Individuals’ and Parents’ of Individuals Living with SMA Type II Perspective of Exercise

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Abstract

Spinal muscular atrophy (SMA) is a genetic – autosomal recessive – degenerative neuromuscular condition. SMA causes proximal muscle weakness and muscle atrophy affecting motor function ability and life expectancy. As SMA is currently incurable individuals living with SMA are reliant on medical and allied health interventions to treat their symptoms and prolong their life. SMA Type II is classified as having symptom onset of 6 - 18 months and sitting as the highest motor function achieved. Exercise is a treatment option recommended by the Consensus Statement for Standard of Care in SMA for individuals living with SMA Type II. An individual’s decision to engage in exercise is influenced by their perceptions about exercise and those held by their social network. There is limited research into perceptions of exercise for individuals and parents of individuals living with a degenerative neuromuscular disease (NMD).

The aim of this research was to explore exercise from the perspective of individuals and parents of individuals living with SMA Type II including perceptions about: 1) the effectiveness of individuals’ past and current exercise engagement to treat the symptoms and slow the progression of SMA Type II; 2) the value of exercise in treating the symptoms of SMA Type II; and 3) exercise determinants. Participation was open to individuals of all ages within Australia diagnosed with SMA Type II and their parents or guardians. An explanatory sequential mixed methods methodology was used. The Theory of Planned Behaviour and the social ecological model of physical activity were used as the theoretical lens. An online questionnaire was distributed through the SMA Association of Australia, Muscular Dystrophy Foundation and Muscular Dystrophy Australia with the option for participants to subsequently engage in semi-structured, in-person interviews.

Twenty participants completed the online questionnaire: eight individuals living with SMA Type II and twelve parents of individuals living with SMA Type II. The age range of individuals living with SMA Type II was 1.6 - 36 years (mean 15.4 ± 10.8 years). Eleven participants were interviewed: three individuals living with SMA Type II (21 - 34 years) and eight parents of children living with SMA Type II (3 - 16 years).

Motor function ability exceeded SMA Type II classification in some participants who achieved independent walking (n=4) and standing (n=3). The focus for many individuals and families living with SMA Type II was to achieve quality of life, optimal health, longevity and independence. Many participants (60%) were currently engaged in
exercise; however, only 30% of participants exercised for more than 90 minutes per week. Swimming (90%) and wheelchair soccer (30%) were the most popular types of exercise. Doctors and physiotherapists predominantly advised swimming or hydrotherapy. Swimming, fine motor skills and respiratory exercises were considered by participants to be the most effective types of exercise to treat the symptoms of SMA Type II. For some individuals and families, significant focus was placed on exercise to treat the symptoms of SMA. Regardless of an individual’s or family’s focus, 80% of participants agreed that exercise was, to varying degrees, beneficial in treating the symptoms of SMA Type II. Participants perceived that exercise improved respiratory function (75%), movement (65%), quality of life (75%), mood (75%) and the ability to cope with SMA Type II (60%). Individual, socio-cultural and environmental determinants influenced an individual’s exercise behaviour. Environmental determinants included access to services, facilities, equipment and disability funding. Socio-cultural determinants included a support system, from family and friends to health professionals, and the knowledge and services that they provide. Individual determinants included knowledge about exercise, equipment and SMA; degeneration of an individual’s condition; motivation and enjoyment of exercise; practices limiting risks to health; and financial ability.

Participants exceeded the clinical classification of SMA Type II, which is consistent with other literature. The different types of exercises participants perceived to be beneficial were shown to have physiological, psychological and/or social benefits. Participants’ perceptions of exercise were consistent with the current understanding that exercise further improves quality of life for individuals living with an NMD and may improve physical and psychological health. Exercise determinants were consistent with those of individuals living with a degenerative NMD. As participants’ condition degenerated lack of accessible facilities and exercise knowledge became prominent barriers to exercise. Government policies could improve accessibility, whilst practitioners and schools could improve individuals’ exercise knowledge. Exercise participation of individuals living with SMA Type II could be increased by implementing strategies that address exercise barriers.

This research suggests that the clinical classification of SMA should be revised to consider current knowledge on motor function ability and life expectancy of individuals living with SMA Type II. Increased communication regarding exercise for individuals living with an NMD may increase exercise engagement. Improved
accessibility of facilities, services and equipment, and development and integration of sports could increase exercise engagement. Improved education about exercise in SMA for physicians, allied health professionals, schoolteachers and individuals living with SMA Type II may increase exercise engagement. Further research is needed into exercise determinants, exercise outcomes and strategies to facilitate exercise behaviour for individuals living with SMA Type II. Addressing exercise perceptions could improve exercise participation and subsequently improve morbidity, mobility and the quality of life of individuals living with SMA Type II.
Declaration of Originality

I, Marissa Jane Hoey, declare as the author of this thesis that it is my own work with contributions from my supervisory team- Jane Taylor, Bec Mellifont and Brendan Burkett. This thesis has not been submitted in any form for another degree or diploma at any university or other institute of tertiary education. Information derived from the published and unpublished work of others has been acknowledged in the text and a list of references has been documented.

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I would like to acknowledge the Spinal Muscular Atrophy Association of Australia, Muscular Dystrophy Foundation and Muscular Dystrophy Australia for their support and help throughout this project.

Finally I would like to thank my family and friends who have supported me through this process.

This thesis is dedicated to Narissa.

28/07/2017
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<thead>
<tr>
<th>Abbreviation</th>
<th>Definition</th>
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<tbody>
<tr>
<td>ADLs</td>
<td>Activities of Daily Living</td>
</tr>
<tr>
<td>CF</td>
<td>Cystic Fibrosis</td>
</tr>
<tr>
<td>COPD</td>
<td>Chronic Obstructive Pulmonary Disease</td>
</tr>
<tr>
<td>Consensus Statement</td>
<td>Consensus Statement for Standard of Care in Spinal Muscular Atrophy</td>
</tr>
<tr>
<td>DMD</td>
<td>Duchenne Muscular Dystrophy</td>
</tr>
<tr>
<td>DOSS</td>
<td>Dysphagia Outcome and Severity Scale</td>
</tr>
<tr>
<td>EP</td>
<td>Exercise Physiologist</td>
</tr>
<tr>
<td>ICF</td>
<td>International Classification of Functioning, Disability and Health</td>
</tr>
<tr>
<td>IMT</td>
<td>Inspiratory Muscle Training</td>
</tr>
<tr>
<td>MDA</td>
<td>Muscular Dystrophy Australia</td>
</tr>
<tr>
<td>MDF</td>
<td>Muscular Dystrophy Foundation</td>
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<tr>
<td>MHFMS</td>
<td>Modified Hammersmith Functional Motor Scale</td>
</tr>
<tr>
<td>MMT</td>
<td>Manual Muscle Test</td>
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<tr>
<td>MS</td>
<td>Multiple Sclerosis</td>
</tr>
<tr>
<td>NAIP</td>
<td>Neuronal Apoptosis Inhibitory Protein</td>
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<tr>
<td>NDIS</td>
<td>National Disability Insurance Scheme</td>
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<tr>
<td>NMD</td>
<td>Neuromuscular Disease</td>
</tr>
<tr>
<td>PCF</td>
<td>Peak Cough Flow</td>
</tr>
<tr>
<td>PD</td>
<td>Parkinson’s Disease</td>
</tr>
<tr>
<td>PE</td>
<td>Physical Education</td>
</tr>
<tr>
<td>Physio/s</td>
<td>Physiotherapist/Physiotherapy</td>
</tr>
<tr>
<td>OT</td>
<td>Occupational Therapist</td>
</tr>
<tr>
<td>RMT</td>
<td>Respiratory Muscle Training</td>
</tr>
<tr>
<td>SeDS</td>
<td>Sedentary Death Syndrome</td>
</tr>
<tr>
<td>SF-36</td>
<td>Short Form (36) Health Survey</td>
</tr>
<tr>
<td>SMA</td>
<td>Spinal Muscular Atrophy</td>
</tr>
<tr>
<td>SMAA AAA</td>
<td>Spinal Muscular Atrophy Association of Australia</td>
</tr>
<tr>
<td>SMN</td>
<td>Survival Motor Neuron</td>
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<tr>
<td>VC</td>
<td>Vital Capacity</td>
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<tr>
<td>VFSS</td>
<td>Videofluoroscopic Swallow Study</td>
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<td>WHO</td>
<td>World Health Organization</td>
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Chapter 1  Introduction

Spinal muscular atrophy (SMA) is a neuromuscular condition that primarily affects motor function ability and life expectancy.\(^1\,^2\) SMA is currently incurable; therefore, individuals living with SMA are reliant on medical and allied health interventions to treat their symptoms and prolong their life.\(^3\,^4\) Exercise and perceptions of exercise for individuals living with SMA may provide insight into the reasons why (or why not) an individual decides to exercise. To date, only limited research about exercise perceptions in more general neuromuscular disease (NMD) exists.

SMA has been established as the second most common autosomal recessive disorder and it has been detected globally in different ethnic groups, including Australians.\(^2\,^5\,^7\) SMA is classified into types based on symptom onset and condition severity.\(^8\,^9\) This research focused on individuals living with SMA Type II, which has a symptom onset of 6 - 18 months, motor function ability of ‘never stands’ and an uncertain life expectancy.\(^4\,^6\,^10\) Occupational therapy, physical therapy, speech therapy, nutrition therapy and exercise are recommended allied health interventions that can provide supportive treatments for individuals living with SMA Type II.\(^4\) Individuals living with an NMD are more likely to live a sedentary lifestyle; however physical inactivity can lead to a plethora of poor outcomes: increased deconditioning, increased loss of muscle mass, reduced stamina, increased muscle weakness, increased levels of pain, increased levels of tiredness, obesity and decreased bone density.\(^11\,^12\) Exercise and physical activity has been shown to improve cardiorespiratory function and muscle strength, prevent or reverse physical deconditioning, reduce pain, improve movement and prolong motor function ability for individuals living with an NMD.\(^11\,^12\) Perceptions of exercise influences an individual’s choice to engage in exercise and their level of participation.\(^13\) The Theory of Planned Behaviour can provide an understanding of an individuals’ behavioural decisions, including their decision to engage in exercise.\(^13\) Currently, there is limited research into the perceptions of exercise for individuals and the parents of individuals living with a degenerative NMD.
Chapter 1: Introduction

1.1 Research context

To gain an understanding of how exercise can provide therapy for individuals living with an NMD, and more specifically SMA Type II, a number of documents were reviewed and formed the premise of this research. These include the Consensus Statement for Standard of Care in SMA (Consensus Statement), exercise interventions conducted with individuals living with SMA Type II, exercise advice published by Muscular Dystrophy UK, the Australian Physical Activity and Sedentary Behaviour Guidelines, and the Theory of Planned Behaviour.

The Consensus Statement was devised to fill a void in practice guidelines for the clinical care of individuals living with SMA. It details the common problems associated with SMA and the supportive treatments that can be used to manage this condition. The Consensus Statement can be viewed by practitioners (specialist physicians, general practitioners and allied health professionals) and provides guidance on the minimum level of care for individuals living with SMA. The Consensus Statement recommendations for orthopedic care and rehabilitation, in relation to individuals living with SMA Type II, states ‘Contracture management and exercise are a major focus of treatment, with implementation of regular stretching... Regular exercise should be encouraged to maintain fitness and endurance and might include swimming and adaptive sports’ (p. 1043). In addition to the Consensus Statement, four articles described research that investigated the effect of exercise in individuals living with SMA Type II, which are outlined in Chapter 2.

