health care systems and to establish a common scientific base to understand health and the consequences of disease (1, 21). In 2001 the ICF was endorsed by the 54th World Health Assembly for international use (19). The ICF is based on an earlier version called the International Classification of Impairments, Disabilities, and Handicaps (ICIDH) from 1980 (20). One of the differences between these versions is that the ICF focus on health components, while the ICIDH had concentrated on the consequences of disease (1). The ICF provides a new, more neutral stand and highlights functioning as a component of health (1, 20). When the ICF was developed, cultural and linguistic aspects were considered, which was a limitation of the earlier version (ICIDH) (20). Thus, the ICF provides a common language to describe health that is used in research to mediate the results amongst different professions in health care and between different countries (1, 19).

The ICF has a base in both a *medical model* and a *social model* (1, 19). The medical and social model has some contradictory features but should be combined for a comprehensive picture of health (1, 19). The *medical model* sees the disability as a problem within the person, which is directly caused by a disease; the health care system has the responsibility to cure or treat the disease. Health care systems have a strong

influence from the political level, which has the obligation to facilitate good healthcare with the mission to cure disease. The *social model* sees the disability as a socially created problem generated by society. The disability is not due to the person but instead to a complex combination of circumstances. From this perspective, the society needs to change to prevent disability (1, 19).

The general view of disability needs to consider these models. According to the ICF, both models have been combined into the *biopsychosocial model*, which views health as a combination of biological, individual, and social perspectives (1, 19). In addition, health care providers have the obligation to involve the patient in the choice of treatment, and the patients have the right to autonomy according to the Swedish Health and Medical Services Act (22).

The different components in the ICF interact with each other according to the following model (Figure 2):

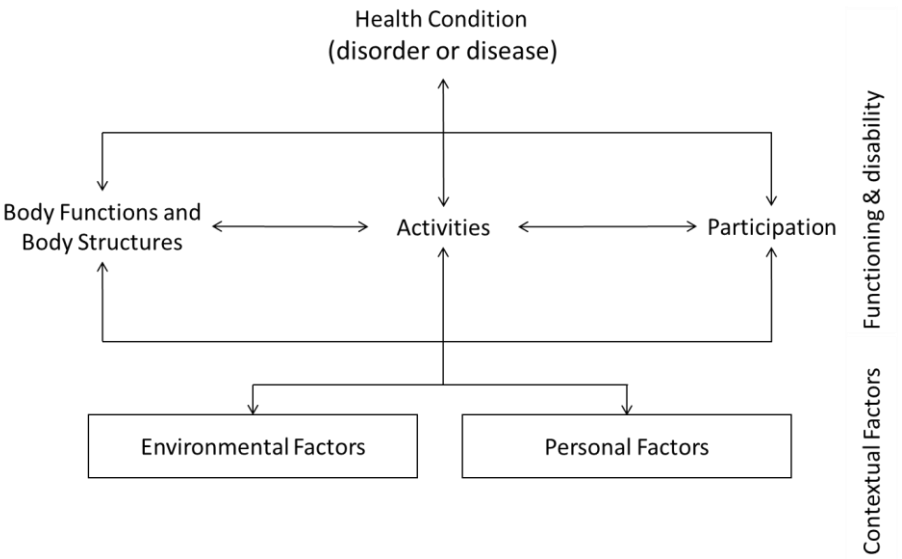

Figure 2. The ICF model and the interactions between the ICF components (1, 19).

In Figure 2, the reverse arrows between the components illustrate that it is not only in the person; the disability can be caused by but also that the environment or personal factors affect disability (1, 19). The ICF can be used to describe how a person is affected by his or her health condition, including the bodily symptoms as well as the possibility for participation. Furthermore, the environment's influence and personal factors can affect a person's ability to handle the health condition (1, 19)

The different components are defined as follows (1, 19):

- **Body functions** are both the physiological and psychological systems in the body.
- **Body structures** are the anatomical part of the body, such as organs, limbs, and their components.
  Disabilities in both body function and body structure are labelled as *impairment*.

- **Activities** are the ability to execute a task or an action by an individual, and *disability* in activity performance is labelled as *activity limitation*.
- **Participation** is defined as involvement in a life situation, and disability is defined as *participation restrictions*.
  Activity and participation involve learning and applying knowledge, carrying out daily routines, mobility, self-care, domestic life, etc.

- **Environmental factors** involve the physical, social, and attitudinal environment in which people conduct their lives.

- **Personal factors** include gender, age, coping styles, social background, education, overall behaviour pattern, and other factors that influence how disability is experienced by the individual.

The different components in the ICF have a hierarchical order with a one to four digit level to describe functioning, disability, and contextual factors, including about 1200 categories altogether that are used to describe patients with different health conditions. The categories in the activity and participation components can be single tasks or multiple, complex compositions of tasks evaluated as either being able to do a task or not but also

if someone is doing the task or not (1). For the level-one classification (chapters), the categories are generally described and are gradually more specific onwards to the level-four classifications. In this thesis, a one-level classification will be applied.
To facilitate clinical application, comprehensive core sets have been developed for different diagnoses (23-25).

1.2.1.1 ICF and chronic conditions

During 2002-2003, both brief and comprehensive core sets were developed for 12 different chronic conditions, including rheumatoid arthritis (RA) and osteoarthritis (23) but not yet for myositis. The purpose of the brief core set was to facilitate a minimum of categories that should be applied in clinical practice and research when assessing health in the different conditions. The comprehensive core set aims to describe a broader picture of health from a multidisciplinary perspective (23). The development of these ICF core sets included a method using a formal decision-making and consensus process, a Delphi exercise, and a systematic review alongside with empirical data collection (23).

In the ICF comprehensive core set for RA, there are 96 categories; the brief version has 39 categories, and both versions include the components of body function and structure, activity and participation, and environmental factors (26). For the ICF core set in RA, studies have been done to validate it to different health care providers (27-29) and to the patient perspective (30, 31). These studies indicate that there are several categories in the ICF core set for RA that are included based on the perspective of both the health professionals and the patient but that there are also areas not included in the ICF core set (27-31). In an article using focus groups to describe the experience of systemic sclerosis, 86 categories were identified and linked to the ICF components of body function, body structure, activities, participation, environmental and personal factors, as well as the health condition itself (32).

For myositis, at present there is no developed ICF core set. In a review article about the disabling nature of idiopathic inflammatory myopathies, links to the ICF were discussed, and it was suggested that all the components in the ICF were represented when describing myositis (33).

Even though there is no ICF connected core set, the International Myositis Assessment and Clinical Studies (IMACS) group has established a core set of measures to use to evaluate disease activity in myositis (34). The six-item core set includes assessments evaluated by the physician, patient-reported measures, and laboratory measures (34) (Table 1).

Table 1. Measures included in the IMACS six-item core set (34).

	Measure
By the physician	Physician Global Assessment of disease activity (PGA) rated on a Visual Analogue Scale (VAS)
	Muscle Strength (Manual Muscle Test in eight muscle groups (MMT-8)
	Extra muscular activity (VAS)
By the patients	Patient's global assessment of health (VAS)
	Physical Functioning (Health Assessment Questionnaire, HAQ)
Laboratory measures	Creatine Kinase (CK)

The IMACS group has also developed a consensus on outcome measures and improvement of disease activity (34). A minimum of 15% improvement in muscle strength and physical function is considered clinically meaningful for patients with PM and DM (34). Another organisation working to establish patient-reported outcomes in rheumatic diseases is the Outcome Measures in Rheumatology (OMERACT). The OMERACT currently has a myositis special interest group that aims to establish patient-reported outcomes in myositis (35). Whether these will be connected to the ICF is not known.

The main areas of this thesis include hand function, activities, work ability, and HRQoL.

1.3 Hand function

In general, hand function is important in many activities and includes the ability to move the hands and fingers and having the strength and endurance to perform daily activities for prolonged periods (36).

Mobility, strength, and the absence of pain are required to use the hands in the most effective way (37). Polymyositis and DM mainly affect the proximal muscles; however, some previous researchers have reported that the distal muscles may also be involved (7, 38, 39). One study referred to patients in the late phases of PM and DM (39), and another was conducted in patients with cancer-associated dermatomyositis (CAD) (38).

There are also indications that arthritis in the wrist and finger joints (metacarpophalangeal (MCP) and proximal interphalangeal (PIP) joints) is more common in patients with inflammatory myopathies than was previously thought (≈20%) (40). Arthritis in the hands is known to lead to deformities, dysfunction, and reduced range of motion (41, 42). However, in the studies included in this thesis, PM and DM patients with arthritis or other comorbid diseases that could affect the hand were excluded. Measures of mobility and strength of the hand are linked to the ICF component of body function, while dexterity is linked to activities and participation (43-46). In this thesis, the power of hand grip will be assessed by two different measures. The Grippit©, which measures grip force, will be used in Papers I and II (in Newton, N) (47). In Paper III, a computer-connected Jamar dynamometry will be used, which measures grip strength (in kilograms, kg) (48). The Grippit© assesses grip *force* by calculating the average force used when the patient squeezes as hard as he or she can for 10 seconds (47). The measure of grip force indicates the endurance to maintain the power of the grip. This differs from the measure of hand grip strength (Jamar) where the patients are asked to squeeze as hard as they can (peak strength) three times, and the average of these three trials is taken as the measure (47, 48). Since these two assessments measure different entities, they are not comparable (48).

1.3.1 Hand exercise

Hand exercise interventions are commonly used as a treatment to improve hand function and to improve or maintain activity performance (49). In rheumatic or muscle-affecting

diseases, studies on hand exercises have indicated improvement in hand function (50-54). These hand exercise studies usually include movements to enhance range of motion (ROM) and/or hand strength (51, 55, 56). An effective prescription (number of repetitions and frequency) for hand exercises to improve hand function has not been established (41). In general, to enhance strength and endurance, the American College of Sports Medicine (ACSM) recommends exercise 2–3 days per week, with every movement repeated 8–12 times (57). To enhance muscle strength and endurance over the exercise period, resistance, frequency, or duration must be increased (the progressive overload principle) (57). Hand exercises included in an occupational therapy programme showed increased activity performance in patients with RA (58). Until recently, no studies had evaluated the feasibility and the effectiveness of a hand exercise intervention in patients with PM and DM.

1.4 Activities

The disability that the myositis causes increases significantly during the disease course (3, 59), and a substantial number of patients experienced reduced ability to function in their environment and manage their activities (2, 4, 60). Thus, patients with PM and DM are affected in their activity performance due to the muscle weakness and reduced muscle endurance in the proximal muscles. Commonly reported difficulties include rising from a chair, walking up stairs, lifting objects, or combing their hair (2, 33, 39). In occupational therapy, *activity* and *occupation* are two central concepts.

1.4.1 The concepts of activity and occupation

There are several different views about what defines activities and occupations in the occupational therapy and occupational science literature (61-66). One elementary way to differentiate activity from occupation is that occupation has to have a goal and to be meaningful (61, 63, 64).

Nelson (61) describes occupation as two things, occupational form and occupational performance (61). Occupational form is the objective environment and sociocultural reality where the actual doing/activity performance take place (61). Nelson sees the occupational form as the task; when meaning, structure, and a goal are added, occupational performance is accomplished (61). Other authors and researchers share this

view. Pierce (62) describes occupation as a subjective event that has a form, a pace, a beginning, and an end. In contrast, activity is a generalizable classification of human actions that is defined by one's culture (62). In Pierce's point of view, occupation is something that has happened and is dependent on the individual, the place, and the time, and can therefore not be repeated (62). An activity becomes an occupation when an individual has performed it (62). Turner et al. (63) defines occupation according to four statements. Occupation is the doing in a person's everyday life; it is driven by the person's needs, aspiration, and environment. It relates to the individual's definition of purposeful use of time; occupations are the means a person uses to create balance and control in life (63). According to Turner et al. (63), activities are the blocks that together create occupation (63). Occupations are based upon a person's style and are influenced and dependent upon that person's roles (64).

The concept of activity as described in the ICF has similarities to the description of activity made by Golledge (64) and Turner et al. (63), where activities are seen as the bricks that together with doing, meaning, need, balance, control, and goals, become an occupation (63, 64). According to Pierce, occupation cannot be objective and generalizable because it is dependent on the person, the setting, and the time in which it is performed (62) and therefore not what the ICF measures in the component activities. In the definition of occupational form by Nelson (61), there are some similarities to the ICF's environmental factors and personal factors. Nelson reflects about the values, norms, and roles that the ICF includes (61). Based on this discussion between occupation and activities, the component activities defined by the ICF are more closely connected to the concept of *activity*, and this concept will be explored in this thesis. One important aspect of activity both for the society and the individual is the ability to work (67).

