

Employment status and work performance in adults with myotonic dystrophy

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Abstract

Aims

The primary aims of this study were to identify the rates of employment (defined as the number of people in paid employment) and factors influencing work performance in adults with myotonic dystrophy. The secondary aim was to determine time loss from work due to their health.

Study Design

This cross-sectional study drew upon data collected from a broader study exploring the prevalence and outcomes of genetic muscle disorders in New Zealand (MD-Prev). This study extracted the demographic, diagnostic and questionnaire data for adults (aged ≥ 16 years) diagnosed with myotonic dystrophy type 1 and 2 from the MD-Prev database.

Participants

The MD-Prev study identified 327 adults with a confirmed diagnosis of myotonic dystrophy in New Zealand. They were all invited to participate in a multi-domain assessment, and 202 participants consented.

Outcome Measures

The Work Limitations Scale (WLQ-25) was used to measure work performance. Personal, occupational and environmental factors were considered in the analysis of work performance. This holistic view included measures to address self-efficacy, mood, cognition, physical functioning, pain, fatigue, sleep and support. A Spearman correlation helped identify factors associated with work performance. A Mann-Whitney U test compared data between those employed and unemployed to determine if there was a significant difference between these groups that could explain employment status. A chi-square test compared age, gender, ethnicity, and type of myotonic dystrophy between participants and non-consenters.

Results

Out of 202 participants, only 69 were employed at the time of the study. Despite a low employment rate, 73% had not taken time off work due to their health in the previous fortnight.

Physical and mental-interpersonal demands of a job affected work performance for 62.1% and 57.4% of the sample. Over 50% of those working reported no difficulty in time management (56.7%), and work-output (55.9%), which indicates participants are performing well in certain aspects of their job demands. Pain and fatigue impacted significantly on meeting work demands followed by depression, cognition, sleep, and physical functioning.

Age, gender and ethnicity did not influence employment status. However, there was a significant difference in ethnicity between those who did and did not participate in the assessments suggesting caution in the representation of findings for people of non-European ethnicity. Participants who completed tertiary education had a greater chance of employment with 56.5% of them in professional roles.

Conclusion

This study builds on previous research and highlights that people with myotonic dystrophy are a minority group in the New Zealand labour sector. Work performance was most disturbed at the physical level, with fatigue and pain being the main factors influencing all domains of work performance. Addressing pain and fatigue management in addition to symptoms of depression, cognition and environmental barriers may facilitate work performance and productivity. Encouragingly many people employed in this study were performing well in most aspects of their role.

These findings may provide insight for health professionals, disability services, and employers when supporting a person with myotonic dystrophy into sustainable careers that will accommodate their long-term capabilities as their condition progresses.

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Attestation of Authorship

I hereby declare that this submission is my own work and that, to the best of my knowledge and belief, it contains no material previously published or written by another person (except where explicitly defined in the acknowledgements), nor material which to a substantial extent has been submitted for the award of any other degree or diploma of a university or other institution of higher learning.

30th May 2019

Signature

Date

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Chapter 1 Introduction

As an occupational therapist (OT) I am passionate about the role my profession has, in enabling individuals with a disability achieve their work, career, or occupational goals. The specialist focus for occupational therapists is occupation which is defined as “everything that we do in life, including actions, tasks, activities, thinking and being” (Law, Baum, & Dunn, 2001, p. 6). In this study “occupation” or “work” will be defined as paid employment whether encompassing part-time or full-time hours.

Myotonic dystrophy is the most common form of adult muscular dystrophy; however, I feel this group are an unrecognised minority group in New Zealand with complex and intricate health and social care needs. I developed a special interest in myotonic dystrophy due to my professional career supporting people with lived experience of this condition. My interest in this research topic developed from my own experience as a practitioner. Employment was an issue amongst many young to middle aged myotonic dystrophy adults’ who were either unemployed, struggling to source suitable work or worried about their future career as their condition progressed. Given the value of purposeful employment on health, social, mental and emotional wellbeing (Lexell, Langdell, & Lexell, 2017), I believe this activity should be an essential consideration in the management of people with myotonic dystrophy, especially those who are willing and able to work.

Employment is the act of being engaged in an activity in exchange for payment (Brookes et al., 2009). The importance of paid employment is central to an individual’s self-worth and provides the most likely route out of poverty and enables participation in the wider society (Polanyi & Tompa, 2004; Stats NZ Tatauranga Aotearoa, 2014). It creates a sense of identity and purpose and contributes to economic self-maintenance, home and family maintenance, service or personal development and can represent our contribution to our community, workplace, or society (McColl, Law, & Stewart, 2015). Everyone has a right to seek paid employment; it is a social determinant of health and enhances social inclusion for people with disabilities (Barnes & Mercer, 2005; Lindsay, Adams, McDougall, & Sanford, 2012). Yet people living with a chronic medical condition or disability remain underrepresented in the New Zealand labour market. Findings from the 2013 New Zealand census showed that only 45% of people living

with a disability are employed on either a part or full-time basis compared with 72% of non-disabled adults. Importantly 74% of those who were not employed said they would like to work if a job was available (Stats NZ Tatauranga Aotearoa, 2014). This resonates in other parts of the world as many individuals with disabilities are unemployed, despite being capable and eager to work (Padkapayeva et al., 2017).

To provide context for this study I will firstly describe the condition of myotonic dystrophy, and the impact it can have on functional performance. Secondly, I will consider the context of New Zealand legislation for employment within disability. Finally, I will introduce the concept of the Person, Environment, Occupation (PEO) model (Law et al., 1996), which is a theoretical model used by occupational therapists to inform clinical practice.

1.1 Myotonic Dystrophy

Myotonic dystrophy, also referred to as myotonia atrophica, dystrophia myotonica, myotonia dystrophica, steinert's disease (type 1) and proximal myotonic myopathy (type 2). Prevalence ranges between 2.1% to 14.3% per 100,000 worldwide (Mathieu, Boivin, & Richards, 2003). A recent epidemiological study in New Zealand found that myotonic dystrophy affects eight in every 100,000 people (Theadom et al., 2019).

Myotonic dystrophy is an autosomal dominant disease caused by an expansion of a DNA triplicate in chromosome 19 or 3, known as CTG. Repeats can vary from 50 to over 1000. Generally, the more repetitions, the more severe the condition. The length of the repetitions is partly correlated with the severity of the disease and the age of onset (Gennarelli et al., 1996).

There are two main types of myotonic dystrophy type 1 and type 2, both caused by different gene mutations. Although the signs and symptoms are similar, type 2 tends to be milder than type 1. Type 1 is the most common form of myotonic dystrophy and muscle weakness predominantly affects the lower legs, hands, neck, and face. Muscle weakness in type 2 primarily involves the muscles of the neck, shoulders, elbows, and hips. Congenital myotonic dystrophy, apparent at birth, is a variation of type 1 but is more severe than the adult manifestation and can be life threatening (Genetics Home

Reference, 2019). As a result, congenital myotonic dystrophy will not be included in the current study.

Myotonic dystrophy is a multi-systemic condition and symptoms can include progressive muscle loss, daytime sleepiness, cataracts, apathy, slurred speech, hair loss, cardiac conduction defects, respiratory, digestive, reproductive and endocrine difficulties (Harper, 2009). One of the distinguishing symptoms in myotonic dystrophy is myotonia, a prolonged muscle contraction when individuals are unable to relax muscles after use, most noticeable in their hands. In comparison to other muscular dystrophies, myotonic dystrophy affects a variety of organs in the body (Harper, 2009) and cognitive and mental functioning (Meola & Sansone, 2007). Fatigue, executive and visuospatial dysfunctions, and anxious personality traits (deteriorating with age) are common manifestations in people with the classic adult forms of myotonic dystrophy (Harper, 2009). Excessive sleepiness, loss of initiative, tendency to postpone actions, and personality traits of stubbornness and avoidance (Timman, Tibben, & Wintzen, 2010) may present an added challenge when seeking employment.

1.1.1 Myotonic Dystrophy and Occupation

The range and severity of symptoms mean that myotonic dystrophy affects many aspects of a person's physical, cognitive, mental, and social wellbeing. It has also been proposed that there are additional under-recognised symptoms that are important and have a critical effect on an individual's health status (Heatwole et al., 2012), which makes this condition even more complex. The secondary implications of myotonic dystrophy extend to core aspects of an individual's life. These include high unemployment rates (Fowler et al., 1997; Gagnon, Mathieu, & Noreau, 2007), poor participation in recreational activities (Gagnon, Mathieu, et al., 2007; Nätterlund & Ahlström, 1999) and lower educational attainment (Gagnon et al., 2008). Gagnon et al (2008) identified that people with myotonic dystrophy had the highest rate of disrupted participation and the lowest levels of satisfaction in employment. Employment is therefore an important area of concern for this population group and most likely an important issue to address for those living with myotonic dystrophy in New Zealand.

Diagnosis can happen at any time across a life span, however symptoms often become noticeable at a working age, during a person's twenties or thirties (Gagnon, Mathieu, et al., 2007). This onset can affect gainful employment due to the progressive disposition of the condition. As with most human beings the ability to undertake, thrive and maintain work is important for adults with myotonic dystrophy (Andries et al., 1997). Work gives meaning to life, creates a sense of purpose and identity and enhances community integration, self-esteem and financial stability (Andries et al., 1997).

However, the progression and severity of symptoms in myotonic dystrophy can vary considerably between different people. As a result, individuals face challenges planning their imminent and future career with uncertainty about how their condition will manifest over time.

There appears to be limited research specific to employment in myotonic dystrophy especially in New Zealand. However, there has been a recent re-focus by the New Zealand government to increase the number of people with disability into paid employment (Office for Disability Issues, 2016a). This is not a new initiative, rather one that has been a priority for many years but gained little traction. To support individuals with myotonic dystrophy, attain, maintain, or regain paid employment one needs to firstly identify how many people are either employed or unemployed. When exploring what factors may influence work performance it is important to consider the impact of the symptoms (physical and mental) and the secondary implications these have on social wellbeing, functional performance, employment rates and education.

My research will focus on employment rates and identify factors that may influence work performance for people with myotonic dystrophy. However, to inform the study it was important to understand the legislation of the New Zealand government and its focus to support individuals with a disability into employment. The next section outlines the historical background of disability in the employment sector and the obligations New Zealanders must adhere to for employing people with disabilities.

1.2 Occupation within the Disability Sector

1.2.1 Historically

Historically, people with disabilities have been socially disadvantaged and more susceptible to poverty due to increased unemployment rates (Lastuka & Cottingham, 2016). In the 19th and some of the 20th century, disability was viewed as a burden on the family and the economy. Solutions to this was often segregating people with a disability and placing them in institutions, not recognising the potential they could add to the labour market. Effectively shaming and hiding them away from the rest of society (Office for Disability Issues, n.d.).

The need to address the disadvantages faced by disabled people in the labour market has for some time been officially recognised both nationally and internationally. Views have changed somewhat in the past 100 years in New Zealand led by the changing attitudes overseas and social policy changes. Sheltered employment opportunities for disabled people began with the Disabled Persons Employment Promotion Act 1960. However, there were still discrepancies as managers of sheltered workshops were exempt from applying the same employment conditions on the open labour market, which created further segregation for the disability sector (Office for Disability Issues, n.d.).

From the 1970s, the government's approach to services for disabled people became more community and rights based (Office for Disability Issues, n.d.). In New Zealand, the Industrial Relations Act (1973) established the under-rate workers' permit. This recognised the need for disabled people to have opportunities to mainstream employment and enabled a person with a disability to work in the labour market and receive a wage that matched their productivity (Office for Disability Issues, n.d.). They did not describe how they measured this productivity. However, this permit was a major milestone as it was one of the first times disabled people were supported into employment.

1.2.2 Current Society

Although reform started in 1970's there is still a level of exclusion from the paid labour market for people with disabilities evident today. Research continues to highlight

social exclusion that people with disabilities face across a range of spheres — political, economic, social and cultural (Appleton-Dyer & Field, 2014). One in six people who acquire a disability while working lose their job within a year and a third of disabled people who acquire work are unemployed again within a year, in comparison to one-fifth of non-disabled people (Burchardt, 2000). This creates an assumption that disabled people are not capable of contributing to the work force or maintaining a career.

In New Zealand, discrimination in employment on the grounds of disability was first prohibited by the Human Rights Act 1993. This was strengthened in 2008 with Article 27 of the Convention on the Rights of Persons with Disabilities which called on countries to “recognise the rights of persons with disabilities to work on an equal basis with others”, and outlined a number of steps to achieve this objective (United Nations, 2008, pp. 19-20).

Despite the prohibition of discrimination by the Human Rights Act in 1993, a study commissioned by the Ministry of Social Development (MSD) demonstrated that discrimination is still an issue in employers’ attitudes towards employing people with disabilities. This research identified the following results:

“Most employers said that they would be unlikely or less likely to employ disabled people if they were moderately intellectually disabled (60 per cent) or had a moderate to high speech impairment (60 per cent). Some were less likely to employ people if they had a mental illness such as depression (47 per cent), had a moderate to high sight impairment (41 per cent), had a moderate to high hearing impairment (41 per cent) or were in a wheelchair (36 per cent)” (Woodley, Metzger, & Dylan, 2012, p. 6).

This implies a ‘hierarchy’ of disability where the type and severity of a person’s condition can have an impact on employers’ perception of their employability, regardless of whether they are trained or experienced to do the job (Woodley et al., 2012). People with myotonic dystrophy experience some of these impediments (speech impairment, mild / moderate intellectual difficulties, sight impairment, use of a wheelchair) and based on these findings there may be a preconceived idea that they will not be suitable for the work force. They are a range of employment services and

resources in New Zealand to support employers hire people with disabilities e.g. Workbridge. Despite this some employers in the 2012 study reported the following:

“Many felt that staff would not be comfortable working alongside disabled people. Similarly, many felt that their customers and clients would not be particularly comfortable dealing with disabled people” (Woodley et al., 2012, p. 6).

“Two-thirds (67 per cent) said they would be influenced by negative reactions from staff. Similarly, three-quarters (75 per cent) said that they would be influenced by the negative reactions of clients and customers” (Woodley et al., 2012, p. 6).

These findings highlight that discrimination and segregation are still very present and noticeable in New Zealand. It is important to consider these societal influences when analysing the literature as they may have a greater influence on a person's employment status rather than the impact of their myotonic dystrophy.

The first New Zealand Disability Strategy was agreed in 2001. Its aim was to eliminate barriers so disabled people could participate fully in the community (Minister for Disability Issues, 2001). Proactively, the 2001 New Zealand Disability Strategy included among its objectives to “provide opportunities in employment and economic development for disabled people” (Minister for Disability Issues, 2001, p. 17). Strategic priorities for implementing both the Disability Strategy and the UN Convention were set out in the Government's Disability Action Plan 2014–2018 (Office for Disability Issues, 2014). This plan included a strategy to “increase the number of disabled people, including long-term unemployed disabled people, into paid employment and self-employment on an equal basis with others” by “building employers' confidence to employ disabled people and provide accessible workplaces, opportunities for work experiences, entrepreneurship, and education achievement and skill development” (Office for Disability Issues, 2014, p. 5). The recent development of the New Zealand Disability Strategy 2016–2026 has continued to identify the importance of disabled adults having a meaningful job and being able to support their families. The Strategy guides the work of government agencies on disability issues and the Disability Action Plan which is currently under review, is the primary driver for implementing this strategy (Office for Disability Issues, 2016a).

Based on decades of legislation and strategies one could argue why is there still discrepancies in employment rates for people with disability in New Zealand. A report by Stats NZ Tatauranga Aotearoa (2017) outlined that disabled people were twice as likely to be either unemployed or not getting as much work as they would like. The 'underutilisation rate' for disabled people in New Zealand was 23.5%, more than double the rate for non-disabled people at 11.4% (Stats NZ Tatauranga Aotearoa, 2017). The number of people who participated in this survey with a rare condition is unknown.

John F Kennedy once said, "no one gains from fair employment law and legislation if there is not employment to be had". He wanted to make sure everyone who wanted to find a job could find a job as it is a fundamental premise upon which society is based (Frost, 2013). Reflecting on this quote, employment law and legislation to support people with a disability into employment has been in place for decades in New Zealand. Employment opportunities have also been available, but this appears to have had little impact for those with a disability. The unemployment rate for disabled people remained the same from 2001 to 2013 whereas the rate for non-disabled people fell slightly over that period (Stats NZ Tatauranga Aotearoa, 2014). The implementation of these policies is questionable since they have not made an impact on increasing the employment rate for people with disabilities in New Zealand. The employment rates continue to remain low and people with disabilities remain disadvantaged in the employment sector.

Healthcare professionals and governments recognise the value of empowering people with a disability by way of gainful employment (Employment New Zealand, 2009; Minis et al., 2010). High profile individuals such as Stephen Hawking, and Michael J Fox have helped shift the stigma associated with disability by demonstrating how disabled people can participate and make a valuable contribution in a working environment. However, employers must also provide opportunities for people with disabilities by giving them a fair opportunity to apply themselves in the labour market.

1.3 Theoretical Model

The Person-Environment-Occupation (PEO) Model (Law et al., 1996) is a theoretical model, widely used in OT practice to analyse performance and will be used in this

study to identify enablers and barriers in occupational performance for people with myotonic dystrophy. When analysing occupational performance, it incorporates the person, their environment and occupation. The person domain incorporates role, self-concept, cultural background, personality, health, cognition, physical performance, and sensory capabilities. The environmental domain includes the physical, cultural, institutional, social, and socio-economic environment. Occupation refers to groups of tasks that a person participates in to meet their intrinsic needs for self-maintenance, expression, and fulfilment (Law et al., 1996). The three domains are interdependent and influenced by each other and their level of overlap determines occupational performance. The temporal aspects of the model recognise that the interaction and influence of these three components vary across time (Law et al., 1996).

One of the reasons the PEO model (Law et al., 1996) was chosen over other OT models was its consideration to time, which is important due to the progressive impact of myotonic dystrophy. Its ability to adapt across changing situations enables therapists to conceptualise and plan interventions which consider human development and change (Strong et al., 1999). This model can also be applied to organisations and communities such as a person's place of employment (Christiansen, Baum, Bass-Haugen, & Bass, 2005). The PEO model (Law et al., 1996) can be manipulated with intervention at any of the three levels and purposeful occupation performance is achieved when this dynamic relationship is effectively managed (McColl et al., 2015).

It is important to note that the capacity of the PEO model (Law et al., 1996) can vary between countries and cultures and may not meet the needs of all clients, especially clients from non-Western cultures (Wong & Fisher, 2015). This is a limitation in its use and should be considered when applying to different ethnic groups.

Based on this conceptual theory it could be hypothesised that work performance is influenced by the individual's personal characteristics and traits (role, self-concept, cultural background, personality, health, cognition, physical performance, and sensory capabilities) their environment (physical, institutional, social, cultural and socio-economic, in which occupations take place), and occupation (type of work, education, and tasks people do in their daily lives). Performance is the act of doing and this may diminish when one of the three components are not in cohesion with the others. The

more an occupation meets the needs of accomplishment, affirmation, coherence, pleasure, companionship, and renewal the more likely it is to be associated with health (Morgan, 2010).

The structure of this thesis will start with a review of the existing literature on employment amongst people with myotonic dystrophy before then outlining the methods and results of the current study. It will conclude with a discussion on how the results can be applied to current knowledge and propose concepts for further investigation and future practice.

Chapter 2 Literature Review

This chapter reviews the existing literature on myotonic dystrophy, employment legislation specific to people with disability, benefits of work on health and wellbeing and factors that relate to employment for people with neuromuscular diseases (NMDs).

To inform the current study a narrative review of the existing literature published in English was conducted. Publications were retrieved following a search on the following resources: EBSCOhost, CINALH, Pubmed, Scopus, Science Direct, Google Scholar, OVID Medline, and Cochrane Library between 1990 – 2019. The search terms used were: myotonic dystrophy OR myotonia atrophica OR dystrophia myotonica OR myotonia dystrophica, OR steinert's disease combined with one or more of the subjects: work, OR career, OR job, OR employment, OR occupation, OR vocation, OR profession, OR perform, OR success, OR capabilities, OR influence, OR effect, OR relate, OR impact.

Due to the limited number of robust publications on employment in myotonic dystrophy, the scope of the literature search was widened to include employment in all “muscular dystrophies” and “neuromuscular diseases”. Employment is a central and time-consuming activity in adult life. A persons experience of work has a great influence on their overall quality of life (Kielhofner, 2009). Therefore, articles addressing quality of life in myotonic dystrophy with reference to employment were also considered. Additional literature on disrupted participation in activities and life habits in myotonic dystrophy with reference to employment were also contemplated to inform this study. A list of search terms can be found in Appendix A. Reference lists from identified publications and publications from common authors were searched to elicit further literature. Finally, the articles selected were critiqued in depth to identify those most appropriate for inclusion in this review. Figure 1 outlines the literature review process and reoccurring authors cited.

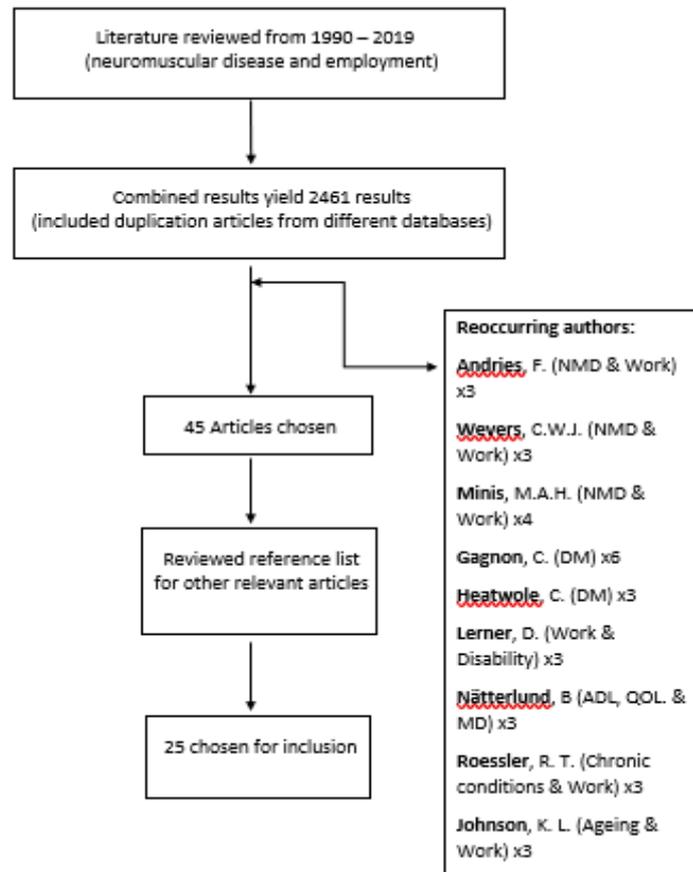


Figure 1: Literature search

DM = myotonic dystrophy; NMD = neuromuscular disease; ADL = activities of daily living; QOL = quality of life; MD = muscular dystrophy.