Furthermore, Muscular Dystrophy UK published a document entitled ‘Exercise Advice for Adults with Muscle-Wasting Conditions’. This document provides general exercise advice for individuals living with an NMD, including the mode, frequency, duration, intensity, precautions and progression of exercise activities. It recommends that individuals living with an NMD should aim to engage in: aerobic exercise for 150 minutes/week; strength training 2 - 3 days/week, completing one set of 8 - 12 repetitions; and stretching daily, completing 2 - 4 sets of 30 - 60 second repetitions. Precautions for exercise include not exercising to exhaustion, avoiding excessive eccentric (muscle lengthening) exercises, awareness of the difference between over-worked muscle weakness and muscle tiredness, protecting joints by exercising in the correct positions and an understanding that exercise should not result in pain. It was recommended that individuals consult their
general practitioner before commencing an exercise regime and seek advice from their physiotherapist or medical team. The exercise advice published by Muscular Dystrophy UK is similar to exercise advice for the Australian population.

The Australian Government Department of Health has published physical activity and sedentary behaviour guidelines for Australians of all ages and abilities. Australian children aged 0 - 5 years are recommended to engage in physical activity intermittently throughout the day for at least 3 hours/day. Australian children aged 5 - 17 years are recommended to engage in physical activity for at least 60 minutes/day, strength training 3 days/week and to minimise sedentary behaviour (time spent sitting), including limiting time spent on electronic devices. Recommendations for adult Australians aged 18 - 64 years are to engage in moderate-intensity physical activity for 150 - 300 minutes/week, vigorous intensity physical activity for 75 - 150 minutes/week, or a combination of the two; strength training 2 days/week; and a reduction in sedentary behaviour. Notably, these exercise recommendations are for Australians of all abilities. Therefore, it is important to further understand and differentiate the exercise perceptions and exercise behaviour of Australian individuals living with SMA Type II, to determine if they are following these guidelines and those set out by the Consensus Statement and Muscular Dystrophy UK.

The decision to engage in exercise, an individual’s exercise behaviour and the effectiveness of exercise to treat the symptoms of SMA Type II is influenced by perceptions about exercise held by the individual and their social network. The Theory of Planned Behaviour shows that attitude, subjective norms and perceived behavioural control impact on an individual’s intentions, which subsequently influences their behaviour. An individual’s exercise beliefs, as well as their evaluation of the likely outcome achieved from exercise engagement will influence their attitude towards exercise. The beliefs held by an individual’s social network (family, practitioners, teachers, peers, etc.) and the individual’s motivation to comply with their social network’s beliefs, creates a subjective norm surrounding exercise behaviour. An individual’s perceptions regarding the level of complexity in engaging in exercise, based on their past experiences and predicted future experiences, influences their perceived behavioural control. Factors that affect exercise behaviour are known as exercise determinants and include facilitators and barriers of exercise. An individual’s perception of facilitators and barriers to exercise and their perceived control over these exercise determinants also contributes to their perceived
behavioural control. This research presents a preliminary exploration of the attitudes, subjective norms and perceived behavioural control, including exercise determinants, that influence the intentions and exercise behaviour of individuals living with SMA Type II.

1.2 Research issue

Exercise is a supportive treatment recommended by the Consensus Statement for individuals living with SMA Type II, and the notion that Australian individuals living with SMA Type II ought to be engaging in exercise is further supported by a variety of other current publications. Engagement and adherence to exercise is dependent upon an individual’s perceptions about the value of exercise and whether they have correctly assessed the risks and benefits associated with conducting exercise activities. Thus, exploring the perceptions of Australian individuals living with SMA Type II, and the parents of these individuals, can help to understand the determinants, facilitators and barriers to exercise for this population. Additionally, understanding can be gained about individuals and parents perceptions of the value and effectiveness of exercise to treat the symptoms and slow the progression of SMA Type II.

1.3 Research aim

This research aims to explore exercise from the perspective of individuals and parents of individuals living with SMA Type II.

1.4 Research objectives

The research objectives of this study are:

1. To explore the perceptions of Australian individuals living with SMA Type II and the parents of individuals living with SMA Type II about the effectiveness of past and current engagement in exercise as a therapy to treat the symptoms and slow the progression of SMA Type II;

2. To explore the perceptions of Australian individuals living with SMA Type II and the parents of individuals living with SMA Type II about the value of exercise in treating the symptoms of SMA Type II; and
3. To explore the perceptions of Australian individuals living with SMA Type II and the parents of individuals living with SMA Type II about exercise determinants.

1.5 Research significance

This research is significant, as limited research has been conducted into the exercise perceptions and exercise behaviour of individuals living with SMA Type II. Degenerative physical conditions are often listed within the exclusion criteria of exercise research into physical disabilities. Currently there is no clear guidance on how children living with an NMD should engage in exercise. It is also unclear if published exercise advice found in the Consensus Statement and ‘Exercise Advice for Adults with Muscle-Wasting Conditions’ is being utilised by individuals living with SMA Type II, their parents and their practitioners.

Individuals living with an NMD, their parents and their practitioners may not know how and how often to engage in exercise. For individuals living with SMA Type II attitudes, subjective norms, perceived behavioural control and exercise behaviour is unknown in Australia and worldwide. Through greater understanding of exercise perceptions and determinants, strategies can be implemented to improve exercise engagement and adherence of individuals living with SMA Type II. This research will also contribute to the limited research conducted into the exercise perceptions of individuals living with an NMD.\(^{17-22}\)

1.6 Research overview

This research used a sequential explanatory mixed methods methodology to explore the exercise perceptions of individuals living with SMA Type II, as well as those amongst their social networks.\(^{23,24}\) An online questionnaire was distributed to individuals and parents of individuals living with SMA Type II through the Spinal Muscular Atrophy Association of Australia (SMAAA), Muscular Dystrophy Foundation (MDF) and Muscular Dystrophy Australia (MDA) associations. Subsequent interviews were conducted with interested participants.

The Theory of Planned Behaviour was used as a framework to analyse the exercise behaviour of individuals living with SMA Type II. Individuals’ attitude can
be influenced by their perceptions of the effectiveness of exercise, value of exercise and exercise determinants. Individuals’ subjective norm can be influenced by exercise determinants. Finally individuals’ perceived behavioural control can be influenced by their perceptions about the effectiveness of exercise and their exercise determinants. The social ecological model of physical activity was used as a framework to analyse the exercise determinants of individuals living with SMA Type II. As individual determinants are so varied, a list of exercise determinants devised from Biddle, Mutrie and Gorely (2015) and Sallis and Hovell (1990) were used to provide clarification. These include socio-demographic, biological, behavioural, cognitive and psychological determinants. Environmental, socio-cultural and individual determinants can influence an individual’s attitude, subjective norm, perceived behavioural control and thus their exercise behaviour. By understanding exercise perceptions and exercise determinants, strategies can be implemented to improve exercise behaviour of individuals living with SMA Type II, which may offer improvements in physical and psychological health.

1.7 Thesis structure

This thesis is comprised of seven chapters. This chapter introduced SMA and the context for this research, including the Consensus Statement, exercise guidelines for individuals living with an NMD, exercise guidelines for Australians, the Theory of Planned Behaviour and the social ecological model of physical activity. The research issue, aim, objectives, significance and overview were also presented.

Chapter Two provides a comprehensive overview of SMA and the effect exercise has on individuals living with SMA Type II. Chapter Three is a literature review presenting the determinants for exercise from the perspective of individuals living with an NMD. Chapter Four outlines the research design, including the research ethics, theoretical framework, methodology, researcher bias, and data collection and analysis methods.

Chapter Five presents the research findings in relation to the research objectives and theoretical framework. Participants’ exercise behaviour, the effectiveness of exercise and the value of exercise to treat symptoms and slow the
progression of SMA Type II are detailed. Participants’ exercise determinants are described and categorised into individual, socio-cultural and environmental determinants, as outlined by the social ecological model of physical activity.

Chapter Six provides a discussion of the research findings, presenting novel insight into the exercise perceptions of Australian individuals living with SMA Type II. The research findings are positioned within the current literature, including the Consensus Statement, the Theory of Planned Behaviour and the social ecological model of physical activity.

Chapter Seven provides an overall conclusion for the research and recommendations for future research, practice and policy.
Chapter 2 Overview of spinal muscular atrophy

2.1 Introduction

The previous chapter presented the thesis context, issue, aim, objectives, significance, overview and structure. This chapter provides an overview of spinal muscular atrophy (SMA), as a foundation for understanding and interpreting this research. The incidence, classification, diagnosis, genetics and life expectancy for individuals living with SMA Type II are outlined. The effects of SMA Type II progression on continence, intellectual ability, fertility and pregnancy, motor function ability, orthopaedic conditions, respiratory conditions, gastrointestinal and nutritional concerns and palliative care are included. The quality of life for individuals living with SMA Type II is explored, incorporating psychological health and interactions with family and friends. Clinical trials and supportive treatments for SMA Type II are detailed, encompassing clinical trials for genetic and drug therapies, drug interventions, respiratory interventions, orthopaedic interventions and gastrointestinal and nutritional interventions. Finally, exercise as a supportive treatment for SMA Type II is highlighted, with a detailed review of exercise interventions that have investigated the effects of exercise for individuals living with SMA Type II.

2.2 Overview of SMA

SMA is a genetic – autosomal recessive – degenerative neuromuscular condition. SMA affects the anterior horn cells of the spinal cord, causing proximal muscle weakness and muscle atrophy. It is a complex condition that affects multiple systems within the body. Signs and symptoms of SMA include low muscle tone and symmetrical proximal weakness with full sensory sensation; the absence or diminishment of tendon reflexes; and leg weakness more prominent than arm weakness. The primary care physician should provide individuals and/or parents of individuals living with SMA with online resources, information about SMA advocacy groups and information about clinical trials and genetic counseling. According to the Consensus Statement, a multidisciplinary practitioner approach, including neurologists, respiratory and orthopaedic specialists, geneticists, general practitioners or paediatricians, nutritionists, occupational therapists and physiotherapists, is required to achieve the best possible outcome for individuals
living with this condition. There is great variation in the care and outcomes for individuals living with SMA, due to differences in the extent of practitioners’ knowledge about SMA and its complexities, family resources and care between regions and cultures. It is reported that 1 in 35 to 1 in 50 people are carriers of SMA, and approximately 1 in 6,000 to 1 in 10,000 live births result in a baby born with SMA. The carrier frequency in Australia and New Zealand, specifically, is estimated to be 1 in 49. Approximately 29% of individuals with the condition have SMA Type II. However, the SMA patient registry is voluntary; therefore, there is no current or accurate record of how many Australians live with SMA Type II. Approximately 100 Australians were known to be living with SMA Type II in 2016, which included both members and non-members of Spinal Muscular Atrophy Association of Australia (SMAAA) (J. Cini, CEO of SMAAA, oral communication, August 2016). Statistically, Australia should have more than 100 individuals living with SMA Type II. However, inaccurate statistical records could mean that Australians living with SMA Type II are at risk of being underserviced and lack the information they need to make informed decisions about their life and medical care.

SMA is classified into categorical types based on age of onset and the highest level of motor function achieved, which can indicate natural age of death (Table 1). SMA Type II has been classified as having an onset of symptoms at between 6 - 18 months of age, where the individual never stands or walks unaided, and the natural age of death occurs after 2 years.