1.4.2 Work ability

Work in general is important both for the society and the individual since work has positive effects on health and the economy (67). Periods of sick leave should be minimized because long-term sick leave affects the individual negatively and may result in reduced self-esteem and self-confidence, which makes a return to work more difficult (67, 68). Work ability has been described as the relationship between the individual's resources and job demands. These demands include what type of work it is, what the work

incorporates, the organization, and the work environment (67, 69, 70). Thus, reduced work ability is associated with more physically demanding jobs and older age (69, 71). If the demands from work are higher than the individual's resources, the work ability is decreased and needs to be restored, improved, or supported (67).
Sandqvist and Henriksson (72) suggest a conceptual framework that is of importance for the assessment of work functioning (72). They divide work functioning into three dimensions: *work participation and society* (Work Participation), which includes both the individual's ability/opportunity to acquire/maintain a work position in the society and to fulfil a work role as well as conditions on the labour market. The second dimension, *work performance and the individual* (Work Performance), involves the individual's ability to satisfactory perform different work tasks necessary for a certain work position. The third dimension, *individual capacity and physiological/psychological functioning* (Individual Capacity), involves the physical and psychological attributes of an individual that enable work activities, such as muscle strength (72). Work Participation is linked to the ICF component participation, Work Performance to the ICF component activities, and the Individual Capacity to the ICF component of body functions and structures (72).

For patients with other rheumatic diseases, such as RA or systemic sclerosis, work ability is affected (73, 74). In one previous study conducted in Hungary, 42% of patients with PM and DM were not able to work at any time from the onset of the disease due to functional impairments and activity limitations caused by the myositis (4). However, limited information is available on whether PM and DM affect work ability or confer a risk factor for sick leave and early retirement, and the risk factors that affect the work ability have not been identified. The symptoms PM and DM may cause might give rise to psychological consequences that negatively affect quality of life (3, 4, 75).

1.5 Health-related quality of life

According to the WHO, health is a fundamental right of every human and is defined as a state of complete physical, mental, and social wellbeing and not merely the absence of disease or infirmity (76). The latter (health=absence of disease) definition is more of a bio-statistical theory of describing health and defining disease as an organ failure leading to disease or ill health. The use of a holistic theory also includes "feeling" and a phenomenon of "ability or disability" when describing health (77). There could be disease

without affecting feelings, for instance, in a person who is in a coma (77). Since the goals of medicine are to save lives and improve quality of life, health is essential in public health care (77).

Several studies have shown that patients with PM and DM rate lower HRQoL compared to the general population (3, 4, 75). However, lower ratings on HRQoL are not uncommon in chronic diseases (3). Patients with DM also report reduced HRQoL due to the skin manifestations, such as pruritis (78).

In this thesis, the HRQoL was measured using the Short Form-36 (SF-36). The SF-36 was developed because of an increasing consensus of the importance of considering the patients' opinion of their health when evaluating medical care or the impact disease cause on health (79, 80). It also was intended to be a generic instrument that could be used to compare chronic diseases to a general population (80). The SF-36 was designed to be used as a measure in the Medical Outcomes Study (MOS) (79). The choice of dimensions included in the SF-36 was based on concepts in the most frequently used health measures (79). The areas chosen were Physical Functioning (PF), Role Physical (RP), General Health (GH), Social Functioning (SF), Role Emotional (RE), and Mental Health (MH). Two additional dimensions, Bodily Pain (BP) and Vitality (VT), were added based on results from empirical studies (79). These dimensions were assumed to be universal and would represent fundamental human functions and wellbeing (80). By comparing the SF-36 to other measurements used to assess general health, the SF-36 has been found to include the eight dimensions that are most commonly included in these generic measures (81). In the general population, there is a gender difference where men rate their HRQoL better than women, especially in the Physical Functioning, Bodily Pain and Vitality dimensions (82).

The ICF components of body function and structures, and activities and participations, including work/employment, are represented in different dimensions of the SF-36 (83, 84). The SF-36 corresponds additionally to a person's wellbeing as it relates to health perception (83, 84). In patients with PM and DM, the HRQoL is rated lower in all dimensions of the SF-36 compared to the population norms, and interestingly, the HRQoL does not correlate to the disease course or muscle function (3, 4).

1.6 Rationale for this thesis

Patients with PM and DM are classically described as having more proximal than distal weakness (7, 10). However, in a study conducted by our research group, we observed that disability due to distal weakness seemed to be a more common problem in patients with PM and DM (85) than previously described. Therefore, we wanted to investigate how and when hand function was affected. Since hand function is of importance when performing daily activities (36) and has been shown to be associated with HRQoL (86, 87), we wanted to correlate these variables with hand function in patients with PM and DM. Hand exercise is a common treatment to improve or maintain hand function and activity in occupational therapy (49); therefore, we aimed to develop a hand exercise programme to test its feasibility and its possible effect on hand function and activity. Furthermore, since work ability is commonly affected in other rheumatic conditions (73, 74) and work is essential for both the individual and society (67, 72) and also because there was limited information about work ability in patients with PM and DM, we investigated work ability.

2 Aim

2.1 General aims

The overall aim of this thesis was to describe and explore hand function, activity limitation, work ability, and HRQoL in patients with PM and DM.

2.2 Specific aims

The more specific aims were as follows:

- To investigate hand function in PM and DM patients and compare it with reference values in healthy individuals and also to determine if hand function was correlated with activity performance and HRQoL (Paper I).
- To compare patients with PM and DM to normative values from the literature regarding grip force and HRQoL at different time points up to six years after diagnosis to investigate how grip force changes over time and to correlate grip force with HRQoL (Paper II).
- To develop a 12-week hand exercise intervention for patients with PM and DM and evaluate adherence, patient opinion of programme design and overall feasibility, hand function, and activity limitation after the intervention (Paper III).
- To investigate the work situation, work ability, work-related risk factors, and influence of the physical and psycho-social work environment in patients with PM and DM (Paper IV).

3 Methods

3.1 Study design

This thesis employed a quantitative method with either cross-sectional, over time, or an intervention pilot design (Figure 3).

Paper I
Design: Observational and cross-sectional design
Data collection: 2001
Study population: n=31 PM/DM patients
Assessments: Functional assessments and questionnaires
Data analysis:
Descriptive (mean, SD, median, IQR, CI)

Comparative (independent-sample t-test, Mann Whitney U-test)
Correlations (Pearson's and Spearman's correlation coefficient)

Paper II
Design: Observational, cross-sectional, and over time design
Data collection: 2003-2012
Study population: n=89 PM/DM patients
Assessments: Functional assessments and questionnaire
Data analysis:
Descriptive (percentage, mean, SD, median, IQR)
Comparative (Wilcoxon Signed-rank test (null hypothesis=no difference))
Correlations (Spearman's correlation coefficient)

Paper III
Design: Pilot intervention study
Data collection: 2010-2012
Study population: n=11 PM/DM patients
Assessments: Functional assessments and questionnaire
Data analysis:
Descriptive (numbers, percentage, mean, SD, median, IQR)
Comparative (Wilcoxon Signed-rank test (null hypothesis=no difference))

Paper IV
Design: Observational and cross-sectional design
Data collection: 2012-2013
Study population: n=48 PM/DM patients
Assessments: Functional assessments and questionnaires
Data analysis:
Descriptive (number, percentage, mean, SD, median, range, IQR)
Comparative (Mann Whitney U-test)
Correlations (Spearman's correlation coefficient)

Figure 3. Methods, participants, and data analysis in Papers I-IV.
n=number, PM=polymyositis, DM=dermatomyositis, SD=standard deviation, IQR=interquartile range.

3.2 Patients

Patients in all papers were recruited through the Swedish Myositis Network (SweMyoNet) Registry at the Rheumatology Department at Karolinska University Hospital in Stockholm. In Paper II, patients from Akademiska University Hospital in Uppsala and Falun Hospital in Falun were also included.

In Papers I-IV, 143 patients were included (women, n=85 and men, n=58) with PM (n= 81) and DM (n=62). Most of the patients were included in one study (n=111, 62%); 56 patients (31%) took part in two studies, and 12 patients (7%) participated in three studies. Patient demographics for Papers I-IV are presented in Table 2.

Table 2. Demographics for the participants in Papers I, II, III, and IV.

Demographics	Paper I n=31	Paper II n=89	Paper III n=11	Paper IV n=48
Age (years), mean (SD)	56 (11)	60 (14)	63 (12)	54 (10)
Disease duration (years), mean (SD)	7 (6)	n/a	8 (7)	9 (9)
Gender, women, n (%)	18 (58)	52 (58)	6 (55)	29 (60)
Diagnosis, PM, n (%)	20 (65)	53 (60)	5 (46)	25 (52)

n=number, SD=standard deviation, n/a=not applicable, PM=polymyositis.

When the study reported in Paper I was conducted, 50 patients diagnosed with PM or DM were identified through the SweMyoNet Registry at the Rheumatology Clinic at Karolinska University Hospital in Stockholm, Sweden, and were informed about the study.

Inclusion criteria were definite or probable PM or DM according to the diagnostic criteria by Bohan and Peter (12). Exclusion criteria for participation were other diseases or injuries that could affect hand mobility and grip force, such as other rheumatic or neurological diseases. The study included 31 patients (Table 2).

In Paper II, the cohort consisted of patients from three rheumatology clinics in Sweden (Falun, Stockholm, and Uppsala) who were identified through the SweMyoNet Registry. Data were collected between June 2003 and February 2012. Inclusion criteria for Paper II were patients diagnosed with definite or probable PM or DM (12) between 2003 and 2012, with at least one recorded value between the time of diagnosis (year 0) and 6 years after diagnosis on either the grip force as measured by Grippit© (47) *or* HRQoL measured by SF-36 (80, 88). Exclusion criteria were other diseases or conditions that could affect hand function, such as arthritis.

A total of 127 patients with PM or DM were identified. Thirty-eight patients were excluded as they did not have any grip force or HRQoL data recorded or because they had a coexisting condition (Figure 4). Finally, the study included 89 patients with PM and DM (Table 2).

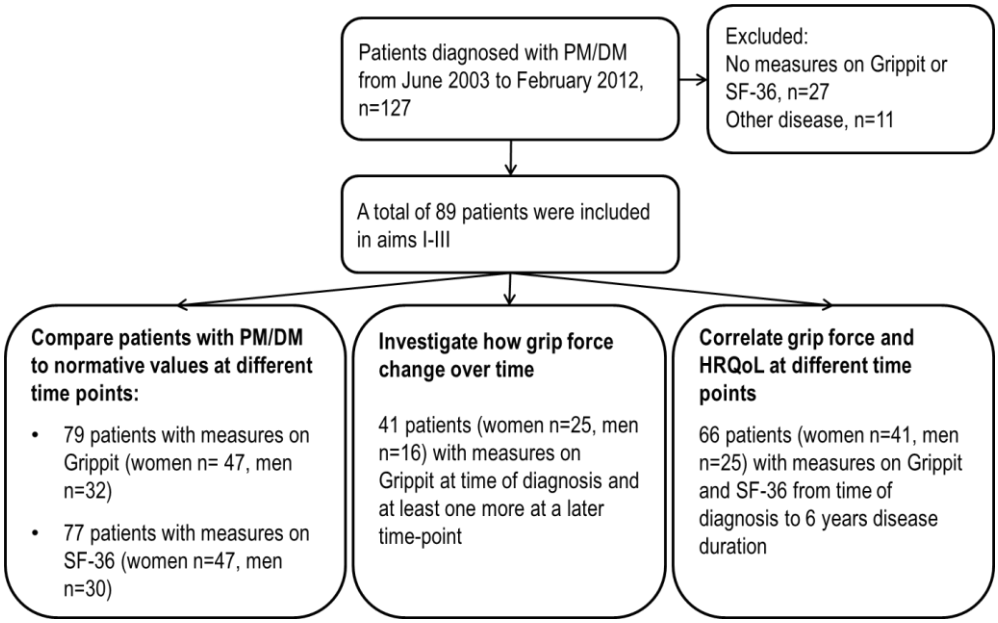

Figure 4. Paper II, flowchart for the patients in the SweMyoNet registry divided into the different aims.
PM=polymyositis, DM=dermatomyositis, n=number, SF-36=Short Form-36.

In Paper III, the hand exercise intervention was introduced to a convenience sample of 15 patients with PM and DM (probable or definite, according to Bohan and Peter's criteria) (12) who were followed at the rheumatology clinic at Karolinska University Hospital in Stockholm and who had the following:

- Reduced hand grip strength (≥20%) when compared to gender- and age-matched normative values from the literature (47) (according to patient records)
- Established disease (> 6 months)
- Low disease activity (with conventional immunosuppressive treatment according to the choice of the treating physician)

A total of 11 patients completed the hand exercise intervention (Table 2).

In Paper IV, 78 patients with PM and DM at working age were identified through the SweMyoNet Registry at Karolinska University Hospital in Stockholm, Sweden, in June 2012 and invited to participate in this study (Figure 5).