The search yielded articles from studies primarily undertaken in the Netherlands, Sweden, United States and Canada. There was no published research from Australasia identified. When reviewing the literature, it was important to note employment opportunities, disability supports, policies and legislation in these countries are different to New Zealand.

Common themes identified in the literature that influenced work opportunities and employment rates for people with a neuromuscular disease (NMD) were gender, age, education level, type of NMD and supports (Andries et al., 1997; Fowler et al., 1997; Gagnon et al., 2008; Minis et al., 2010). Very few studies focused solely on myotonic dystrophy. Rather they included myotonic dystrophy with a combination of other neuromuscular diseases (NMDs) such as facioscapulohumeral muscular dystrophy (FSHD), hereditary motor and sensory neuropathy (HMSN), limb-girdle muscular dystrophy (LGMD). Reasons for combining multiple NMDs was due to low prevalence rates of each separate condition, and the similarities between some of the conditions

in relation to their symptoms and subsequent effect on functional tasks and quality of life. As a result, literature relating to quality of life issues, performance, and employment in other long-term conditions was inferred to when collating the review. The findings from this review are presented under the categories of the PEO model (personal, environmental and occupational factors) outlined in the introduction (Law et al., 1996).

2.1 Personal Factors

The personal domain of the PEO model includes role, self-concept, cultural background, personality, health, cognition, physical performance, and sensory capabilities (Law et al., 1996). This section will describe themes in the literature, classified under the personal domain of the PEO model (Law et al., 1996).

2.1.1 Health / Physical Performance / Role

A review of the literature by Minis, Heerkens, Engels, Oostendorp, and Van Engelen (2009) identified 10 factors associated to employment in three NMDs. These factors related to body function (physical functions, muscle power functions), socioeconomic factors (age, gender, education) and work-related factors (type of occupation, interest in employment). In addition to the 10 factors, 84 other factors identified in their review were not considered since they were only present in one study. These factors were not outlined, despite some of them yielding significant results. Due to the variability of condition progression in most NMDs excluding some of these significant factors from the results may have limited the depth of understanding in employment status and work profiles for this population group.

Despite this limitation to not report on all the significant factors in their study Minis et al. (2009) recognised that disease related factors influenced employment in people with a NMD as opposed to other chronic conditions such as chronic obstructive pulmonary disease (COPD) (Orbon et al., 2005). They also recognised that the type of NMD was a factor when examining employment status, however it was not obvious if this related to one muscular dystrophy condition more than another. To answer this question Fowler et al. (1997); Jensen et al. (2008); Minis et al. (2010) all identified that the type of NMD had an impact on employment status especially people with myotonic dystrophy. The literature also indicated work performance may differ

between these groups (Andries et al., 1997; Minis et al., 2010). The symptoms of myotonic dystrophy often manifest or progress during ones working life and their progressive impact can affect performance in activities of employment. Additionally, the uncertainty around progression timeframes may concern potential employers as noted in a report commissioned by Ministry of Social Development (Woodley et al., 2012). This suggests the need for research to look at these NMDs separately, given their unique presentation, progressive symptom profiles and differences in employment status and work performance.

Duration of symptom onset has also been found to be an important factor in work performance as it correlates with the rate of severity especially in mobility limitations, cognitive impairment, and activity performance (Heatwole et al., 2012). It is therefore likely that an earlier onset of symptoms may lead to poorer work outcomes. In a study by Heatwole et al. (2012) the greatest increase in symptom prevalence and severity for people with myotonic dystrophy occurred between the ages of 21 – 30 years. Physical limitations increased the most from 33.3% to 82.7% between the ages of 31- 40 years (Heatwole et al., 2012). Based on these literature results and the low employment rates for people with myotonic dystrophy it was hypothesised that people with myotonic dystrophy retire at a young age in comparison to the general population. Some individuals with a NMD never sought paid employment, or their career trajectory was shortened due to the severity of their symptoms (Fowler et al., 1997). This raises concerns for the financial, social, physical, emotional, and mental wellbeing of this population given the importance of productive work on health and wellbeing.

However, the sample selection in these studies may have influenced the results. For example, in Heatwole et al. (2012) study, participants were selected from a registry database which are optional for people to opt in. Consequently, it is unlikely to be a source of participants representative of the broader myotonic dystrophy population. In addition, everyone in this registry did not participate which could lead to the potential for sampling bias. Therefore, it was likely many participants had an expressed interest in research since they were already on the registry and may have had the cognitive and physical ability to take part. Similar to Heatwole et al. (2012), physical symptoms associated with the condition also had a negatively influence on work continuation and potential work performance in a study by Andries et al. (1997). The survey used in

Andries et al. (1997) study was not accurately described which was a limitation as the validity and reliability of the study was not transparent. As this study is 22 years old its credibility today has lessened due to changes in support, attitudes and opportunities within the employment sector e.g. increase in female employment, immigration, and flexible work environments.

Symptoms that significantly affected the lives of people with myotonic dystrophy in the literature were explored as they could also have an impact on work performance. These symptoms included fatigue, mobility, an inability to do specific activities, problems with the use of hands or arms and impaired sleep or daytime sleepiness (Heatwole et al., 2012; Laberge, Bégin, Montplaisir, & Mathieu, 2004).

Fatigue is defined as an overwhelming sense of tiredness, lack of energy, and a feeling of exhaustion that goes beyond muscular weakness (Krupp, 2003). The literature is abundant on the impact of fatigue and pain in NMDs (Gagnon et al., 2008; Jensen, Abresch, Carter, & McDonald, 2005; Kalkman et al., 2005; Minis et al., 2010; Wokke, 2007). Fatigue had the greatest life impact in Heatwole et al. (2012) study and both sleepiness and fatigue constitute as major complaints in myotonic dystrophy type 1 and type 2 (Giubilei et al., 1999; Tieleman et al., 2010). Interestingly in Kalkman et al. (2005) physical functioning and social functioning contributed significantly to fatigue severity for people with myotonic dystrophy. There were limitations in their study with the methodology chosen as the cross-sectional design meant they were unable to draw conclusions on the direction of the association. In addition participants who were not diagnosed in the research hospital did not have their diagnosis checked to ensure accuracy (Kalkman et al., 2005).

Fatigue was reported in a study by Minis et al. (2010) amongst both people employed and unemployed with a NMD. Despite high levels of fatigue its impact did not affect employment status for some of this sample, whose employment rate was also high (Minis et al., 2010). However, this was not reflective for people with myotonic dystrophy, as there was a significant difference in fatigue, activity levels, motivation, and concentration between employed and unemployed individuals (Minis et al., 2010). This study did not determine if these differences in symptom severity came before or

after the person was employed or if being employed influenced an exacerbation in symptoms.

In another study 45% of those surveyed with myotonic dystrophy had experienced disruption in their work due to disability related factors such as strength, fatigue, and pain (Gagnon et al., 2008). Muscle strength decline disrupted participation in daily activities and accomplishment of social roles such as employment (Gagnon et al., 2008). Individuals who had to leave their jobs as their condition progressed often found it difficult to acquire a new job and signed onto government benefits (Gagnon, Noreau, et al., 2007). A limitation to Gagnon et al. (2008) study was the high burden placed on participants; it took 5 full days of evaluation. This duration may have been a barrier for people working and others may have declined due to fatigue and pain implications. Despite this limitation their study also supported the idea that symptoms and type of NMD were a factor on employment status and performance, and it was therefore important to consider these elements in the current study.

This literature identifies that the condition type and associated symptoms e.g. fatigue, pain, weakness, reduced quality of communication and the effect on facial expression due to muscle weakness, are important considerations on work performance but are not solely decisive to employment duration (Andries et al., 1997; Fowler et al., 1997; Madej-Pilarczyk, 2014). Myotonic dystrophy presents a range of unique symptoms and the impact of these symptoms on work performance have not been explored in Australasia nor have the barriers and facilitators to employment. Due to the uniqueness of the condition the factors that influence work performance for people with myotonic dystrophy may be different to other neuromuscular and chronic conditions. However, there has been limited research on the emotional and cognitive aspects of living with myotonic dystrophy and their impact on occupation.

2.1.2 Self-concept / Personality / Psychological

Andries et al. (1997) and Fowler et al. (1997) described that the physical impairments of a NMD are not an obstacle for work continuation but rather the individual's intellectual capacity to adjust psychologically. Social support might also be an important resource in making that adjustment but was not considered in Andries et al. (1997); Fowler et al. (1997); Madej-Pilarczyk (2014) studies.

The onset of the condition may influence work performance and career trajectory. As previously stated, those diagnosed at a young age can often have more severe symptoms which include both physical and mental elements, and therefore they may have difficulty seeking and maintaining a job (Harper, 2009). In addition, there is the psychological adjustment to consider which may vary depending on age of diagnosis. However, an opposing argument is those diagnosed at a young age have a greater opportunity to adapt to the functional and psychosocial demands of their condition and set realistic goals early in life (Andries et al., 1997). In comparison, those who acquire myotonic dystrophy at an older age may have more difficulty in adapting from their normal patterns of behaviour. They may be qualified and working in a job that is no longer sustainable as their condition progresses. Therefore, their work performance and career trajectory may be disrupted by a later onset diagnosis and their psychological ability to accept and alter work goals and life aspirations. As with an interest in employment, psychosocial adjustment may be both the cause and result of employment (Fowler et al., 1997).

High motivation to continue working and high self-esteem were factors in a recent study by Lexell et al. (2017) that participants expressed as a resource for work continuation. A positive attitude towards the progression of their condition helped them manage work situations. However, the sample size of this study was small (nine people), with restricted age criteria and inclusion of seven different NMDs. In addition, the sample was male dominant with one female and all participants were in paid employment thus not reflective of the myotonic dystrophy population.

Disclosure of a condition to employers can have a positive or negative impact (Minis et al., 2014). Some people hide symptoms and postpone disclosure due to the risk on their social, economic, and emotional benefits of work. Instead of feeling fearful to disclose information about their condition people with myotonic dystrophy should feel empowered to communicate their work problems early so timely support can be provided (Minis et al., 2014). This study only interviewed 16 people with a type of NMD, who were working at least 12 hours a week. People who were working less hours and those no longer working were not included in the study and may have had different perspectives which would have added depth to the results.

There was conflicting evidence in the literature on individuals' levels of interest to seek work. Many studies identified people willing and able to work and the percentage of participants who had never worked was low (Fowler et al., 1997; Lindsay, McDougall, Menna-Dack, Sanford, & Adams, 2015; Minis et al., 2009). However, a study by Eggers and Zatz (1998) identified that more than half of the people unemployed with progressive muscular dystrophy did not want a job. A possible explanation sought was the muscular strength required for many jobs and restricted workplace adjustment. Fowler et al. (1997) also identified individuals not interested in employment frequently reported having a physical disability as their barrier in comparison to people who were interested in seeking employment. Interestingly people with myotonic dystrophy reported physical limitations as their barrier to employment however their physical limitations and employment rate was lower when compared to other muscular dystrophies. People with myotonic dystrophy perceived their physical capabilities to be lower than measured ability (Fowler et al., 1997). This may infer the influence of one's self-confidence, self-esteem and self-efficacy in their physical capabilities. It may also be due to greater incidence of anxiety and depression in this population (Harper, 1989) and their difficulty with abstraction and new concept formation (Gagnon, Noreau, et al., 2007). Fowler et al. (1997) identified that people with myotonic dystrophy regardless of employment status tended to be more discouraged, pessimistic, worried, shy, passive, and lacking in ambition than those with other NMDs (Fowler et al., 1997). This lack of motivation and apathy could impact on their ability to perform and attend to work tasks.

Fowler et al. (1997) was one of the early studies exploring employment within NMDs. Although this study did highlight the need to consider self-efficacy, emotional wellbeing, and mood it did not describe the sample sufficiently and relevant employment measures were not utilised. The inclusion of six different NMDs may have diluted the impact of results for the myotonic dystrophy population. Due to these limitations and duration since data collection occurred the results cannot be generalised to people with myotonic dystrophy in New Zealand.

Despite the limitations in these studies (small sample sizes, sample not representative of the myotonic dystrophy population and inclusion of multiple NMDs), it was apparent that physical and psychological capabilities should be considered when

exploring employment status and performance in this population. However, the potential role of self-efficacy in myotonic dystrophy has not been explored in depth.

2.1.3 Cognition

An issue for some people with a disability is the additional time and effort it takes to perform daily tasks and activities that non-disabled people can take for granted, such as getting out of bed, going to work, personal cares and making a meal (Barnes & Mercer, 2005). The daily activities of transfers, personal cares, and transportation may be more cumbersome and exerting as their condition progresses, leading to reduced work hours and eventually leaving the work force (Mitchell, Adkins, & Kemp, 2006). The additional effort and time will vary for each individual and the severity of symptoms will be of importance. In some situations, the mental effort to attend and perform at work is even more strenuous than the physical aspects of the job. Wevers, Brouwer, Padberg, and Nijboer (1993) concluded that maintaining a job demands more effort for a person with a neuromuscular condition in comparison to those who do not have the condition. This is due to the additional effort in getting ready for work and commuting to their job. The physical exertion required for these self-care tasks means their ability to focus, concentrate and attend to work may be depleted before they arrive at their workplace.

In comparison to other NMDs, myotonic dystrophy is a multisystem condition that affects cognitive impairment, executive dysfunction and causes avoidant personality traits (Meola et al., 2003; Modoni et al., 2008; Weber et al., 2010). Cognitive rates fluctuate between individuals with the condition and may influence work performance and employment status as identified in a study by Minis et al. (2010). In their study employed people with myotonic dystrophy reported significantly fewer concentration problems and higher motivation in comparison to those unemployed (Minis et al., 2010). This indicates cognitive functioning is likely to be more of a factor in work performance and employment rates for people with myotonic dystrophy than other NMDs.

This review of personal factors based on the PEO model revealed a lack of research specifically exploring the potential role of fatigue, cognition, pain, emotional wellbeing

and self-efficacy on work performance and employment status for people with myotonic dystrophy.

2.2 Environment

The social model of disability recognises that the environment (social, physical, cultural, and institutional) can create barriers to participation for people with a disability not associated to their condition (Oliver, 1996). Therefore, it was important to consider the role of the environment as either an enabler or barrier to individuals work performance. These societal and environmental factors may be classified as attitudinal, communication, physical, policy, programmatic, social and transportation (Centers for Disease Control and Prevention, n.d.). To my knowledge there are no services in the New Zealand disability sector that continually follow and support an individual throughout their career journey and from job to job. As a result, some people end up in the “disability trap” where their only viable option is associated with disability income subsidy as they begin to struggle in their work performance or source a suitable job (Johnson, Brown, & Knaster, 2010). This section will describe themes in the literature, classified under the environmental domain of the PEO model which are physical, institutional, social, cultural and socio-economic environment (Law et al., 1996).

2.2.1 Physical / Institutional environment

Workplace accommodations addressing physical, social and attitudinal barriers are an important means of ensuring equal opportunity for employment among persons with disabilities (Gates, 2000). These include physical and technological modifications such as assistive devices, job modifications and time flexibility, development of the work environment such as workstations, assistance of other people, workplace culture, supports, partnerships, communication, inclusive recruitment and hiring practices, work location and transportation (Padkapayeva et al., 2017).

Workplace accommodations can help increase work performance and retain staff for longer (Ministry of Social Development, 2016). In addition, legislation supports work place accommodations for people with disabilities to increase equality, enabling all employees to do their jobs as well as possible and participate in society (Minister for Disability Issues, 2001; Office for Disability Issues, 2014; United Nations, 2008).

However, the low employment rate for people with disabilities appears to be a worldwide problem, despite recognised legislature to support this population group into employment.

To understand the reasons for this low employment rate Nevala, Pehkonen, Koskela, Ruusuvoori, and Anttila (2015) conducted a systemic review of the literature to identify the barriers and facilitators for work place accommodation and their effectiveness in supporting work performance. This review included research from various countries and identified moderate evidence that workplace adaptations can increase employment amongst people with physical disabilities (Nevala et al., 2015). The workplace adaptations included career counselling and guidance, education in self-advocacy, support from others, flexible work schedules, and special transport (Nevala et al., 2015). Some of these enablers such as access to transport, vocational counselling, career guidance and education in self-advocacy are not readily available in all parts of New Zealand. Accessible transportation is a problem especially in rural areas. Lack of awareness of the needs of people with myotonic dystrophy and limited specialist services providing support due to the rarity of the condition are also a factor for people with this condition.

Literature on the key facilitators and barriers to employment for people with a disability were identified. These include assistive products, timely work adjustments, self-advocacy from the disabled persons, support of the employer and community, level of training and counselling disabled people receive, and flexibility with respect to work schedules and work organisations (Minis et al., 2014; Nevala et al., 2015). People with a neuromuscular disease were not represented in Padkapayeva et al. (2017) or Nevala et al. (2015) studies and therefore those results cannot be compared to the myotonic dystrophy population.

Wevers et al. (1993) identified that almost half the people with FSHD in their study experienced physical problems in their work performance due to the environment. Workplace adjustments had not been made for many people in the study. However, this study is 26 years old and the current range of environmental aids and supports may not have been available when the study was conducted. Additionally, there are considerable differences in the symptom profiles of myotonic dystrophy and FSHD.

Work adaptations can consist mainly of reducing time pressure demands (Minis et al., 2012). The most frequent workplace adjustments in studies with NMDs included adaptations of working conditions, terms of employment, and task content (Andries et al., 1997; Minis et al., 2012). Andries et al. (1997) and Lexell et al. (2017) also found the importance of flexible work environments including flexible work hours, realistic physical expectations, and autonomy at work as a precondition for maintaining employment for individuals with a neuromuscular disease. Without workplace adaptations the risk of early retirement was four times greater for people with a neuromuscular disease including myotonic dystrophy (Andries et al., 1997). None of these studies focused solely on myotonic dystrophy and access to these employment facilitators may be different in New Zealand. Therefore, they are limitations in applying these results to the myotonic dystrophy population and NMD groups in New Zealand.

Existing evidence on the use of personal assistance services to support someone in their work environment with daily activities and tasks concluded that people with disabilities who had access to personal assistance services at the workplace had increased their work ability from being substantially limited to not limited at all (Solovieva, Wallsh, Hendricks, & Dowler, 2010). Although personal assistance services were effective in this study there was limited information on the questions asked, type of disabilities included, and the sample size was small and predominately white females. Individuals had to self-refer to these services which may have been a limitation for some who were less proactive in seeking workplace accommodations and those who did not have the knowledge or resources to make these enquiries. This study on personal assistance services was conducted in America, and the funding for employment support in America is not comparable to New Zealand as the medical and vocational rehabilitation services are mainly funded by private insurance.

Gagnon et al. (2008) recognised that access to mobility aids and technology was another obstacle for performance and participation in work activities. In New Zealand individuals with a chronic condition can access aids from the Ministry of Health. Although the majority of these aids are funded the range and individualisation of equipment to meet the specific needs of an individual are limited especially for outdoor pursuits. People have less control over their choice of aid and may have to conform to an off the shelf product. Not having access to the most suitable aid may

limit a person from releasing their full potential especially in the work environment. Gagnon et al. (2008) found that the environment was a leading contributing factor towards participation in activities such as employment for people with myotonic dystrophy however certain limitations should be considered when interpreting these results. The many statistical tests in their study could have led to type 1 error.

Factors external to the individual such as employer attitudes, support and willingness to modify working conditions or the physical environment (Saeki, 2000) have been suggested to be the main cause of unemployment rather than the physical disability (Andrén & Grimby, 2004; Murphy, Molnar, & Lankasky, 2000). The attitudes of co-workers, employers and their understanding of the disability were key factors in facilitating or preventing employment in studies by Minis et al. (2014) and Lexell et al. (2017). Employers willingness to support participants with a neuromuscular disease depended on their ability to manage company policies and their relationship with their employee (Minis et al., 2014). In some instances, it influenced opportunities for job retention or the possibility for alternative employment whereas in other circumstances it led to termination of one's contract (Minis et al., 2014). This was the perception of the participants. Employers perspectives were not gathered, and company policies were not analysed to add depth to or confirm these findings. The policies related to employment in the Netherlands will therefore differ to New Zealand employment laws and company policies. In comparison to other employees, employers are more likely to question the work ethic of disabled workers and their aspirations for career advancement. There is a preconceived idea that people with a disability are more prone to work absenteeism, less committed to their work and less capable to work alongside others (Cunningham, James, & Dibben, 2004). These assumptions were noted in the report on 'employer attitudes towards employing disabled people', in New Zealand (Woodley et al., 2012). It is not known if these employers had experience in hiring or working alongside someone with a neuromuscular disease.

Condition specific symptoms can also affect work performance as previously discussed. Nätterlund and Ahlström (1999) found people with muscular dystrophy had difficulties with heavy work due to muscular weakness. Despite this knowledge few people accepted help or avoided heavy work. They proposed that these individuals may have felt it was not socially acceptable to ask for help and avoided certain actions, so their

weakness was not visible to the public. People can be reluctant to disclose their condition for fear of been put in an unfavourable position such as dismissal (Minis et al., 2014). The measures used in Nätterlund and Ahlström (1999) study are not readily available, one was developed inductively in an interview study and its validity to the myotonic dystrophy population has not been tested. In addition, the instrument tools were not used over time for repeated measurement prior to this study.

There is clear evidence of vertical and horizontal segregation such that disabled employees are disproportionately represented in semi-skilled and unskilled occupations. Stevens (2002) surveyed 120 companies and found contradictions between the appreciable numbers of companies with a published policy on the Disability Discrimination Act and the small numbers of disabled people employed by these companies. Companies were willing to employ disabled people for branch level work e.g. junior administrative, but not develop them for work at higher management levels. The results from this telephone survey are only indicative because the numbers of existing disabled employees were small amongst the four employment sectors (Stevens, 2002).

Segregation was also apparent in a study by Hyde (1998) as organisational decision makers consider disabled employees to be less capable than others to perform tasks that were visible and or critical and as a result did not assign them to these projects (Hyde, 1998). This has an impact on a person's sense of belonging and self-efficacy. Lexell et al. (2017) identified that a feeling of belonging to a context is an important aspect for work continuation even if they sometimes experienced difficulties during the actual work. Their results were based on a small sample and only one participant had myotonic dystrophy. However, it is likely these feelings of inclusion could be representative to many different groups of people with and without a disability.