Table 1. Clinical classification of SMA detailing symptom onset, motor function ability and life expectancy

<table>
<thead>
<tr>
<th>SMA Type</th>
<th>Symptom Onset</th>
<th>Highest Motor Function Achieved</th>
<th>Life Expectancy</th>
</tr>
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<tbody>
<tr>
<td>Type I</td>
<td>&lt;6 months</td>
<td>Never sits</td>
<td>&lt;2 years</td>
</tr>
<tr>
<td>Type II</td>
<td>7 - 18 months</td>
<td>Never stands</td>
<td>&gt;2 years</td>
</tr>
<tr>
<td>Type III</td>
<td>&gt;18 months</td>
<td>Stands and walks</td>
<td>Adult</td>
</tr>
<tr>
<td>Type IV</td>
<td>Second or third decade</td>
<td>Walks during adult years</td>
<td>Adult</td>
</tr>
</tbody>
</table>

The Consensus Statement states of individuals living with SMA Type II:

“Some learned to achieve independent sitting, whereas others need help to sit up. The defining characteristic is an ability to maintain a sitting position unsupported. At the strongest end of this category are
SMA is a condition with a continuous spectrum scale, due to the intricate complexities and unique genetic manifestations within individuals. Currently, there are five recognised types of SMA, in addition to several other forms of the condition, such as autosomal dominant SMA, x-linked SMA and lower extremity dominant SMA. Literature often cites the clinical classification of SMA and does not include recent research which has found improvements in motor function ability and life expectancy for individuals living with SMA Type II (Table 2).

Table 2. Impact of recent research on the classification of SMA

<table>
<thead>
<tr>
<th>SMA Type</th>
<th>Symptom Onset</th>
<th>Highest Motor Function Achieved</th>
<th>Life Expectancy</th>
</tr>
</thead>
<tbody>
<tr>
<td>Type 0</td>
<td>Prenatal</td>
<td>None</td>
<td>≤ 6 months</td>
</tr>
<tr>
<td>Type I</td>
<td>&lt;6 months</td>
<td>Sitting with support</td>
<td>≤ 2 years, 6% - 30% live to 4 years, 0% - 30% live to 20 years, oldest reported 35 years</td>
</tr>
<tr>
<td>Type II</td>
<td>6 - 18 months</td>
<td>≈ 75% sit independently, ≈ 25% stand, 29 reported cases of walking</td>
<td>&gt;2 years, ≈ 90% live to 20 years, ≈ 50% live to 40 years, oldest reported 69 years</td>
</tr>
<tr>
<td>Type III (a &amp; b)</td>
<td>&gt;12 months</td>
<td>Independent ambulation, frequent falls and loss of function &gt;2 - 4 years</td>
<td>Normal</td>
</tr>
<tr>
<td>Type IV</td>
<td>Adult onset 20 - 40 years</td>
<td>Independent ambulation, will lose some function</td>
<td>Normal</td>
</tr>
</tbody>
</table>

The vast genetic variation seen between individuals, even those classified within the same type of SMA, could impact the exercise perceptions and behaviour of these individuals. Diagnosis and severity of SMA can be determined through genetic testing by examining the survival motor neuron (SMN) gene, which produces SMN protein. SMN protein is essential for the survival and maintenance of muscles and motor neurons. Muscle weakness and atrophy occur before motor neuron degeneration, indicating that the SMN protein is particularly essential to muscle maintenance and survival.
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The SMN gene has two copies: SMNT (SMN1) and SMNC (SMN2). In healthy populations, the SMNT gene successfully expresses SMN protein 90 - 100% of the time, whereas the SMNC gene is 20 - 30% successful in producing the SMN protein. In approximately 95% of cases, SMA occurs as a result of the deletion or mutation of the SMNT gene (Figure 1). Therefore, individuals diagnosed with SMA are dependent upon the reduced capacity of the SMNC gene to produce the SMN protein.

![Figure 1. Comparison of SMN genes and synthesis of SMN protein (%) in healthy & SMA populations](image)

The quantity of SMNC gene that an individual possesses determines the SMN protein concentration, and, thus the severity of SMA symptoms. A higher number of SMNC genes results in increased production of the SMN protein and a mild severity of SMA symptoms are displayed, whereas a lower number of SMNC genes results in decreased production of protein and more severe symptoms. It has been evaluated that 68 - 82% of individuals with SMA Type II have three copies of the SMNC gene. Typically, individuals with Type I have less than three copies of the gene and individuals with Type III have more than three copies.

The severity of SMA is not solely determined by SMNC gene number. Other factors include interactions of proteins (SMN, follistatin etc.) in neuron axon transport, at the neuromuscular junction and/or at the muscle. Also, other genes, such as the Neuronal Apoptosis Inhibitory Protein (NAIP) gene and XS2G3 cDNA gene can impact on SMA severity. This explains the phenomenon of differences between individuals who have been diagnosed with the same SMA type and same number of SMNC genes, but who present with varying forms of severity. These genetic differences can influence individuals’ life expectancy, motor function and
their physiological response to exercise intervention, which may impact upon the individual’s exercise perceptions and participation.

Four studies have investigated life expectancy for individuals living with SMA Type II (Table 3). Zerres and colleagues (1997) conducted two rounds of data collection, one in Poland starting in 1960 and one in Germany starting in 1985. All four studies used the Kaplan-Meier method to calculate life expectancy, which has several assumptions that can increase or decrease survival probabilities compared with actual life expectancy data. The extent of actual data recorded in these four studies compared to the employment of the Kaplan-Meier equation to calculate survival probabilities is unknown. Additionally, the Kaplan-Meier method addresses survival rate, but does not account for prospective medical interventions that can be implemented to keep individuals alive or alter the quality of life experienced by individuals living with SMA Type II.

**Table 3. Published life expectancies of SMA Type II individuals**

<table>
<thead>
<tr>
<th>Study</th>
<th>Sex</th>
<th>Participant</th>
<th>Survival Probabilities (%) of SMA Type II Individuals at a certain age (years)</th>
<th>Combined*</th>
</tr>
</thead>
<tbody>
<tr>
<td>Zerres</td>
<td></td>
<td>Poland (1960)</td>
<td>107 M:F 48:59 20 97.8% 96.1% 77.5% 68.2% 59.1%</td>
<td>402 205:197 43 100% 100% 98.25% 95.77% 84.84% 78.11% 67.91% 52% 4.84% 30% 0%</td>
</tr>
<tr>
<td>Zerres</td>
<td></td>
<td>Germany (1985)</td>
<td>133 M:F 70:63 11 99.1% 99.1% 87.1% 79.6% 75.4% &lt;30%</td>
<td>105 62:43 3 100% 100% 97% 92% 92% 92%</td>
</tr>
<tr>
<td>Chung</td>
<td></td>
<td>China (2004)</td>
<td>26 M:F 9:17 4 100% 100% 100% 92% 92% 92%</td>
<td>31 16:15 5 100% 100% 97% 93% 93% 93% 52% 0%</td>
</tr>
<tr>
<td>Ge</td>
<td></td>
<td>China (2012)</td>
<td>105 M:F 62:43 3 100% 100% 97% 92% 92% 92%</td>
<td>105 62:43 3 100% 100% 97% 92% 92% 92%</td>
</tr>
<tr>
<td>Farrar</td>
<td></td>
<td>Australia (2013)</td>
<td>31 M:F 16:15 5 100% 100% 97% 93% 93% 93% 52% 0%</td>
<td>31 16:15 5 100% 100% 97% 93% 93% 93% 52% 0%</td>
</tr>
</tbody>
</table>

Zerres results have been separated due to differences in life expectancy

*Combined data is calculated from published figures, not raw data

Medical advances and higher levels of multidisciplinary care have improved life expectancy of individuals living with SMA Type II. Notably, the study by Zerres and colleagues (1997) included data from 1960, while Ge and colleagues
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(2012) states that individuals living with SMA Type II residing in China receive lower standards of care compared to those living with the condition in Europe and the United States of America. These two studies account for 85.4% (n= 345) of the combined results for life expectancy of SMA Type II. Farrar and colleagues (2013) present a more recent Australian study, and, hence, may be deemed more relevant for comparison in first world countries, despite smaller participant numbers. With improved life expectancy, more focus is placed on reducing morbidity and improving the quality of life of individuals living with SMA. Exercise is a supportive treatment that could reduce morbidity and improve mobility and the quality of life for individuals living with SMA Type II. Furthermore, exercise participation could be improved by implementing strategies that address the exercise perceptions of individuals living with SMA Type II.

2.3 The effects of SMA Type II progression on functional abilities

Individuals face numerous challenges and conditions that manifest from the muscle degeneration and atrophy caused by SMA. For the majority of individuals living with SMA Type II, continence, intellectual ability and fertility are not affected. However, individuals living with SMA Type II encounter the following challenges and conditions:

1. Delayed ability to achieve motor function, followed by loss of function later in life resulting in issues (e.g. inability to sit unsupported);
2. Orthopaedic complications (e.g. kyphoscoliosis);
3. Respiratory problems (e.g. sleep disordered breathing); and
4. Gastroenterological and nutritional concerns (e.g. swallowing problems).

Individuals living with SMA Type II eventually need palliative care, unless circumstances dictate otherwise. The effects of SMA Type II progression on functional abilities can be biological determinants for exercise behaviour. Strategies that address exercise determinants and perceptions of individuals living with SMA Type II could improve their exercise participation and further research is needed in this area.
2.3.1 Continence

Urinary and bowel incontinence can be more prevalent in individuals living with SMA Type I and II. However, for the majority of these individuals, this condition is more likely due to the misconception that incontinence is a symptom of SMA and/or the reliance on others to go to the bathroom, than an actual result of muscle weakness.

2.3.2 Intellectual ability

SMA does not affect the intellectual ability of individuals. In fact, many individuals living with SMA have normal cognitive abilities and may have a high intellectual ability, particularly verbal intelligence, compared to their peers. A contributing factor for this may be that the compromised physical abilities of individuals living with SMA leads them to choose intellectual pursuits, rather than physical pursuits. However, research has found that SMA can cause atrophy in the brain, irrespective of hypoxia, and can affect visual and sensory nerves.

2.3.3 Fertility and pregnancy

Fertility is not impaired for individuals living with SMA. However, all children born to individuals living with SMA will be carriers of the condition. The probability of an individual living with SMA having a child with SMA is one in 1,340, provided that the reproductive partner has at least two copies of the SMN gene. There are several documented cases of women living with SMA Type I, II and III who have become pregnant and successfully carried their babies to term. The effects of pregnancy on women living with SMA Type I, II or III are varied; some have no permanent change to their condition, whilst the condition of others declines.