Inclusion criteria in Paper IV were patients (a) diagnosed with probable or definite polymyositis and dermatomyositis (12) (b) who were 18-67 years of age, (c) and still alive in June 2012. Exclusion criteria eliminated patients who were (a) unable to read and understand questionnaires in Swedish, (b) on sick leave due to other causes than myositis, (c) studying, or (d) unemployed.

The study included 48 patients with PM and DM (Table 2, Figure 5).

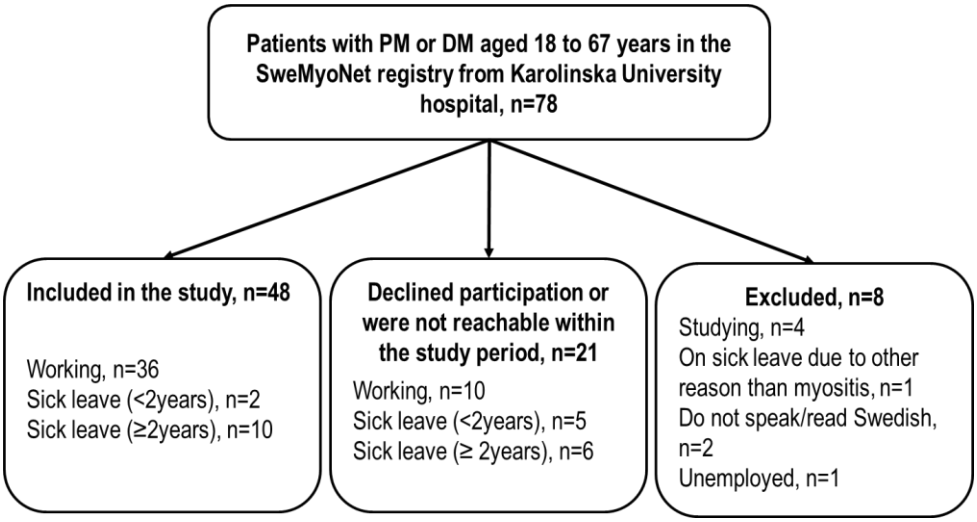

Figure 5. Paper IV, inclusion and exclusion of patients.
PM=polymyositis, DM=dermatomyositis, n=number.

3.3 Data collection procedure

Patients included in this thesis have been identified through the SweMyoNet Registry, which is a national quality of care registry used in clinical practice. Patients are followed longitudinally, and measures are registered once a year, including disease activity, disease damage, and HRQoL, as suggested by the IMACS group (34). The measures are collected by a team made up of an occupational therapist, a registered nurse, a physical therapist, and a physician. There is also an international network regarding myositis that collects data in a registry with the aim to enable research on genetics and epidemiology.

3.3.1 Paper I

In Paper I, the data collection was done at one occasion. The same occupational therapist (M Regardt) assessed hand function, and the patients answered the self-assessment questionnaires.

3.3.2 Paper II

In Paper II, the data were retrieved from the SweMyoNet Registry. The data included follow-up measurements, grip force and HRQoL, patient characteristics at the time of diagnosis (age, diagnosis, and gender), medical treatment, and IMACS recommended outcome measures of disease activity (34). Missing information was retrieved from patient records when possible. As registry data were collected from clinical practice, a time lapse was discovered in some cases between diagnosis and the first measure (year 0). In 23% of the patients, there were no data on grip force or HRQoL from year 0.

3.3.3 Paper III

In Paper III, patients were assessed before and after the 12–week hand exercise intervention (Figure 6). The physician evaluated disease activity (according to the IMACS six-item core set (34)), the occupational therapist (Y Axelsson) assessed hand function (Jamar dynamometer, pinch meter, Grip Ability Test (GAT), Purdue Pegboard), and the patients answered the Disability of the Arm, Shoulder, and Hand (DASH) questionnaire to evaluate activity limitation (89, 90) (Figure 6).

The hand exercise programme was introduced by a second occupational therapist, the author (M Regardt). The occupational therapist observed the patients performing the exercise to ensure that they were able to follow the programme and had understood how the exercises should be carried out. Patients were given the option of exercising at the hospital once a week and follow-up visits were done throughout the study either by phone, at the hospital, or both and were planned jointly by the patient and occupational therapist (M Regardt). The occupational therapist supervised the patients at the follow-up visits and checked the exercises to make sure that the programme was performed correctly. Patients were asked not to change their lifestyles or to start any other form of exercise during the study period, and this was confirmed at the end of the study.

3.3.3.1 Hand exercise programme

Since there is limited information about hand function and whether it affects activity in patients with PM and DM, a general hand exercise programme that included hand strengthening movements involved in accomplishing daily activities was designed.

The programme was based on hand- and finger-movements using personally adapted resistance putties (standardized doughs) {Royal putty [medium, light or x-lite (Mediroyal Nordic AB©, Stockholm, Sweden)] and Jura putty [medium soft (JURA Medical©, Glasgow, United Kingdom)]}. The putties were tested so that each patient could fully flex his or her fingers through the dough (91), and the patients were asked to evaluate subjectively whether the putty was “too soft”, “too hard”, or “just right”. If the dough were too hard or too soft, it was changed to another with more or less resistance or was mixed until it felt “just right”. When the patient could flex his or her fingers through the dough and it felt “just right”, he or she was asked to rate exertion on one of the movements in the programme [finger flexion, 30 repetitions (Figure 6)] using the Rating of Perceived Exertion (RPE) Borg Category Ratio (CR) 10 scale of exertion (92, 93). The occupational therapist (M Regardt) chose the putties based on a lower limit on the RPE Borg CR 10 scale, “moderate exertion” (≥ 3) (92, 93). The mean rating on the scale was “strong exertion” 5 (± 1.8), with the range being from 3 (“moderate”) to 8 (“more than strong exertion”). The hand exercise programme was intended to be performed three times a week for 12 weeks (36 total times) and was illustrated by pictures from the Mobilus Professionals program (Mobilus Digital Rehab AB Sweden©, Gothenburg, Sweden). To facilitate improvement in strength, the number of repetitions increased every fifth week (91). Figure 6 provides further information about the hand exercise programme, the various movements, and the number of repetitions performed throughout the intervention. Patients kept diaries of their exercise, documenting each session performed, their ratings on the RPE Borg CR 10 scale (92, 93), and any comments they had about the programme.

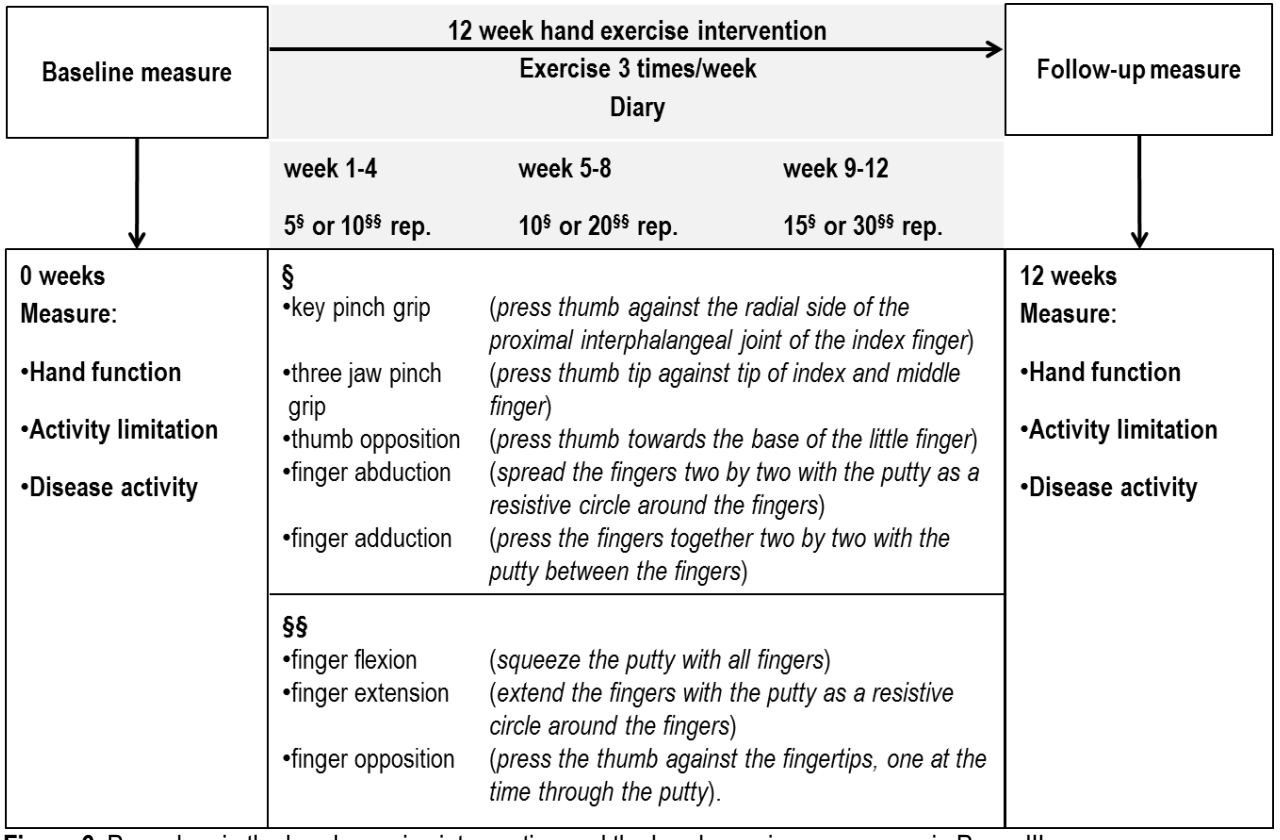

Figure 6. Procedure in the hand exercise intervention and the hand exercise programme in Paper III.

Figure 6 legend. Two sets of movements performed at every exercise session (three times per week for 12 weeks).
§= 5 repetitions performed during week 1-4, 10 repetitions performed during week 5-8, and 15 repetitions performed during week 9-12.
§§= 10 repetitions performed during week 1-4, 20 repetitions performed during week 5-8, and 30 repetitions performed during week 9-12, rep=repetitions.

3.3.4 Paper IV

In Paper IV, an information letter was sent to eligible patients who were then contacted by phone. Patients who were working or had been on sick leave <2 years and who consented to participate in the study were interviewed in person at Karolinska University Hospital by an occupational therapist (M Regardt). Those individuals who had been on sick leave ≥2 years had the questionnaires and consent form sent to them by mail with a prepaid return envelope. If there were any unanswered questions, the occupational therapist (M Regardt) contacted patients by phone to give them the opportunity to complete the questionnaires. Information on disease activity at the time of the study was captured from the SweMyoNet Registry and assessed according to the International Myositis Assessment and Clinical Studies Group (IMACS) (34).

3.4 Outcome measures

In this thesis, outcome measures to assess hand function, activity limitation, work ability, HRQoL, and evaluation of a hand exercise intervention were used (Table 3).

Table 3. Outcomes used in this thesis and their connection to the ICF.

Measure	Characteristics of measure	Paper	ICF components and chapters			
			Body functions	Activities and participation	Environmental factors	Personal factors
Grippit©	Grip force	I-II	b7			
Jamar dynamometry	Hand grip strength	III	b7			
Pinch meter	Pinch grip strength	III	b7			
GAT	Grip ability	III		X		
Purdue Pegboard	Dexterity	III		d4		
EPM-ROM scale	Range of motion	I	b7			
DASH	Physical function and symptoms of the upper extremity	III	b1, b2, b7	d1, d2, d4, d5, d6, d7, d8, d9		X
MAP	Difficulty and importance of specific activities	I		X		
WAI*	Self-reported work ability	IV	b1, b2, b4, b5, b8	d8		
Work-related risk factors*	Occurrence of work-related risk factors are present	IV	b7	d4		
WEIS*	Perception of performance, satisfaction and wellbeing	IV	b1, b2, b7	d1, d2, d7, d8, d9	e1, e2, e3, e4	
SF-36	HRQoL	I-II	b1, b2	d2, d4, d5, d6, d8, d9		
PF				d4, d5, d9		
RP				d2, d8		
BP			b2	d8, d8		
GH						
VT			b1			
SF			b1	d9		
RE			b1	d2, d8		
MH			b1			

GAT=Grip Ability Test, EPM-ROM= Escola Paulista de Medicina-Range of Motion, DASH=Disabilities of the Arm, Shoulder, and Hand, MAP=Myositis Activities Profile, WAI=Work Ability Index, WEIS=Work Environment Impact Scale, SF-36=Short Form-36, PF=Physical Functioning, RP=Role Physical, BP=Bodily Pain, GH=General Pain, VT=Vitality, SF=Social Functioning, RE=Role Emotional, MH=Mental Health, b1=Mental functions, b2=Sensory functions and pain, b7= Neuromusculoskeletal and movement-related functions, d1= General tasks and demands, d2=Learning and applying knowledge, d4=Mobility, d5= Self-care, d6= Domestic life, d7= Interpersonal interactions and relationships, d8=Major life areas, d9=Community, social, and civic life, e1=Products and technology, e2= Natural environment and human-made changes to environment, e3=Support and relationships, e4=Attitudes, X= the measure has a link to the component, *=In the column of measures subjective linking to the ICF component by the author M Regardt.