Some of the studies in the literature (Andries et al., 1997; Fowler et al., 1997; Gagnon et al., 2008; Hyde, 1998; Minis et al., 2009; Stevens, 2002) are over a decade old and the results may be indicative of societal barriers at the time. There is a likely chance that the availability of resources and supports were more limited for people to attain or return to work after their diagnosis / condition progression in comparison to the

current era. It is hoped that acceptance and support for hiring people with disabilities is evolving since these earlier studies.

The literature above demonstrated that factors such as employer attitudes, discrimination, inadequate transportation and support, low self-esteem, inaccessible jobs, and non-accommodating environments are barriers for disabled people when seeking and performing paid work.

2.2.2 Social/ Cultural

Due to the low levels of employment amongst people with disability, poverty and social exclusion are frequently observed social factors (Perron, Veillette, & Mathieu, 1989). It is therefore not surprising that one of the significant risks associated with disability is poverty. It has been suggested that the progressive social deterioration of families with myotonic dystrophy occurs over several generations as more severe forms of the disease manifest (Gagnon, Noreau, et al., 2007). Future generations are therefore born into these environments which leads to further progression of this social deterioration. These factors plus environmental barriers at home, work and in the community; family responsibilities; role of caring for young children or other family members; and available social supports may influence work participation and performance for this group.

Support networks include family, friends, community / disability services, employers, employees, and community groups. In the literature, support provided by family and friends was perceived as an obstacle (Gagnon et al., 2008; Lindsay et al., 2015). This result was surprising as the main source of support for individuals is often their family. The reasons for family support been an obstacle may be due to the genetic element of myotonic dystrophy and therefore other affected family members might not be able to provide the necessary support. Myotonic dystrophy has a profound effect on the families of individuals diagnosed with the condition (Harper, 2009). The sample in Lindsay et al. (2015) was based on youth 16-19-year-olds, none of them had myotonic dystrophy and therefore it was not reflective of the current study population. Due to the sample age in this study, parents may have been more protective and a leading voice in their teenager's decisions. Despite this Gagnon et al. (2008) study was

representative of people with myotonic dystrophy but did not take into consideration the role of family in different cultures.

Gagnon et al. (2008) also identified from their sample that a large percentage of participants were single and lived alone, which further diminished natural supports. This is consistent with a previous study by Cardol et al. (2002) and indicated that a person's social environment can be an important factor in their ability to participate in work. These are important aspects to consider when exploring the potential factors influencing work performance.

In the literature, work environments that were more accepting to support the individual with their work demands yield better productivity and outcomes rates. An important aspect to work continuation was how participants felt they contributed towards sharing knowledge about disability with their colleagues (Lexell et al., 2017). However other people with a known disability may experience discrimination and negative work ethos in the work environment (Saal, Martinez, & Smith, 2014). They may find themselves in a situation where a (potential) employer focuses on their disability rather than the skills and experience they can bring to the job.

The importance of having support from family and good relationships with colleagues, and managers at work are crucial for work retention especially when the disease is progressive (Lexell et al., 2017). This support from family conflicted to earlier studies (Gagnon et al., 2008; Lindsay et al., 2015) however Lexell et al. (2017) only had one participant with myotonic dystrophy in their sample. Problem solving and negotiating first within personal relationships and later in work relationships with the aim to maintain their job was a strategy used by many in a study by Minis et al. (2014). To gather a better understanding of the impact support networks, have on people with myotonic dystrophy in New Zealand, it was important to include this factor in the current study as it may influence work performance and employment status.

Service providers can play an important role in overcoming some of these society and workplace barriers by acting as a coordinate between employers and people seeking work and offering individuals information so they can make informed decisions. Participating in decision making can rebuild an individual's sense of self-determination,

leading to improved participation patterns and overall quality of life (Light & Gulens, 2000). Due to the slow progression of myotonic dystrophy people may need continuous support and input from employers, managers and or vocational rehabilitation therapists to maintain employment (Minis et al., 2010). Community and government services should be committed to identify work performance factors early so they can support the person through the process and obtain work efficiency. Unfortunately, this ongoing monitoring and support throughout a person's career is not currently available in New Zealand.

The location of these studies described above should be considered, as it could make a difference on the employment rate. Cultural differences and acceptance of women and disabled people in paid employment differ in western and non-western countries. In addition, the available supports, and incentives for people with disability to seek employment in these countries may have influenced results.

2.2.3 Socio-economic

Personal factors such as age, gender and condition type were common themes associated with employment in the literature.

Numerous studies indicated that young males with a NMD had an increased chance of being gainfully employed (Andries et al., 1997; Brown & Johnson, 2014; Fowler et al., 1997; Lexell et al., 2017; Madej-Pilarczyk, 2014; Minis et al., 2010; Nätterlund & Ahlström, 2001; Wevers et al., 1993) but the reasons for this age and gender gap is less well known.

In one study the rate of employment for males and females started to drop from 34 years of age. This rate of decline was more rapid for females and continued as they got older (Andries et al., 1997). It was not surprising age correlated with employment status amongst this population group due to the progressive nature of symptoms especially between the ages of 30 – 45 years, which for many people is the prime stage in their career. In comparison employment rates for the general public declined later in life and at a more gradual pace (Andries et al., 1997). Andries et al. (1997) estimated a total career span of 31 years for men and 26 years for women with a neuromuscular disease based on age, gender, and education level. Reasons for an earlier decline in

employment amongst women could include women of a maternal age, who may have taken time off work to have a family; affected women whose children also have myotonic dystrophy and may struggle to participate in social activities over and above looking after their children and family. These figures were an accumulation of results from all four NMDs included in the study. The progression of symptoms and decline in function can vary between each neuromuscular disease and therefore it is difficult to ascertain from this study if the rate of decline in employment was reflective of myotonic dystrophy and not skewed by other NMDs included in the study (Andries et al., 1997).

Fowler et al. (1997) also found that employment decreased as individuals with neuromuscular diseases became older. However, this stage of decline happened at an earlier age for people with a neuromuscular disease than the general population in New Zealand. The Organisation for Economic Cooperation and Development (OECD) reported employment rates for the general public between 55 -64 years of age were 83.6% for male and 72.4% for females (Organisation for Economic Cooperation and Development, 2017). This indicates the general population (people who do not have muscular dystrophy) have longer career spans. Of the 154 people with a NMD interviewed in Fowler et al. (1997) study, 40% were employed at the time the study was conducted and 50% had previously been employed. However, it is not clear if people in employment were more likely to take part in a study of this nature incurring selection bias and if those unemployed had retired or were unemployed but able to work.

Research continues to highlight that disabled people are under-represented in managerial and professional positions and over-represented in lower paid service jobs (Barnes & Mercer, 2005). Barriers to employment exist at several levels including individual, societal, occupational and environmental (Lindsay et al., 2015). However, what we do not know is how this influences work performance for people with myotonic dystrophy in New Zealand and the level of time loss from work due to their health or medical care.

2.3 Occupation

Occupation is defined as a group of self-directed, functional tasks and activities that a person engages in to meet their intrinsic need for self-maintenance, expression, and fulfilment (Law et al., 1996). In this study occupation is referred to as paid employment or work.

2.3.1 Employment Rate

As previously outlined the type of NMD has an impact on employment status. To explore this concept further Fowler et al. (1997) found that people with myotonic dystrophy had lower employment rates than hereditary motor sensory neuropathy (HMSN), spinal muscular atrophy (SMA), facioscapulohumeral muscular dystrophy (FSHD), becker muscular dystrophy (BMD), and limb girdle muscular dystrophy (LGMD) (Fowler et al., 1997). Reasons for this reduced rate of employment in myotonic dystrophy included associated symptoms such as lower levels of motivation and cognition, impaired intellectual functioning, and poor psychological adjustment, (Fowler et al., 1997), which have been clinically linked to myotonic dystrophy type 1 (Meola et al., 2003; Van der Werf et al., 2003). However, this study is over 20 years old and the sample group was ambulatory patients, which is not representative of the true myotonic dystrophy population. Consequently, the validity of these findings can be questioned when applying them to people with myotonic dystrophy.

This correlation of NMD to employment status was strengthened by a later study that showed the employment rate for people with myotonic dystrophy (47.8%) was the lowest amongst two other NMDs (HMSN (63.7%) and FSHD (70.3%)) (Minis et al., 2010). These employment rates were higher than previous studies (Andries et al., 1997; Fowler et al., 1997), which may have been due to advancements in technology and increased supports for people to seek employment over the decade. However a later study by Minis et al. (2012) showed a reduction in the percentage of people working with a NMD from 61% (Minis et al., 2010) to 43% (Minis et al., 2012). Reasons for this decline in employment rates may be due to the study methodology which in the later study was based on a secondary analysis of multidisciplinary team reports. Therefore, it was not purposefully formatted in comparison to the earlier study, which used more than one assessment tool and included multivariate analysis. In addition,

the sample size for the later study was much smaller and taken from only one outpatient clinic in comparison to the 2010 study, which had 591 participants.

In the literature there was limited information on the number of hours people worked and some studies did not take into consideration people who worked less than 8 hours per week (Minis et al., 2010; Minis et al., 2014). This may have excluded some older people and individuals who were working less hours either due to their age, household responsibilities or performance limitations of their condition. The number of work hours will be considered in the present study when deciding inclusion criteria.

As described above the rates of employment are low for people with myotonic dystrophy in comparison to other NMDs. Due to the limited number of studies and fluctuating employment rates it is difficult to predict the employment rate for people with myotonic dystrophy in New Zealand based on current literature.

2.3.2 Education Level & Occupation Type

One factor influencing this lower level of employment amongst people with NMDs was education, which can impact on a person's work and career opportunities. On average employed individuals with a NMD are significantly higher educated in comparison to those not employed (Minis et al., 2010). In the literature people with myotonic dystrophy appeared to have lower levels of education in comparison to other groups of neuromuscular and long-term conditions (Fowler et al., 1997; Minis et al., 2010).

Minis et al. (2010) suggested that employed people with myotonic dystrophy may have lower educational levels due to the multi-organ impact of the condition which also affects cognitive functioning. This finding was strengthened in a later study where less than 15% of participants had college or university degrees and only one fifth did administrative or intellectual work (Peric et al., 2013). The impact of cognitive functioning may explain the lower educational levels and thus employment rates related to the nature of this pathology. However, a limitation in Minis et al. (2010) study was the data selection. Relevant predictive factors such as the environment, terms of employment, social support and work demands that influence employment were not referred to and therefore may not have been considered. Eleven factors were found to influence gainful employment for certain patient groups of NMD.

Although these may be helpful in the development of treatment strategies, only six of these were relevant using multivariate. This explained 42.4% of the variance, meaning 57.6% was left unexplained (Minis et al., 2010). Caution must also be applied when applying Peric et al. (2013) results as ethnicities were not reported and the study was conducted in Serbia, where the culture is different to New Zealand.

Gagnon et al. (2008) identified in their study that people with myotonic dystrophy who had a lower level of education were also at risk of presenting disrupted participation in other activities such as recreation, housing, and mobility. In their study education was a significant factor in univariate analysis of employment as previously shown in an earlier study by Fowler et al. (1997) but not in the multivariate model. As a result, many people attained employment that required lower levels of education such as clerical positions or manual labour. However physically, strenuous jobs are not sustainable for individuals with myotonic dystrophy, leading to people being unemployed at a younger age in comparison to other groups of NMDs (Andries et al., 1997; Kalkman et al., 2005). People who had gained a higher level of education were more likely to be employed in professional type roles which were less physically strenuous. They had more career opportunities and were able to maintain work for longer or have more flexibility in their work environment with structural adaptations and flexibility in work hours (Fowler et al., 1997).

It is important to consider that a person with myotonic dystrophy might get diagnosed in early to mid-adulthood as previously discussed. As a result, they may have chosen a physically strenuous career pathway without having the knowledge that an underlying condition may affect the long-term sustainability of their career. It is worth noting that the study protocol in Gagnon, Mathieu, et al. (2007) relied on self-reported measurements which may be different from objective measurements of participation, especially if the person has cognitive impairment. This difference in self-reported outcomes and measured outcomes were identified in a study by Fowler et al. (1997). There was also a greater number of women in Gagnon, Mathieu, et al. (2007) studies and therefore it was not a fair representation of the true myotonic dystrophy population which affects male and females equally. Their study did not focus primarily on employment, rather it looked at participation in a range of activities, therefore employment was not examined in depth.

The type of occupation, which could be determined by education was also an important factor in work performance. Fowler et al. (1997) found that fewer people with myotonic dystrophy worked in professional, management and technical roles in comparison to people with other neuromuscular diseases. Only 19% of the 41 individuals with myotonic dystrophy were professional/ management / technical workers compared with 42% of the 98 individuals with one of the other five types of neuromuscular diseases (Fowler et al., 1997). A far greater number of people with myotonic dystrophy (80% of the remaining 33 people) were industrial or service/clerical/sales workers, compared with individuals who had a different neuromuscular disease (58% of the 98 people) (Fowler et al., 1997). Individuals who held professional/ management/technical roles remained employed later in life in comparison to the other occupational groups. However, the length of employment in these professional roles were still lower for people with myotonic dystrophy in comparison to people with one of the other four NMDs (Fowler et al., 1997).

In a study by Gagnon, Mathieu, et al. (2007) holding down a paid job was the second most disrupted life activity for people with myotonic dystrophy. They were also dissatisfied with their ability to participate in this activity. People with adult myotonic dystrophy type 1 reported greater levels of disruption in comparison to those with mild myotonic dystrophy type 1, 51.9% to 16.7% respectively. Almost half of the participants (44.5%) either needed human assistance or were not able to engage in employment (Gagnon, Mathieu, et al., 2007). This low level of satisfaction could impact on wellbeing, self-esteem, and confidence to apply for future jobs. This contrasted with a study by Nätterlund and Ahlström (1999) where participants experienced most problems and least satisfaction in activities of mobility and transportation. However, their subjects were composed of a range of muscular dystrophies and not solely myotonic dystrophy. Both studies did not detail the type of employment people held, hours worked and environmental adaptations that may have been implemented.

In comparison to participants with other NMDs a larger number of people with myotonic dystrophy had lower levels of education which may be attributed to the symptoms discussed above. Both condition specific symptoms and education levels are likely the reason for low employment rates amongst this group in comparison to other

NMDs. Based on the above literature one could hypothesize that people with myotonic dystrophy are more likely to be employed if they are younger males with higher education and activity levels and a willingness to adapt to changing circumstances.

2.3.3 Time off work

One of the gaps in the current literature is the impact of myotonic dystrophy on time loss from work. This time loss would have an associated effect on cost of illness. To recognise the indirect and direct cost of productivity loss both presenteeism and absenteeism need to be considered.

A study by Fouad et al. (2017) identified the effect of work productivity for people with a chronic condition. The more severe the condition and quantity of morbidities experienced the greater the level of productivity loss (Fouad et al., 2017; Lenneman, Schwartz, Giuseffi, & Wang, 2011). Workers with chronic diseases were more likely to have increased absenteeism and presenteeism rates at 6.34 and 2.36 times the rate of people with no chronic diseases (Fouad et al., 2017). Presence of multi-morbidity showed more significant increases in absenteeism and presenteeism rates as well as an increased probability in negative critical work incidents (Fouad et al., 2017).

Myotonic dystrophy is associated with co-morbidities and therefore based on the results of Fouad et al. (2017) individuals with this condition may be more likely to have work performance issues and higher levels of time loss in comparison to other NMDs or milder forms of myotonic dystrophy. However findings in Fouad et al. (2017) study were based on a population group of predominantly male dominated employees working in a shipping yard in Egypt. The chronic conditions experienced were similar to secondary symptoms and co-morbidities of myotonic dystrophy such as cardiac and respiratory complaints but none of the participants had myotonic dystrophy. It therefore does not reflect the current study population but instead builds on research knowledge already known.

Time pressures and low job autonomy were unfavourable work conditions reported by a quarter of workers with a neuromuscular disease (Andries, Kremer, Hoogendoorn, Wevers, & Van Putten, 2004). Many of these individuals were physically disabled and had jobs that were physically demanding. These studies did not explore time off work

or measure different domains of work performance and therefore this knowledge remains unknown.

2.4 Conclusion

Potential factors identified in this review that could have an important influence on work performance and employment status have been classified into personal, environmental, and occupational domains. The literature identified that physical and psychological capabilities; work adaptations; age, gender, education; type of occupation and condition; social support, and employer's acceptance can have an influence on employment status, career longevity and work performance for people with neuromuscular diseases.

The implications of living longer with disabilities and progression of symptoms in people with myotonic dystrophy may influence work performance and one's ability to remain in the workforce (Johnson et al., 2010). It is therefore important to gain a perspective of factors affecting work performance specifically for people with myotonic dystrophy. The number of people seeking disability allowance will increase unless there is more investment in resources and support to enhance the work status of people with myotonic dystrophy or NMDs in New Zealand.

Although many studies have investigated the employment situation of people with a neuromuscular disease, very few of those studies have focused on the recent rates of employment, time loss and factors affecting work performance specific to myotonic dystrophy. Previous studies had not exhausted all recruitment options to capture the full myotonic dystrophy population of a country / state and therefore an accurate and reliable employment rate for this condition could not be confided. Additionally, there is a lack of research exploring psychological factors such as the potential influence of mood, self-efficacy, and support, that are likely to influence work performance. More importantly none of the articles focused specifically on myotonic dystrophy and employment. Most studies included more than one NMD and therefore these results could not be generalised solely to the myotonic dystrophy population.

There was no current literature identified from Australasia and it remains unclear how relevant the existing literature is to the New Zealand context. To the best of my

knowledge time loss at work for people with myotonic dystrophy has not previously been explored. To better understand the employment status of people with myotonic dystrophy in New Zealand it is important to identify the current employment rate of this population group in comparison to the overall population. Factors affecting their work performance need to be identified in order to effectively support and facilitate people living with myotonic dystrophy overcome employment barriers in New Zealand.

To address some of these gaps in the literature the aims of the present study were to identify the environmental, personal, and occupational factors that influence work performance for people with myotonic dystrophy and time loss at work due to their condition. It is hoped this study will bring a new dimension to current knowledge for people in Australasia. This information may be crucial when planning supports to help maintain careers and reduced early retirement rates for people with myotonic dystrophy in New Zealand.

2.5 Hypotheses

Based on the literature above the following hypothesis was established for this study:

- Given the low rates of employment for disabled people in New Zealand, it would seem reasonable to predict that the ability to obtain and maintain work with myotonic dystrophy is harder due to the rarity of the condition and progressive rate of deterioration. I therefore hypothesis that the employment rate for people with myotonic dystrophy will be lower than the rate for people with disability in New Zealand.
- Less than 50% of people with myotonic dystrophy in New Zealand will be in paid employment.
- Personal traits (self-efficacy, mood, cognition, physical functioning, pain, fatigue, education, sleep) environmental factors (social, physical, institutional, cultural) and occupational criteria (type of work, education level) will be associated with work performance for people with myotonic dystrophy.
- Time loss from work over a two-week period is likely to be higher than other conditions due to the multifaceted and progressive nature of myotonic dystrophy.

Chapter 3

3.1 Aims

The primary aims of this study were to:

- identify the rates of employment (defined as the number of people in paid employment) in adults diagnosed with myotonic dystrophy.
- determine time loss from employment (defined as the number of half or full days the person has taken off work) in adults diagnosed with myotonic dystrophy.
- explore the factors influencing work performance in adults with myotonic dystrophy.

This is the first known study in New Zealand examining work performance and time loss in people with myotonic dystrophy. This chapter describes the methods employed to achieve these aims; the study design, participants, ethical considerations, outcome measures, data management and statistical analysis for this study.

3.2 Study Design

3.2.1 MD-Prev Study

This Masters project draws upon data collected from a larger study exploring the prevalence and outcomes of genetic muscle disorders in New Zealand (MD-Prev study). The parent study was funded by the Health Research Council. Details of how it was conducted and the data collection instruments have been published elsewhere (Theadom et al., 2019).

Ethical approval for the MD-Prev study was received from the Northern Y Regional Ethics Committee of NZ (Reference: 14/NTB/118 Appendix **Error! Reference source not found.**) and the Auckland University of Technology Ethics Committee (Reference: 14/296 Appendix **Error! Reference source not found.**). Additional ethical approval was not required for the data extracted for this study. Cultural appropriateness for the questionnaire measures were checked by cultural advisors on the MD-Prev team.

In summary, as part of the MD-Prev study all people (including children and adults) diagnosed with a genetic muscle disorder on 01/04/2015 were identified through hospital and medical notes, the New Zealand Neuromuscular Disease Registry, the Muscular Dystrophy Association of New Zealand (MDANZ), disability services and self/family referral. Medical records of the participants were accessed to confirm their diagnosis and thus eligibility to be part of the study. This study was broader than the current study and included many genetic muscle disorders. Genetic muscle disorders were defined as disorders that primarily affected the muscles and included nine types of muscular dystrophy, some types of myopathies (e.g. congenital myopathy), ion channel myopathies and pompe disease. This vigorous process of recruitment meant all avenues were used to reach the myotonic dystrophy community throughout New Zealand.

All identified participants were invited by phone call or letter to complete an assessment about the impact of their condition on their lives and everyday functioning which included, access to healthcare, symptoms, education, community participation, quality of life and impact on relationships (Appendix D). A questionnaire exploring on-the-job work demands (Work Limitations Scale (WLQ-25)) (Lerner et al., 2001) was included as part of the parent study, however, analysis of this measure for people with myotonic dystrophy was not part of the main analyses, so this current study had a unique and novel focus.

The assessments were piloted in advance and estimated to take approximately 90 minutes to complete. This was deemed acceptable as they had been well tolerated in other studies of people with neurological conditions (Theadom et al., 2012). The assessments could be completed in written or verbal forms, with further support provided by the researchers where necessary. People with restricted communication and physical impairments were taken into consideration with the option to complete the assessment in person with a trained researcher or to administer the assessment over several sessions to accommodate fatigue and time constraints. The option of a proxy was also available for people with severe mental or physical impairments. Most assessments were completed with the researcher assistant in person to help facilitate understanding of the questions and to prevent issues of literacy or physical limitations

affecting validity of the data collection. Telephone assessments were also conducted if preferred by the participant or due to logistical reasons to facilitate data completion.

This flexible mode of assessment administration was designed to account for participant preference, enhance recruitment, facilitate data completion/response rate, and increase feasibility if practical difficulties made an in-person assessment too challenging (Bowling, 2005).