2.3.4 Motor function ability

As the condition of individuals living with SMA Type II degenerates, a concurrent gradual decrease in motor function ability is experienced. Motor function ability is a common concern for parents and individuals living with SMA Type II, as individuals living with SMA Type II require self-care and mobility assistance. There are documented cases of individuals living with SMA Type II who have the ability to walk independently or with assistance; however, this is uncommon. Standing was the highest motor function achieved for 24.4% (n=41)
of participants living with SMA Type II in Zerres and colleagues (1997) research and 45% (n= 76) experienced weakness in the distal segment of their arm before 5 years of age. The ability to sit independently was lost before 9 years of age in 11.4% (n= 12) of participants living with SMA Type II in Ge and colleagues (2012) research. The loss of mobility increases sedentary behaviour, which is associated with increased morbidity and mortality.\textsuperscript{74}

### 2.3.5 Orthopaedic conditions

Limitations to the motor function ability of individuals living with SMA Type II result in the formation of joint contractures, spinal deformations (such as kyphoscoliosis), pelvic obliquity, hip dislocation, increased risk of pain, osteopenia and fractures.\textsuperscript{4,75} Kyphoscoliosis can also contribute to poor respiratory function.\textsuperscript{4}

### 2.3.6 Respiratory conditions

Cardiorespiratory illness should be prominently considered when exploring conditions that manifest in individuals with SMA Type II, as it is a major cause of morbidity and mortality.\textsuperscript{4} Weakness of the respiratory muscles, ineffective cough and swallowing dysfunction (which can cause aspiration) may all contribute to chest infections.\textsuperscript{4} Zerres and colleagues (1997) reported that 52% (n=125) of participants living with SMA Type II experienced pneumonia before 5 years of age and Ge and colleagues (2012) found that 24.7% (n=26) of participants, all less than 9 years of age, were hospitalised due to pneumonia. Sleep disordered breathing is also prominent in individuals living with SMA Type II, and eventually develops into daytime respiratory failure.\textsuperscript{4} The cause of death for eight out of nine individuals was attributed to cardiorespiratory illness between two studies examining survival rate of SMA Type II.\textsuperscript{2,6} Exercise may improve the cardiorespiratory function of individuals living with SMA Type II, as it has shown to improve respiratory function of individuals living with SMA Type III.\textsuperscript{76,77}

### 2.3.7 Gastrointestinal and nutritional concerns

Muscle weakness, particularly of facial and respiratory muscles, leads to swallowing, feeding and gastrointestinal problems.\textsuperscript{4} These include: fatigue with feeding, difficulty or inability to self-feed, limited ability to open the mouth, weak bite force and poor swallowing coordination. These difficulties can lead to choking, aspiration and reflux, and may eventuate into pneumonia and/or potential death;
constipation, which can impact respiratory function for individuals using abdominal muscles for respiration; prolonged meal times, increasing fatigue and reducing time spent on other activities; and becoming under or over weight, which can have health and mobility implications.\textsuperscript{4}

2.3.8 Palliative care

Supportive treatments may prolong the life and independence of individuals living with SMA Type II and whilst some individuals may die suddenly from causes such as pneumonia, others may need to access palliative care services. It is desirable for interventional supportive care, such as gastrostomy tube and tracheotomy, to be undertaken early rather than in emergency situations.\textsuperscript{4,78} Tracheotomy is a treatment option that individuals and parents should be aware of; however, the Consensus Statement raises concerns about the potential of the individual and their quality of life for this particular treatment.\textsuperscript{4} The Consensus Statement advises that tracheotomy should be avoided if possible, and carefully discussed if parents raise it as a treatment option.\textsuperscript{4} Palliative care services and trained staff may be better suited to provide care than untrained support workers during periods of acute illness and terminal care.\textsuperscript{78} However, it should be noted that hospital and hospice care staff are generally not well informed about the care needs for individuals living with SMA.\textsuperscript{78} Following the death of an individual who lived with SMA, families are best supported in their bereavement by counseling professionals who have had an established rapport with the family whilst the individual was still alive.\textsuperscript{78} In part this may be due to misconceptions that practitioners, researchers and the general population often have about the quality of life experienced by individuals living with SMA Type II.

2.4 Quality of life

Adults and adolescents living with SMA Type II have stated that they have a high quality of life.\textsuperscript{44,79} Whilst children living with SMA may develop behavioural or psychological disorders, the risk of this is low and no different to the risk of any child developing such a disorder.\textsuperscript{80} However, siblings of individuals living with SMA Type II do have a statistically higher risk of developing behavioural problems than their peers or the sibling living with SMA Type II.\textsuperscript{80} The majority of individuals living
with SMA Type II complete secondary education, participate in tertiary education, find employment and actively direct their own life.44,81

Individuals living with SMA Type II are not precluded from making friends, and family and friends are considered to be an important aspect of life for individuals living with SMA.44 Adult individuals living with SMA Type II have, on average, seven close friends.44 In 2007 adults within the general population typically had five to eleven friends that they contacted weekly.82 While individuals living with SMA Type II are statistically more likely to be single compared to the general population, the majority of adult individuals living with SMA Type II have engaged in romantic relationships, some marry or cohabitate with their partner and some become parents.44

Friendships, family relationships and social acceptance influence the perceptions of, and participation in, exercise for individuals living with a disability.20,83 Exercise has been shown to be beneficial in improving quality of life and psychological health for individuals living with an neuromuscular condition.22,84,85 By increasing exercise participation through improved understanding of exercise perceptions, the quality of life and psychological health of individuals living with SMA Type II could be further improved.13,22,84,85

2.5 Clinical trials and supportive treatments

SMA is currently incurable; however, clinical trials are being conducted to find an effective treatment or cure for this condition.3,4,34,86-88 The focus of clinical trials are drug and genetic therapies that target either SMN \textsuperscript{T} (SMN1) gene replacement, modulation of the SMN \textsuperscript{C} (SMN2) gene, neuroprotection of motor neurons or muscle protection.89 In the interim, sub-optimal drug therapies, respiratory therapies, orthopaedic therapies, nutrition and exercise are supportive treatments that have been shown to improve the lives of individuals living with SMA.3,4,34,86-88

Two clinical trials are currently investigating stem cell therapy as a treatment for individuals living with SMA, but results are yet to be published.90 Seven clinical trials are currently investigating drug therapies, including Valproate, Levocarnitine, Pyridostigmine, Celecoxib and LM1070, as treatments for SMA.90 SPINRAZA is the first drug designed to specifically treat SMA Type I, II and III; however, not all participants were responsive to the drug in the clinical trial.89,91 SPINRAZA was approved by the United States Food and Drug Administration on 23\textsuperscript{rd} December 2016.
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and is currently seeking approval from the European Medicines Agency; it is not yet available for Australians. Individuals in the trial who responded to treatment showed improved motor function ability and life expectancy. The side effects of the drug, however, include increased risk of respiratory infections, atelectasis (collapsed lung), constipation, thrombocytopenia (low platelet count) and coagulation abnormalities, as well as potential renal toxicity. SPINRAZA will therefore have a dual effect upon individuals exercise behaviour. Improved functional ability may increase exercise choices and improve exercise engagement however the side effects of SPINRAZA may place restrictions on exercise engagement.

Respiratory interventions for individuals living with SMA Type II include supportive treatments for ineffective cough, daytime and sleep disordered breathing, airway clearance techniques and limiting risk factors to health. Manual and mechanical cough assistance is recommended, and can be utilised daily, if needed, to treat ineffective cough. Non-invasive ventilation is recommended to treat daytime and sleep disordered breathing. Tracheotomy is another a treatment option, as previously discussed. Secretion mobilisation techniques, such as chest physiotherapy, postural drainage and oral suctioning, are recommended for airway clearance. Vaccinations and early use of antibiotics are also recommended to limit preventable health risks. Exercise is also a supportive treatment that may improve the respiratory function of individuals living with SMA Type II, as it has been shown to improve the respiratory function of individuals living with SMA Type III.

Orthopaedic interventions for individuals living with SMA Type II include surgery, orthotics, adaptive aids and exercise. Surgery for scoliosis improves sitting balance, endurance and may improve or slow the rate of decline in pulmonary function. Serial casting and regular stretching to address joint contractures in conjunction with knee-ankle-foot orthotics, standing frames and walking aids can also assist individuals living with SMA Type II who have sufficient strength to stand and walk. Manual or power mobility and assistive technology, including exoskeletons, can also improve functional ability, independence and social ability of individuals living with SMA Type II. Exercise ability could also be improved by orthopaedic interventions.

Physiotherapists, occupational therapists, speech therapists and/or nutritionists should be consulted when implementing gastrointestinal and nutritional interventions. These may include changes to food consistency and thickened liquids.
to reduce the risk of choking and aspiration. Limiting high fat foods may reduce the risk of reflux. A semi-solid diet may reduce muscle fatigue, improve mastication and reduce time spent eating. Inserting a gastronomy tube is a treatment option if oral feeding is not meeting nutritional needs or is unsafe. Whilst nutritional and other interventions have improved the quality of life and life expectancy of individuals living with SMA Type II, new research into the effects of exercise is showing that it is a supplementary treatment that is its own medicine as detailed in section 2.6.

2.6 Exercise as a supportive treatment

Exercise can improve muscular strength, endurance, cardiovascular function and motor function in healthy populations. However, 11 Cochrane Reviews investigating the effect of exercise in individuals living with NMDs all determined that more research is needed before it can be definitively established that exercise is beneficial as a therapy.

To date, research has demonstrated that exercise can make physiological changes to the human body, and the type of exercise influences the physiological adaptation. Strength training enhances muscle hypertrophy, which increases DNA production and, subsequently, increases protein production within muscles. Therefore, strength training may reduce the severity of SMA through increased production of SMN protein, which is deficient in individuals living with SMA and vital for muscle maintenance and the prevention of muscle atrophy. Additionally, strength training may help to combat muscle weakness experienced by individuals living with SMA, allowing them to maintain motor function abilities for longer. Aerobic training increases cardiac output and capillary density, improving oxygen delivery to muscles, thereby increasing an individual’s capacity to produce energy. Individuals living with SMA have reduced capillary numbers within their muscles, reducing their body’s capacity to deliver oxygen and remove waste. Therefore, aerobic training may improve energy levels and improve cardiorespiratory output for individuals living with SMA. Compared to healthy individuals, individuals living with SMA Type II have higher levels of adipose tissue, which increases morbidity and decreases motor function. Exercise can change body composition by reducing adipose tissue and increasing lean muscle mass. Therefore, individuals living with SMA Type II may increase the longevity of their life and improve motor function by engaging in exercise through improved cardiorespiratory
output, improved motor function, increased protein synthesis, increased energy levels and reduced morbidity.

To date four exercise interventions have investigated the effect of exercise in individuals living with SMA Type II. Three investigated the effect on motor function ability and one on respiratory function. Motor function ability is a prominent concern for individuals living with SMA Type II, as decreased function leads to loss of independence, a restriction on social opportunities, muscle atrophy and can increase pain levels. 

Respiratory function is also a prominent concern for individuals living with SMA Type II, as previously discussed. The four exercise interventions are outlined and evaluated to understand the effect exercise has on individuals living with SMA Type II.

The first intervention occurred over a two year period and examined the effects of physiotherapy and hydrotherapy as a supportive treatment for SMA in 50 participants; 30 with SMA Type II and 20 with SMA Type III. Physiotherapy occurred once per week, while hydrotherapy in a 30°C pool took place twice per week for 30 minutes in children and 45 minutes in adults. Despite the two year intervention, orthopaedic conditions, including scoliosis, kyphosis and the degree of obliquity in hips, knees and ankles, worsened. Still, the authors concluded that muscular strength stabilised and psychological benefits occurred in participants living with SMA Type II. However, these were subjective conclusions, as muscular strength and psychological health was not actually measured. The authors cited the unreliability of manual muscle testing (MMT) in children, and psychological assessments were not reportedly conducted before, during or after treatment. According to the Barthel Ladder scale, a measure of functional independence for personal care and mobility, improvements in the performance of daily activities were seen in participants living with SMA Type II. Nine participants gained motor function abilities during the intervention, five gained the ability to sit and four gained the ability to stand; however four lost the ability to walk. It could be concluded from this study that exercise can improve motor function, but does not arrest the progression of the condition. This intervention did not impact the progression of bone obliquities and, due to lack of formal testing, strength and psychological progression were not scientifically determined. It is important to note, though, that only 30 participants with SMA Type II were recruited for this intervention, which is a small
sample size compared to interventions with other populations. However, this is still the largest group of participants living with SMA Type II in all reported interventions.