3.4.1 Hand function

3.4.1.1 Grip force and hand grip strength

Grip force and hand grip strength were measured separately in the right and left hands by either the Grippit© (Detektor AB, Göteborg, Sweden) or the computer-connected Jamar dynamometer (Biometrics E-link H 500 hand kit) (47, 48). The former measure (Grippit©) has been suggested to be a reliable measure of grip force in patients with myositis (94). These measure of grip force and hand grip strength are linked to the ICF component body functions (95, 96) (Table 3). In the general population, there is a difference in grip force and hand grip strength between women and men; therefore, the analysis was done on women and men separately (47, 48).

The Grippit© was used in Papers I and II and is an electronic measure that gives maximum, mean, and final values after a period of 10 seconds and measures in Newtons (N) (47). Measurements are recorded every half second during the 10-second test, and the higher the score, the better the grip force. Mean values over 10 s have been used in the analysis. Normative values on grip force based on gender and age were available from a Swedish cohort of healthy individuals (47).

The Jamar dynamometer is a computer-connected device used to measure hand grip strength in kg, and it was used in Paper III (Biometrics E-link H 500 hand kit) (48). The average of three measures for each hand was used in the analysis. Normative data from a population-based study for women and men in different age groups are available for comparison (48). A minimal significant change of at least 6 kg indicated a clinically meaningful improvement (97).

3.4.1.2 Pinch grip strength

The computer-connected Biometrics pinch meter was used to measure pinch grip strength separately in the right and left hands (kg) (Biometrics E-link H 500 hand kit) in three positions: *key* (lateral), *three-jaw* (tri-pod), and *thumb to index finger* opposition (tip-to-tip) (Paper III). Three trials were performed per position, and the average value per position for each hand was used in the analysis. The literature contains no comparable normative values or guidelines on what could be considered a clinical improvement in the

Biometrics pinch meter (Biometrics E-link H 500 hand kit) since it has a thinner profile design than a regular pinch gauge meter. Therefore, the definition that IMACS suggested for clinically meaningful improvement in muscle strength and physical function (≥15%) was used in Paper III (98). Pinch grip strength has been linked to the ICF component body function (95, 96) (Table 3).

3.4.1.3 Grip ability

The Grip Ability Test (GAT) (99) was used to measure grip ability in Paper III. The GAT includes three grips that patients perform at one time. The faster they complete the test, the better their score and grip ability. The three grips include putting a sock over the non-dominant hand, putting a paperclip on an envelope, and pouring water from a 1-litre jug into a cup. In the literature, the average mean for healthy controls was 16.5 s, and a value <20 was regarded as a normal grip ability (99, 100). The GAT is linked to the ICF component activities (100) (Table 3). There is no recommendation on what is considered a clinically meaningful improvement in GAT. Therefore, the definition suggested by IMACS for clinically meaningful improvement in muscle strength and physical function (≥15%) was used in Paper III (98).

3.4.1.4 Dexterity

The Purdue Pegboard was used in Paper III to measure dexterity (101). The Purdue Pegboard includes two parts; in the first part, patients put as many pegs as possible on a board in 30 seconds using only one hand at a time. In the second part, patients have 60 seconds to manipulate pegs, collars, and washers (*assembly*) onto the board. The more pegs, collars, and washers the patient places on the board, the better the dexterity (101). The test was done three times, and the average of the three trials was used in the analysis. For comparison, the literature contains normative values based on convenience sampling (101). A repeatability test on the Purdue Pegboard has been conducted on another muscle-affecting diseases (muscular dystrophy), suggesting a true difference of two or three pegs (102). Based on these results, improvement considered to have a clinically meaningful difference was set to ≥3 in Paper III. Dexterity measured by the Purdue Pegboard is linked to the ICF component activities (95) (Table 3).

3.4.1.5 Hand joint mobility

The Escola Paulista de Medicina-Range of Motion (EPM-ROM) scale was used to evaluate joint mobility in Paper I (103). The EPM-ROM scale measures active joint mobility with goniometry in seven joints. Only joints involving the hand were measured (wrist, thumb, and the MCP joints). The range of mobility varies from 0 to 3, where 0 is equivalent to full mobility and 3 indicates severe joint mobility limitations. The final score is calculated through the sum of the values on the right and left sides and is then divided by 2. This calculation gives a scale from 0 to 21, where 0 indicates good joint mobility and 21 indicates severe joint mobility limitation (103). Joint mobility is linked to the ICF component of body function (96) (Table 3).

3.4.2 Activity limitation

Activity limitation has been evaluated by either the DASH questionnaire (Paper III) (89, 90) or the Myositis Activities Profile (MAP) (Paper I) (104).

The DASH is a 30-item questionnaire designed to measure physical function and symptoms in people with any or several musculoskeletal disorders of the upper limb (105). Patients self-rate their ability on a five-grade scale ranging from no difficulty (1) to impossible to do (5). Scores were calculated with a range from 0–100, and a higher score indicated greater activity limitations. For comparisons, the literature contains normative values from the general population (90). In the DASH questionnaire, a minimal important change of at least 10 points was considered a clinically meaningful improvement in Paper III (105). The questions in DASH are linked to the ICF components of body function, activities, and participation and also to personal factors (106) (Table 3).

The MAP is a disease-specific questionnaire measuring difficulty in performing the activities of daily life during the past week (104). Difficulty is defined as how hard the activity is to perform and how important the activity is for the person to manage. Each activity is scored on a 7-point scale (where 1 indicates no problem and 7 indicates impossible to do). The activities are divided into four sub-scales: movement activities, activities of moving around, self-care activities, and domestic activities. The questions are ordered in rank, and the median value and quartiles for every sub-scale are used in

the results. Four additional single items are included in the MAP regarding social activities, avoiding over-exertion, work/school work, and leisure activities, which are presented in the results as the actual score (104). The MAP was developed based on the International Classification of Impairments, Disabilities, and Handicaps (ICIDH)-2 Beta-2 draft and has connections to the ICF component of activities and participation (104) (Table 3).

3.4.3 Work Ability

Self-rated work ability was assessed by the Work Ability Index (WAI) in Paper IV (107, 108). The WAI is a questionnaire that contains seven items (10 questions) that are rated on different scales. One example of the questions concerns the principal demands at work (psychologically demanding, physically demanding, or a combination of both) (107). A sum score can be calculated by summing up the seven items to a total WAI score in the interval of 7-49. A total WAI score of 7-27 indicates poor work ability (*restore* work ability). A score from 28-36 suggests moderate work ability (*improve* work ability), while a score in the range of 37-43 indicates good work ability (*support* work ability). Excellent work ability is represented by a score of 44-49 (*maintain* work ability) (107, 109). The WAI has not been linked to the ICF components in a standardized way. Based upon a subjective linking to the ICF components by the author (M Regardt), the WAI seems to have links to body functions and structures and in the ICF component of activities and participation, which includes work (21) (Table 3).

3.4.4 Work-related risk factors

To assess work-related risk factors in Paper IV, the Swedish Work Environment Authority's statute book (AFS) provision AFS 1998:1, *Ergonomics for the Prevention of Musculoskeletal Disorders,* was used (110). Eleven questions were selected based on hand function and the symptoms of proximal muscle weakness and reduced endurance that are common in PM and DM (7, 10, 111). Self-rating questions concerning work-related risk factors were addressed by asking *how often* prolonged or recurrent work is done with the arm and shoulder, with the neck and back bent, doing fatiguing leg work, with exhaustive manual lifts, and carrying or repetitive work movements. The questions were answered by the patients on a four-point scale with rating steps ranging from "never" to "very often". Based upon a subjective linking, by the author (M Regardt), the

questions regarding work-related risk factors seems to have links to the ICF components body functions and activities and participation (21) (Table 3).

3.4.5 Work Environment

The semi-structured interview instrument Work Environment Impact Scale (WEIS) was used in Paper IV to assess how the individuals experience and perceive their work environment (112). When using the WEIS, it is mandatory to have a job to relate to. Therefore, we chose to perform this measure on the patients who had been working at some point within the last two years (n=38). The WEIS contains 17 items, which reflect upon the client's own perception of opportunities and constraints in the work environment related to social groups, physical spaces, objects, and tasks (113). There is an interview guide to follow, and complementary questions are used when necessary. After the interview, the assessor rates the 17 items on a four-point rating scale. Ratings 1 and 2 indicate that the item "strongly interferes" or "interferes" with the individual's work performance, satisfaction, and physical/emotional/social wellbeing, while 3 and 4 imply that the item "supports" or "strongly supports" the individual's work performance, satisfaction, and physical/emotional/social wellbeing (112). The environmental factors that interfere and/or provide support are concluded in a summary based on the 17 items. The WEIS was developed from an occupational therapy model, the Model of Human Occupation (MoHO) (114). According to the model, the environment gives both opportunities for behaviour but also presses for certain demands of an individual (70). The WEIS does not assess the environment but instead evaluates its effect on a person's performance (114). The environment's impact on a person depends on the person's values, interests, personal caution, habits, roles, and performance (70). There has not been any standardized linking between the items in the WEIS and the components in the ICF. Based upon subjective linking by the author (M Regardt), the WEIS seem to have links to the ICF components of body functions, and activities and participation as well as in the environmental factors (21) (Table 3). However, there were questions that did not appear to be connected to the ICF, such as questions related to time and expectations of demands.

3.4.6 Health-related quality of life

In Papers I and II, the HRQoL was evaluated by the patient-reported questionnaire SF-36 (80, 88). The SF-36 contains questions relating to both physical and mental aspects of self-experienced HRQoL in eight dimensions. The eight dimensions are Physical Functioning (PF), Role Physical (RP), Bodily Pain (BP), General Health (GH), Vitality (VT), Social Functioning (SF), Role Emotional (RE), and Mental Health (MH) (80, 88). The dimensions are scored on a scale ranging from 0 to 100, with 100 indicating excellent HRQoL. Gender- and age-matched values for the Swedish general population in the literature were used for comparisons (80). The questions in SF-36 are linked to the ICF components body function and activities and participation (84, 115, 116) (Table 3).

3.4.7 Evaluation of the hand exercise programme

The hand exercise intervention (Paper III) was evaluated using adherence, exertion, and patients' opinion about the design of the hand exercise programme. Adherence was defined as the completed number of exercise sessions performed compared with the expected number (36). This information was collected from patients' exercise diaries. An acceptable adherence was ≥75% (≥ 27 sessions). The hand exercise programme was evaluated based on patients' exertion rating using the RPE Borg CR 10 scale after every session (92, 93). In addition, patients were asked their opinions about the programme, the frequency, and the overall feasibility of undertaking the hand exercises.

3.5 Data analysis

A summary of the different statistical analyses performed in this thesis is presented in Table 4.

Table 4. Summary of statistical analyses performed in Papers I-IV.

Methods	Paper I	Paper II	Paper III	Paper IV
Mean (SD)	X	X	X	X
Mean (CI)	X			
Mean difference (CI)		X		
Median (IQR)	X	X	X	X
Percentage	X	X	X	X
Number	X	X	X	X
Range				X
Independent sample t-test	X			X
Mann-Whitney U-test	X			X
One sample t-test 95% CI	X			
Pearson's correlation coefficient	X			
Spearman's correlation coefficient	X	X		X
Wilcoxon Signed-rank test		X	X	
Mixed linear model		X		
Bonferroni after test		X		
Effect size			X	

SD=standard deviation, CI= Confidence intervals, IQR=Interquartile Range.

3.5.1 Paper I

To describe grip force, hand mobility, activity limitation, HRQoL, and disease activity, mean, standard deviations (SD), median, Interquartile Range (IQR), confidence intervals (CI), independent-sample t-test, and Mann Whitney U-tests have been used. Mean values, one-sample t-test, and CIs were employed to compare patients with PM and DM to gender- and age-matched normative values regarding HRQoL using SF-36 (80) and grip force using Grippit© (47). The mean value for the normative values was determined by collecting a gender- and age-matched mean value for each patient and

then calculating a new mean value for the reference group. The differences between patients with PM and DM and also between women and men regarding grip force, hand mobility, activity limitation, and HRQoL were analysed by mean values, independent sample t-tests, and Mann Whitney U-tests. Pearson's correlation coefficient, respective Spearman's r mean values, and CIs were used to calculate correlations between hand mobility and grip force with regard to the variables' activity limitation, HRQoL, disease activity, and disease duration. All statistical calculations were made using the Statistical Package for the Social Sciences (SPSS; Chicago, IL, US) version 15. The level of significance was set to ≤0.05.