To prevent possible selection bias in the outcome assessment, multiple proactive steps that have proven to be effective in previous outcome studies, were undertaken including; i) collecting multiple contact details from referrers to facilitate connection with participants; ii) designing study materials that were engaging and presented in a manner to facilitate completion; iii) explaining the study clearly and concisely so it was meaningful to participants; iv) offering koha in acknowledgement of participant's time (total of \$20 food/fuel voucher following completion of the assessment).

The primary researcher of the current study gathered data for the MD-Prev study in the lower North Island. Other research assistants were employed to conduct assessments in the remainder of the country.

3.2.2 Current Study

Demographic, diagnostic information and questionnaire data for adults (aged ≥ 16 years) who had been diagnosed with myotonic dystrophy type 1 and 2 were extracted from the MD-Prev database for the purpose of this study. This was to enable a specific focus of the impact of myotonic dystrophy on employment status and performance, distinct from the broader focus of the MD-Prev study.

Drawing data from the MD-Prev study helped to enhance the quality of this study through its robust case ascertainment procedures, diversity of recruitment strategies and number of participants involved (where recruitment can be particularly challenging for this relatively rare condition) than if this analysis had been conducted in isolation. This ensured all eligible participants throughout New Zealand were invited to participate in the assessment increasing accessibility and unbiased nationwide representation to minimise the risk of systematic error resulting from selection bias.

3.3 Study Participants

Participants were eligible to take part in this study if they met the following inclusion criteria:

- Living adult (aged 16+) resident in NZ,
- Diagnosed with myotonic dystrophy genetically or clinically by a neurologist,
- Completed the impact assessment on functioning and quality of life from the MD-Prev study.

3.3.1 Recruitment

All participants' who had given written informed consent (Appendix) according to the regulatory and legal requirements for the MD-Prev study and met the inclusion criteria above were included in the current study. Participants were notified that they were free to discontinue their participation at any time and skip questions they did not want to answer. As a result, it was predicted that some participants may not complete some of the questionnaires in full which may impact on the current study results. Basic data (age, gender, ethnicity, type of DM) was gathered on those who did not consent to the full impact study but were included in the prevalence part. This data was included in the current study to identify any potential bias between consenters and non-consenters.

3.4 Measures

This section describes and discusses the assessments used to answer the research question. Measurement instruments used in the MD-Prev study that were likely to be associated with work performance and employment status (based on previous literature) were selected for this study. These measurement instruments and questions can be viewed in the appendices (Appendix F). Data from these measures were extracted to address work performance, self-efficacy, wellbeing and mood, cognition, physical functioning, pain, fatigue, sleep, and social supports. Data from two separate questions in the study were also extracted which identified what changes participants had to make to enable them to continue in their role at work and if having myotonic dystrophy affected their employment in any way.

Demographic data including age, gender, ethnicity, employment status, occupation type and education level for participants with myotonic dystrophy was also extracted from the MD-Prev dataset. As shown in Table 1 mTable 1: Participant characteristics marital status was dichotomised as single or domestic union / married and ethnicity was grouped into four categories. Occupations were categorised under eight groups and are outlined in Table 2.

Results were compared between employed and unemployed participants using a Mann-Whitney *U* test to identify if there was any significant difference that may have influenced employment status or work performance. Available demographic data (age, gender, ethnicity, condition subtype), on those who did not complete the full MD-Prev study was compared with those who did using a chi-square test to identify if there was any significant difference between the two groups. The assessment measures used in this study are described below in further detail.

3.4.1 Work Limitations Scale (WLQ-25)

The Work Limitations Scale (WLQ-25) (Lerner et al., 2001) is an easy to use self-reported questionnaire, measuring the degree to which employed individuals are experiencing limitations on-the-job due to their health problems. The Work Limitations Scale (Lerner et al., 2001) contains 25 items that are categorised into four scales. Each scale score is interpreted as the percentage of time in the previous two weeks that a person was limited on the job in performing specific job tasks (Lerner et al., 2003).

1. The time management scale contains five items addressing the ability to work the required number of hours and scheduling work demands.
2. The six-item physical demands scale covers a person's ability to perform job tasks that involve bodily strength, movement, endurance, coordination and flexibility (e.g. lifting).
3. The mental-interpersonal demands scale has nine items addressing cognitive job tasks and on-the-job social interactions (e.g. concentrating on work).
4. The output demands scale contains five items concerning diminished work quantity and quality (e.g. handling the workload).

Participants rated the 25 items on their level of difficulty or ability to perform the specific job demands in the previous two weeks. Each scale was interpreted as the percentage of time that a person was limited in performance. Scales were scored from 0 (limited none of the time) to 100 (limited all of the time) (Lerner et al., 2001).

Two additional questions were added for the purpose of this study to measure time loss. "In the past two weeks, how many full workdays did you miss because of your health or medical care?" and "In the past two weeks, what was the total number of days you missed part of a workday because of your health or medical care?". A full workday missed was given the value of 1.0 and a part day 0.5. The accuracy of self-reported absence days was established by Revicki, Irwin, Reblando, and Simon (1994). These questions were included to measure the impact of time loss at work for people with myotonic dystrophy, an aspect that to my knowledge has never been previously investigated.

Features of the WLQ-25 (Lerner et al., 2001) that made it suitable for this study were its utilisation in a variety of jobs; it addressed the mental-interpersonal and physical demands which interfere with worker performance; and it was client centered therefore addressing factors important to the worker (Schultz & Edington, 2007). It also demonstrated strong reliability and validity measuring the impact of chronic conditions on an individual's job performance (Lerner et al., 2001).

The WLQ-25 (Lerner et al., 2001) looks at different aspects of work performance to identify some of the key factors that contribute to employee performance and productivity loss. Understanding both the impact of an illness on employee performance and the variables that contribute to this problem can help employers and other stakeholders set priorities for intervention (Lerner et al., 2001). Thus, this study's results may provide insight into work performance and ways to enhance it amongst individuals with myotonic dystrophy.

3.4.2 New General Self-Efficacy Scale

Self-efficacy can be defined as a person's level of confidence in their ability to perform a specific action or achieve a desired outcome (Bandura, 1997). It is the foundation of human motivation, performance accomplishments, and emotional well-being

(Bandura, 1997). Research has found that self-efficacy predicts several important work-related outcomes, including job attitudes (Saks, 1995), competency (Martocchio & Judge, 1997), and work performance (Stajkovic & Luthans, 1998). Myotonic dystrophy type 1 has been associated with a lack of motivation and a decrease in persistence, self-directedness and cooperativeness (Winblad, Lindberg, & Hansen, 2005). These personality traits are associated to self-efficacy and performance which was why this measure was included in the study.

The New General Self-Efficacy Scale (NGSE) by Chen, Gully, and Eden (2001) was developed from the General Self-Efficacy (GSE) scale (Schwarzer & Jerusalem, 1995). The NGSE was chosen for this study due to its high test-retest reliability, unidimensional construction, internal consistency, and stability over time. This contrasted with the GSE scale which was multidimensional and had lower predictive validity.

The NGSE is composed of eight statements. It asked participants to rate their self-efficacy on a five-point Likert scale from strongly disagree (1) to strongly agree (5). These scores were summed to produce a global score (ranging from 8 to 40), with higher scores indicating higher levels of self-efficacy.

3.4.3 Hospital Anxiety and Depression Scale

Individuals with a progressive chronic condition can be vulnerable to mood disorders including depression and anxiety (Buist-Bouwman, De Graaf, Vollebergh, & Ormel, 2005; Härter, Conway, & Merikangas, 2003; Katon & Ciechanowski, 2002; McWilliams, Cox, & Enns, 2003; Scott et al., 2007). This is especially true for people with myotonic dystrophy as the condition is multi-systemic, genetic and typically presents when individuals are working and creating lifelong goals and aspirations. A diagnosis such as myotonic dystrophy can have a devastating effect on a person and their family as they deal with the grief and trauma of their genetic condition. As discussed in the introduction employment has been shown to have a positive impact on emotional, physical, and psychological health for people with and without chronic conditions. It was therefore important to establish if a correlation existed between work performance, employment rate and psychological mood in the myotonic dystrophy population.

The Hospital Anxiety and Depression Scale (HADS) (Zigmond & Snaith, 1983) was used to gather information on the participants emotional mood and wellbeing over the previous seven days. The HADS provides clinicians with an acceptable, reliable, valid, and easy to use practical tool for identifying and quantifying depression and anxiety (Michopoulos et al., 2008).

As its name suggests the HADS was designed for use in a medical practice. However further validation studies have been undertaken in a variety of settings and centres confirming that the HADS performs well in assessing severity and caseness of anxiety disorders and depression in primary care patients and the general population (Bjelland, Dahl, Haug, & Neckelmann, 2002).

The HADS is composed of fourteen questions; seven addressing depression and seven addressing anxiety. Each question is scored on a four-point scale from 0 – 3. Results range from 0-21 for depression and 0–21 for anxiety. Total independent scores for depression and anxiety are rated as normal 0 -7; borderline abnormal 8 - 10; abnormal 11-21.

3.4.4 Cognitive Function and Executive Function Subscales – Neuro-QoL

The Neuro-QoL is a reliable and valid standardised questionnaire to measure health-related quality of life (Cella et al., 2012). It was developed, validated and tested against specific neurological conditions in adult and paediatric populations. Muscular dystrophies were one of these targeted neurological diseases included in the validation process.

The feasibility of the Neuro-QoL (Cella et al., 2012) makes it a preferred option in clinical research specific to neurological conditions. The researcher chose the validated short form cognition test to identify if it was a predictor factor in work performance. Items were selected from the respective item banks to enhance estimation of a patient's health status. It took approximately two minutes to complete which was important to reduce impact of fatigue over the assessment process.

The scale was composed of eight items. Respondents rated the frequency they experienced each statement in the past seven days e.g. "In the past seven days I had trouble keeping track of what I was doing if I was interrupted". These statements were

scored on a five-point Likert scale ranging from one = “never” to five = “very often (several times a day)”.

3.4.5 ACTIVLIM

The ACTIVLIM (Vandervelde, Van den Bergh, Goemans, & Thonnard, 2007) was developed specifically for people with NMDs. It is a unidimensional scale that only measures activity limitations without other characteristics (Vandervelde et al., 2007). The 22-item scale explores difficulty in performing daily activities that require the use of upper limbs or/and the use of lower limbs without support (aids or human assistance). Individuals score their perceived difficulty for each item, on a three-category rating scale: “impossible”/ “difficult”/ “easy”, scored as zero, one or two, respectively. Activities unfamiliar to the participant or not attempted in the last three months are scored as missing (Vandervelde et al., 2007).

The measure was developed based on the RASCH measurement model to enable conversion of ordinal scores to a linear scale. The ACTIVLIM has excellent test retest reliability (0.93), content validity, concurrent validity with the functional independence measure ($p=0.85$) with evidence of responsiveness to change (Vandervelde, Van den Bergh, Goemans, & Thonnard, 2009).

Physical functioning can impact on a person’s functional performance at home, in their community and at work. This measure was therefore included in the study to identify if physical functioning (not specific to on-the-job physical demands, measured by the WLQ-25) was an influential factor in work performance or employment rates for people with myotonic dystrophy.

3.4.6 Short-form McGill Pain Questionnaire

The Short-form McGill Pain Questionnaire (SF-MPQ-2) (Dworkin et al., 2009) measures the major sensory and affective symptoms of both neuropathic and non-neuropathic pain. It has proven excellent reliability and validity and is a recommended assessment tool in clinical research for neuropathic and non-neuropathic pain conditions (Dworkin et al., 2009).

The SF-MPQ-2 (Dworkin et al., 2009) has 22 descriptors describing different qualities of pain and related symptoms. Participants rate their level of pain intensity and related

symptoms experienced during the past week on a numerical zero to ten scale; zero = “no pain” and ten = “worst possible”.

Pain is common among individuals with slowly progressive NMDs and is often a symptom for people with myotonic dystrophy (George, Schneider-Gold, Zier, Reiners, & Sommer, 2004; Meola & Moxley, 2004; Parmova, Voháňka, & Strenková, 2014). The impact of pain can lead to fatigue, isolation and reduced quality of life for people with myotonic dystrophy (Parmova et al., 2014). This could also lead to reduced performance by hindering mood, functional capabilities, and sleep. It was therefore important to identify if there was an affiliation between participants self-reported pain levels and their work performance.

3.4.7 Adult Fatigue Subscale – Neuro-QoL

Central fatigue is one of the main symptoms experienced in myotonic dystrophy (Chaudhuri & Behan, 2004) and is more commonly experienced in myotonic dystrophy type 1 than in other NMDs (Harper, 2009; Kalkman et al., 2005). Fatigue can often be a restricting factor in performance and lead to increased risk of social isolation, depression, apathy and cognition difficulties.

In this study fatigue was measured on the adult fatigue subscale extracted from the Neuro-QoL (Neuro-QoL Executive Committee, 2010). As previously explained the Neuro-QoL (Neuro-QoL Executive Committee, 2010) is a reliable and valid standardised questionnaire for neurological conditions including muscular dystrophies. The adult fatigue subscale is composed of nine statements which asked participants how fatigue impacted on specific elements in their life e.g. “fatigue interferes with carrying out certain duties and responsibilities”. Individuals rated their level of agreement with each statement on a seven-point scale; one being “completely disagree” and seven being “completely agree”.

3.4.8 Pittsburgh Sleep Quality Index

As was previously stated myotonic dystrophy is a multisystemic condition. Weakness and myotonia of the upper airway and other respiratory muscles may be responsible for sleep and breathing disorders in myotonic dystrophy e.g. sleep apnea; daytime fatigue; and hypersomnolence (Hansotia & Frens, 1981).

Excessive Daytime Sleepiness has been referred to as the most frequent non-muscular symptom in myotonic dystrophy (Hilton-Jones, 1997) and could impact on an individual's sleep regulation. Quality sleep is important for optimal performance as it impacts on concentration, mood, health, energy and overall quality of life (Killgore, 2010; Pilcher & Huffcutt, 1996). Due to the association of sleep disorders in myotonic dystrophy it was important to identify its association to work performance.

The Pittsburgh Sleep Quality Index (PSQI) (Buysse, Reynolds, Monk, Berman, & Kupfer, 1989) is a self-rated, effective assessment used to measure quality, disturbances, and patterns of sleep in adults. It differentiates "poor" from "good" sleep quality by measuring seven areas (components): subjective sleep quality, sleep latency, sleep duration, habitual sleep efficiency, sleep disturbances, use of sleeping medications, and daytime dysfunction over the last month (Buysse et al., 1989).

The PSQI (Buysse et al., 1989) is a valid measurement and has obtained internal homogeneity, consistency (test-retest reliability), and validity (Buysse et al., 1989). A global PSQI score > 5 yielded a diagnostic sensitivity of 89.6% and specificity of 86.5% ($\kappa = 0.75$, $p < 0.001$) in distinguishing good and poor sleepers (Buysse et al., 1989).

3.4.9 Multidimensional Scale of Perceived Social Support

Social support was measured due to its importance in building resilience during times of stress and traumatic events. Individuals with a progressive genetic condition such as myotonic dystrophy will experience periods of grief and trauma throughout their life and external support can be important during these times.

The Multidimensional Scale of Perceived Social Support (MSPSS) (Zimet, Dahlem, Zimet, & Farley, 1988) captures the subjective adequacy of social support from family, friends, and significant others. In comparison to other social support scales the MSPSS (Zimet et al., 1988) is shown to be psychometrically sound with good reliability (internal .88 and test-retest .85), factorial validity and construct validity ($r = -.25$, $p < .01$) (Zimet et al., 1988). The MSPSS contains 12 items rated on a seven-point Likert scale ranging from one "very strongly disagree" to seven "very strongly agree" (Zimet et al., 1988).

A hypothesis in this study was that support from family could be different to other support groups (friends / significant others) given the genetic impact of myotonic dystrophy on the family unit. A study by Gagnon et al. (2008) identified that the role of support provided by family and friends, was an obstacle to participation for 21% to 27% of people with myotonic dystrophy. This could be due to the genetic association of myotonic dystrophy being an autosomal dominant disorder. Family members are likely to be affected by the condition, resulting in reduced support from the latter. Due to the influence social support can have on participation (positively and negatively) it was important to consider it as a predicting factor in work performance for people with myotonic dystrophy.

3.5 Data Management

All data received for this study had been anonymised under the conditions of the MD-Prev study, thereby ensuring that the participant's identity remained unknown (Theadom et al., 2019).

No identifying data was shared for the current study. Questionnaire data in the form of an electronic database was only identified by a registration number. All hard copy data was locked in a cabinet and stored in the study office at AUT.

3.6 Data Screening

Data checking was carried out in a robust manner for the MD-Prev study and the extracted data for this study was further examined for potential input errors and outliers using SPSS frequency and distribution tests. The internal consistency, patterns of missing data, skewness and kurtosis and score distribution (to identify any potential floor or ceiling effects) of each outcome was checked prior to analysis to reduce the risk of measurement issues distorting the study findings.

The Kolmogorov-Smirnov test was significant for all subscales of the WLQ-25 (Lerner et al., 2001) and each subscale also had a kurtosis of >3 indicative of a non-normal distribution of the data on this primary outcome measure.

3.7 Data Analysis

Data analysis was performed using SPSS statistical software package version 24 (SPSS 12.0.1 for Windows, SPSS Inc., Chicago, USA).

Questionnaires were scored according to the standardised instructions of each measure. Data did not meet the parametric criteria for normal data and therefore non-parametric testing was completed except for the ACTIVLIM measure which yield data with a normal distribution.

Demographic and diagnostic information were summarised in a table to describe the sample characteristics. Numbers and percentages, median and interquartile ranges or means and standard deviations (depending on the type of data and nature of data distribution) were used. Differences between those consenting to the study and those who did not were determined using a chi-square test.

The second data analysis identified the number (%) of the sample who were in part or full-time employment and their types of occupations. Time loss was analysed on frequency and distribution over a two-week period.

To explore which factors were associated with work performance, data from the WLQ-25 (Lerner et al., 2001) were entered in a Spearman correlation with data on self-efficacy, wellbeing and mood, cognition, physical functioning, pain, fatigue, sleep, and social support.

A Mann-Whitney U test was employed to test for statistical differences between the variable factors (self-efficacy, wellbeing and mood, cognition, physical functioning, pain, fatigue, sleep, and social support) on those employed and unemployed that could influence employment status. The Mann-Whitney U test ranks the medium values of the variables across the two groups and the statistical difference is reported as U .

Qualitative data on the changes made to enable participants continue in their role at work and if their myotonic dystrophy affected their employment in any way was classified into categories using conventional content analysis and ordered in level of frequency (Hsieh & Shannon, 2005).

Chapter 4 Results

This chapter will present the findings from this cross-sectional questionnaire study. Firstly, it presents descriptive statistics on consent rates into the study and describes the participant characteristics and their employment status. It explores the rates of employment in adults diagnosed with myotonic dystrophy in New Zealand. Differences between those consenting to the study and those who did not will be displayed in a table. Secondly, the impact of participants physical and emotional health or medical care on their levels of work performance and time taken off work over a two-week period will be examined. Thirdly, the results of the Mann-Whitney *U* test and Spearman correlation coefficients test will be compared to identify any sociodemographic and condition-related factors that could influence work performance between those employed and unemployed. Lastly, this chapter will report some participant comments on how myotonic dystrophy affects their work performance to augment the quantitative findings.

4.1 Recruitment and Retention

The MD-Prev study identified 343 individuals with a confirmed diagnosis of myotonic dystrophy in New Zealand of whom 327 were adults (> 16 years of age) and invited to participate. There were 202 (62%) participants who completed the full MD-Prev study and their data was included in this study. Figure 2 outlines how this sample was selected from the main MD-Prev study.

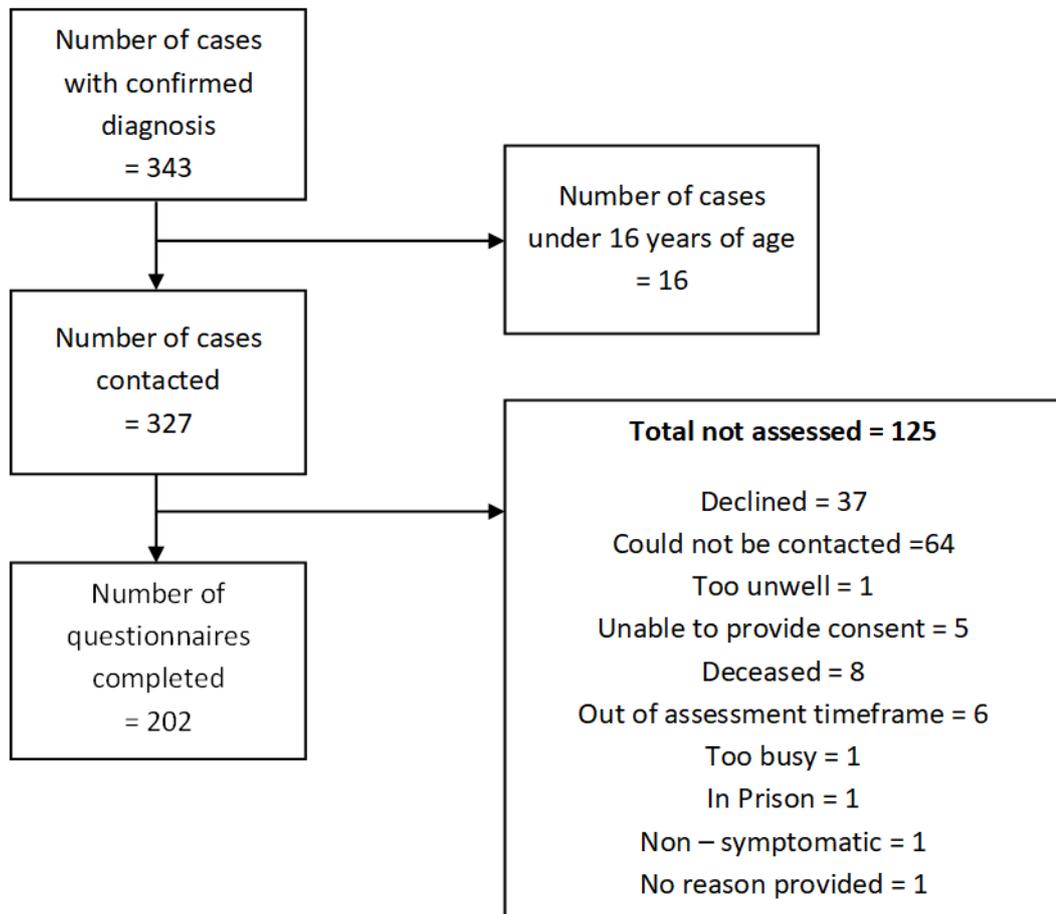


Figure 2: Total participant flow diagram

4.1.1 Sample Characteristics

The sample ranged between 16–77 years of age, with a median age of 48 years (IQR 22). The mean age for male participants was 44 years (SD 14.57) and female 47 years (SD 15.24). Most of the sample were in early adulthood with 57.4% under the age of 50 years. A slightly higher percentage of participants were female, but this difference was not of significance nor was age between the two genders. Most participants were of European ethnicity with type 1 myotonic dystrophy. Comparisons between those who had myotonic dystrophy type 1 and type 2 were not explored due to the small number of participants with myotonic dystrophy type 2. A higher percentage of people who had completed tertiary education were in paid employment (56.5%). The characteristics of the full cohort and the study sample are presented in Table 1 below.