The second intervention was a clinical trial that implemented a 12-week home-based strength training program supervised by physiotherapists. 30 Nine participants aged between 5 - 21 years, diagnosed with SMA Type II (n=6) and III (n=3) completed the intervention. 30 Adherence to the intervention was high at 90.4%, no adverse events occurred and participants in 99.5% of cases felt no pain. 30 Significant increases in participants’ mean amount of weight lifted without change in perceived exertion, and significant improvements for strength using MMT measurements were found. 30 A non-significant increase in strength was found using quantitative muscle analysis and no change in strength was found using hand-held dynamometry. 30 A significant increase in motor function using the Modified Hammersmith Functional Motor Scale (MHFMS) was found; however, this improvement was still within the range of variability for this test, leading to ambiguous clinical relevance. 30 Five participants’ MHFMS scores increased, one participant had no change and two participants’ scores decreased. 30 Lewelt and colleagues (2015) stated that some participants had ‘meaningful improvements’ in motor function ability, including one participant who was able to independently and safely climb four stairs after the exercise intervention; a task that could not be accomplished previously. However, observable functional ability was not considered to be a measurable outcome prior to the implementation of the intervention, so was not included in baseline testing. 30 This study highlights that progressive resistance strength training can be beneficial for individuals living with SMA Type II although results can be variable between individuals. This may be an indication of the genetic variability amongst individuals living with SMA Type II and/or that individuals’ exercise determinants could influence the outcome of their exercise performance.

The third intervention was a case study of a 25 year old man living with SMA Type II who was treated for swallowing difficulties three times per week for 30 minutes over seven months. 28 Treatment included flexibility exercises for the neck and temporomandibular joint, together with passive and active oropharyngolaryngeal exercises. 28 The dysphagia outcome and severity scale (DOSS), which rates functionality of swallowing between 1 (lowest functionality) and 7 (highest functionality), was used on the individual to determine the effectiveness of the intervention. 28 The participant’s DOSS score was 1 pre-treatment and improved to 3
A videofluoroscopic swallow study (VFSS) showed the participant’s inability to swallow without aspirating due to an incomplete closure of the velopharyngeal port before treatment. After treatment, VFSS showed that complete closure of the velopharyngeal port was achieved, resulting in the participant swallowing without aspiration. It can be concluded that in this case study, exercise improved swallowing function; however, more participants are needed before a proper evaluation of this supportive treatment can be made.

The fourth intervention was a study of eleven participants living with SMA Type II aged 6 - 16 years, who were recruited to investigate glossopharyngeal pistoning, a breathing technique used to improve vital capacity (VC) of the lungs. While eleven participants were recruited, only five participants were able to learn the glossopharyngeal pistoning technique. The five participants were treated 4 days per week for 8 weeks, but one participant lost motivation throughout the intervention. VC and chest expansion did not reportedly improve post-intervention although discrepancies were found in the study. Firstly the authors reported, ‘The training compliance for all four children who learned the technique was 100%.’ (p. 1327); however, five of eleven participants learnt the technique. Secondly, the authors stated in the results section ‘There were no changes in VC’ (p. 1326) and ‘The median GIV (Glossopharyngeal Insufflation Volume = VC + Glossopharyngeal pistoning) for the five children who learned the technique was 0.28L... ’ (p. 1326); however, in the discussion section, it was stated ‘...and they increased VC (median change 0.28L)’ (p. 1327). Based on these results, it would appear that glossopharyngeal pistoning is not an effective supportive treatment for individuals living with SMA Type II. However this research highlights that exercise perceptions and determinants may influence an individual’s exercise participation as loss of motivation was cited as a factor to discontinue treatment for one participant.

The current research shows that for individuals living with SMA Type II, exercise can improve gross motor skills and swallowing function; however, the participant groups in the four exercise interventions outlined are very small compared to research participation in other populations. Two studies observed improvements in motor function ability, but did not use valid, measurable tests in their research. Therefore, observable functional task testing should be considered for future studies amongst this population group.
2.7 Conclusion

SMA Type II is a complex genetic condition that affects life expectancy, motor function ability, orthopaedic health, respiratory function, gastrointestinal function and the relationships of individuals living with this condition. Supportive treatments for this condition include drug, respiratory, orthopaedic, and nutritional and exercise interventions. These are all factors that could influence an individual’s exercise perceptions and participation. Published literature on exercise perceptions and exercise determinants of individuals living with an NMD provides a foundation to understand exercise perceptions and participation of individuals living with SMA Type II, and this is explored further in Chapter Three.
Chapter 3 Literature review: Exercise determinants for individuals living with a neuromuscular disease

3.1 Introduction

The previous chapter provided an overview of spinal muscular atrophy (SMA), the effects of SMA Type II progression on functional abilities, the quality of life of individuals living with this condition, current clinical trials and supportive treatments, exercise as a supportive treatment and palliative care. This chapter reviews the published literature about exercise determinants for individuals living with an neuromuscular disease (NMD). Research has shown that exercise determinants, attitude, subjective norms and perceived behavioural control influence an individual’s decision to exercise.\textsuperscript{13,27} By understanding this effective interventions can be developed and implemented for individuals living with an NMD, including SMA Type II. Implementing effective exercise interventions and increasing individuals’ participation in physical activity and exercise could improve motor function ability in individual’s living with SMA Type II, and may offer other health benefits.\textsuperscript{13,22,28-30}

This literature review explores the exercise and physical activity perceptions of individuals living with an NMD to gain a better understanding of their exercise determinants. Rather than exploring all individuals living with an NMD, literature on individuals living with a degenerative NMD who were phenotypically similar to individuals living with SMA Type II was included. It was presumed that this body of literature would best inform exercise determinants of individuals living with SMA Type II. The literature review method is outlined, including the search strategy, selection of literature, review strategy and search results. The literature review findings detail the exercise determinants for individuals living with an NMD, study conclusions and research gaps.

3.2 Literature review method

3.2.1 Search strategy

A systematic search for the literature was implemented using PsycINFO, SPORTDiscus, CINAHL, Scopus, PubMed and the Cochrane Library databases. The
search terms used were (chronic disease or chronic condition or disability) AND (physical activity or exercise or sport) AND (barrier or facilitator or determinant or perspective or view or opinion or attitude). Inclusion criteria allowed for the inclusion of studies on children, adolescents and adults, parents perspectives and any neuromuscular condition or disease that primarily affects muscular function or motor control. The exclusion criteria excluded animal studies and any study that did not include participants with an NMD or condition.

### 3.2.2 Final selection of literature to review

Upon review of the literature, articles that exclusively studied participants with either Multiple Sclerosis (MS; n=17) or Parkinson’s disease (PD; n=4) were excluded as these neuromuscular conditions do not have a similar phenotype to SMA Type II. The researcher deemed that the quantity of articles relating to these conditions could influence the results of the literature review to a degree where it would no longer be tailored to individuals living with SMA Type II. Articles with participants comprised of various neuromuscular conditions that included MS and PD were included. Where possible, filters were applied to exclude studies with participants over the age of 50 years. Due to the life expectancy of individuals living with SMA Type II, it was deemed that studies investigating participants over 50 years of age would not be relevant to this research. However, not all articles with a participant age of over 50 years were filtered out, and the author deemed that to exclude the remaining articles would have left limited review literature.

### 3.2.3 Review strategy

Rychetnik and Frommer’s (2002) schema was applied as a system to review the literature. A table was created using this schema, which outlined details about publications, participants, research design, research methods, research interventions, exercise determinants (facilitators and barriers), clinical implications, quotes and other notes. Exercise determinants were then categorised in reference to the social ecological model of physical activity which specifies the environmental, socio-cultural and individual determinants that impact physical activity. To clarify the individual determinants within the social ecological model of physical activity a list of exercise determinants was used that have been outlined by Biddle, Mutrie and Gorely (2015) and Sallis and Hovell (1990). Agreed determinants of exercise included environmental determinants (weather, access to facilities, safety of the
environment), social determinants (friends, family, colleagues) and psychological determinants (confidence, attitude). Determinants of exercise outlined only by Biddle, Mutrie and Gorely (2015) included socio-demographic determinants (age, gender, socio-economic status), biological determinants (fitness, obesity, disability) and behavioural determinants (smoking, diet, previous exercise). Determinants outlined only by Sallis and Hovell (1990) included cognitive determinants (health and exercise knowledge, decision-making skills).

3.3 Literature search results

From the search results, 18 articles were identified for review (Figure 2). Of the 18 articles reviewed, seven examined exercise perceptions \(^{17-20,22,116,117}\), one was a literature review regarding exercise perceptions, \(^{21}\) and ten reviewed quality of life. \(^{79,83-85,112,118-122}\) Five articles reviewed participants diagnosed with a juvenile onset NMD exclusively, but only two of these specifically explored individuals’ perceptions of exercise. \(^{17,116-118,120}\) Four articles included participants with juvenile and adult onset NMDs, and two of these articles specifically explored exercise perceptions. \(^{18,21,79,112}\) Participants detailed in five articles had adult onset NMDs, of which only one specifically explored exercise perceptions. \(^{22,85,119,121,122}\) Finally, four articles detailed participants with mixed populations of both NMDs and other physical disabilities; two of which specifically explored the participants’ exercise perceptions. \(^{19,20,83,84}\) A summary of the literature reviewed is outlined in Appendix 1.
Chapter 3: Literature Review

Figure 2. Summary of literature review search

* NMD, Neuromuscular disease; MS, Multiple Sclerosis; PD, Parkinson’s disease
3.4 Literature review findings

Many determinants can influence an individual’s perceptions of, and participation in, physical activity.\textsuperscript{13,27} The literature in this review found that individuals living with a slowly progressive NMD have additional determinants that influence their perceptions of, and participation in, physical activity compared to individuals living without an NMD.\textsuperscript{19,21,22} These include environmental determinants, social determinants, and individual determinants, specifically, psychological, biological, socio-economic and cognitive determinants.\textsuperscript{13,17-22,79,83-85,112,116-122} Time and quality of life were themes that also emerged from the reviewed literature.\textsuperscript{13,17-22,79,83-85,112,116-122}

3.4.1 Environmental determinants

Environmental determinants, including transport, weather, equipment and accessible facilities have been identified as barriers to physical activity for individuals living with an NMD\textsuperscript{17,18,22,83,116,118} and individuals living with a physical disability.\textsuperscript{123-127} Whilst some of these determinants are applicable to all individuals,\textsuperscript{128} it is important to note that the difficulties faced by individuals living with an NMD can be greater than for those who do not.\textsuperscript{124} For instance, an individual living with an NMD may not only need access to reliable transportation but a person to drive them, exposure to bad weather may result in illness for individuals living with an NMD leading to more serious consequences that could take months to recover from or threaten their life, etc.\textsuperscript{18,83,118} Environmental determinants can impact an individual’s immediate and established exercise perceptions, attitude, perceived behavioural control and behaviour.

3.4.2 Socio-cultural determinants

Individuals living with an NMD rely on their social networks, including family, friends, work colleagues, support workers, doctors, health professionals, etc. to retain their autonomy.\textsuperscript{20,118-120} Social support of family and friends is a common determinant of participation in exercise for individuals living both with and without an NMD and can either be a barrier or facilitator of exercise, depending on the support received.\textsuperscript{18,20,22,116,118,120,121,123,125-130} However, the literature identified in this review demonstrated that the perceptions of individuals living with an NMD and their
exercise behaviour were heavily influenced by their social network, arguably to a far greater extent than for individuals living without an NMD.

*When a person’s support network promoted segregated program involvement, then that was the type of recreation he or she participated in most often. Many times segregated participation was chosen because the support network thought this was the only real option for recreation involvement.* (p. 159)

There are advantages and disadvantages to participating in segregated and integrated physical activity. Segregated physical activity often has appropriate levels of competition for individuals living with an NMD and promotes fun and enjoyment in physical activity. However, social isolation could also be increased through participation in segregated physical activity as it excludes the participation of friends and family. Integrated physical activity promotes independence and learning social norms. Physical education programs can also promote socialising and possible friendships for individuals living with an NMD.