3.5.2 Paper II

The study used number, percentage, median, Interquartile Range (IQR), mean, and standard deviations (SD) to describe the characteristics of patients at disease onset. The Wilcoxon Signed-rank test was applied to assess cross-sectionally if there were differences in grip force (47) and HRQoL (80) among patients diagnosed with PM and DM at different times (time of diagnosis (Year 0) to 6 years after diagnosis) as compared to normative values for healthy individuals or the Swedish population in general in the literature (null hypothesis = no difference between PM and DM distribution and normative values). For each patient, a gender- and age-matched value was collected from the literature based on healthy individuals (for Grippit©) or from the general population (for SF-36) (47, 80). These values represent the sample of normative values used in this study. Observed values for patients with PM and DM were standardized using gender- and age-specific normative values (mean and standard deviation) in the literature (47, 80). The significance level was set to ≤0.05, and in the analysis on grip force, it was adjusted by the Bonferroni after test. To describe how many of a patient's grip force values were lower than normative values, the study compared them to standard deviation (47) and error of measurement (94) adjusted values and presented as percentages. The mixed linear model was used to investigate how grip force changed over time with year 0 (time of diagnosis) and years 1, 2, 3, 4, 5, and 6 after diagnosis as repeated measurements. The mixed linear model enabled comparisons longitudinally when subjects had missing values at any given time-point (117). The study analysed potentially influential factors (disease duration, gender) for main effects and possible interaction effects. Comparisons among factors over time were adjusted by the Bonferroni after test.

Mean difference and CI were used to describe differences. The significance level was set at ≤ 0.05.

The Spearman correlation coefficient (r_s) was used to assess correlations between grip force and HRQoL at different times (year 0 and years 1, 2, 3, 4, 5, and 6 years after diagnosis). The correlation coefficient was set to $r_s \geq 0.6$, and a Bonferroni-adjusted significance level of ≤ 0.0036 was considered significant. According to Munro (1997), correlation coefficients between 0 and 0.25 are considered to have no or very low correlation, coefficients between 0.26–0.40 have a low correlation, coefficients between 0.41–0.69 have a moderate correlation, coefficients between 0.70–0.89 have a high correlation, and coefficients between 0.90–1.0 have a very high correlation (118).

All analyses on grip force (Grippit©) (47) were performed on both the right and left hands. All statistical calculations were made using the IBM Statistical Package for the Social Sciences (SPSS) version 19 (Armonk, NY, USA).

3.5.3 Paper III

Patient characteristics, adherence, disease activity, medical treatment, measures of hand function (hand grip strength, pinch grip strength, grip ability, and dexterity) and activity limitation are presented as numbers, percentages, means, standard deviations (SD), median (md) and interquartile ranges (IQR). The Wilcoxon Signed-rank test was used to analyse differences at baseline between patients with PM and DM and normative values from the general population or healthy individuals from the literature regarding hand grip strength (Jamar) (48), grip ability (GAT) (99), dexterity (Purdue Pegboard) (101), and activity limitation (DASH) (90) (null hypothesis = no difference between PM and DM distribution and normative values). The observed values for patients with PM and DM were standardized using gender- and age-specific normative values from the general population or healthy individuals (mean and standard deviation) from the literature (48, 90, 99, 101).

The Wilcoxon Signed-rank test was used to analyse differences between baseline and follow-up in hand function (hand grip strength, pinch grip strength, grip ability, and dexterity), activity limitation, disease activity, and medical treatment. Effect size was used to evaluate the responsiveness defining values from 0.2–0.5 as a low level of

responsiveness, values from 0.51–0.8 as moderate, and values > 0.81 as a high level of responsiveness (97).

Individual differences between baseline and follow-up have been described and meaningful change evaluated based on respective measures and, if not applicable, the IMACS-suggested definition of clinically meaningful improvement in muscle strength and physical function (≥15%) has been used (98). The significance level for all analyses was ≤0.05. All statistical calculations were made using the IBM Statistical Package for the Social Sciences (SPSS) version 19 (Armonk, NY, USA).

3.5.4 Paper IV

To describe demographics, work situation, self-rated WAI, work-related risk factors, and the WEIS, this study used mean, standard deviation (SD), median, IQR, range, number, and percentage.

The Mann Whitney U-test or independent sample t-test was employed for group comparisons regarding demographics, work situation, self-rated WAI, and work-related risk factors. The Spearman's rho was used for calculations of correlation between self-rated work ability (total WAI score) and percentage of full-time employment, work-related risk factors, and also the work environment (WEIS). Spearman correlation coefficients between 0 and 0.25 were considered as no or very low correlation, 0.26–0.40 as low correlation, 0.41–0.69 as moderate correlation, 0.70–0.89 as high correlation, and 0.90–1.0 as very high correlation (118). For all statistical calculations, the Statistical Package for the Social Sciences (SPSS; Chicago, IL) version 19 was used. A statistical significance level of ≤ 0.05 was defined.

3.6 Ethical considerations

All papers were approved by the regional ethical review board. In Paper I, patients gave oral consent to participate, which was the requirement of the ethical committees in Sweden at the time of the study. In Papers II-IV the patients gave written consent to participate, in accordance with the Declaration of Helsinki (119). The patients were reassured that if they chose not to participate, their care at the Karolinska University Hospital would not be affected.

4 Results and discussion

4.1 Hand function in patients with PM and DM (Papers I, II, and III)

In this thesis, we aimed to investigate and explore different aspects of hand function in patients with PM and DM.

Results from Paper I demonstrate that both women and men with PM and DM have reduced grip force compared to gender- and age-matched healthy individuals from a Swedish cohort ($p<0.001$) (120). The measure of hand grip strength or grip force is linked to the ICF component of body function. Patients with PM and DM are typically described as having proximal muscle weakness in the upper and the lower extremities, whereas measures of strength in the distal muscles are rarely included in recommended outcome measures, such as the manual muscle test in eight muscle groups (MMT-8) proposed by IMACS (7, 34). In this study (Paper I) we could, for the first time, demonstrate that hand grip strength and grip force may be negatively affected in up to 77% of patients with PM and DM (120). This was a cross-sectional study, and we used the conventional measure of grip force (Grippit©) (47, 120). This study raised the question of whether low grip force was already present at the time of diagnosis or if the impaired grip force develops during the disease course, as well as if it is reversible with conventional treatment. This was the background for Paper II, in which we did indeed confirm low grip force at the first visit at the time of diagnosis in about 92% of women and 93% of men with PM and DM when compared to normative values from gender- and age-matched healthy individuals from a Swedish cohort (Paper II) (47). This is novel information that can be added to the previously suggested effect on fine motor skills and distal muscles in the late phase of PM and DM disease (39, 121). In over time analysis in Paper II, the total group of patients improved in grip force from time of diagnosis to one or two years after (Paper II).

In our studies (Paper I-III), we could also confirm reduced strength in the hand muscles by using two different measures: hand grip force (Grippit©) and hand grip strength (Jamar) (47, 48). Both measures gave comparable results, thus confirming reduced strength in the distal muscles of the upper extremities (Paper II) (120, 122).

The presence of weakness in distal muscles has previously been suggested in both the wrist flexion/extension and the ankle dorsi/plantar flexion by using the MMT, which suggested the proximal weakness was more pronounced than the distal weakness in patients with PM and DM (123). In that study, most of the patients (82-94%) had no or only mild limitation in wrist flexion and extension (123). In this thesis, the measures of strength in the distal muscles were limited to the hands, as assessed by measuring the grip force (Grippit©) or the hand grip strength (Jamar) (47, 48) (Paper II) (120, 122). The hand grip strength was measured by asking the patients to squeeze (flexing fingers) as hard as they could on a cylinder-shaped measure either three times (Jamar) (peak strength) (48) or by using the grip force by holding the grip for 10 seconds (Grippit©) (endurance to maintain power) (47). Even though some muscles are involved in both finger flexion (Grippit© and Jamar) and wrist flexion (MMT), these measures are not comparable since they measure different entities. Furthermore, MMT may be a less sensitive measure to identify mild distal impairment (98). Muscle weakness in the distal muscles of the lower extremity (heel and toe lift) have also been shown in patients with PM and DM (94). Since there were no significant differences in grip force between patients with PM and DM, these diagnoses were analysed together (120).

When we assessed grip force at different time-points after diagnosis we found that the women with PM and DM had reduced grip force not only at diagnosis but also six years after diagnosis compared to the normative values for healthy individuals in the literature (Paper II) (47) (Table 5). In contrast to the women, the men had low grip force in both hands at diagnosis and up to two years after diagnosis (Paper II). In men, there were no additional significant differences at other time-points after diagnosis (Table 5) (Paper II). These observations suggest that grip force might have different patterns over time in women and men. Somewhat contradictory, the results of the over time analysis on grip force (Paper II) did not show any significant interaction between gender and disease duration in patients with PM and DM in relation to grip force, suggesting that women and men with PM and DM did not change differently over time (Figure 7) (Paper II). However, the number of observations after two years was low, particularly in the male group, making data after two years less reliable.

Table 5. Grip force in women and men with PM and DM in the registry compared to gender- and age-matched normative values of healthy individuals from the literature (47). Grip force is presented for both the right and left hand, respectively, and years since the diagnosis.

Disease duration (years)	Grip force (N) in women with PM/DM, n=47		Grip force (N) in women normative values, n=105[a]		Percentage of normative values (%)
Right hand	Md	IQR	Md	p-value	Md
0, n=36	109	72-154	206	**<0.001**	54
1, n=20	130	103-184	206	**0.001**	60
2, n=20	112	79-164	206	**0.001**	57
3, n=16	134	81-144	206	**0.001**	60
4, n=11	161	75-201	197	**0.013**	69
5, n=5	148	51-225	197	0.138	69
6, n=7	111	75-168	197	**0.018**	56
Left hand					
0, n=36	85	59-134	194	**<0.001**	49
1, n=21	137	86-202	194	**0.010**	61
2, n=20	100	70-148	194	**<0.001**	51
3, n=16	107	77-128	194	**0.001**	56
4, n=11	113	48-173	173	**0.008**	61
5, n=5	114	32-176	173	0.080	53
6, n=7	93	83-125	173	**0.018**	51
Disease duration (years)	**Grip force (N) in men with PM/DM, n=32**		**Grip force (N) in men normative values, n=64[a]**		**Percentage of normative values (%)**
Right hand	Md	IQR	Md	p-value	Md
0, n=26	211	138-276	386	**<0.001**	52
1, n=10	311	253-398	418	**0.017**	78
2, n=11	329	196-366	386	**0.016**	85
3, n=7	261	250-368	386	0.028	65
4, n=4	257	192-347	402	0.068	63
5, n=4	328	192-416	402	0.068	79
6, n=3	319	n/a	418	0.109	83
Left hand					
0, n=27	203	133-251	334	**<0.001**	57
1, n=10	324	231-368	386	0.037	88
2, n=11	290	169-333	334	**0.013**	83
3, n=7	220	187-313	334	0.043	56
4, n=4	238	121-354	360	0.144	64
5, n=4	289	182-424	360	0.144	81
6, n=3	301	n/a	386	0.285	80

PM=polymyositis, DM=dermatomyositis, N=Newton, n=number, Md=median, IQR=Interquartile Range, [a]=Nordenskiold and Grimby 1993 (13), p-value=probability value, n/a=not applicable