Table 1: Participant characteristics

Participant Characteristics	Consenting Participants n (%)	Non-consenting participants n (%)	Test of difference
Gender			
Male	85 (42.1)	58 (46.4)	$\chi^2 = 0.67$
Female	117 (57.9)	67 (53.6)	$p = 0.41$
Ethnicity			
European	195 (96.5)	100 (80.0)	$\chi^2 = 25.25$
Maori/Pasifika	4 (2.0)	10 (8.0)	$p < 0.0001$
Asian	3 (1.5)	12 (9.6)	
Unknown	0 (0.0)	3 (2.4)	
Condition subtype			
Type 1	190 (94.1%)	121 (96.8)	$\chi^2 = 1.15$
Type 2	12 (5.9%)	4 (3.2)	$p = 0.28$
Education level			
Primary School	6 (3.0%)		
High School	110 (54.4%)		
Polytechnic / College	46 (22.8%) 40 (19.8%)		
University			
At least one Co-Morbidity			
Yes	94 (46.5%) 107 (53.0%)		
No	1 (0.5%)		
Missing			
Marital Status			
In a relationship	111 (55%)		
Not in a relationship	91 (45%)		
Children			
Yes	110 (54.5%)		
No	92 (45.5%)		

4.1.2 Potential Bias

The age range for those who did not consent to the study was 16–89 years. The median age was one year higher than the consenting group at 49 years (IQR 24) and the mean age was 47 years (SD 16.48). The 202 respondents were compared to the 125 non-respondents using a chi-square test and no significant difference was found in relation to age, gender, and type of myotonic dystrophy. As shown in Table 1 there was a significant difference in ethnicity between those consenting and non-consenting with a greater number of Maori, Pacific and Asian ethnicities that did not participate. The unknown ethnicities for those who did not consent may have had a minor influence on the level of significance between the two groups. Data gathered on non-consenters was limited to age, gender, ethnicity and type of myotonic dystrophy. It would have been good to know the difference in employment status between consenters and non-consenters as people in paid employment may not have had the time to participate and thus a significant cohort of non-participants may have been employed.

4.2 Employment Rates

Out of 202 participants, 69 (34.2%) were employed at the time of the study. There was 25 (29.4%) males and 44 (37.6%) females and a greater number of females worked part-time (n=22) in comparison to males (n=3). The mean age in the working group was 44 years (SD12.98). The different categories of occupations are outlined in Table 2 with the highest rate of employment in professional roles. Due to the small sample size the types of occupations between genders were not explored.

Table 2: Employment status

Participant Characteristics	No of Participants	Percentage (%)
Employment Status		
Unemployed	133	65.8%
Beneficiary / unemployed	82	41.0%
Homemaker /carer	21	10.5%
Student	6	3.0%
Retired	22	10.9%
Other	2	0.8%
Employed	69	34.2%
Full time	44	21.8%
Part-time	25	12.4%
Occupation Category		
Professional	19	27.9%
Clerical & Administrative	13	19.1%
Community service	11	16.2%
Trade / Technician	8	11.8%
Manager	6	8.8%
Sales	6	8.8%
Labourer & Driver	4	5.9%
Other	2	1.5%

4.3 Work Performance and Time Loss

4.3.1 Work Performance

68 of the 69 (98.6%) people in employment completed the WLQ-25 (Lerner et al., 2001). The median scores and interquartile ranges for each of the four subscales of the WLQ-25 (Lerner et al., 2001) are shown in Table 3. Health impacted to varying degrees on work performance for most respondents in one of the four subscales (physical, time management, mental-interpersonal, output).

Over 50% of the sample reported no difficulty in time management (56.7%), and work output (55.9%), which indicates some participants are performing well in aspects of their job demands. Work performance was impacted mostly by the physical demands of a job affecting 62.1% of the population. This was followed by the mental-

interpersonal demands affecting 57.4% of those working. These rates of impact varied from mild to more severe. The mean figure below for time management and output demands does not represent that overall fractionally more people struggled to meet the mental-interpersonal demands of their role albeit in milder ways. This indicates that although time management and output are the second and third areas of most difficulty it is not reflective for everyone. The margins between three of these scales were minimal and therefore, all four areas with vigilance to physical demands need to be carefully considered when assessing work performance in a person with myotonic dystrophy. Table 3 outlines the scores for each of the four work demands.

Table 3: Work Limitations Subscale scores

WLQ-25 Subscale	Median	(IRQ)	Mean (SD)	(SD)
Time management	0.00	(16.25)	12.06	(20.54)
Physical demands	20.00	(47.50)	29.85	(34.93)
Mental-Interpersonal demands	2.78	(9.03)	8.29	(13.90)
Output demands	0.00	(15.00)	11.26	(19.99)

WLQ-25 Subscale range 0-100. Difficulty none of the time = 0 Difficulty all the time = 100

4.3.2 Time Loss

Time taken off work within a two-week period due to health or medical care ranged between zero to ten days. 97% of participants employed completed these questions and 73.0 % had not taken any time off work for their health in the previous two weeks. The mean number of days off work over a two-week period was 0.57 days (SD 1.50). It is not known if this was due to factors associated with myotonic dystrophy or if it was due to other health issues. Table 4 below outlines the frequency of time off work over the previous two-week period.

Table 4: Time off work over 2 weeks

Days off work	Frequency	Valid %
0	49	73.0
0.5	4	6.0
1.00	6	9.0
2.00	3	4.5
3.00	3	4.5
5.00	1	1.5
10.00	1	1.5
Total	67	100

4.4 Work Performance Factors

Differences between those employed and unemployed were found through non-parametric testing on sociodemographic and illness-related factors. An exception was the ACTIVLIM, which produced normal data and therefore parametric testing was used for this measure.

The results showed that unemployed individuals experienced greater levels of muscle weakness, respiratory difficulties, and imbalance in comparison to those who were employed. Those who were not in employment had increased levels of pain, depression, fatigue and lower levels of social support, self-efficacy, physical and cognitive functioning.

Support from family, friends and significant others are combined and represent a mean score in tTable 5. The number of individuals who sought support from friends was significantly higher among the working population (13%) in comparison to people who were unemployed (6.3%). This may be due to more friendship networks formed through work relations. The results and test difference between the two groups are outlined below in Table 5above.

Table 5: Differences between participants employment and unemployed

Variable	Total Sample (n=202)		Employed (n = 69)		Unemployed (n=133)		Test Difference
	Mean	(SD)	Mean	(SD)	Mean	(SD)	
ACTIVLIM Patient Measure (-6.59 – 6.27)	2.43	(2.57)	3.81	(2.05)	1.72	(2.53)	t = 6.328, p < 0.000 *
	Median	(IRQ)	Median	(IRQ)	Median	(IRQ)	Test Difference
Global PSQI (0-21)	5.00	(5.00)	5.00	(4.00)	6.00	(2.00)	U = 3973.50 p < 0.136
Mc Gill Pain (0-10)	0.36	(2.00)	0.00	(1.64)	0.55	(2.11)	U = 3580.50 p < 0.015 *
HADS Anxiety (0-21)	3.00	(5.00)	3.00	(5.00)	4.00	(5.00)	U = 4420.50 p < 0.668
HADS Depression (0-21)	4.00	(5.00)	3.00	(3.00)	5.00	(4.00)	U = 2836.00 p < 0.000 *
Neuro-QoL Cognitive Function (8-40)	35.00	(8.00)	38.00	(8.00)	34.00	(9.00)	U = 3132.50 p < 0.004 *
Social Support (1-7)	5.50	(1.17)	5.83	(1.13)	5.42	(1.42)	U = 3437.50 p < 0.007 *
Generalised Self Efficacy (1-5)	3.50	(1.22)	4.00	(1.31)	3.25	(1.38)	U = 2483.50 p < 0.000 *
Fatigue severity (9-63)	50.00	(20.00)	37.00	(20.00)	50.12	(14.00)	U = 1115.00 p < 0.000 *

*Significant difference p < 0.05

4.5 Correlations

To further explore if the differences between those who were employed and unemployed were linked to work performance, a Spearman correlation coefficients test was used. This two-tailed test is a measure of correlation for non-parametrical

data (Field, 2018) which was appropriate for the current data set. As shown in Table 6 higher levels of pain and fatigue were significantly associated with lower work productivity across all four work domains (physical, time management, mental-interpersonal, output).

Depression, cognition, sleep and physical functioning impacted on three of the four work scale demands. There was a significant difference in these factors between those employed and unemployed as outlined in Table 5, except for sleep which was similar in both groups.

Employment status correlated with education, depression, physical functioning, fatigue, self-efficacy, support, and cognition. As previously shown in Table 5 these factors were significantly different for both groups in this study and they impacted on work performance as shown by the Spearman correlation. Age and gender were not significantly linked to work performance nor was age on physical demands. The strongest correlations are highlighted in Table 6 below.

Table 6: WLQ-25 Correlations

	Age	Education	Gender	ACTIVLIM	Sleep	Anxiety	Depression	Cognitive	Pain	Support	Fatigue	Self-Efficacy
Age	1.000											
Education	0.038	1.000										
Gender	0.103	0.032	1.000									
ACTIVLIM	-.448**	.217**	0.018	1.000								
Sleep	0.022	-0.097	0.018	-.338**	1.000							
Anxiety	-0.132	-0.098	.175*	-0.101	.236**	1.000						
Depression	0.089	-0.098	-0.040	-.395**	.260**	.322**	1.000					
Cognitive	0.060	.225**	0.065	.285**	-.377**	-.477**	-.455**	1.000				
Pain	0.021	-0.110	.145*	-.378**	.374**	.294**	.423**	-.391**	1.000			
Support	-0.030	0.046	0.078	.217**	-.143*	-0.002	-.416**	0.134	-.143*	1.000		
Fatigue	0.142	-0.118	0.013	-.485**	.289**	0.145	.391**	-.344**	.341**	-0.136	1.000	
Self-Efficacy	-.173*	.163*	0.038	.625**	-.312**	-.259**	-.543**	.400**	-.349**	.302**	-.399**	1.000
Time Management	0.017	-0.038	0.106	-.492**	.279*	0.186	.549**	-.433**	.419**	-.365**	.603**	-.450**
Physical Scale	0.186	-0.035	0.144	-.442**	0.208	0.050	0.150	-0.106	.315**	.315**	.358*	-0.241
Mental-Interpersonal	-0.134	-0.171	-0.120	-0.112	.358**	.393**	.446**	-.663**	.345**	-0.171	.438**	-0.237
Output	0.089	-.285*	-0.045	-.416**	.443**	.333**	.483**	-.573**	.451**	-0.207	.639**	-.404**

4.6 Participant Experiences

Participants were asked open ended questions as to whether they felt that their myotonic dystrophy affected their employment in any way and if changes had to be made to enable them to continue in their role. Their responses were analysed and categorised into groups.

4.6.1 Work Adaptations

Participants (62%, n = 202) reported that their genetic condition directly affected their employment either by creating difficulty seeking employment or maintaining employment. The main causes were difficulty working the required number of hours and withstanding the physical demands of the job (standing tolerance, tiredness, physical role demands, heavy lifting, speech, and dexterity due to myotonia).

Out of 69 working individuals 46 (67%) of them did not have to make changes in their work to continue in their role at present but many were mindful that they may need to make adaptations in the future.

“I am going to have to find a less physical job fairly soon. Currently, I get very tired and I know that as my condition gets worse, I will not be able to continue my current job.” (Male 35 years)

There were 17 (25%) participants who disclosed the type of adaptations made to their work to enable them to continue in their role. The most common reported change was a reduction in work hours mainly due to fatigue and daytime sleepiness.

“I have had to reduce my hours and responsibilities due to reduced energy (fatigue) and motivation.” (Female 49 years)

Flexibility in job arrangements enabled individuals to work from home and manage their workload to suit their needs. This was one of the most beneficial changes allowing people to remain in their job for longer. Low-cost, simple modifications such as flexible working conditions and support, supportive managers, and greater understanding from colleagues can enable a person with a disability remain in employment (Employment New Zealand, n.d).

Some participants had to find new ways to complete their job and problem solve to overcome barriers. The highest reported adaptations were to the physical

environment. Changes included the provision of seating, specialised equipment (raised toilet seats, magnifying glasses), mobility aids and reducing physical tasks.

Individuals who were self-employed explained it was easier to tailor the job to their needs by working from home, having flexible work hours, and adapting their environment. However, financial sustainability and the long-term security of being self-employed was an issue as they had to seek additional help or sell their business when their condition progressed.

4.6.2 Symptoms

Speech disturbance is a symptom of myotonic dystrophy (slurred speech, jaw lock). Vocal communication is an important skill in many work environments even to get through the interview stage. These speech impediments were perceived as a barrier to seeking employment for some participants.

Myotonia, is the classic symptom of myotonic dystrophy and affects hand coordination and dexterity. Loss of hand function was an issue for those working and a barrier to work for others.

"I can get a job, but my hands lock so it is difficult to do things."
(Female 31 years)

"I cannot do jobs requiring dexterity and strength or things that require a certain amount of physical activity. I am unable to stand for long periods of time or pick up objects." (Male 38 years)

Fatigue was also perceived as a barrier to finding employment by individuals who were not working.

"The excessive tiredness of my condition impacts on my ability to be a reliable employee." (Male 42 years)

4.6.3 Work Culture

As per the policies and legislation mentioned earlier employer expectations and employees' needs should be considered to supply adequate provision of support, as necessary. This was not always the case as one participant stated

“If I make or ask for changes, they will ask questions. I am currently managing most of the time at work.” (Female 61 years)

An inclusive, respectful working environment is favourable. A small percentage of participant’s employers were aware of their limitations and did not expect them to do work that was too demanding. Support from colleagues was also important for some participants to help them overcome physical work challenges.

People who were no longer able to meet demands in their role struggled to seek new employment due to their condition or lack of experience in other fields. Society acceptance and prejudices of living with a long-term disability were perceived as an employment barrier. Previous work experiences led to feelings of rejection, lack of confidence and self-esteem to apply for future jobs.

“I feel employers do not want to hire me as my condition may affect how I work. When I told them about my condition, they stopped giving me work. This happened on two occasions. It knocked my confidence and self-esteem.” (Male 21 years)

As a result of these experiences and difficulties maintaining work, many participants sought early retirement or discontinued work from an early age (before the age of 50). Some never achieved work status due to perceived or environmental barriers and past negative experiences trying to secure a job.

4.7 Summary

This chapter presented the finding from this cross-sectional questionnaire study. It identified characteristics of people with myotonic dystrophy working and living in New Zealand and described the types of occupations most frequently sought in this cohort. It was revealed that rates of employment were low amongst people with myotonic dystrophy and that many in employment experienced physical limitations that affected their work performance and career sustainability. Fatigue, pain, cognition, and mental wellbeing were variables influencing the level of work performance and this was supported with respondents’ narratives as to how myotonic dystrophy impacted on their employment. Environmental barriers and enablers to work were also identified through participant narratives. Interestingly the average amount of time off work was low over a two-week period.

Chapter 5 Discussion

This chapter will discuss the results of the study and compare them to previous literature. It will examine limitations to the study design and methodology, and the potential effects these may have on the findings. Finally, it will contemplate how these results can enhance vocational support for people with myotonic dystrophy and opportunities for future research on this topic.

The primary purpose of this cross-sectional study was to explore the rates of employment and to identify the factors influencing work performance in adults diagnosed with myotonic dystrophy. The employment rate for people with myotonic dystrophy was significantly lower at 34.2% in comparison to the national rate at 45% of all disabled adults employed in New Zealand (Stats NZ Tatauranga Aotearoa, 2014).

This study identified that the physical demands of a job had the greatest impact on work performance for people with myotonic dystrophy. This was mainly due to symptoms of fatigue, functional limitations, and pain. There was a significant difference in cognition, depression, self-efficacy, and physical functioning between the two groups (employed and unemployed) and these factors were also shown to influence work performance.

The People Environment Occupation (PEO) model (Law et al., 1996) outlined in the introduction will be used to frame this discussion.

5.1 Personal Factors

5.1.1 Health / Physical Performance / Role

Research has shown that a person's type of impairment can impact on their ability to engage in paid employment (Stats NZ Tatauranga Aotearoa, 2014). The multisystemic nature of myotonic dystrophy means individuals in this study had a wide variety of symptoms including agility impairments, sight, speech, and cognitive impairments that could impact on employment. People who were employed had a higher level of physical functioning in comparison to those unemployed. Physical limitations e.g. standing for periods of time (>20 mins), muscle weakness, slurred speech, fatigue, and an inability to lift and carry items were some of the main reasons people ceased

employment or were having difficulty at work. Due to the multisystemic nature of the condition, participants often had more than one complexity and those who did were more likely to be unemployed or have lower levels of work performance.

Although over half of the participants (66%) reported that their genetic condition had directly affected their employment at some stage, those who were working appeared to be performing well. Albeit the numbers employed were small.

Fatigue was found to be the most debilitating factor in work performance. Fatigue was identified in a high proportion of those employed and unemployed. Higher levels of fatigue were associated with poorer work outcomes and performance. Fatigue levels were greater amongst the unemployed group and was the most cited reason in the participant narratives of how the condition affected their employment. Previous literature supports this finding that fatigue is an important variable affecting individuals everyday lives with myotonic dystrophy (Gagnon et al., 2008; Heatwole et al., 2012; Minis et al., 2010). This study adds to previous work by revealing its impact on work performance.

Fatigue impacts on a person physically, psychologically, and socially regardless of having a disability (Craig, Tran, Wijesuriya, & Boord, 2006; Williamson, Feyer, & Friswell, 1996). In the literature people with other NMDs also had high levels of fatigue but continued to maintain employment except for those with myotonic dystrophy (Gagnon et al., 2008; Minis et al., 2010). The multisystemic impact of myotonic dystrophy and cognitive changes associated with the condition may be a reason that participants in this study struggled to manage the impact of fatigue on work performance. Thus, treating this symptom (independent variable) in therapy may have a knock-on effect on many performance areas such as cognition, mood, physical functioning, pain, sleep, and self-efficacy.

Understanding the levels of fatigue between various NMDs may explain why certain groups can still perform well despite having high levels of fatigue and lead to timely interventions for other groups. To gain a greater understanding of the impact of fatigue on people with myotonic dystrophy it would be good to measure it through the course of a week or a month rather than a point in time. Identifying periods when people are less fatigued and thus more productive could increase their ability to

manage workload and lead to better performance. This ongoing assessment of fatigue could be a tool used in future therapy.

5.1.2 Sensory Capabilities

Pain is a common symptom in myotonic dystrophy and can have a direct impact on functional performance, mood and cognition (Chapman & Gavrin, 1999). The literature supports my findings that pain may be in part, related to fatigue (Jensen et al., 2008; Stanos & Houle, 2006) as pain and fatigue were the only measures that correlated with all areas of work performance – time management, output, interpersonal-mental and physical. Pain and fatigue also correlated with an increase in the number of problems with physical functioning, mood, sleep, and cognition. In this study pain was assessed over a short time frame (within a week) and thus a broader knowledge of the severity, type (affective/sensory), location of pain and longevity would provide greater insight into its impact on work performance.

5.1.3 Psychological Attributes

The emotional and mental impact of unemployment appears to be greater among those with disabilities (Turner & Turner, 2004). This study recognised that a person's psychological functioning can impact greatly on their work performance and ability to seek and maintain employment. The relationship between occupation and health as described by Wilcock (1999) can be seen in this study. Those who were engaged in an occupation (employment) had higher levels of wellbeing in a social, mental, emotional, and physical way. The higher incidence of depression amongst the non-working population correlates to the importance of "doing" and "being" in occupations, which can lead to higher levels of mental stability (Wilcock, 1999).

Despite the results showing a significant link between depression and work performance it is important to note that the data are slightly skewed. Only a small number of participants scored within the clinical depression range, and most of the sample fell within the normal range. Thus, depression and anxiety may only be an issue for some people with myotonic dystrophy and when experienced can have an important impact on functioning. This suggests that those affected may benefit from tailored intervention and support. Consequently, it is important that mental wellbeing is assessed and monitored for people with myotonic dystrophy throughout their

lifespan and a referral made if necessary, to a medical practitioner or health care professional as levels of depression and anxiety are likely to change over time. For example, clinicians need to be aware that people with myotonic dystrophy may be especially susceptible to symptoms of depression and anxiety when their condition progresses. Ongoing monitoring will ensure adequate support and intervention is provided in a timely manner to limit the long-term impact on work performance and employment status.

5.1.4 Cognitive Capability

There was a significant difference in self-reported cognitive ability between those employed and unemployed which indicates it may be a perceived or actual barrier for some when seeking paid employment. Myotonic dystrophy is one of the few muscular dystrophies that affect the brain and can cause changes in cognition over time (Meola & Sansone, 2007). Individuals with an adult onset of myotonic dystrophy type 1 commonly experience daytime sleepiness, fatigue, executive and visuospatial dysfunctions, and anxious personality traits (deteriorating with age) (Meola & Sansone, 2007) which may lead to early and sudden retirement. This reinforces the importance of having access to periodic cognitive assessments in addition to the traditional physical and medical assessments.

Cognition can impact aspects of functional performance (Velligan et al., 1997) and in this study, it impacted on work performance in the areas of output, time management, and mental-interpersonal demands. Cognition correlated positively with education and employment and its presence may be a reason that people with myotonic dystrophy have lower rates of employment in comparison to other NMDs that do not have an association with cognitive function. Those who had reduced cognitive functioning may have struggled to cope and adapt to physical challenges in comparison to others who were better able to problem solve and seek appropriate support/resources.