The role of doctors and health professionals in influencing the decision of individuals living with an NMD to exercise could be an important determinant. Phillips, Flemming and Tsintzas (2009) found that approximately two-thirds of their participants (n≈9) felt that engaging in exercise would be beneficial; however, only one-third of participants were advised to exercise by their doctor and only one participant was given a specific exercise program. Additionally, participants stated that one of their barriers to exercise was their lack of confidence in exercise instructors. Participants felt that exercise instructors lacked knowledge of their condition and the impact that exercise would have on them.

Exercise advice and prescription from a doctor is a strategy that has been used to improve physical activity levels within the general public. Literature has also found that doctors can hinder the exercise engagement of individuals living with a physical disability. Therefore, doctors of patients living with an NMD could greatly enhance exercise participation by prescribing exercise and/or referring them to exercise professionals, including physiotherapists and exercise physiologists, who have the expertise to train an individual living with an NMD.

The ‘disability movement’ was instigated to combat the oppression and stigma associated with disability by advocating for social change, equality and social acceptance. It promotes the civil rights, anti-discrimination, representation and
inclusion of individuals living with a disability to ensure their equal participation in society.\textsuperscript{135-137} The issues faced by the disability movement include workforce participation, reproduction rights, the right to live independently and have access to adequate personal care provisions.\textsuperscript{135-137} The ‘disability movement’ was a facilitator in socialising and leisure activities for individuals living with an NMD.\textsuperscript{18} The stigma of disability was a barrier mentioned by several studies for individuals living with an NMD or physical disability, which could lead to withdrawal of participation in activities of daily living (ADLs), let alone physical activity.\textsuperscript{18,20,119,125,126}

Integrated physical activity can combat stigma by allowing others to view individuals living with a disability as individuals and not as their disability.\textsuperscript{20} Sensitivity to disability is crucial to the formation of successful relationships between individuals living with and without disabilities.\textsuperscript{83} Role models were an important facilitator of exercise for individuals living with a physical disability, but were often absent.\textsuperscript{124,126,127,134} Some individuals living with an NMD felt ‘alone and forlorn’ when their support network continued to participate in activities that they could no longer accomplish.\textsuperscript{18} Additionally individuals missed the activities that either had to be adapted or completely stopped as their condition degenerated.\textsuperscript{18}

Individuals living with an NMD are reliant on others, such as support workers, family or friends to perform ADLs, including body movements, which can create a barrier to physical activity participation.\textsuperscript{20,118-120} Participation in exercises such as Qigong could be an avenue for individuals living with a NMD to investigate.\textsuperscript{117} Qigong was found to reduce stress and could be adapted to include visualisation if an individual was unable to perform the movements, thus negating the need for others to perform body movements for them.\textsuperscript{117}

Finally individuals living with an NMD are known to conceal the emotional and physical impacts of NMD symptoms from family, friends and health professionals.\textsuperscript{18,85,118,119} This can impact on the support given to an individual living with an NMD from their social network, as well as contributing to any psychological distress the individual may be experiencing. Thus, an individual’s concealment of the severity of their disability would create additional barriers to participation in life, as well as physical activity. An individual’s support network, and the degree to which an individual is motivated to comply with their support network’s beliefs, creates a subjective norm. It is apparent from the literature reviewed that a person’s support network greatly influences their perception of exercise, participation in exercise and
their degree of social isolation. This should be considered when engaging with an individual who has an NMD.

3.4.3 Individual determinants

Individual determinants for exercise outlined in the literature review included psychological, biological, socio-demographic and cognitive determinants. Individual determinants could influence an individual’s attitude and perceived behavioural control towards their exercise behaviour.

3.4.3.1 Psychological determinants

All articles reviewed acknowledged the relationship between psychological determinants and physical activity or quality of life. Motivation, enjoyment, self-perception, self-esteem and self-confidence or self-consciousness are all determinants that influence physical activity, for individuals living both with and without an NMD. However, fear is mentioned by several studies as an obstacle faced by individuals living with an NMD or physical disability, including fear of equipment failure, fear of falling, fear of injury and fear of illness. These fears can be attributed to individuals living with an NMD fearing their loss of autonomy, their fear of becoming bedridden or institutionalised. Ultimately, individuals living with an NMD fear that participation not just in physical activity, but in certain ADLs, will result in their condition worsening and/or the possibility of further shortening their already reduced lifespan. Other determinants, such as the weather, can also impact on the level of prominence an individual’s fear will contribute to their perceptions of exercise and their decision to participate in exercise.

Ng, Talman and Khan (2011) reported that 48% (n=21) of participants experienced ‘emotional disturbance’ including depression and anxiety. Furthermore, 31.8% (n=14) were prescribed antidepressants, but only one participant received counselling. Depression and anxiety were inversely correlated with participation in recreational activities and physical activity, for individuals living with a NMD and physical disability. Psychological determinants could influence individuals’ attitude and perceived behavioural control and thus their exercise behaviour. It has been recommended that increased psychological support be offered to individuals living with an NMD, particularly if they are experiencing depression or issues with motivation, self-perception and/or self-esteem. Increasing psychological support
for individuals living with an NMD is a strategy that can also be used to positively influence an individual’s perceptions of exercise and decision to participate in physical activity.

3.4.3.2 Biological determinants

Multiple biological determinants create barriers to participation in physical activity for individuals living with an NMD. These include muscle weakness, loss of functional muscle tissue, muscle disuse, overuse injuries, pain, fatigue, reduced energy levels, cardiopulmonary problems, increased adipose tissue, mobility limitations and increased energy expenditure while engaging in motor functions. Literature has found that individuals living with a physical disability also cite their disability as an exercise determinant, often a barrier to their exercise behaviour. Individuals living with an NMD or physical disability are reported to be less physically active compared to individuals living without an NMD or physical disability. Additionally, individuals living with an NMD or physical disability had more perceived barriers to exercise than individuals living without an NMD. Individuals living with an NMD have a high risk of developing hypokinetic diseases and sedentary death syndrome (SeDS). Hypokinetic diseases and SeDS are diseases associated with muscle disuse, physical inactivity and sedentary life-style choices that increase an individual’s probability of premature disability and/or death. Conditions associated with SMA Type II include sleep apnoea, obesity and respiratory problems, which are all listed as hypokinetic diseases and contribute to SeDS.

Individuals living with an NMD have significantly lower forced vital capacities and significantly higher activity thresholds. This demonstrates that individuals living with an NMD have to work harder to supply oxygen to their body than individuals that do not have an NMD. McCrory and colleagues (as cited by McDonald, 2002) found that individuals living with an NMD spent significantly less time with their heart rate in the active range and exercised at lower intensity rates, compared to controls. Thus, physiological impairments contribute to motor function limitations. Engaging in physical activity was significantly inversely correlated with mobility limitations, pain intensity, pain interference and depressive symptoms for individuals living with an NMD.
Muscle weakness, poor balance and hand and foot problems are barriers to physical activity for individuals living with an NMD.\(^{17}\) McDonald and colleagues (1995) found that by 6 years of age, participants living with Duchene Muscular Dystrophy (DMD) had a 50% reduction in tension for knee extension, compared to controls (as cited by McDonald, 2002).\(^{21}\) Lower limb strength is an important factor in determining whether an individual will gain and retain the ability to walk.\(^{21}\) Abresch and colleagues noted that 36% (n= 292) of individuals living with an NMD had moderate to very significant difficulty controlling their weight (as cited by McDonald, 2002).\(^{21}\) Munn (2010) deliberately maintained a BMI of 18, which is classified as underweight, in order to improve his balance and mobility. Lack of mobility and obesity are barriers to exercise participation and influence an individual’s exercise perceptions, thus increasing the probability of individuals living with an NMD living a sedentary lifestyle.

Pain, fatigue and energy levels were listed in multiple studies as barriers to physical activity or ADLs for individuals living with an NMD\(^{17,18,21,22,85,112,116-118}\) or physical disability.\(^{124,125,140}\) Pain was found to be both a barrier and facilitator to exercise by Anens, Emtner and Hellstrom (2015). No significant difference has been found between bodily pain experienced by individuals living with SMA Type II or III and the United States population norms.\(^{112}\) However, a weak association between pain and the extent that health limits physical activity (physical function) was found in individuals living with SMA.\(^{112}\) Additionally, individuals living with SMA had a moderate association between pain and the extent that health limits work or daily activities (role-functioning physical), personal evaluation of health (general health), interference with normal social activities (social function) and vitality.\(^{112}\) A perpetuating cycle between pain, physical activity, physical condition and quality of life has been acknowledged.\(^{112}\) Lack of physical activity leads to deconditioning, which is associated with decreased pain tolerance.\(^{112}\) Subsequently, pain and biological deterioration can negatively impact quality of life.\(^{112}\)

Biological determinants, including pain, sleep apnoea, fatigue, obesity, deconditioning, muscle weakness and cardiovascular system inefficiency create barriers to exercise for individuals living with SMA Type II. However, engagement in exercise can improve quality of life and may reduce pain levels through pain acceptance and physiological changes. Biological determinants and an individual’s
perceived power over these determinants contribute to perceived behavioural control, and influence intention and exercise behaviour.

### 3.4.3.3 Socio-demographic determinants

Socio-economic constraints have also been identified as barriers to physical activity for individuals living with an NMD\(^{17,18,22,83,116}\) and individuals living with a physical disability.\(^{124-127,134}\) Whilst lack of financial resources are applicable to all individuals,\(^{128}\) it is important to remember that individuals living with an NMD would have additional costs, such as employing support workers, and are likely to be reliant on a more limited income, such as a disability pension, to support themselves and their lifestyle choices.\(^{18,22,85,121}\) Thus, socio-economic status can have a significant impact on an individual’s exercise perceptions, perceived behavioural control and exercise behaviour.

### 3.4.3.4 Cognitive determinants

Knowledge about exercise is an exercise determinant for all individuals,\(^{128,132,133}\) however lack of knowledge is often cited as a barrier to the exercise behaviour of individuals living with an NMD\(^{18,20-22,83,116}\) or physical disability.\(^{124-126,129}\) Individuals living with an NMD or physical disability are often restricted in their choices, including exercise options, and limitations are imposed upon them.\(^{17,20,84,116-118,120,124-126,134}\) The physical and emotional effects of DMD restricted choices and the independence of individuals from their parents.\(^{118}\) This is particularly concerning, as individuals living with DMD and their parents had different views on the obstacles they faced and how their condition should be managed.\(^{118}\)

Two studies highlighted individuals’ living with an NMD perseverance and ingenuity through their ability to adapt new ways of performing tasks, such as constructing unusual body contortions in order to stand from a wheelchair.\(^{18,119}\) Adaption of physical activity is a facilitator for exercise participation in individuals living with an NMD.\(^{17,117}\) The use of assistive devices were also found to facilitate exercise for individuals living with an NMD,\(^{17,18,85}\) and exoskeletons may be useful as they become more readily available.\(^{92,142}\) Therefore, an individual’s ability and willingness to be creative, in combination with a physical activity program that is tailored to meet the needs of the individual, should be considered when developing exercise programs for an individual living with an NMD.
Several studies noted high or complete unemployment rates for individuals living with an NMD. Bostrom and Ahlstrom (2004) stated that individuals living with an NMD had limitations to the work they could engage in and that the majority of those studied who were employed worked with computers. However, Munn (2010) notably contrasted these statistics. His case study, which presented his own experience of living with an NMD, detailed his achievements, which included a PhD in physics, being chief scientist and director of research, an adjunct professor and author of books and scientific articles. An individual’s participation in employment influences their socio-economic status and can, among other things, influence the type and amount of physical activity that they can then participate in.