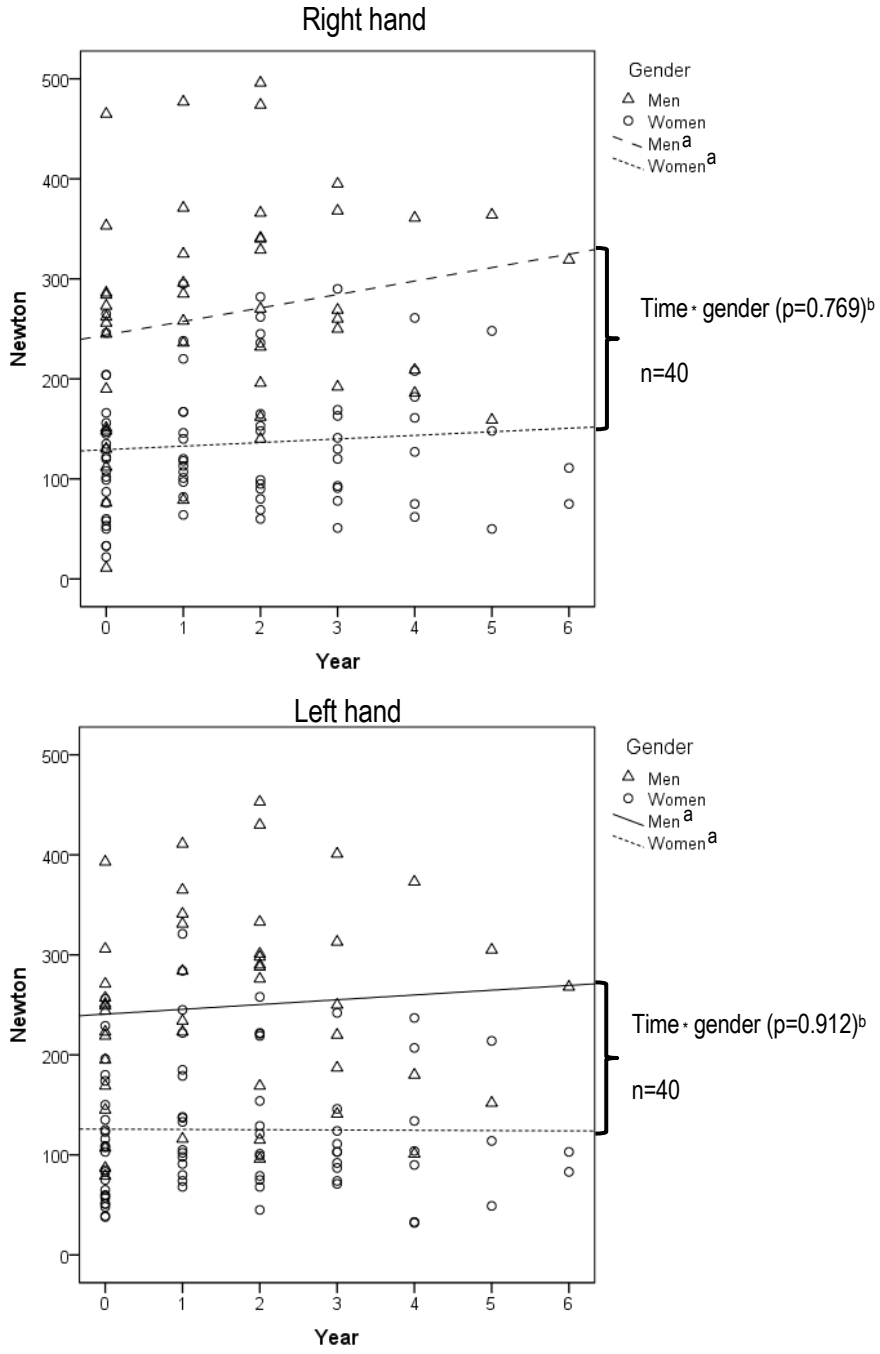

Figure 7. Grip force in the right and left hand in women (o) and men (Δ) with PM and DM over time. [a]=regression line, [b]=There was no interaction between gender and time, indicating that the grip force did not change differently over time in women and men with PM and DM.

There could be several explanations for the reduced grip force in patients with PM and DM. One possibility is that a high disease activity affects general health in the early active disease phase, causing an impact on grip force (87). Another possibility is that muscle weakness in the proximal muscles of the arm could contribute to the lower grip force, suggesting that the proximal weakness may lead to a changed activity repertoire in the daily life of patients with myositis. Such a change in activity performance may result in reduced use of the whole arm, which can have a negative influence on grip force as a consequence. This hypothesis could not be tested in our studies since we did not include assessments of strength in the proximal arm muscles. Yet another possibility that cannot be excluded is that the disease may impact the distal muscles that affect grip force, but this hypothesis requires further explorations.

In this thesis, hand functions were investigated using hand grip strength or hand grip force on a body function level as well as measures of pinch grip strength, grip ability, dexterity, and hand joint mobility with links to the ICF components body function, and activity and participation. Our results indicated only a small limitation in joint mobility in patients with PM and DM by the score of 3 out of 21 (0=good joint mobility) (120). In the hand exercise intervention study (Paper III), the patients with PM and DM had reduced dexterity compared to normative values from the literature at baseline (101, 122). This pilot study included a small convenience sample of patients with reduced hand grip strength, which may have caused a selection bias that could have influenced the result (122). However, in a previous study with the aim to evaluate the associations between grip strength and dexterity in healthy individuals, only a low correlation was found between these two measures (124). In that study, only 16% of the variety in grip strength was related to the variation in dexterity (124), indicating that patients with PM and DM may have impaired dexterity (122). However, this finding needs to be explored further in a larger cohort of patients.

One recent report indicates that arthritis in the wrist and finger joints is more common in patients with PM and DM than earlier described (40); arthritis in the hands is known to impair and limit hand function (41, 42). Therefore, since the aim was to describe and evaluate impaired hand function due to muscle weakness, patients with arthritis or other

conditions that could affect hand function were excluded from our studies to avoid a potential influence of arthritis on hand function in Papers I-III (120, 122).

4.2 Activity limitations in patients with PM and DM (Papers I, III, and IV)

Next we investigated what activities patients with PM and DM perceived as difficult and if they correlated with grip force.

Patients with PM and DM rated their activity performance from slightly difficult to very difficult in Paper I (120). Activities of moving around, work, and leisure were reported as the most difficult to perform (120). These results were confirmed in the hand exercise intervention (Paper III) where the participants had significantly more activity limitation (regarding the upper extremity), as measured by DASH, compared with normative values based on the general population from the literature (90, 122).

The measures of activity limitation in this thesis have links to the ICF components body function, and activities and participation, as well as to personal factors. In patients with PM and DM, the reduced grip force was associated to domestic activities measured by MAP (Paper I) (120). Thus, a reduced grip force may increase activity limitation in domestic activities in patients with PM and DM (120).The results on activity limitation in the hand exercise study (Paper III) are based on patients with reduced hand grip strength, which is known to affect activity performance in other rheumatic diseases (45, 125). In general, patients with PM and DM are affected in their activities due to the muscle weakness and reduced muscle endurance in the proximal muscles, leading to difficulties rising from a chair, walking up stairs, or combing their hair (33).

Our results add knowledge that the activity performance may be affected by muscle weakness in the distal muscles as well (122). Activity limitation has been studied to evaluate pharmacological treatment in patients with PM and DM (126). That study showed that patients with PM and DM had limitations in their daily activities with a value of approximately 20 points on a scale from 0-45, where 45 points indicates no difficulty (126). In Paper I, work was one of the activities rated as most difficult by the patients with PM and DM and is in general an important aspect of activity both for the individual and for

society (67, 120). Therefore, we aimed to investigate the work ability in patients with PM and DM in Paper IV.

4.2.1 Work ability in patients with PM and DM

In paper IV we investigated work ability in 48 patients with PM and DM. Most of the patients had been working during the past two years and 36 out of 38 were working at the time of the study. The 10 patients who had been on sick leave ≥2 years had a range of sick leave between 34−162 months. In the whole group, 44% were working full-time (40 hours/week), 31% worked part time, and 25% were on full-time sick leave. Even though a large proportion of the patients with PM and DM were working, 62% reported poor or less good self-rated work ability (Table 6) (Paper IV).

Table 6. Self-rated work ability measured by WAI in patients with PM and DM.

Work Ability Index (WAI)	Patients, n=48	Patients (working or on sick leave <2 years) n=38	Patients (on sick leave ≥2 years) n=10	Mann-Whitney U-test	
	n (%)	n (%)	n (%)	z	p-value
Total WAI score				-2.5	0.01
Poor (Restore)	16 (33)	6 (16)	10 (100)		
Less good (Improve)	14 (29)	14 (37)	0 (0)		
Good (Support)	11 (23)	11 (29)	0 (0)		
Excellent (Maintain)	7 (15)	7 (18)	0 (0)		
Sickness absences during the last 12 months (question 6)					
None	10 (21)	10 (26)	0 (0)		
1-7 days	5 (10)	5 (13)	0 (0)		
8-24 days	13 (27)	13 (34)	0 (0)		
25-99 days	5 (10)	5 (13)	0 (0)		
100-365 days	15 (31)	5 (13)	10 (100)		

n=number, %=percent, p-value=probability-value

No previous studies have addressed the self-rated work ability in patients with PM and DM. In other rheumatic diseases, work ability is often impaired (127-129). The measures used in this thesis to assess different aspects of work have links to the ICF components of body function, and activities and participation as well as environmental factors. A poor self-rated work ability predicts disability pension (71) and might therefore be important to assess and possibly include in a rehabilitation programme for patients with PM and DM.

Patients with PM and DM who had been on sick leave ≥2 years had more work-related risk factors than those who were still working. Work-related risk factors, such as heavy and prolonged or non-varied loads, can generate work-related musculoskeletal disorders (130) caused by a repetitive overload of the body (110). The areas of questioning were selected based on both the use of hand strength as well as the symptoms of proximal muscle weakness and reduced endurance that are common in PM and DM (7, 10, 111). We found that patients on sick leave ≥2 years had more work tasks that included prolonged or recurrent work with *arm/hand stretched forward*, *the use of hand-held machines, working with pedals,* and *walking long distances* (Table 7) (Paper IV). These results indicated that the patients were affected in their ability to work by the manifestations of the disease and suggested that patients with these types of work-related tasks may benefit from an analysis of their work situation and specific support to enhance their ability to continue working (Paper IV).

Table 7. Work-related risk factors (based on Authority's statute book (AFS) provision 1998:1[a]) in patients with PM and DM.

Work-related risk factors	Patients n=48	Patients working or on sick leave <2 years, n=38	Patients on sick leave ≥2 years, n=10	
	Quite to very often %	Quite to very often %	Quite to very often %	p-value[b]
Repeating similar working movements	79	79	80	0.94
Prolonged or recurrent work:				
Arm/hand				
Above shoulder height	38	21	66.7	0.22
Stretched forward	56	47	90	**0.02**
Forceful movements and uncomfortable hand position/grips	40	34	60	0.18
Hand-held machines or tools	23	18	40	**0.05**
Legs				
Repeated raising to ladder and squatting	38	34	50	0.37
Work with pedals, walking stairs and long distances	38	26	80	**0.002**
Neck and back				
Neck bent forward, backwards, sideways, or/and twisted repeatedly or for long periods of time	54	50	70	0.26
Back bent forward, backwards, sideways, or/and twisted repeatedly or for long periods of time	40	35	60	0.14
Lifting and carrying				
Repeated exhaustive manual lifts	44	39	60	0.25
Uncomfortable carrying, pushing, or pulling of loads	27	24	80	0.31

PM=Polymyositis, DM=Dermatomyositis, n=number, %=percentage, p=probability, [a]= Ergonomics for the Prevention of Musculoskeletal Disorders, (1998) (http://www.av.se/dokument/inenglish/legislations/Models_for_assessment.pdf), [b]= Mann-Whitney U-test was used to test for statistically significant differences between patients working or on sick leave <2 years and patients on sick leave ≥2 years.

We also investigated how the work environment may impact the work ability in patients with PM and DM. Using the semi-structured interview instrument WEIS, patients with PM and DM reported that the work environment both "supported" them and "interfered" (Paper IV) (Figure 8). The items in the WEIS instrument perceived as "interfering" with

work performance, satisfaction, and physical/emotional/social wellbeing were *Task Demands, Rewards,* and *Time Demands* (Paper IV). This result is in agreement with an earlier Swedish study that included patients with a broad range of musculoskeletal system and connective tissue disorders as well as mental and behavioural disorders that had taken sick leave (131). Further, the WEIS items *Interaction with Others* and *Meaning of Work* were seen as "support" in both the previous and the present study (Paper IV) (131). The highly rated "support" in the work environment regarding *interaction with co-workers and others* might explain why so many of the patients working or on sick-leave for less than two years were working full-time and only had used a few days of sick leave (Table 6) (Paper IV). These results emphasise the importance of the "social" working environment for patients with PM and DM. Support at the workplace has been described as important in improving work participation in patients with musculoskeletal conditions (132), and this might be true for patients with PM and DM as well.