Adequate testing and monitoring of cognitive abilities at an early stage of diagnosis could help identify any concerns. This early intervention and therapy could lead to higher levels of education and increased employment opportunities for some people. It is recommended that people with cognitive complaints be referred to a mental healthcare professional for testing and follow up (Ashizawa et al., 2018). These

complaints should be looked at seriously and appropriate support and therapy provided to enhance the person's ability to manage any difficulties and reduce the potential impact on quality of life, work performance, and career opportunities.

5.1.5 Self-concept / Cultural background / Personality

A job creates a sense of identity and purpose (Unruh, 2004) and it is an important activity throughout one's life regardless of their NMD (Minis et al., 2014). Higher levels of self-efficacy were comparable to higher levels of physical functioning in this study. Those who had a greater sense of belief in their capabilities were better able to perform activities of daily living. Although increased self-efficacy is likely to be a by-product of performing an activity successfully it may also be a precursor to do the activity.

Based on these findings in the personal domain of the PEO model (Law et al., 1996) it is not surprising that a person's functional capability correlated to their employment status and performance. Characteristics of the condition and their personal capabilities had an impact on most people either seeking work or their ability to meet work demands. Fatigue and pain impacted the most on work performance, followed by cognition, depression, physical functioning, and sleep, (which could be classified as dependent variables). These factors were previously identified to have a significant effect on the lives of people with myotonic dystrophy type 1 (Heatwole et al., 2012) and this study identifies more specifically how they relate to work performance. Targeting therapy for pain and fatigue may help enhance the outcome of other factors in the personal domain that are compounded by the impact of pain and fatigue.

Increasing the number of disabled people into employment is not only the role of the individual preparing for work, it is also the role of society and the work environment accommodating them (Weisenstein & Koshman, 1991). Therefore, to fully understand work performance, the interaction between the person and environment will now be explored. When there is a person-environment fit in supporting the valued occupation, success in occupational performance eventually leads to participation and wellbeing (Baum, Christiansen, & Bass, 2015). The environmental factors included in this study were physical, cultural, institutional, social, and socio-economic.

5.2 Environmental Factors

5.2.1 Institutional / Physical Environment

The literature identified that a range of physical and technological modifications can support a person to remain in work and/or enhance their work performance (Padkapayeva et al., 2017).

In this study participants reported that changes to work schedules and demands were of most benefit. Additionally, flexible work hours and the ability to work from home were major advantages in maintaining employment. These environmental changes helped people to manage the impact of fatigue, pain, and cognition on work performance. Participants who did not have to make changes to their workplace, were mindful they would need to consider this as their condition progressed. Support to make adaptations in their job or seek alternative occupations would reduce the occurrence of early retirement when they were no longer able to perform the demands of their current job but wanted to maintain a career. For this to happen people need to feel supported and safe to disclose their condition and have a good relationship with their manager and colleagues (Lexell et al., 2017; Minis et al., 2014). These findings support those identified in the literature review that specific workplace accommodations such as changes in work schedules, work organisation ethos, and help from others promoted employment amongst people with physical disabilities (Nevala et al., 2015).

Studies in the United States have found that interventions to preserve employment are more successful than those aimed at returning people to work (Brown & Johnson, 2014). This finding was reflected in the current study data. Those who were no longer employed spoke about the difficulties in finding alternative sources of employment. In addition, their self-efficacy to obtain work had diminished. Therefore, early support and intervention in the workplace are critical to prevent early retirement. The supports people sought to maintain, or gain employment were not explored in this study and should be considered in future research studies.

5.2.2 Social / Cultural Environment

The participant narratives provided mixed experiences of disclosing information about their condition to an employer. Support from employers and colleagues and their

attitudes and understanding of the persons needs were key factors for some people in maintaining their job. However, the disclosure of their condition was a barrier for others who felt they would be confronted by their ability to perform and thus continued to hide their concerns and learnt to adapt to new challenges. This was also reflected in the literature that people with a neuromuscular disease strived to stay employed without disclosing their illness to avoid risk to their employment (Andries et al., 1997; Heatwole et al., 2015; Minis et al., 2009; Minis et al., 2014). The negative attitude of employers was also an area of concern for some in seeking employment in this study. Further research could explore employers' perspectives of supporting people with myotonic dystrophy at work.

The New Zealand Government recognise the impact of social prejudice on employment (Minister for Disability Issues, 2001; Ministry of Social Development, 2016; Office for Disability Issues, 2014, 2016b). Their priority is to increase the number of disabled people, including long-term unemployed disabled people, into paid employment and self-employment on an equal basis with others. The government plans to increase the number of employers who are confident in employing disabled people through paid internships in the public sector (Office for Disability Issues, 2016b). Giving employers support and information on the benefits of employing disabled people and demonstrating how to support disabled people in the workplace as their condition deteriorates may address some of the long-standing stigma. Applying positive discrimination policies within the work environment should also be enforced.

An interesting finding in this study was the difference in individuals social support due to their employment status. Those employed had significantly more support from friends than those who were unemployed. This may be due to more opportunities to forge networks through work interactions. Greater levels of support impacted positively on depression but did not correlate with mental-interpersonal demands of work. However, it did impact to a lesser degree on time management and physical work demands, which may be reflective of support from colleagues to meet these demands and responsibilities. Support from family members and a good relationship with colleagues and managers were important in a previous study to obtain and continue in work (Lexell et al., 2017).

People with chronic conditions usually want to remain active and productive. However, the progressive episodic nature of many conditions practically guarantees that workers will experience employment problems at some stage (Lerner et al., 2003). In this study over half of the participants recognised their condition affected their employment. Access to vocational therapists at an early stage could help alleviate some stress for the employer and employee by implementing timely strategies and adaptations to the work environment to enhance the PEO fit.

In the narratives, some unemployed participants expressed their desire to work but faced difficulties in looking for jobs to suit their capabilities. These limitations were less prevalent amongst those who had a job, and overall those who were working appeared to be performing well. A smaller percentage had difficulty performing some aspects of the job however, these limitations did not necessarily translate into a need for modifications to the workplace or aids/equipment. This reflects the 2013 New Zealand disability survey where only a small proportion of disabled people needed such modifications (Stats NZ Tatauranga Aotearoa, 2014). Education, and awareness of the impact myotonic dystrophy has on employment and work performance is needed to achieve better work outcomes for this population group.

5.2.3 Socioeconomic

Factors such as age, gender, and ethnicity may compound the disadvantages associated with seeking employment with a disability (Andries et al., 1997; Brown & Johnson, 2014; Fowler et al., 1997; Lexell et al., 2017; Minis et al., 2010). This study is different to findings from other studies (Andries et al., 1997; Fowler et al., 1997; Madej-Pilarczyk, 2014; Minis et al., 2010; Nätterlund & Ahlström, 2001; Wevers et al., 1993) in that age and gender were not found to significantly influence employment status. A greater percentage of females to males were employed, which differed from the literature (Andries et al., 1997; Wevers et al., 1993). This may be due to a larger percentage of females participating in the prevalence study from which the data was extracted. Another explanation is an increase in the profile of woman creating long term careers since the 1990's, which was when these previous studies occurred. The impact of age on work performance was not apparent in this study. This was interesting as one would imagine the older one got the more physically demanding the task became. A possible explanation is the age of this study group whom were

relatively young and over half the number of people unemployed were between 18 - 65 years of age. In addition, people with myotonic dystrophy tended to retire earlier in their working careers and thus the impact of condition progression rather than ageing factors impacted more on performance. These results show condition related factors were more influential to work outcomes than sociodemographic factors such as age and gender.

Despite reaching out to a variety of ethnic groups Maori, Pacific and Asians were underrepresented in this study. The main reason was difficulty contacting individuals in these ethnic group while others declined consent. Consequently, the data may not be generalisable to these ethnicities. The method of recruitment through District Health Boards and disability organisation's may have missed some ethnic groups that do not engage in the public health sector and disability services. Thus, future studies should look at other options to increase connectivity with these minority communities.

Creating a supportive work environment that provides an opportunity to grow and develop despite having a disability is key (Lingard & Francis, 2006). Work environments that are flexible to employees needs and adaptable to alternative ways of working e.g. remote working, flexible work hours, and ergonomic assessments may enable more people to remain in employment for longer. The focus should be placed on how we can support this population group overcome the environmental, societal, and perceived challenges so they are given opportunities to participate in the daily occupation of work.

5.3 Occupational Factors

Occupational factors refer to characteristics of seeking and maintaining employment. The interactions of a person's capabilities in combination with the environment can have a positive or negative influence on occupational performance (Baum et al., 2015). This section will discuss this relationship in terms of employment rates, types of occupation and their relationship to education levels and time off work.

5.3.1 Employment Rate

The study findings support the primary hypothesis that the ability to obtain and maintain work with myotonic dystrophy is harder than other disabled groups. The

predicted rate of less than 50% of participants would be in paid employment was accurate. In New Zealand 327 adults were identified as living with myotonic dystrophy, and 87.46% of these were adults of a working age (18 to 65 years old). Despite this only 32.4% of the sample (n=202) were employed which is lower than the national rate of 45% for all disabled adults or 61% of disabled people under 65 years of age (State Services Commission, 2016; Stats NZ Tatauranga Aotearoa, 2014).

In comparison to previous literature the rates of employment in this study were marginally higher than Fowler et al. (1997) study and significantly lower than Minis et al. (2010) study. However, what was similar was that people with myotonic dystrophy had lower rates of employment in comparison to other types of NMDs (Fowler et al., 1997; Jensen et al., 2008; Minis et al., 2010). This implies people with myotonic dystrophy are at a higher risk of unemployment and thus more susceptible to the secondary offset of this such as lower socioeconomic rates, education, financial instability, loss of self-worth and poverty.

Reasons for this low employment rate in comparison to other disability groups may be due to the physical and cognitive impacts of myotonic dystrophy and a lack of awareness about the condition and resources to support these people into employment. This study has highlighted the significance of cognition, pain, and fatigue on work performance, and thus resources and support to enhance work opportunities should take these factors into consideration.

Disabled people are not only less likely than non-disabled people to be in employment, but if employed they are more likely to work part-time (fewer than 30 hours a week). In 2013, 30% of employed disabled people worked part-time, compared with 22% of non-disabled people. The rate of part-time employment was particularly high for disabled women, of whom 42% worked fewer than 30 hours, compared with 19% of disabled men (Stats NZ Tatauranga Aotearoa, 2014). Comparable to the current study 3.5% of males worked part-time in comparison to 19.1% of females. The rate of females in part and full-time work were equal but there was a significant difference in males working part and full-time. Exploring reasons for this low part-time rate in males may indicate opportunities to progress from full to part-time employment and provide career longevity rather than early retirement as their condition progresses.

5.3.2 Occupation Type and Education

Disabled workers have a similar pattern of occupational distribution to non-disabled workers, with the largest concentrations being in professional and managerial occupations. However, disabled workers were less likely than non-disabled workers to be either professional (19%, compared with 25%) or managers (16%, compared with 19%). This study corroborated with the 2013 Disability Survey (Stats NZ Tatauranga Aotearoa, 2014) that more women worked as clerical and administrative workers, professional or managerial roles and more men worked as labourers, or machinery operators and drivers. The figures for manual labour were lower in this population group than the larger disability group likely due to the physical limitations of myotonic dystrophy. This variation in careers between genders may explain why the employment rate and option of part time work was higher in females as they chose professions, which were less physically strenuous or with more flexibility.

In this study physical demands in a job were reported as most challenging. Some of these could be minimised with the correct support, intervention, and equipment such as environmental adaptations or flexible work conditions. People diagnosed at a younger age are able to make career choices that are less physically demanding and more sustainable to their long-term needs. Early career guidance is important for this group. Time management in work also had an impact on performance. These challenges could be overcome with flexible and innovative ways of working which is one of the prime reasons people with a disability are able to work (Equality and Human Rights Commission, 2012). Flexible work hours and working from home could be one option for this population group. The performance level of those working in this study should be encouraging for people who are willing to seek employment. It indicates that with suitable environmental conditions and support a paid occupation could be pursued.

Education was a significant factor in determining rates of employment in this study. Low levels of education correlated with lower employment rates. Even if a significant disability is present those with a higher level of education have an increased chance of employment (Madej-Pilarczyk, 2014). People who engaged in tertiary education were more likely to secure jobs and maintain paid employment into the future. This may be due to a larger range of suitable jobs available following tertiary education such as

professional and managerial roles. As education has been found to have a strong correlation with future opportunities in the labour market (Minis et al., 2010), advice on education and job counselling should be provided timely and appropriately after the diagnosis.

5.3.3 Time Off Work

This study is different from other studies on myotonic dystrophy and employment as it looked at time loss from work due to health issues. In the current study, people on average took 0.57 days off work over a two-week period. This was lower than the mean number of work absences over a two-week period for people with depression, rheumatoid arthritis and dysthymia which was 1.1 days (Lerner et al., 2004). Some absences were due to people taking sick leave up to three days which could have been due to health issues not associated directly with their myotonic dystrophy. It is however fair to say that absenteeism was relatively low in this group, which will be of interest to employers.

It was hard to compare these results to national statistics as they were measured over a short time frame rather than a 12-month period. We know that the average rate of absence over a year in 2016 was 4.4 days per employee, compared with 4.7 days in 2014 and 4.5 days in 2012 (Business New Zealand and Southern Cross Health Insurance, 2017). However, there are relatively few sources of information available on the number of days away from work due to illness and injury in a New Zealand context and none that compare time off work for people with and without a disability. This study did not address the reason for time off work but things to consider would be caring for an unwell family member or dependent and attending medical appointments. These were the most common causes of work absences from a New Zealand survey in 2016 (Business New Zealand and Southern Cross Health Insurance, 2017).

People also reported that they would need to reduce their work hours as their condition progressed. Difficulty in maintaining work performance for the set amount of time could be a risk for higher absenteeism over a longer time frame. Disclosing their condition to their employer can support early intervention and timely

adjustments to work issues which may help reduce this risk of absenteeism and presentism as their condition progresses.

5.4 Summary

This study adds to what we already know by identifying that people with myotonic dystrophy who are in paid employment are performing relatively well but a concern is that employment rates are low for this group. In addition, work performance is most disturbed at the physical level. This is not surprising given that a main symptom of myotonic dystrophy is progressive muscle weakness and fatigue. My findings point to problems directly caused by myotonic dystrophy that lead to reduced work capacity including fatigue, pain, depression, and cognition as well as problems at an environmental level such as a lack of support at the workplace or the need for workplace accommodations. These variables also correlated with work performance and should be considered by employers, work agencies, vocational therapists, medical professionals, and people with myotonic dystrophy when seeking or maintaining a job.

5.5 Limitations and Strengths

There are several strengths and limitations to this study, which must be considered. This is the first known study in Australasia to examine factors influencing work performance, employment rates and time loss in people with myotonic dystrophy. Previous studies focused on the influence of physical functioning and demographic data rather than a holistic approach. These earlier studies were mainly conducted in the Netherlands and America between the 1980s – 2000s which is not comparable to the current work economy and cultural society in New Zealand. There were very few studies that focused specifically on employment in individuals with myotonic dystrophy. Many included two or more NMDs in their sample reducing the specificity and depth of the results.

This study has helped to highlight the working needs of people with myotonic dystrophy. This will hopefully feed into some of the New Zealand Disability Action Plan 2019 -2022, which is currently in development following a review of the 2014-2018 action plan (Office for Disability Issues, 2018). This action plan is derived from the New Zealand Disability Strategy 2016 – 2026 (Office for Disability Issues, 2016a) and is a set of actions that the government commits to progress over the next four years of which

employment is one of their priorities (Office for Disability Issues, 2018). Information gathered in this study will help support and empower people to engage in employment and tailor vocational rehabilitation programmes specific to the needs of the myotonic dystrophy population.

Providing the study interviews by phone and in paper allowed people the opportunity to complete them at a time convenient for them. The main difference between the data collection methods was time; face-to-face interviewing took longer since it included more social interactions. Young and Murphy (2002) found that there were very few differences in the responses when comparing face-to-face interviewing and telephone interviewing (Young & Murphy, 2002). All participants seemed to answer honestly with more quantitative information collected in face-to-face meetings. The trustworthiness of this study has been considered by using quotes to illustrate the perception of the participants.

This cross-sectional study extracted data from a larger national study on the prevalence and impact of genetic muscle disorders (MD-Prev) funded by the Health Research Council of New Zealand (Theadom et al., 2019). This process of data gathering enhanced the quality and credibility of the proposed project through its robust case ascertainment procedures, diversity of recruitment strategies and number of participants involved. It ensured all people identified with myotonic dystrophy in New Zealand were invited to participate and enabled comparisons between consenters and non-consenters to be explored. The participants in this study included both male and female with a wide variety of ages and professions from across New Zealand. This strengthened the credibility of the study since it is representative of the myotonic dystrophy population. However, caution should be regarded in generalising these results to other populations and ethnic groups as outlined below.

Despite engagement with Spasifik and Mana health publications, culturally specific health providers and employment of a cultural liaison officer, people of Asian, Pacifica, and Maori ethnicities were still underrepresented. Therefore, the results of this study need to be applied with caution when considering these ethnic groups. Capturing a wider range of ethnicities in future studies will be more reflective of the myotonic

dystrophy population in Australasia and allow comparisons to be made between these groups on employment rates and types of occupation.

A strength of this study was the range of measures utilised. These captured a wider perspective on how myotonic dystrophy affects a person physically, mentally, socially, and functionally. However, one of the drawbacks was that the questionnaires were not designed specifically for the myotonic dystrophy population. The questions had to apply to a range of neuromuscular diseases and were therefore more generic. Questions specific to symptoms and management of myotonic dystrophy would have added depth to the results. It would have strengthened the findings that the issues identified were specific to myotonic dystrophy given the physical impact was measured by a generic measure of functioning, the ACTIVLIM. Most of the data collected in this study was not normal, restricting the analysis that could be undertaken.

The scope of this study to ask in-depth questions about work adaptations was limited as the analysis of work performance was not part of the main analyses, in the parent study. As an occupational or vocational rehabilitation therapist it would be good to understand peoples experience in getting a job, years of work experience, employer and co-worker support, and reasons for ceasing work and retiring early. A qualitative methodology approach would have provided more insight into the meaning of work and identified more specific employment issues and factors impacting on work performance for this group.

Although the participation rate was high, people in paid employment may not have had the time to respond to the questionnaire and thus a significant cohort of non-participants may have been working. Volunteer work and students were not included in the employment section. Some participants were in caring roles for family members or volunteered time in community services after ceasing paid employment. Not including these participants narrowed the scope of the findings.

The population group was not defined by location which meant I was unable to ascertain if employment opportunities, social exclusion and isolation were a barrier to employment for those living in rural areas. It would be good to explore employment

rates by location and include access to transport as this could impact on employment opportunities if accessible transport is not readily available.

This study was a snapshot in time with many questions referring to the previous week or fortnight. Although this increased the reliability of recall a longitudinal study would create a better understanding of the performance rate of individuals with myotonic dystrophy at various stages throughout the year. Environmental conditions such as climate can have an impact on symptoms and thus functional performance.

5.6 Future Practice

This study adds to the knowledge already known about employment amongst people with myotonic dystrophy by looking at the impact of a range of possible factors on different aspects of work performance. Unique to this study is it offers an understanding and insight into how people with myotonic dystrophy in New Zealand are performing. Therapists and employers may use this information when enforcing the New Zealand Disability Strategy 2016 – 2026, where a priority is to increase employment amongst people with a disability (Office for Disability Issues, 2016a).

Early intervention is key for physical, mental, and cognitive issues such as fatigue management, pain management, and cognitive training. This support should be ongoing with vocational rehabilitation oversight as their condition changes. Over half the people in this study had not made work adaptations but expected that they would need to in the future. Therefore, people may only need input at various stages of their career due to the slow progressive impact of symptoms, especially in midlife (30- 40 years of age). Ongoing monitoring should occur to ensure timely intervention when required.

The provision of career counsellors for teenagers or adults at an early stage in their diagnosis may help them make realistic and sustainable career choices. Based on the results of this study supporting young people with myotonic dystrophy to gain an education and understand the long-term benefits this can have on their employment opportunities is important. A range of strategies such as volunteering, internships, work placements, and approaching organisations in person for a work trial, might enable a person to re-enter the workforce and build up their work experience and résumé. The findings from this study could empower therapists, teachers, and career

guidance officers to support and encourage people with myotonic dystrophy into suitable professions, identify their strengths and help them overcome barriers. This level of support may give them opportunities to engage in a career with longevity throughout their lifespan.

Forward planning and timely input are valuable. Knowing who to go to for support may empower individuals to reach out for assistance and overcome work demands. Exploring work challenges and available supports is an important conversation starter that should be revisited throughout the persons work career. Educating employers about the skills a person can offer and initiating an early conversation about adaptations and modifications to enhance work performance is important.

Since the introduction of individualised funding in New Zealand and more recently enabling good lives and system transformation people with disabilities have more flexibility and choice in how they utilise their allocated funding from the government (Ministry of Health, 2017). This funding can now be used in the area of employment to help a person search for work (develop CVs), prepare for work (interview techniques and training) and ongoing support in the logistics around maintaining a paid job. Providing choice and control to disabled people on their levels of support may enable more of them to source out employment and volunteer opportunities in their community.

This study provides insight into what factors to consider when supporting a person with myotonic dystrophy maintain or obtain employment. These findings may help support a young person with myotonic dystrophy into more sustainable career opportunities that will match their long-term skills. Interventions should be patient-centred and timely e.g. cognitive therapy and fatigue management. Professionals must realise that not everyone with myotonic dystrophy will present the same. It depends on the severity of their gene mutation, the age of manifestation and affected body systems. Therefore, the impact of symptoms can range from mild to severe and individuals may experience varying problems in their work performance due to differences in personal capabilities, work demands and environmental components in their community and workplace. A positive outcome of this study was that most of the

participants working are performing well and it is encouraging to know that with the correct support and input many more could gain employment.

Chapter 6 Conclusion

This cross-sectional study explored the employment rate, time loss from work and factors that influence work performance in people with myotonic dystrophy. The research revealed that this population group have an increased risk of unemployment in comparison to other people with a disability in New Zealand. Whilst employment rates are low, it was encouraging that the study revealed that those in employment are performing well in most aspects of their role and have limited time loss off work due to their health. The impact of time loss is an important area in research as it is often a key concern of employers.

Pain and fatigue were found to have the greatest influence on work performance, with depression, reduced cognition and sleep also playing a role. The physical demands in a job were reported by participants to be the most difficult. The multisystemic nature of the condition and the effects it has both physically and mentally on a person's functional performance may be a reason this population group are underrepresented in the labour market. The unknown rate of progression, impact of negative employer and employee attitudes and lack of timely support and intervention are also worth considering.