Individuals’ cognitive processes and their perceptions and decisions to participate in employment, physical activity and many other facets of life are influenced by many determinants. These include the individual’s psychological wellbeing, their social network’s promotion or suppression of their independence and life goals, their biological wellbeing, environmental determinants, socio-economic determinants, time and quality of life. Individuals’ cognitive determinants can impact upon their attitude, perceived behavioural control and exercise behaviour.

### 3.4.4 Time

Time emerged as a strong theme from this review of literature in terms of the impact of time on an individual’s NMD condition, amount of time taken for an individual living with an NMD to engage in exercise, and the impact of living with an NMD condition on psychological wellbeing over time. Bolstrom and Ahlstrom (2004) felt that the International Classification of Functioning, Disability and Health (ICF) model for participation in physical activity should be adapted to include time (Appendix 2 & Appendix 3).

The first adaptation showed the impact of time on an individual’s deteriorating condition and the subsequent effect on exercise participation. This adaptation is supported by the literature, which shows that as time goes by, the condition of an individual living with an NMD will worsen and their physical health will decline as they lose functional abilities. This, in turn, places additional barriers on the individual’s social, financial and environmental determinants to perceptions of, and participation in, ADLs and physical activity behaviour.
The second adaptation Bostrom and Alhstrom (2004) proposed to the ICF model was to show the impact of time regarding an individual’s past (memories) and future (plans) experiences with exercise participation. This change is also supported by the literature, which showed that as time progresses, an individual’s coping skills improve and their psychological distress decreases. Therefore, their coping skills will become a facilitator for their exercise perceptions, including attitude and perceived behavioural control, as well as their exercise participation.

Finally, lack of time, due to work and family commitments, was found to be a barrier common to individuals living with and without an NMD. However, individuals living with an NMD or a physical disability need more time than others to plan and perform their ADLs as they may need to investigate facility accessibility and organise support workers or family to support them in the activity. Therefore, individuals’ perceptions of the time needed for participation in physical activity could impact on their perceived behavioural control and decision to engage in exercise.

3.4.5 Quality of life

Multiple studies have found that individuals living with an NMD rate their quality of life either equal to or greater than individuals living without an NMD. In the words of Munn ‘there have also been so many good things that have come because of my condition that I would not give those up for anything’ (p. 153). Exercise has been shown to improve quality of life for individuals living with a disability, including improved physical, psychological and employment outcomes. Participation in exercise was the main influencing determinant in quality of life for individuals living with a spinal cord injury. Studies have also found that engagement in exercise or physical activity improved quality of life for individuals living with MS and adults over 65 years of age. For individuals living with amyotrophic lateral sclerosis (ALS) the most important determinant to quality of life was the extent of care and support provided to the individual by their social network, as they continued to engage in their social and recreational activities. Therefore, the perceptions of individuals living with an NMD regarding exercise, and their consequent decision to engage or not engage in exercise, can impact on their quality of life. Conversely, quality of life is also a determinant for exercise participation.
3.5 Research gaps

Only 18 articles were found that sufficiently related to the perspectives on exercise or quality of life in individuals living with an (non-MS or PD) NMD. Of these 18 studies, only two specifically explored individuals’ perceptions of exercise with a participant group comprised of individuals living with a juvenile onset neuromuscular disease. This highlights the importance for more research into individuals’ exercise perceptions within this population. Moreover, the 18 articles reviewed researched vastly different subjects within the broader topic of exercise from the perspective of individuals living with an NMD. For instance, one article examined individuals’ perceptions regarding the differences between integrated and segregated physical activity, whilst another examined the Transtheoretical Model of Behaviour Change in relation to individuals living with an NMD to determine if it was a reliable model in this population. Research into exercise perceptions and activities in individuals living with an NMD are restricted to two studies (at most).

Several articles consulted for background information regarding physical activity for individuals living with a physical disability excluded degenerative physical conditions from their criteria. This indicates that individuals living with a degenerative neuromuscular condition are an under-represented population within this research area. As so little research has been conducted with this population regarding exercise perceptions, corroboration in results between the articles evaluated in this literature review were not possible. Furthermore, facilitators of physical activity were not addressed in many of the articles. This is consistent with Shields and colleagues (2012) findings that there is a gap in the research regarding facilitators of physical activity for children living with a disability.126

No published research has investigated the perceptions of individuals living with an NMD regarding differences between types of physical activity, apart from the differences between integrated and segregated physical activities. Also, no research has been conducted that specifically explores the perceptions of parents who have children living with an NMD or condition, regarding the benefits, risks and determinants of exercise for their children. Finally, little research has been conducted with individuals who have a juvenile onset degenerative NMD regarding their perceptions of exercise.
Chapter 3: Literature Review

The reviewed literature led to a number of research questions about individuals living with SMA Type II:

1. What are the past and current exercise habits of individuals living with SMA Type II?
2. How effective do individuals living with SMA Type II think exercise is?
3. What value do individuals living with SMA Type II place on exercise?
4. What are the exercise determinants for individuals living with SMA Type II?

These research questions informed the development of a questionnaire and subsequent in-person interviews used in this research to provide evidence about perceptions of exercise within this population as well as further evidence about perceptions of exercise for individuals living with an NMD.

3.6 Conclusion

Limited research has been conducted into the exercise perceptions and determinants of individuals living with an NMD. This literature review found that environmental, socio-cultural and individual, specifically, psychological, social, biological, socio-demographic and cognitive determinants, time, and quality of life can influence exercise perceptions and behaviour within this population. The determinants found in this literature review can influence individuals’ attitude, subjective norm, perceived behavioural control and subsequent exercise behaviour. To add to the body of knowledge regarding exercise for individuals living with a NMD this research has been designed to explore exercise from the perspective of individuals and parents of individuals living with SMA Type II. These include perceptions about: 1) the effectiveness of individuals’ past and current exercise engagement to treat the symptoms and slow the progression of SMA Type II; 2) the value of exercise in treating the symptoms of SMA Type II; and 3) exercise determinants.
Chapter 4  Research Design

4.1 Introduction

The previous chapter reviewed the published literature relating to exercise determinants of individuals living with a neuromuscular disease (NMD). This chapter describes the research design used to answer the three research objectives. It includes the research ethics; theoretical framework; methodology; researcher bias; participant description; recruitment strategy; data collection and analysis methods; and research validity, reliability, trustworthiness, credibility and dependability.

4.2 Research ethics

Ethics approval for the conduct of this research was sought through the University of the Sunshine Coast Human Research Ethics Committee (S/14/603) (Appendix 4). Informed consent was sought from participants prior to the administration of the online questionnaire and the conduct of in-person interviews. Consideration was given towards potential physical (discomfort and/or exhaustion), psychological (unintentional upset or remembrance of painful memories) and social (identifiability) risks to participants. These risks were deemed to be negligible and were further minimised by the researcher providing participants with information about these risks and ensuring that they were aware of their right to withdraw from the study at any time.

4.3 Theoretical framework

Theories provide an explanation as to how and why independent variables cause, influence or affect outcomes, or dependent variables.\(^23\) The Theory of Planned Behaviour and the social ecological model of physical activity were used in this research.\(^{13,25}\) These theories provided a theoretical lens that helped to shape the collection and analysis of data, presentation of the findings and the discussion.\(^23\)

The Theory of Planned Behaviour explains the link between attitude, subjective norms and perceived behavioural control to an individual’s intentions and their subsequent behaviour (Figure 3).\(^{13}\) However the Theory of Planned Behaviour does not account for numerous determinants that influence attitude, subjective norms,
perceived behavioural control, intention and/or behaviour, thus, weakening the theory.

Firstly, the Theory of Planned Behaviour is criticised for its one directionality and lack of pathways that factors can take (e.g. that beliefs can directly affect behaviour). Secondly, the Theory of Planned Behaviour does not distinguish an individual’s autonomy and motivation for engaging in a behaviour, which can influence attitude, perceived behavioural control and intention. Types of motivation include intrinsic motivation (pleasure), identification (personal values e.g. learning a new skill), introjection (shame or guilt derived from external sources), and external regulation (forced with rewards or punishment by external sources).

Finally, intention cannot fully explain or predict behaviour. Past behaviour, environmental and individual determinants, emotions and unconscious influences, self-efficacy, conscientiousness, anticipated regret and intentional stability are not accounted for with intention. Self-efficacy refers to a person’s belief in their ability to perform a desired action and different types of self-efficacy can be implemented at different stages of behaviour. Conscientiousness refers to a person’s drive to achieve a behaviour and can be developed through planning the behaviour, recording behaviour, setting goals and receiving feedback on progress. Anticipated regret refers to engaging in a behaviour to avoid feeling guilty or regretful. Intentional stability refers to how stable one’s intentions are, and if one’s stated intention is corroborated by their actual behaviour.

However, the Theory of Planned Behaviour is widely used in observational research and has been extensively tested and proven effective in predicting intention and behaviour, which is why it was chosen for this research. Whilst there are gaps in the Theory of Planned Behaviour it was deemed to be the most suitable theory for this research as it described influences that affect behaviour, which was lacking in other theories. Furthermore, critics of the Theory of Planned Behaviour do not dispute that intention, attitude, subjective norm and perceived behavioural control affect behaviour and can provide understanding of behaviour. In this research, the Theory of Planned Behaviour assists in explaining the link between exercise perceptions (beliefs regarding behaviour) of the effectiveness and value of exercise to an individual’s attitude and intention to exercise and their subsequent exercise behaviour. Furthermore, exercise determinants (control variables) and an individual’s past exercise experiences and their predictions for future exercise experiences influence their perceived behavioural control and, ultimately, their current and future
Exercise behaviour.

The social ecological model of physical activity was also used to ensure that influences beyond those addressed in the Theory of Planned Behaviour were taken into consideration. The social ecological model of physical activity is a model that identifies multiple levels of determinants, individual, socio-cultural and environmental, that can influence an individual’s health behaviour (physical activity) (Figure 4). This research used the social ecological model of physical activity to distinguish participants’ exercise determinants, explain their exercise behaviour and tend to some of the Theory of Planned Behaviour criticisms. This model demonstrates that environment and policy, in addition to socio-cultural and individual factors, like motivation, have an influential role on an individual’s exercise behaviour. The social ecological model of physical activity does not identify the most influential determinants, or explain how determinants interact between the individual, socio-cultural and environmental/policy levels. This creates difficulties in specifying what determinants to target when implementing an intervention. However, by focusing on multiple levels of influence, a range of options for interventions can be implemented, particularly at the environmental/policy level, which can impact the whole population. The social ecological model has been widely used to understand

Figure 3. Theory of Planned Behaviour

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multiple determinants by prominent authorities worldwide, including the World Health Organization, which is why it was chosen for this research. The social ecological model of physical activity encompasses all of the determinants that influence exercise behaviour. However, it does not provide an explanation of the process behind an individual’s decision to engaging in exercise behaviour as detailed in the Theory of Planned Behaviour.