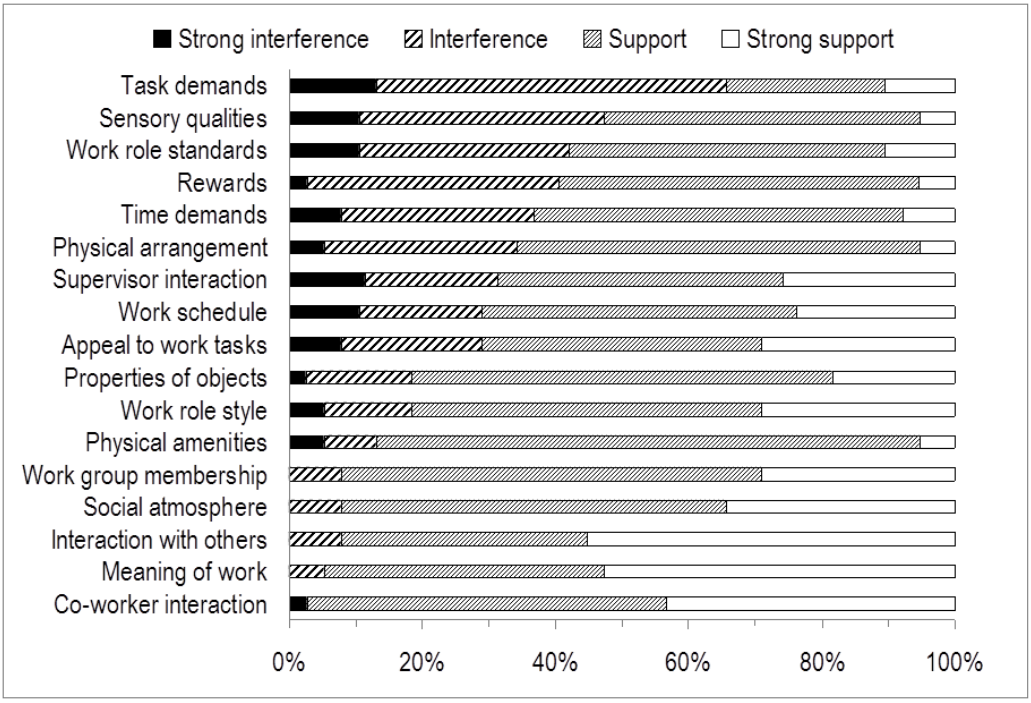

Figure 8. The distribution of the Work Environment Impact Scale (WEIS) in patients with PM or DM (n=38).
PM=Polymyositis, DM=Dermatomyositis, n=number, %=percentage.

A lower self-rated work ability, according to the WAI, correlated well to a lower percentage of full-time employment (r_s 0.8, p<0.01 (Figure 9) (Paper IV). Accordingly, a large proportion of the patients (n=27) had a need to restore or improve their work ability (Paper IV) (Figure 9).

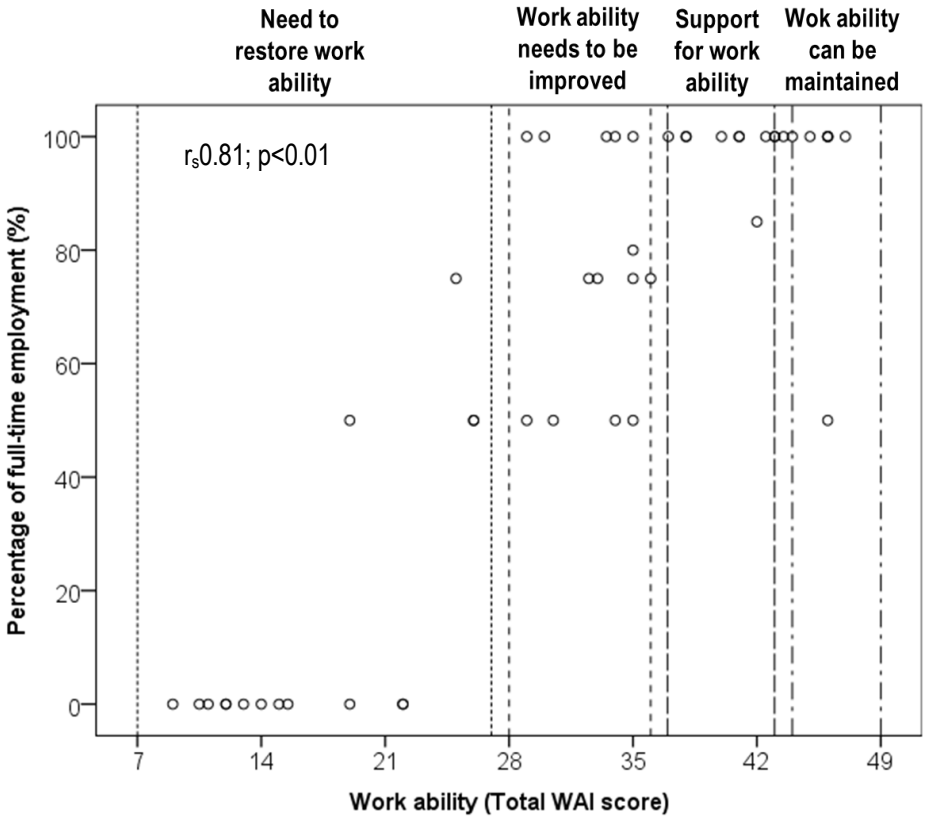

Figure 9. Percentage of full-time employment in relation to self-rated work ability (total WAI score) n=48.
n=number, r_s= Spearman correlation coefficient, p=probability value.

The self-rated work ability additionally correlated to the following work-related risk factors: more frequent occurrence of *work with arm/hand stretched forward* (r_s 0.79, p<0.001), *forceful movements* and *uncomfortable hand positions/grips* (r_s 0.43, p=0.003) and *back bent forward, backwards, sideways, or/and twisted* repeatedly or for long periods of time (r_s 0.48, p=0.001) (Paper IV).

Moderate to high positive correlations were seen between self-rated work ability and five items of the WEIS (Paper IV). For example, a lower self-rated work ability (total WAI score) was associated with having tasks that were too demanding to handle (*Task Demand;* r_s 0.7, p<0.01), not having enough time to manage the workload (*Time Demand;* r_s 0.5, p<0.01), not having enough challenges in their work (*Appeal to Work Task;* r_s 0.5, p<0.05), excessive expectations on work performance (*Work Role Standard;* r_s 0.5, p<0.01), and not having the opportunity to influence working hours (*Work Schedule;* r_s 0.4, p<0.05) (Paper IV). These results emphasise the importance of the physical and psycho-social work environment for the self-rated work ability (Paper IV). In the future, a study with a qualitative approach would enhance the understanding of the factors that led to permanent sick leave.

4.3 Health-related quality of life in patients with PM and DM (Papers I and II)

Next we wanted to investigate how the disease affects HRQoL and how HRQoL correlates to the reduced grip force.

In Paper I, the patients with PM and DM rated the HRQoL lower in all dimensions of SF-36 compared to the general population in Sweden from the literature (Paper I) (120). Health-related quality of life measure used in this thesis was linked to the ICF components of body function, and activities and participation.

In the general population, there is a gender difference where the men rate their HRQoL better than the women (82). However, this difference was not detected in our study (Paper I) (120). In Paper II, we wanted to further investigate whether the lower ratings of HRQoL were present at different time-points. In that study (Paper II), we found that the HRQoL was rated lower in both women and men with PM and DM compared to the general population at the time of diagnosis (Paper II).

The women with PM and DM also had lower values at disease duration 1, 2, 3, and 6 years, while the men reported lower HRQoL only after 2 years disease duration (Paper II). The dimensions of General Health and Vitality were more commonly rated lower in women with PM and DM compared to the general population (Paper II), suggesting that women with PM and DM perceive the disease differently from men or are more affected by the disease than the men with PM and DM. This could, however, not have been answered by the study design used in this study (Paper II). It would also have been interesting to explore how the HRQoL changed over time in patients with PM and DM and if it changed differently between women and men. However, the HRQoL data in Paper II did not permit an analysis over time. Other studies have described the negative effect of myositis disease on a patient's HRQoL (4, 75). Also, it is well known that the disease negatively affects the HRQoL when other rheumatic conditions are present (133).

Quality of life is of importance for patients' health and is therefore central to address in clinical practice. Since HRQoL in patients with PM and DM is affected at the time of diagnosis, it is important to confront this issue at the beginning of the care. In rheumatic diseases, there are factors that promote HRQoL, such as having a strong sense of coherence, feeling rested after sleep, and having work capacity (134). It has yet to be explored whether these factors would be promoting for patients with PM and DM. Promoting factors could be the base of an intervention to increase HRQoL in patients with PM and DM.

The HRQoL dimension Vitality and Mental Health (SF-36) in women with PM and DM was associated with the grip force (Paper I) (120). In Paper II, we found that impaired grip force in patients with PM and DM had associations as well to the dimensions Role Physical, Mental Health, and General Health (SF-36) up to 4 years. Although these correlations were present only at one time point each in Paper II, this observation is in agreement with the results in Paper I (120). Interestingly, grip force has been found to predict poorer general health (87) and mortality, especially in those with reported disease compared to those who were healthy in a general population of elderly participants (86, 87). One possible explanation could be that with poorer health, the activities performed during a day may be less than usual due to the health condition; the reduced use of the hands could be a consequence, leading to a lower grip force.

Despite conventional treatment, patients with PM and DM have reduced grip force, and it seems to affect both activity and HRQoL, which might indicate a need for targeted therapies, such as hand exercises.

4.4 Hand exercise as an intervention (Paper III)

As a result of the observed low grip force despite pharmacological treatment (Papers I and II), we found it of clinical importance to develop an intervention with a focus to improve hand function and additionally have an effect (improve or maintain) on activity performance (120, 122). Therefore, we developed a hand exercise programme to test its feasibility and the preliminary results of its effect on hand function and activity. The results in Paper III showed that the hand exercise programme was feasible but had a limited effect on hand function and activity performance (122). The measures used to evaluate hand function and activity limitation in Paper III have links to the ICF components body function, and activities and participation

We could demonstrate that patients with PM and DM had good adherence (> 75%) in completing a hand exercise programme (122). Feasibility, adherence, and relevant outcome measures are of importance when designing a new therapeutic intervention. Aspects that are known to positively influence adherence are supervision, the use of an exercise diary, the number of movements prescribed, and if the patient perceives the intervention as beneficial (135-138).

The hand exercise intervention in Paper III was primarily home-based with an average of four follow-ups conducted either by phone or at the hospital (122). As recommended, all participating patients filled in the exercise diary. A low number (≤3) of movements predict good adherence in patients with neck and low-back pain (138). The number of movements in our study was between six and eight, but this did not seem to influence adherence (122) (Table 8).

At baseline, all patients had reduced hand grip strength by approximately 32% in the right hand and 30% in the left hand, but whether this reduced handgrip strength was perceived as reduced or if it affected the patients' ability to perform daily activities was not addressed in the current study (Paper III) (122).
However, in other rheumatic diseases, reduced hand grip strength has been shown to be associated with activity limitations (125, 139, 140).

Table 8. Measures of adherence as a total and based on exercise location and measures of exertion at baseline and at different time-points in the hand exercise intervention.

Patient ID	Adherence (0-36)			PRE Borg CR 10 scale (0-10)			
	Total exercise sessions n	Exercise sessions at the hospital n	Exercise sessions at home n	Baseline	Week 1-4 median (IQR)	Week 5-8 median (IQR)	Week 9-12 median (IQR)
1	31	0	31	5	5 (4.8-5.3)	5 (4-5)	5 (4.3-5.8)
2	36	0	36	5	3 (3-3)	3 (3-3)	3 (3-3)
3[a]	33	1	32	8	6 (6-7)	6 (6-6)	6 (6-6)
4	34	1	33	7	5 (4-5)	5 (4-7)	4 (4-5)
5	34	5	29	7	3 (3-3)	3 (2.8-4)	3 (2-3)
6	30	7	23	5	4 (3-4)	1 (0-1.3)	0 (0-0.5)
7	28	1	27	3	No reported measures		
8	36	5	31	4	3 (3-4)	4 (3.3-4)	3 (3-4)
9	36	3	33	3	2.5 (2-3)	4 (3.3-4)	4 (4-4)
10[a]	35	1	34	7	3 (2-5.5)	2 (2-3)	3 (2-3.8)
11	31	1	30	3	3 (2.3-4)	4 (3-4)	4 (3-4)

n=number, RPE= Rating of Perceived Exertion, CR= Category Ratio, IQR= Interquartile Range,
[a] = left hand dominance

The patients rated their exertion on a moderate level (3 or 4) on the PRE Borg CR 10 scale throughout the 12-week intervention (122) (Table 8). There are recommendations to have this level of exertion as an initial load and then increase the load to strong exertion (5 or 6) (41). These results indicate that there would be a possibility to increase the

resistance in the hand exercise programme. The patients thought that 30 repetitions were too many and took too long to perform (122). With a more appropriate hand training dough that has a higher initial resistance, the number of repetitions could be reduced since there are new exercise dough types that have recommendations based on hand strength.

We demonstrated that patients with PM and DM can improve in hand function and activity performance using a hand exercise programme (122). However, improvement was limited using the current exercise regimen, indicating a need to modify the programme. The hand exercise programme consisted of different movements to strengthen the muscles of the hand. The movements were done with up to 30 repetitions, which indicate that the programme might improve muscular endurance. Another reason for the limited result might be that the assessment used to evaluate muscle endurance (Jamar and pinch grip strength) measures peak strength. This may indicate that the programme trained one aspect (endurance), and the measures assessed another (peak strength). However, in other rheumatic and muscle-affecting diseases, hand exercise is commonly used as a treatment in occupational therapy; studies on hand exercise have shown improvements in hand function (51, 52, 54, 141).