Given that work performance levels were generally good for those who were employed, it is a concern that the employment rate is low in comparison to the national rate for people with disability. The study revealed that people who left employment for whatever reason, found it difficult to obtain other sources of employment. If the reason for leaving employment was due to difficulties associated with their condition, there may be scope for a person to remain in their role with ongoing vocational support from a trained health care professional. A holistic approach should be applied, and factors of cognition, pain, fatigue, emotional well being and physical functioning explored. In addition, self-efficacy, sleep, and social support ought to be considered. Additionally, if the reason for leaving employment was unrelated to their myotonic dystrophy for example to have children, people may benefit from a supported return to work. The responses to the open-ended questions highlighted the importance of flexible work environments, options to work from home, reduced work hours, supportive employers and employees and ongoing vocational rehabilitation.

These environmental factors could facilitate work performance and enable a person with myotonic dystrophy maintain their career for longer.

This research is timely due to the current review of the New Zealand Disability Action Plan (Office for Disability Issues, 2018). This is the Government's commitment to implement the New Zealand Disability Strategy (2016-2026) where a priority is to increase the rates of employment amongst the disabled population (Office for Disability Issues, 2016a). This could be an opportunity for the New Zealand Government to review their current supports and recognise the underutilisation rate of this population group in the labour sector. Early career guidance and timely intervention and adaptations could enable a person to obtain or maintain a job with longevity at the point of diagnosis and symptom progression.

The results of this research also highlight the need to look at personal, occupational, and environmental factors when analysing work performance. For example, the findings of this study support the idea that education has a significant influence on one's ability to obtain and maintain employment even as their condition progresses. Tertiary level education or retraining should be encouraged. Early career guidance and necessary adaptations could enable a person to obtain or maintain a job with longevity. Cognitive capacity is especially relevant for this group as myotonic dystrophy is one of the few muscular dystrophies that can disturb cognitive functioning. Further research using more comprehensive neuropsychological assessment batteries to explore different domains of cognitive functioning (e.g. memory, ability to process new information and make decisions) is needed. It would be good to analyse the difficulties experienced in this population and to examine the potential impact on work performance in more detail. More information on specific cognitive difficulties could help design targeted interventions to facilitate people's work performance.

This study benefitted from the recruitment process and nationwide sample of people living with myotonic dystrophy across New Zealand. However, was limited by low engagement of people with non-European backgrounds impacting on the generalisability of the findings. Despite this key limitation, the study has confirmed findings from studies done in other countries (Andries et al., 1997; Fowler et al., 1997; Minis et al., 2010), highlighting the issue of low employment rates in this population. It

has added to existing literature by increasing our understanding of the role of pain, fatigue, and other key areas where targeted interventions could facilitate work performance and career longevity in people with myotonic dystrophy. The insights gained from this study may be of assistance to health care professionals, disability services, family, recruitment agencies and employers when supporting people with myotonic dystrophy in their current and future careers.

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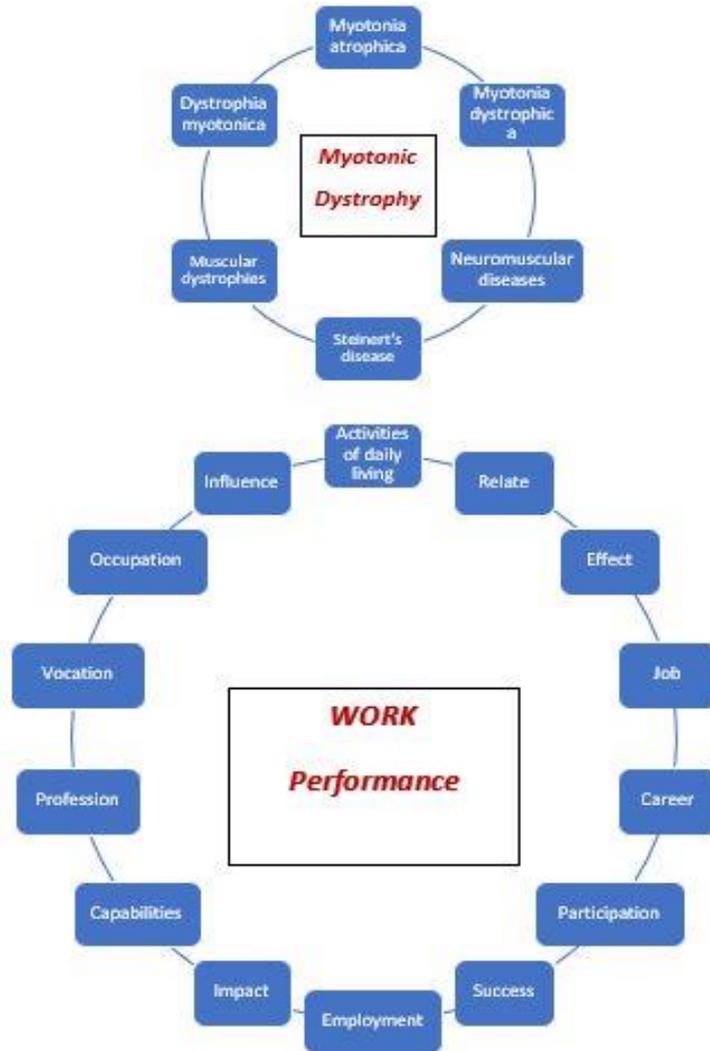
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Appendix A: Literature Search Terms



Appendix B: Northern Y Regional Ethics Committee of New Zealand Ethics Approval



Health and Disability Ethics Committees
 Ministry of Health
 C/- MEDSAFE, Level 6, Deloitte House
 10 Brandon Street
 PO Box 5013
 Wellington
 6011

0800 4 ETHICS
 hdec@moh.govt.nz

09 September 2014

Dr Alice Theadom
 AUT North Shore Campus, AA254C
 Auckland 1142

Dear Dr Theadom

Re: Ethics ref:	14/NTB/118
Study title:	Prevalence and impact of genetic muscle disease

I am pleased to advise that this application has been approved by the Northern B Health and Disability Ethics Committee. This decision was made through the HDEC-Full Review pathway.

The main issues considered by the HDEC in giving approval were as follows.

- The Researchers explained that the study aims to identify the frequency and impact of genetic muscle disorders in New Zealand. The Committee queried if there was any existing literature on the prevalence of MD in New Zealand. The Researchers explained that earlier work on this topic was not of a high quality.
- The Committee confirmed that disclosing health information for screening was justified, as it is the only way to identify the patient population. The Committee suggested being clear about how potential participant information was sought.
- The Committee noted that some adults will not be able to consent. Please explain why?
- The Researchers explained that some of adult participants have severe MD which affects the nervous system, reducing competency. There will not be very many of these cases.
- The Researchers justified the subgroup's involvement because they represent an important subset of people who experience the severe impact of disease. The Committee was satisfied that there was a need to include this group.
- The Committee confirmed that the legal representative would, in all cases, be providing consent for any adults, as a legal guardian. Welfare guardians or advance directives were suitable due to the observational nature of the research.
- The Committee added that participants must be given the opportunity to dissent, in which case they should not be enrolled.
- (R.7.1) Please explain how risks to researchers will be managed. The Researchers explained that risks posed to researchers out in the field were mitigated by having access to crisis team numbers, as well as having an awareness of participants who may have high levels of anxiety or depression. Referrals will not occur unless with the consent of the participants.
- The Researchers further explained that researchers out in the field will have an electronic tracking system which they must check in and out of, which is monitored externally. The Committee was satisfied with the response.
- The Committee confirmed written consent will be obtained before conducting the in-person interview or telephone assessment

- Please explain what the reasoning is behind using data for a larger study.
- The Committee requested the following changes to the Participant Information Sheet and Consent Form:
 - Please review PIS/CFs for correct use of 'you' and 'they', currently inconsistent.
 - The Committee noted that consent form 'yes/no' options should only be available if the bullet point is truly optional. Please review and remove the yes/no for those which are not optional.
 - Simplify the child assent form language where possible.

Conditions of HDEC approval

HDEC approval for this study is subject to the following conditions being met prior to the commencement of the study in New Zealand. It is your responsibility, and that of the study's sponsor, to ensure that these conditions are met. No further review by the Northern B Health and Disability Ethics Committee is required.

Standard conditions:

1. Before the study commences at *any* locality in New Zealand, all relevant regulatory approvals must be obtained.
2. Before the study commences at a *given* locality in New Zealand, it must be authorised by that locality in Online Forms. Locality authorisation confirms that the locality is suitable for the safe and effective conduct of the study, and that local research governance issues have been addressed.

After HDEC review

Please refer to the *Standard Operating Procedures for Health and Disability Ethics Committees* (available on www.ethics.health.govt.nz) for HDEC requirements relating to amendments and other post-approval processes.

Your next progress report is due by 09 September 2015.

Participant access to ACC

The Northern B Health and Disability Ethics Committee is satisfied that your study is not a clinical trial that is to be conducted principally for the benefit of the manufacturer or distributor of the medicine or item being trialled. Participants injured as a result of treatment received as part of your study may therefore be eligible for publicly-funded compensation through the Accident Compensation Corporation (ACC).

Please don't hesitate to contact the HDEC secretariat for further information. We wish you all the best for your study.

Yours sincerely,



Mrs Raewyn Sporle
Chairperson
Northern B Health and Disability Ethics Committee

Encl: appendix A: documents submitted
appendix B: statement of compliance and list of members

Appendix A
Documents submitted

<i>Document</i>	<i>Version</i>	<i>Date</i>
Protocol	1	13 June 2014
CV for CI	1	26 June 2014
Evidence of scientific review	1	26 February 2014
Evidence of scientific review	1	26 February 2014
Evidence of scientific review	1	26 February 2014
Evidence of scientific review	1	26 February 2014
Evidence of scientific review	1	07 March 2014
CVs for other Investigators	1	02 July 2014
CVs for other Investigators	1	02 July 2014
CVs for other Investigators	1	02 July 2014
PIS/CF	1	30 June 2014
PIS/CF	1	30 June 2014
PIS/CF	1	30 June 2014
PIS/CF	1	30 June 2014
PIS/CF	1	30 June 2014
Survey/questionnaire	1	30 June 2014
Survey/questionnaire	1	14 June 2014
Survey/questionnaire	1	19 July 2014
Survey/questionnaire	1	19 July 2014
Survey/questionnaire	1	19 July 2014
Survey/questionnaire	1	13 June 2014
PIS/CF	1	19 July 2014
PIS/CF	1	19 July 2014
PIS/CF	1	19 July 2014
PIS/CF	1	19 July 2014
PIS/CF for persons interested in welfare of non-consenting participant	1	19 July 2014
PIS/CF	1	15 August 2014
Covering Letter	1	14 July 2014

Appendix B
Statement of compliance and list of members

Statement of compliance

The Northern B Health and Disability Ethics Committee:

- is constituted in accordance with its Terms of Reference
- operates in accordance with the *Standard Operating Procedures for Health and Disability Ethics Committees*, and with the principles of international good clinical practice (GCP)
- is approved by the Health Research Council of New Zealand's Ethics Committee for the purposes of section 25(1)(c) of the Health Research Council Act 1990
- is registered (number 00008715) with the US Department of Health and Human Services' Office for Human Research Protection (OHRP).

List of members

<i>Name</i>	<i>Category</i>	<i>Appointed</i>	<i>Term Expires</i>	<i>Present on 02/09/2014?</i>	<i>Declaration of interest?</i>
Mrs Raewyn Sporle	Lay (the law)	01/07/2012	01/07/2015	Yes	No
Mrs Maliaga Erick	Lay (consumer/community perspectives)	01/07/2012	01/07/2015	Yes	No
Mrs Phyllis Huitema	Lay (consumer/community perspectives)	19/05/2014	19/05/2017	Yes	No
Miss Tangihaere Macfarlane	Lay (consumer/community perspectives)	19/05/2014	19/05/2017	Yes	No
Mrs Kate O'Connor	Non-lay (other)	01/07/2012	01/07/2015	Yes	No
Mrs Stephanie Pollard	Non-lay (intervention studies)	01/07/2012	01/07/2015	Yes	No
Dr Paul Tanser	Non-lay (health/disability service provision)	01/07/2012	01/07/2015	Yes	No
Ms Kerin Thompson	Non-lay (intervention studies)	01/07/2012	01/07/2015	Yes	No

<http://www.ethics.health.govt.nz>

Appendix C: Auckland University of Technology Ethics Committee



11 September 2014

Alice Theadom

Faculty of Health and Environmental Sciences

Dear Alice

Ethics Application: **14/296 Prevalence and impact of genetic muscle disorders in New Zealand (MD-Prev).**

Thank you for submitting your application for ethical review to the Auckland University of Technology Ethics Committee (AUTECSecretariat). I am pleased to confirm that the Chair and I have approved your ethics application for three years until 11 September 2017.

As part of the ethics approval process, you are required to submit the following to AUTECSecretariat:

- A brief annual progress report using form EA2, which is available online through <http://www.aut.ac.nz/researchethics>. When necessary this form may also be used to request an extension of the approval at least one month prior to its expiry on 11 September 2017;
- A brief report on the status of the project using form EA3, which is available online through <http://www.aut.ac.nz/researchethics>. This report is to be submitted either when the approval expires on 11 September 2017 or on completion of the project;

It is a condition of approval that AUTECSecretariat is notified of any adverse events or if the research does not commence. AUTECSecretariat approval needs to be sought for any alteration to the research, including any alteration of or addition to any documents that are provided to participants. You are responsible for ensuring that research undertaken under this approval occurs within the parameters outlined in the approved application.

AUTECSecretariat grants ethical approval only. If you require management approval from an institution or organisation for your research, then you will need to obtain this.

To enable us to provide you with efficient service, we ask that you use the application number and study title in all correspondence with us. If you have any enquiries about this application, or anything else, please do contact us at ethics@aut.ac.nz.

All the very best with your research,

A handwritten signature in black ink, appearing to read 'K O'Connor', written in a cursive style.

Kate O'Connor

Executive Secretary

Auckland University of Technology Ethics Committee

Appendix D: Participant Information Sheet



Adult Participant Information Sheet

Study title:	Prevalence and impact of genetic muscle disorders in New Zealand		
Locality:	New Zealand	Ethics committee ref.:	14NTB/118
Lead investigator:	Alice Theadom	Contact phone number:	0800 MDPREV

You are invited to take part in a study that is looking at how many people are affected by genetic muscle disorders in New Zealand. We would also like to find out how people and their significant others are affected by these conditions.

If you do not want to take part, you do not have to give a reason, and it will not affect the care you receive. If you do want to take part now, but change your mind later, you can pull out of the study at any time.

This information sheet will help you decide if you would like to take part. It sets out why we are doing the study, what your participation would involve, what the benefits and risks to you might be, and what would happen after the study ends. We will go through this information with you and answer any questions you may have. You do not have to decide today whether or not you will participate in this study. Before you decide, you may want to talk about the study with other people, such as family, whānau, friends, or healthcare providers. Feel free to do this.

If you agree to take part in this study, you will be asked to sign the Consent Form on the last page of this document. You will be given a copy of both the Participant Information Sheet and the Consent Form to keep. This document is 7 pages long, including the consent form. Please make sure you have read and understood all the pages.

WHAT IS THE PURPOSE OF THE STUDY?

We are a team of researchers who work in universities, hospitals and community organisations across New Zealand with an interest in supporting people with neuromuscular conditions. The purpose of this study is to find out how many people are affected by genetic muscle disorders in New Zealand. We would also like to find out how peoples' everyday lives are affected and to identify where there are unmet needs.

We hope that by finding out this information, will help us to improve the support and treatment people and their significant others receive. Even if you are not noticing any effects from your condition, this is just as important for us to know as if you do.

This study is being funded by the Health Research Council of New Zealand.

If you have any questions about the study please contact the Study Manager,

Kerry Walker:

Telephone: 0800 MDPREV (637738)

Mobile: 021 2458597

E mail: kwalker@aut.ac.nz

This study has been approved by the Health and Disability Ethics Committee reference: 14/NTB/118

WHAT WILL MY PARTICIPATION IN THE STUDY INVOLVE?

You have been identified as someone who may have a genetic muscle disorder. We are inviting everyone with a genetic muscle disorder in New Zealand to participate in this study (about 1200 people). This will enable us to explore the services and supports people receive across the country.

If you would like to take part in this study, we would like to arrange a time to come and talk to you. You will be asked questions about how you find completing everyday activities such as study/work, socializing, your quality of life and about any symptoms you experience. All researchers who will be asking these questions have been specially

trained for this project. We can arrange for a member of the team to visit you at your home or other convenient location such as at your local GP surgery or a private room in a public library or you can complete the questionnaire over the phone or by yourself in your own time.

When a researcher comes to visit you or talks about the study with you, you will have the opportunity to ask any questions you may have. If you are happy to take part, you will be invited to sign the consent form. The researcher will ask you some questions about how you have been doing and your ability to take part in everyday activities. The assessment should take about one and half hours. You will be able to have support people with you during the assessment and we can complete the assessment over several sessions if you prefer.

We aim to finish collecting data for this study by the end of March 2016.

WHAT ARE THE POSSIBLE BENEFITS AND RISKS OF THIS STUDY?

Taking part in this study will take some of your time and require you to answer a series of questions. There are no known risks caused by this study, however you may feel uncomfortable or embarrassed by some of the questions. You do not have to answer any questions you do not wish to do so. All our researchers have received training in administering these assessments and working with people who have a genetic muscle disorder.

Your usual medical care will not be affected in any way by participating in the study or withdrawing from the study at any stage. Your participation in this study will be stopped should you experience any harmful effects or if the doctor feels it is not in your best interests to continue. Similarly, your doctor may at any time provide you with any other treatment he/she considers necessary.

As part of the study we will be working with health care and service providers about the findings of the study. We aim to identify areas of unmet need for people living with these conditions to improve service delivery. We will also be in contact with your GP or neurologist about your diagnosis and if any information that may be of benefit to you emerges during the study, we will let them know.

WHO PAYS FOR THE STUDY?

There should be no direct costs to you in taking part in this study.

A \$20 food/fuel voucher will be provided to you after completion of the assessment to acknowledge your contribution to this research.

Assessments will be completed at your home or other accessible location. If for some reason you need to travel for the assessment, your mileage, or costs (receipt/ticket required) will be reimbursed. Some questions can be asked over the phone.

WHAT IF SOMETHING GOES WRONG?

It is unlikely that you will be at risk of harm from taking part in this study. If something goes wrong, please contact the study manager as soon as possible 0800 MDPREV (637738)

WHAT ARE MY RIGHTS?

Your participation is entirely voluntary, and you will be able to withdraw from the study at any time without experiencing any disadvantage.

The study files and all other information that you provide will remain strictly confidential, unless information is revealed that indicates you, your child or someone else is at risk. The answers to your questions will be stored separately to any document that has your name and contact details on.

No material that could personally identify you will be used in any reports or discussions about this study.

You will be able to access your information collected as part of the study if you wish to do so. If any information that may be of benefit to you emerges during the study, we will contact you to let you know.

WHAT HAPPENS AFTER THE STUDY OR IF I CHANGE MY MIND?

Upon completion of the study your records will be stored for 16 years in a locked cabinet. The cabinet will be based at AUT University in Auckland by the lead investigator (Dr Alice Theadom). All computer records will be password protected. Any identifying information will not be shared outside of the research team without seeking your permission.

As there will be a lot of valuable information collected as part of this study, we would like to make anonymized data collected as part of the study available to other

international researchers on completion of the study. However, we will only share your anonymized data if you wish us to do so, otherwise your data will be deleted before being shared with other researchers.

After 16 years all electronic information will be deleted, and paper forms will be shredded and destroyed with the university confidential waste.

After we have looked at all the data, we will send you a summary of results if you would like to receive them. This will be in early 2016.

WHO DO I CONTACT FOR MORE INFORMATION OR IF I HAVE CONCERNS?

If you have any questions, concerns, or complaints about the study at any stage, you can contact:

Kerry Walker, Study Manager

Telephone number: 0800 MDPREV (637738)

Email: kwalker@aut.ac.nz

If you want to talk to someone who is not involved with the study, you can contact an independent health and disability advocate on:

Phone: 0800 555 050

Fax: 0800 2 SUPPORT (0800 2787 7678)

Email: advocacy@hdc.org.nz

For Māori health support please contact:

Te Puna Oranga (Waikato DHB Māori Health Unit),

Hockin Building, Level 1, Pembroke St, P.O.Box 934, Hamilton.

Ph: 07 834 3644. Fax: 07 834 3619.

You can also contact the health and disability ethics committee (HDEC) that approved this study on:

Phone: 0800 4 ETHICS

Email: hdecs@moh.govt.nz

Please keep this for your information.

Thank you for interest in this study.

Appendix E: Adult Consent Form

Registration Number:	Participant Initials:	Date of Birth:
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Adult Consent Form

If you need an INTERPRETER, please tell us.

I have read or have had read to me in my first language, and I understand the Participant Information Sheet dated 03/09/2014.

I have been given sufficient time to consider whether or not to participate in this study.

I have had the opportunity to use a legal representative, whānau/ family support, or a friend to help me ask questions and understand the study.

I am satisfied with the answers I have been given regarding the study and I have a copy of this consent form and information sheet.

I understand that taking part in this study is voluntary (my choice) and that I may withdraw from the study at any time without this affecting my medical care.

I consent to the research staff collecting and processing my information, including information about my health from medical records and health databases.

If I decide to withdraw from the study, I agree that the information collected about me up to the point when I withdraw may continue to be processed.

I consent to my GP or treating clinician being informed about my participation in the study and of any significant results obtained during the study.

I agree to an approved auditor appointed by the New Zealand Health and Disability Ethic Committees, or any relevant regulatory authority or their approved representative reviewing my relevant medical records for the sole purpose of checking the accuracy of the information recorded for the study.

I understand that my participation in this study is confidential and that no material, which could identify me personally, will be used in any reports on this study.

I know who to contact if I have any questions about the study in general.

I understand my responsibilities as a study participant.

I wish to receive a summary of the results from the study. Yes No

I agree to my anonymised data being shared with researchers overseas for further research into neuromuscular conditions Yes No

Declaration by participant:

I hereby consent to take part in this study.

Participant's name:

Signature:

Date:

Declaration by member of research team:

I have given a verbal explanation of the research project to the participant and have answered the participant's questions about it.

I believe that the participant understands the study and has given informed consent to participate.

Researcher's name:

Signature:

Date:

Error! Not a valid bookmark self-reference. **Appendix F: MD-Prev Questionnaire**



Registration number:

Participant initials:

Date of birth:

d d m m y y

MD-Prev Adult Participant Questionnaire

Welcome to the MD-Prev Questionnaire

Thank you for participating in our study. Your feedback is important.