Patients with PM and DM did not have reduced grip ability as measured by GAT (Paper III) (122). The reason for this finding could be that patients with PM and DM have good grip ability or that this measure did not include grips that may be impaired in patients with PM and DM since the GAT was developed for patients with arthritis (99). In both rheumatoid arthritis and systemic sclerosis, reduced joint mobility and impaired grip ability are present (139, 142-144). The results from Paper I showed only small limitations in joint mobility (120).

Therefore, a hand exercise programme focusing on increasing grip strength, dexterity, and activity performance with increased resistance in the movements and limiting the duration of each exercise session would be preferable in a future study.

4.5 Methodological discussion

The results from this thesis will add to the knowledgebase on hand function, activity, and participation in patients with PM and DM. To describe the methods and results in the

thesis, the ICF level-one classification was used. The use of a level-one classification gives a broad description about what ICF components and categories in the ICF might be impaired in patients with PM and DM. Further studies would be of interest to enable disease-specific core sets for patients with myositis.

A general limitation of the studies in Papers I-III is that the measures of grip force, grip strength, and HRQoL were compared to reference values from the literature. There is a limitation of using literature-derived control groups. To handle this, a gender- and age-matched value was collected from the literature for every patient.

In Paper I, the mean value for this control group was used in the one-sample t-test analysis. In Papers II and III, to address the deficiency of a control group, the analyses were more appropriately done by standardizing the observed values (mean values and standard deviations) and testing the null hypothesis (=there are no differences) by using the Wilcoxon Signed-rank test. Even though there was the limitation of using normative values from the literature, the comparable values along with patient-reported assessments provided a direction of whether the patients had disabilities or not.

The E-link pinch meter has a thinner profile than the regular pinch gauge meter and could therefore not be compared to normative values. In the future, a study to derive normative values for the E-link pinch meter would give valuable information about the pinch grip strength.

The measures of activity and participation (MAP, DASH) used in this thesis enabled a description of what limitations and restrictions are present in patients with PM and DM (89, 90, 104). However, these measures did not evaluate if the activity is meaningful (occupation), whether the participants actually perform it, or if it negatively affects their lives (61, 63, 64). Only a few articles have assessed activity limitation in patients with PM and DM (2, 33, 39, 145). Therefore, the results from this thesis will add to the knowledgebase of activity and participation in patients with PM and DM.

One previous study showed that the MAP covers some of the activities that are important to improve for patients with PM and DM (146). However, some areas are still missing in the MAP, such as sexual activity, sleep, and bicycling (146). In the future, it would be of interest to investigate which activities limit patients and which are of importance.

A patient-derived activity limitation measure would be preferred both in research and in clinical practice.

In this thesis, the SF-36 was chosen to evaluate HRQoL. As with the MAP and DASH measures, there might be areas of HRQoL that are important to the patient but are not covered by the SF-36. Therefore, it is necessary to perform qualitative studies to explore areas where the patients need support so that the health care providers can help the patients in the most appropriate way.

The WAI was able to detect reduced work ability in patients with PM and DM. The WAI has been used to predict the use of health care and rehabilitation and has also been shown to have associations to HRQoL (147), suggesting the use of WAI as a screening tool to evaluate self-rated work ability in patients with PM and DM in clinical practice.

In Paper II, the SweMyoNet Registry was used to collect data on grip force and HRQoL over time from the time of diagnosis. Missing data in registries based on data collected through clinical practise are a known general limitation of registries. Other explanations for the missing values may include the medical condition of the patient or potentially missed values from patients with the most severe or mild disability. To handle missing values and small sample sizes in the analysis, the Bonferroni after test was used to modify significance levels. The mixed linear model was used to enable an analysis over time despite the missing values (117). Still, we cannot exclude that the missing values may have affected the results in either a or negative manner. Furthermore, the lower number of observations after two years makes data from the later time points less reliable, particularly when subgrouping in women and men.

The aim of Paper III was to develop a hand exercise programme and to primarily test its feasibility in patients with PM and DM. Even though information about its effect on hand function and activity performance would have been beneficial for clinical practice guidelines, these results were not to be expected with this small convenience sample. However the study did give directions on how to improve the programme design and what measures to include in a future study. A limitation in Paper III was that at the time when the hand exercise study was initiated there were no guidelines on which resistance putty to use based on hand grip strength which nowadays are available. This resulted in a more

subjective estimation of the resistance in the dough and for the participants a too time-consuming programme.

Paper IV included patients both working and on sick leave for more than two years, enabling a possible comprehensive picture about work ability in patients with PM and DM. However, there might be a recall bias since the patients that had been on sick leave for more than 2 years had a range up to 162 months (≈14 years) of sick leave.

4.6 Summary

This thesis has used outcome measures that in some cases have links to the ICF components of body function and structures, activities and participation, and environmental and personal factors. The DASH and SF-36 are represented in several of the ICF components (84, 106). Regarding disabilities in body functions, the major results from these studies were reduced grip force and hand grip strength in patients with PM and DM (Figure 10). The impairment was present at different time points and had associations to both activities and HRQoL (Papers I, II, and III) (120, 122).
In the components of activities and participation, patients with PM and DM perceived the activities of moving around, work, and leisure as most limiting. Health-related quality of life was rated low and was linked to both body functions and activities and participation.

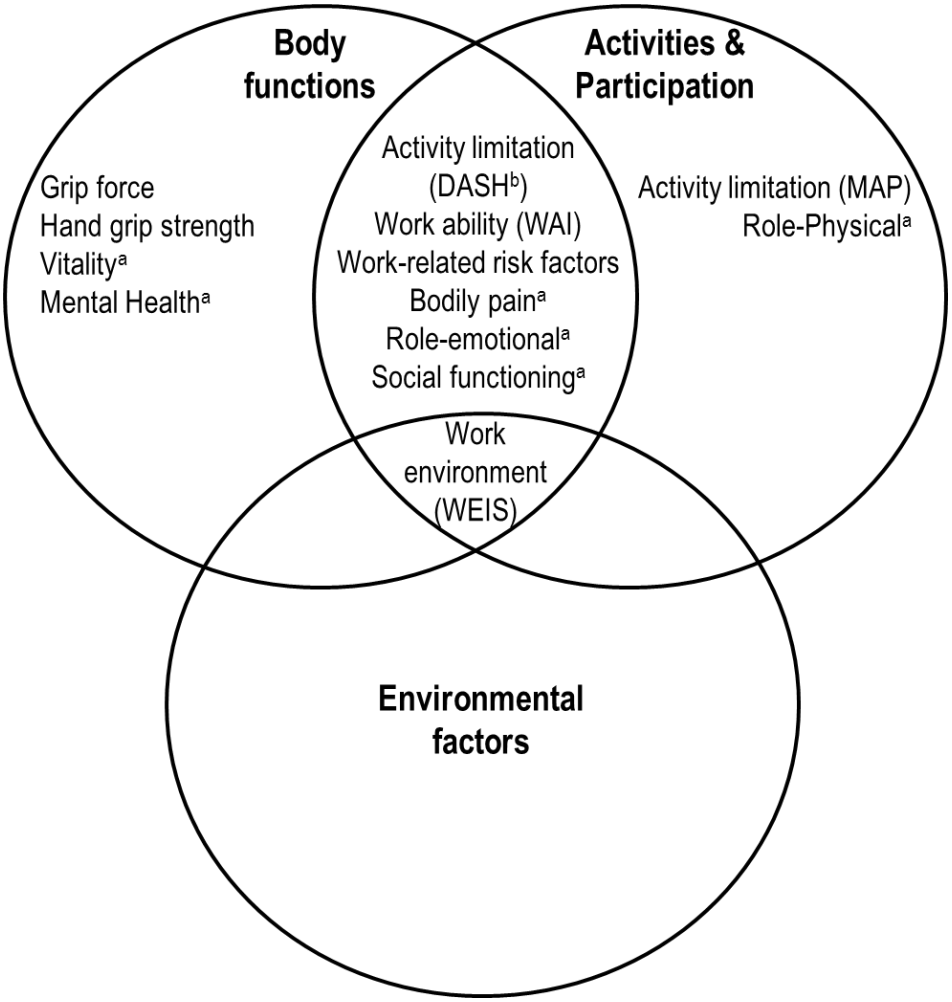

Figure 10. A summary of the results appears in this thesis and the results of disabilities in patients with PM and DM are linked to these ICF components.
a=Health-Related Quality of Life dimensions measured by the SF-36, b=DASH has also been linked to personal factors.

4.7 Concluding remarks

In conclusion, I have in this thesis shown that grip force may be affected in patients with PM and DM at different time-points from the time of diagnosis. In men with PM and DM the reduced grip force was more commonly present within the first one or two years after the myositis diagnosis. The reduced grip force could have an impact on activity performance and HRQoL. Therefore, it is important to use this new knowledge and refer the patients to specific hand therapy and occupational therapists, and to measure the grip force in patients with PM or DM as well as to assist patients with assistive devices or ergonomic advice when needed. A hand exercise programme seems to be feasible to use in patients with PM and DM, but the design of the programme needs to be further evaluated to offer improvements in hand function and activity performance.
Furthermore, work was rated as one of the most difficult activities in patients with PM and DM. In these patients, poor self-rated work ability was common, and there were interfering factors in the work environment that may be related to disease-specific impairment of muscle performance that affect the work performance and indicate a need to identify these interfering factors and to support the patients when required to enhance their work performance.
There is also a need to increase the national and international cooperation regarding research in this rather rare disease. Quality registers are important for research and ultimately for the care and treatment of patients with myositis. It is also important to include the patient in the study design so that the research questions that are being asked and hopefully answered consider the patients' perspective.

4.8 Clinical implications

The results from this thesis have added knowledge about hand function, activity limitation, work ability and HRQoL in patients with PM and DM. The following suggestions are based on the results in this thesis and the author's opinion:

- The hand grip strength or grip force are important to evaluate in the clinical care of patients with PM and DM. Both the Jamar and the Grippit© seem to be able to detect impairments in this body function. I suggest that the grip force or hand grip strength is assessed at the time of diagnosis and at the yearly follow-up visits to

enable appropriate care and for further research on the impact of hand grip strength or grip force in patients with PM and DM.

- If the hand grip strength or grip force is reduced, patients should be referred to specific hand therapy and occupational therapy for ergonomic advice or assistive devices to reduce the disability and maintain independence in activity performance.
- The results from this thesis did not give a conclusive answer on how to perform a hand exercise intervention in patients with PM and DM. It did, however, indicate that a hand exercise intervention is feasible to perform and did not increase the disease activity in patients with PM and DM. A hand exercise programme should be led by a hand therapist or occupational therapist who has experience with this rare disease. The hand exercise should aim to maintain or increase hand function and ultimately improve activity performance and HRQoL if possible.
- There is a need to include a self-reported assessment on work ability in addition to the percentage of full-time employment, such as the WAI, in patients with PM and DM. My suggestion is to add this measure both in clinical practice and at the yearly follow-ups. If added to the SweMyoNet Registry, it would enable a longitudinal aspect of work ability and the ability to identify possible sub-group characteristics of patients who are more at risk for sick leave and therefore in greater need of rehabilitation.
- The disease-specific symptoms in myositis seemed to affect the ability to remain at work, and there was a large proportion of patients with poor or less good self-rated work ability who still worked. The occupational therapist would be able to do jobsite analysis of work demands to adapt the work tasks or the work environment to increase work ability. There are assessments that would be applicable to use, e.g. the Assessment of Work Performance (AWP) and the Assessment of Work Characteristics (AWC) (148).
- Interaction with co-workers and others was seen as a support to remain at work for patients with PM and DM and could be a possible promoting factor to stay at work.

4.9 Future studies

As in all research, these results bring out new questions to address:

- The hand exercise intervention assessed in Paper III seemed to be feasible with good adherence but with limited effect on hand function and activity performance. To further explore the effect of a hand exercise programme in patients with PM and DM, a repeated-measure design, single case methodology or statistical process control should be undertaken (149). The hand exercise programme should have limited movements with a higher initial resistance. In addition, it would also be interesting to investigate whether a hand exercise programme may have a positive effect on HRQoL.
- The dexterity was reduced in a small convenience sample of patients with PM and DM. This aspect of hand ability needs to be addressed in a larger cohort, preferably with patients with PM and DM that have and have not experienced reduced hand grip strength. Adding the measure of dexterity in the SweMyoNet Registry would enable this kind of study.
- The results indicate the importance of evaluating the work ability in patients with PM and DM. Future studies should aim to investigate different patient characteristics that are risk factors for sick leave in myositis. This type of study would be possible if a measure of self-rated work ability were added to the SweMyoNet Registry.
- It would also be interesting to investigate, using qualitative methodology, what the facilitators or barriers are for remaining at work or not.
- In the future, a study investigating the effects of work-related interventions such as the use of adaptation, assistive technology or work place redesign would be interesting.
- In qualitative studies, activity limitation and HRQoL should be explored in PM and DM patients to investigate the extent to which hand function affects activity and HRQoL.

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