Please try to answer every question. However, if there are questions you do not want to answer, you can simply leave them blank.

If you are unsure of how to answer a question, please make a note of this in the text box on the last page of the questionnaire.

If you fill in an answer which does not seem correct (for example; an incorrect date format), you will be prompted to re-think your answer.

Please give one answer per line unless prompted otherwise.

Demographic details

1.1	What is the persons gender	Male, Female
1.2	What ethnicity does the person most associate with?	New Zealand European Other European Maori Samoan Cook Island Maori Tongan Niuean Chinese Indian Unknown Other
	If other, please specify	Text
1.3	What is your current marital status? (tick one only)	Married, civil union, de facto Separated/divorced/widowed Never married (single) Other
	If other, please specify	Text
1.4	Do you have any children?	Yes, No
1.5	What is the highest level of education you completed	Primary school High School Polytechnic or college University

Diagnosis & Co morbidities

2.1	What is your diagnosis?	
2.2	Have you been diagnosed with any other conditions?	Yes No
2.3	If yes, please specify	<p>Hypertension, high blood pressure</p> <p>Alzheimer's disease</p> <p>Dementia</p> <p>Arthritis</p> <p>Cancer</p> <p>COPD</p> <p>Developmental disability</p> <p>Attention deficit</p> <p>Multiple sclerosis</p> <p>Parkinson's disease</p> <p>Mood disorder</p> <p>Schizophrenia</p> <p>Diabetes</p> <p>Epilepsy</p> <p>Kidney or liver disease</p> <p>Other</p>
2.4	If other, please specify	Text

Activity Limitations – Activlim

Please estimate the difficulty or ease with which you would find performing each of the following activities **without any aids or human help** (even if you actually use help in daily life).

Three responses are presented. These assess the perception of the difficulty/ease depending on whether the activity is “impossible”, “difficult” or “easy”. Activities not attempted in the last 3 months are not scored and entered as missing responses (tick the question mark).

So, for any activity, the four potential answers are:

- **Impossible:** You are unable to perform the activity without using any other help
- **Difficult:** You are able to perform the activity without any help but experiences some difficulty
- **Easy:** You are able to perform the activity without any help and experience no difficulty
- **Question mark:** You cannot estimate the difficulty of the activity because you have never done the activity

Watch out!! If the activity was never attempted because it is impossible, then it must be scored “impossible” rather than “question mark”.

How difficult are the following activities?		Impossible	Difficult	Easy	?
1	Putting on a T-shirt				
2	Washing one's upper body				
3	Dressing one's lower body				
4	Taking a shower				
5	Sitting on the toilet				
6	Taking a bath				
7	Walking downstairs				

8	Stepping out of a bathtub				
9	Opening a door				
10	Walking outdoors on level ground				
11	Washing one's face				
12	Hanging up a jacket on a hat stand				
13	Wiping one's upper body				
14	Walking upstairs				

How difficult are the following activities?		Impossible	Difficult	Easy	?
15	Carrying a heavy load				
16	Getting into a car				
17	Standing for a long time (\pm 10 min)				
18	Walking more than 1 kilometre				

Activity Limitations - Upper and lower extremity functioning

What is your ability to do the following activities without support?

	Without any difficulty	With a little difficulty	With some difficulty	With much difficulty	Unable to do
Are you able to turn a key in a lock?					
Are you able to brush your teeth?					
Are you able to make a phone call?					

Are you able to pick up coins from a table top?					
Are you able to write with a pen or pencil?					
Are you able to open and close a zipper?					
Are you able to wash and dry your body?					
Are you able to shampoo your hair?					
Are you able to get on and off the toilet?					
Are you able to step up and down curbs?					
Are you able to get in and out of a car?					
Are you able to get out bed into a chair?					
Are you able to push open a heavy door?					
Are you able to get up off the floor from lying on your back without help?					
Are you able to run errands and shop?					
Are you able to go for a walk of at least 15 minutes?					

Occupation

Has having a genetic muscle disorder affected your employment in any way?	Text
If yes, please explain	Text
What is your current employment status?	<p>Full time paid work (>30 hours)</p> <p>Part time paid work</p> <p>Homemaker / carer</p> <p>Unemployed or redundant</p> <p>Student</p> <p>Beneficiary</p> <p>Other</p>
If other, please specify	Text
What is your occupation?	Text
Which of the following categories does your occupation fall into?	<p>Manager</p> <p>Professional</p> <p>Technician or trade worker</p> <p>Community/personal service worker</p> <p>Clerical and administrative worker</p> <p>Sales worker</p> <p>Machinery operators and driver</p> <p>Labourer</p> <p>Other</p>
If other please specify	Text
How many hours per week do you currently work?	2 digits
Have you had to make any changes to enable you to continue in your role due to your condition to help you to manage?	<p>Yes</p> <p>No</p>
If yes, please specify	Text

Work Limitations (WLQ-25)

Health problems can make it difficult for working people to perform certain parts of their jobs. We are interested in learning about how your health may have affected you at work during the past 2 weeks.

The questions will ask you to think about your physical health or emotional problems. Emotional problems may include feeling depressed or anxious.

Most of the questions are multi-choice. Please answer by placing a mark in a box.

In the past 2 weeks, how much of the time did your physical health or emotional problems make it difficult for you to do the following? *(Mark one box on each line)*

	Difficult all of the time (100%)	Difficult most of the time	Difficult some of the time (about 50%)	Difficult a little bit of the time	Difficult none of the time (0%)	Does not apply to my job
Work the required number of hours	<input type="checkbox"/> ₁	<input type="checkbox"/> ₂	<input type="checkbox"/> ₃	<input type="checkbox"/> ₄	<input type="checkbox"/> ₅	<input type="checkbox"/> ₆
Get going easily at the beginning of the workday	<input type="checkbox"/> ₁	<input type="checkbox"/> ₂	<input type="checkbox"/> ₃	<input type="checkbox"/> ₄	<input type="checkbox"/> ₅	<input type="checkbox"/> ₆
Start on your job as soon as you arrived at work	<input type="checkbox"/> ₁	<input type="checkbox"/> ₂	<input type="checkbox"/> ₃	<input type="checkbox"/> ₄	<input type="checkbox"/> ₅	<input type="checkbox"/> ₆
Do your work without stopping to take breaks or rests	<input type="checkbox"/> ₁	<input type="checkbox"/> ₂	<input type="checkbox"/> ₃	<input type="checkbox"/> ₄	<input type="checkbox"/> ₅	<input type="checkbox"/> ₆
Stick to a routine or schedule	<input type="checkbox"/> ₁				<input type="checkbox"/> ₅	<input type="checkbox"/> ₆

These questions ask you to rate the amount of time you were
able to handle certain parts of your job without difficulty.

In the past 2 weeks, how much of the time were you able to.....

(Mark one box on each line)

	All of the time (100%)	Most of the time	Some of the time (about 50%)	A little bit of the time	None of the time	Does not apply to my job
Walk or move around different work locations (for example, go to meetings), without difficulty?						
Lift, carry or move objects at work weighing <u>more than 5 kilos</u> , without difficulty?						
To sit, stand, or stay in one position for more <u>than 15 minutes</u> while working without difficulty?						
Repeat the same motions over and over again while working without difficulty?						
Bend, twist, or reach while working, without difficulty?						
Use hand-held tools or equipment (e.g., a phone, pen, keyboard, computer mouse, drill, hairdryer, or sander), without difficulty?						

These questions ask you to rate the amount of time you were able to handle certain parts of your job without difficulty.

In the past 2 weeks, how much of the time did your physical health or emotional problems make it difficult for you to do the following? *(Mark one box on each line)*

	Difficult all of the time (100%)	Difficult most of the time	Difficult some of the time (about 50%)	Difficult a little bit of the time	Difficult none of the time (0%)	Does not apply to my job
Keep your mind on your work	<input type="checkbox"/> ₁	<input type="checkbox"/> ₂	<input type="checkbox"/> ₃	<input type="checkbox"/> ₄	<input type="checkbox"/> ₅	<input type="checkbox"/> ₆
Think clearly when working	<input type="checkbox"/> ₁	<input type="checkbox"/> ₂	<input type="checkbox"/> ₃	<input type="checkbox"/> ₄	<input type="checkbox"/> ₅	<input type="checkbox"/> ₆
Do work carefully	<input type="checkbox"/> ₁	<input type="checkbox"/> ₂	<input type="checkbox"/> ₃	<input type="checkbox"/> ₄	<input type="checkbox"/> ₅	<input type="checkbox"/> ₆
Concentrate on your work	<input type="checkbox"/> ₁	<input type="checkbox"/> ₂	<input type="checkbox"/> ₃	<input type="checkbox"/> ₄	<input type="checkbox"/> ₅	<input type="checkbox"/> ₆
Work without losing your train of thought	<input type="checkbox"/> ₁	<input type="checkbox"/> ₂	<input type="checkbox"/> ₃	<input type="checkbox"/> ₄	<input type="checkbox"/> ₅	<input type="checkbox"/> ₆
Easily read or use your eyes when working	<input type="checkbox"/> ₁	<input type="checkbox"/> ₂	<input type="checkbox"/> ₃	<input type="checkbox"/> ₄	<input type="checkbox"/> ₅	<input type="checkbox"/> ₆

The next questions ask about difficulties in relation to the people you came in contact with while working. These may include employers, supervisors, co-workers, clients, customers or the public.

In the past 2 weeks, how much of the time did your physical health or emotional problems make it difficult for you to do the following? *(Mark one box on each line)*

	Difficult all of the time (100%)	Difficult most of the time	Difficult some of the time (about 50%)	Difficult a little bit of the time	Difficult none of the time (0%)	Does not apply to my job
Speak with people in person, in meetings or on the phone	<input type="checkbox"/> ₁	<input type="checkbox"/> ₂	<input type="checkbox"/> ₃	<input type="checkbox"/> ₄	<input type="checkbox"/> ₅	<input type="checkbox"/> ₆
Control your temper around people when working	<input type="checkbox"/> ₁	<input type="checkbox"/> ₂	<input type="checkbox"/> ₃	<input type="checkbox"/> ₄	<input type="checkbox"/> ₅	<input type="checkbox"/> ₆
Help other people to get work done	<input type="checkbox"/> ₁	<input type="checkbox"/> ₂	<input type="checkbox"/> ₃	<input type="checkbox"/> ₄	<input type="checkbox"/> ₅	<input type="checkbox"/> ₆

These questions ask about how things went at work overall.

In the past 2 weeks, how much of the time did your physical health or emotional problems make it difficult for you to do the following? *(Mark one box on each line)*

	Difficult all of the time (100%)	Difficult most of the time	Difficult some of the time (about 50%)	Difficult a little bit of the time	Difficult none of the time (0%)	Does not apply to my job
Handle the workload	<input type="checkbox"/> ₁	<input type="checkbox"/> ₂	<input type="checkbox"/> ₃	<input type="checkbox"/> ₄	<input type="checkbox"/> ₅	<input type="checkbox"/> ₆
Work fast enough	<input type="checkbox"/> ₁	<input type="checkbox"/> ₂	<input type="checkbox"/> ₃	<input type="checkbox"/> ₄	<input type="checkbox"/> ₅	<input type="checkbox"/> ₆
Finish work on time	<input type="checkbox"/> ₁	<input type="checkbox"/> ₂	<input type="checkbox"/> ₃	<input type="checkbox"/> ₄	<input type="checkbox"/> ₅	<input type="checkbox"/> ₆
Do your work without making mistakes	<input type="checkbox"/> ₁	<input type="checkbox"/> ₂	<input type="checkbox"/> ₃	<input type="checkbox"/> ₄	<input type="checkbox"/> ₅	<input type="checkbox"/> ₆
Feel you have done what you are capable of doing	<input type="checkbox"/> ₁	<input type="checkbox"/> ₂	<input type="checkbox"/> ₃	<input type="checkbox"/> ₄	<input type="checkbox"/> ₅	<input type="checkbox"/> ₆

The following questions ask about time missed from work during the **past two weeks**. If you are self-employed or work at home, think about the time you missed on days you expected to work. Report on your **main** job only.

1. In the past 2 weeks, how many **full workdays** did you miss because of your health or medical care? Health includes physical health and emotional problems.

(Mark one box.)

<input type="checkbox"/>											
None	1	2	3	4	5	6	7	8	9	10	11 or More

2. In the past 2 weeks, what was the total number of days on which you missed **part of a workday** because of your health or medical care (*for example, you came in late or left early*)? Health includes physical health and emotional problems.

(Mark one box.)

<input type="checkbox"/>											
None	1	2	3	4	5	6	7	8	9	10	11 or More

Mood and Well-being (HADS)

Read each item below and select the reply which comes closest to how you have been feeling in the past week.

Do not take too long over your replies, your immediate reaction to each item will probably be more accurate than a long, thought-out response.

1.	I feel tense or 'wound up' (tick one only)	Most of the time [3] A lot of the time [2] From time to time, occasionally [1] Not at all [0]
2.	I still enjoy the things I used to enjoy (tick one only)	[Definitely as much [0] Not quite as much [1] Only a little [2] Hardly at all [3]
3.	I get a sort of frightened feeling as if something awful is about to happen (tick one only)	Very definitely and quite badly [3] Yes, but not too badly [2] A little, but it does not worry me [1] Not at all [0]
4.	I can laugh and see the funny side of things (tick one only)	As much as I always could [0] Not quite so much now [1] Definitely not as much now [2] Not at all [3]
5.	Worrying thoughts go through my mind (tick one only)	A great deal of the time [3] A lot of the time [2] From time to time, but not too often [1] Only occasionally [0]
6.	I feel cheerful (tick one only)	Not at all [3] Not often [2]

		Sometimes [1] Most of the time [0]
7.	I can sit at ease and feel relaxed (tick one only)	Definitely [0] Usually [1] Not often [2] Not at all [3]
8.	I feel as if I am slowed down (tick one only)	Nearly all the time [3] Very often [2] Sometimes [1] Not at all [0]
9.	I get a sort of frightened feeling like 'butterflies' in the stomach (tick one only)	Not at all [0] Occasionally [1] Quite often [2] Very often [3]
10.	I have lost interest in my appearance (tick one only)	Definitely [3] I do not take as much care as I should [2] I may not take quite as much care [1] I take just as much care as ever [0]
11.	I feel restless as if I have to be on the move (tick one only)	Very much indeed [3] Quite a lot [2] Not very much [1] Not at all [0]
12.	I look forward with enjoyment to things (tick one only)	As much as I ever did [0] Rather less than I used to [1] Definitely less than I used to [2] Hardly at all [3]
13.	I get sudden feelings of panic (tick one only)	Very often indeed [3] Quite often [2]

		Not very often [1] Not at all [0]
14.	I can enjoy a good book or TV programme* (tick one only)	Often [0] Sometimes [1] Not often [2] Very seldom [3]

DSM –IV item

	Which of the following best describes the way you have been feeling over the last two weeks: (tick one only)	1 = "I do not have any thoughts of killing myself," 2 = "I have thoughts of killing myself, but I would not carry them out," 3 = "I would like to kill myself," and 4 = "I would kill myself if I had the chance."
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About your sleep (Pittsburgh Sleep Quality Index)

The following questions relate to your usual sleep habits over the past month. Your answers should indicate the most accurate reply for the majority of days and nights in the past month.

1.	What time have you usually gone to bed?	Hh:mm			
2.	How long (in minutes) has it taken you to fall asleep each night?	3digits			
3.	When have you usually gotten up in the morning?	Hh:mm			
4.	How many hours of actual sleep do you get a night (may be different than hours spent in bed)	3 digits (one decimal)			
5.	How many hours do you usually spend in bed?	3 digits (one decimal)			
6. During the past month how often have you had trouble sleeping because you.....					
		Not during the past month	Less than once per week	Once or twice a week	Three or more times a week
	Cannot get to sleep within 30 minutes				
	Wake up in the middle of the night or early morning				
	Have to get up to use the bathroom				
	Cannot breathe comfortably				
	Cough or snore loudly				
	Feel too cold				
	Feel too hot				
	Have bad dreams				

	Have pain				
7.	Other reason(s) please indicate how often you have had trouble sleeping because of this reason 1. 2.				
8.	During the past month how often have you taken medication (prescribed or 'over the counter') to help you sleep?				
9.	During the past month how often have you had trouble staying awake while driving, eating meals, or engaging in social activity?				
10.	During the past month how much of a problem has it been for you to keep up enthusiasm to get things done?				
		Very good	Fairly good	Fairly bad	Very bad
11.	During the past month, how would you rate your sleep quality overall?				

Short-Form McGill Pain Questionnaire

This questionnaire provides you with a list of words that describe some of the different qualities of pain and related symptoms. Please put an X through the numbers that best describe the intensity of each of the pain and related symptoms you felt during the last week. Use 0 if the word does not describe your pain or related symptoms.

1. Throbbing pain *none*

0	1	2	3	4	5	6	7	8	9	10
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worst possible

 2. Shooting pain *none*

0	1	2	3	4	5	6	7	8	9	10
---	---	---	---	---	---	---	---	---	---	----

worst possible

 3. Stabbing pain *none*

0	1	2	3	4	5	6	7	8	9	10
---	---	---	---	---	---	---	---	---	---	----

worst possible

 4. Sharp pain *none*

0	1	2	3	4	5	6	7	8	9	10
---	---	---	---	---	---	---	---	---	---	----

worst possible

 5. Cramping pain *none*

0	1	2	3	4	5	6	7	8	9	10
---	---	---	---	---	---	---	---	---	---	----

worst possible

 6. Gnawing pain *none*

0	1	2	3	4	5	6	7	8	9	10
---	---	---	---	---	---	---	---	---	---	----

worst possible

 7. Hot-burning pain *none*

0	1	2	3	4	5	6	7	8	9	10
---	---	---	---	---	---	---	---	---	---	----

worst possible

 8. Aching pain *none*

0	1	2	3	4	5	6	7	8	9	10
---	---	---	---	---	---	---	---	---	---	----

worst possible
-

9. Heavy pain *none*

0	1	2	3	4	5	6	7	8	9	10
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worst possible

10. Tender *none*

0	1	2	3	4	5	6	7	8	9	10
---	---	---	---	---	---	---	---	---	---	----

worst possible

11. Splitting pain *none*

0	1	2	3	4	5	6	7	8	9	10
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worst possible

12. Tiring – exhausting *none*

0	1	2	3	4	5	6	7	8	9	10
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worst possible

13. Sickening *none*

0	1	2	3	4	5	6	7	8	9	10
---	---	---	---	---	---	---	---	---	---	----

worst possible

14. Fearful *none*

0	1	2	3	4	5	6	7	8	9	10
---	---	---	---	---	---	---	---	---	---	----

worst possible

15. Punishing – cruel *none*

0	1	2	3	4	5	6	7	8	9	10
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worst possible

16. Electric-shock pain *none*

0	1	2	3	4	5	6	7	8	9	10
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worst possible

17. Cold-freezing pain *none*

0	1	2	3	4	5	6	7	8	9	10
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worst possible

18. Piercing *none*

0	1	2	3	4	5	6	7	8	9	10
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worst possible

19. Pain caused by light touch *none*

0	1	2	3	4	5	6	7	8	9	10
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worst possible

20. Itching *none*

0	1	2	3	4	5	6	7	8	9	10
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worst possible

21. Tingle or 'pins and needles' *none*

0	1	2	3	4	5	6	7	8	9	10
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worst possible

22. Numbness *none*

0	1	2	3	4	5	6	7	8	9	10
---	---	---	---	---	---	---	---	---	---	----

worst possible

Self-Efficacy (sense of perceived personal control)

Please indicate the extent you agree or disagree with the following statements

	Strongly disagree (1)	2	3	4	Strongly agree (5)
I will be able to achieve most of the goals that I have set for myself					
When facing difficult tasks, I am certain that I will accomplish them					
In general, I think that I can obtain outcomes that are important to me					
I believe I can succeed at most any endeavour to which I set my mind					
I will be able to successfully overcome any challenges					
I am confident that I can perform effectively on many different tasks					
Compared to other people, I can do most tasks very well					
Even when things are tough, I can perform quite well					

Fatigue (Neuro-QoL – Fatigue)

Please choose the number on the 7-point scale below that best represents your level of agreement with each of the statements below;

	Completely Disagree 1	2	3	4	5	6	Completely agree 7
My motivation is lower when I am fatigued							
Exercise brings on my fatigue							
I am easily fatigued							
Fatigue interferes with my physical functioning							
Fatigue causes frequent problems for me							
My fatigue prevents sustained physical functioning							
Fatigue interferes with carrying out certain duties and responsibilities							
Fatigue is among my 3 disabling symptoms							
Fatigue interferes with my work, family and/or social life							

Cognitive Function (Neuro-QoL Cognitive Function)

In the past 7 days.....

	Never	Rarely (once)	Sometimes (2-3 times)	Often (Once a day)	Mildly agree
I had to read something several times to understand it					
I had trouble keeping track of what I was doing if I was interrupted					
I had difficulty doing more than one thing at a time					
I had trouble remembering new information, like phone numbers or simple instructions					
I had trouble thinking clearly					
My thinking was slow					
I had to work really hard to pay attention or I would make a mistake					
I had trouble concentrating					

Cognitive Function (Neuro-QoL Executive Function)

How much difficulty do you currently have.....?

	None	A little	Somewhat	A Lot	Cannot do
Checking the accuracy of financial documents (e.g. bills, check book, or bank statements)					
Counting the correct amount of money when making purchases					
Reading and following complex instructions (e.g. directions for a new medication?)					
Planning for and keeping appointments that are not part of your weekly routine (e.g. a doctor's appointment)					
Managing your time to do most of your daily activities					
Taking care of complicated tasks like managing a checking account or getting finances fixed					
Keeping important personal papers such as bills, insurance documents and tax forms organised					
Learning new tasks or instructions					

Social support (Multidimensional Scale of Perceived Social Support)

We are interested in how you feel about the following statements. Please read each statement carefully. Indicate how you feel about each statement

	Very strongly disagree	Strongly disagree	Mildly disagree	Neutral	Mildly agree	Strongly agree	Very strongly agree
There is a special person who is around when I am in need							
There is a special person with whom I can share my joys and sorrows							
My family really tries to help me							
I get the emotional help and support I need from my family							
I have a special person who is a real source of comfort to me							
My friends really try and help me							
I can count on my friends when things go wrong							
I can talk about my problems with my family							
I have friends with whom I can share my joys and sorrows							
There is a special person in my life who cares about my feelings							

My family is willing to help me make decisions							
I can talk about my problems with my friends							

Thank you very much for completing this questionnaire and for participating in the study.