Figure 4. Social ecological model of physical activity

*SES, Socio-economic status; PA, Physical activity

In this research, the individual determinants of the social ecological model of physical activity were sorted into five categories outlined by Biddle, Mutrie and Gorely (2015) and/or Sallis and Hovell (1990): biological, psychological, behavioural, cognitive and socio-demographic determinants. Biological determinants included motor function ability, respiratory ability, strength, etc.; psychological determinants included motivation, enjoyment, pain, etc.; behavioural determinants included taking measures to prevent illness or injury, diet, avoidance of sedentary behaviour, etc; cognitive determinants included knowledge, cost-benefit ratio, prioritizing, etc.; and socio-demographic determinants included age and finance.
4.4 Methodology

This research utilised a sequential explanatory mixed methods methodology, which is characterised by two phases. Through the implementation of this methodology, general understanding of the research aims and objectives was gained through the collection and analysis of quantitative data (phase one), whilst a deeper understanding is gathered from the subsequent collection and analysis of qualitative data (phase two). The sequential explanatory mixed methods methodology gives consideration to integration and priority of the quantitative and qualitative approaches. Integration refers to the combination of quantitative and qualitative approaches and where in the research design these components occur, such as during conceptualisation of the research questions or in the presentation of research findings. Priority refers to whether equal emphasis is placed on both the quantitative and qualitative data, or if more emphasis is placed on one over the other. The sequential mixed methods methodology was chosen for this research, as there is a small population of individuals living with SMA Type II, dispersed throughout Australia. Therefore, initial contact could reach a wider range of individuals living with SMA Type II, whilst subsequent contact could provide a deeper understanding of individuals’ perspectives.

Implementation of this methodology occurred through the collection and analysis of an online questionnaire to collect quantitative data, which provided a foundation for the subsequent collection and analysis of qualitative data through in-person interviews. Integration of this research occurred in forming research objectives, the design of the online questionnaire and compilation of the research findings. Priority was given to quantitative data in relation to participant characteristics, and detailing participants’ past and current exercise behaviour. Qualitative data was used to ascertain participants’ perceptions of the effectiveness of their exercise engagement to treat the symptoms and slow the progression of SMA Type II and their exercise determinants. Equal priority was given to detailing the value of exercise to treat the symptoms of SMA Type II.

4.5 Researcher bias

Identifying researcher bias contributes to the validity of research. The researcher has a Bachelor of Clinical Exercise Science and 8 years of experience as a
disability support worker. The researcher’s introduction to disability support work was supporting two individuals, over a 3 year period, that self-directed their own care and educated the researcher about the social model of disability and person-centred care. As an accredited exercise physiologist, the researcher’s philosophy upholds that exercise is inherently beneficial to all individuals, and that exercise is a form of medicine.

4.6 Participants

Participation in this research was open to any individual living in Australia that had been diagnosed with SMA Type II and their parents or guardians, regardless of their age.

4.7 Recruitment strategy

The Spinal Muscular Atrophy Association of Australia (SMAAA), Muscular Dystrophy Foundation (MDF) and Muscular Dystrophy Australia (MDA) were engaged to distribute the online questionnaire to their clientele with SMA Type II. All three organisations were concerned about advertising the online questionnaire on their websites. The concern raised by all three organisations was that their clientele might not recognise which type of SMA they were diagnosed with. To address this concern, the organisations contacted their clientele living with SMA Type II directly, via email, letter or Facebook, to inform them of the research.

4.8 Methods

As a sequential explanatory mixed methods design was used, phase one utilised an online questionnaire and phase two in-person interviews.

4.8.1 Online questionnaire

The purpose of the online questionnaire was to collect quantitative data about participants’ exercise behaviour and exercise beliefs, as well as qualitative data regarding exercise advice participants had received from health professionals.
4.8.1.1 Questionnaire design

A questionnaire was developed for this research as no validated questionnaires existed that explored exercise participation and exercise determinants applicable to individuals living with SMA Type II (Appendix 5). The online questionnaire was informed by the Theory of Planned Behaviour and multiple health and wellbeing related sources and instruments. These included the Exercise and Sports Science Australia adult pre-exercise screening system, Disability Sports Australia, Oswestry Low Back Disability Questionnaire, Sport Psychology, Short Form (36) Health Survey (SF-36), World Health Organization Quality of Life – BREF, World Health Organization Quality of Life Spirituality, Religiousness and Personal Beliefs and World Health Organization International Classification of Functioning, Disability and Health checklist.157-164

The online questionnaire collected both quantitative and qualitative data and was created using SurveyMonkey software.165 Quantitative data comprised of questions that could be answered either categorically or continuously.23 Categorical questions included those related to: the first research objective- individuals exercise engagement- including the type and amount of exercise that individuals’ incorporate into their life; and third research objective- individuals exercise determinants- including individuals’ perceptions of their practitioners’ attitudes towards exercise and individuals’ perceptions of physical activity. Continuous questions employed a 1-5 Likert scale of strongly disagree to strongly agree. These questions related to the second research objective- the value of exercise- including the effect of exercise on individuals’ health and body and specifically muscular strength, respiratory function and motor function. Whilst categorical and continuous questions are easier to interpret and complete, they restrict participants’ viewpoints.166 Alternatively, qualitative open-ended questions can be misinterpreted by participants, and can be harder for the researcher to analyse and draw succinct conclusions from.166

The online questionnaire provided anonymity, thus, potentially limiting the social desirability bias (the tendency for participants to respond to questions in a way that is viewed favourably by others).166 However, individuals who participated in the questionnaire may not accurately and fully represent the perceptions of all Australian individuals living with SMA Type II.166 Respondent bias towards or against exercise, or alternatively a lack of response from individuals who do not participate in exercise, may have been generated from the online questionnaire invitation.
The online questionnaire was pilot tested to ensure that all links to, and within, the questionnaire were accurate, the estimated time of completion was correct, and that the questions could be easily understood from a participant’s perspective. Testers included two managers of disability support organisations and colleagues of the researcher. One tester commented that some questions were repetitive; however, repetition was deliberately included to ensure participants’ perspectives were correctly interpreted. The pilot group did not include any individuals living with SMA Type II or their parents, so as to not exclude any potential participants from the actual research study.

### 4.8.1.2 Questionnaire data collection

The online questionnaire was administered on two separate occasions between June - July 2014 and March - April 2015. SMAAA and MDF Queensland were recruited to administer the 2014 release of the questionnaire and SMAAA, MDF and MDA all administered the 2015 release of the online questionnaire. SMAAA and MDF sent an email to perspective participants, with the research invitation letter and a link to the survey attached (Appendix 6). SMAAA also contacted perspective participants via Facebook in 2014. MDA mailed the research invitation letter, which included the web address for the questionnaire, by post. A reminder email or letter was sent several weeks prior to the closing date of the questionnaire in both 2014 and 2015.

Participants provided informed consent prior to starting the online questionnaire through an informed consent page included at the start of the online questionnaire. Data from incomplete questionnaires were not included in the final data set.

### 4.8.1.3 Questionnaire data analysis

Data from the online questionnaire was analysed using SPSS statistics software (IBM Statistics 22) and NVivo research software (Version 11.1.1). Due to the small sample size, analysis was limited to descriptive statistics, including mean, standard deviation and range of scores. Qualitative data was coded and categorised into themes using the social ecological model of physical activity as a theoretical framework to detail individual, socio-cultural and environmental exercise determinants. Analysis of the online questionnaire data helped to shape the direction of subsequent in-person interviews.
4.8.2 Validity and reliability

Several validity and reliability strategies were implemented to ensure the trustworthiness of the online questionnaire. Validity is defined as accuracy in measuring data.\textsuperscript{167} Reliability is defined as reproducibility, or the capacity to measure consistently.\textsuperscript{167} Construct validity was established by asking a wide variety of questions related to the aim, objectives, and theoretical framework.\textsuperscript{168} Face validity and content validity were established through pilot testing with managers of disability support organisations and peer review from university lecturers.\textsuperscript{168} Content validity was also established through thorough research of exercise perceptions of individuals living with an NMD, and use of the Theory of Planned Behaviour and the social ecological model of physical activity.\textsuperscript{168} A copy of the online questionnaire has been included to enhance the reliability of this research and so that it may be reproduced for future studies (Appendix 5).\textsuperscript{23,167} Reliability of the online questionnaire was also enhanced through the use of the Likert scale and categorical scales.\textsuperscript{23} Asking the same question in different ways reduced the risk that participant errors may have occurred, and by asking simple, clearly worded, unambiguous questions reduced misinterpretation and inability to answer.\textsuperscript{168} These strategies were utilised to further enhance reliability.\textsuperscript{168}

4.8.3 In-person semi-structured interviews

The purpose of the in-person semi-structured interviews was to collect in-depth qualitative data about individuals’ perceptions regarding exercise and to explore specific exercise programs that participants have engaged in.

4.8.3.1 Interview design

Interviews revolved around the first and third research objectives. Participants’ perceptions of their practitioners, their past and current exercise behaviour and the perceived benefits, limitations and risks they associated with exercise were further explored. Participants awareness about exercise policies outlined by the Consensus Statement for Standard of Care in SMA (Consensus Statement), exercise advice published by Muscular Dystrophy UK and the Australian Physical Activity and Sedentary Behaviour Guidelines were also explored (Appendix 7). Interviews were designed to be conducted in-person over approximately 2 hours. Research into in-person interviews has shown that participants are more likely to respond openly and accurately as they are in a more natural communicative setting.
that includes small talk, politeness, joking and non-verbal communication. They are also more likely to give greater detail, thoughtfulness and development to their answers. The power of the interaction between the interviewer and interviewee is more equal, allowing for greater clarification, depth and breadth of the topics being discussed. There is also a greater probability of responders participating and participants may be more at ease when discussing sensitive questions. For these reasons, in-person interviews were deemed beneficial despite incurring substantial costs in travelling to the researcher; increasing the time required to interview participants; reducing the standardisation of the interview questions; presenting a greater potential for interviewer impact on participant response; and increasing the risk to the researcher’s safety.

Interview questions were shaped by the questionnaire findings, as established by the sequential mixed methods methodology. To address the first and second research objectives interview participants detailed what exercises they felt impacted movement, strength, respiratory function, and how these exercises impacted on their quality of life thus expanding on the Likert scale answers they had provided in the questionnaire. The first research objective was also addressed by asking interview participants about the benefits, risks and limitations of exercises they had detailed in the questionnaire. To address the third research objective interview participants were asked about their experiences in school physical education (PE) to further investigate participation in school PE that arose from the online questionnaire. Interview participants were also asked if exercise recommendations were achievable and applicable to provide a deeper understanding of exercise behaviour outlined in the questionnaire. Whilst the questionnaire provided a general understanding of participants’ exercise perceptions, the interviews allowed for greater depth of understanding.

4.8.3.2 Interview data collection

Prospective interview participants were sent an email with the research invitation letter prior to the interview (Appendix 8). All interviews were conducted in-person, at the participant’s home (n=8), at a café (n=2) or at the MDA office (n=1). Participants signed an informed consent form prior to the interview. Interviews took between 43 minutes and 225 minutes. Interviews were voice recorded and transcribed.
Participants were emailed their transcript to read and make any adjustments they felt necessary.

### 4.8.3.3 Interview data analysis

Qualitative data from the interviews were analysed using NVivo research software (Version 11.1.1). Data was first read and organised for general understanding, prior to being coded into themes. Data was organised by grouping answers to questions, which enabled the researcher to understand the general consensus of the participants. Data was then coded into themes of environmental, socio-cultural and individual determinants, according to the social ecological model of physical activity. A mixture of predetermined codes (environmental, socio-cultural and individual determinants) and codes that emerged from the data were used.

### 4.8.4 Trustworthiness, credibility, and dependability

The trustworthiness of interview data was accomplished using several credibility and dependability strategies. Credibility is the qualitative counterpart of validity, and is defined as an accurate portrayal of data through assurance that the findings have been described truthfully. Dependability is the qualitative counterpart of reliability, and is defined as a consistent research process that follows the rules of qualitative research methodology.

The credibility measures taken in this research included providing rich, thick description; acknowledging researcher bias; presenting negative or discrepant information; time spent with participants and in participants home environment; peer debriefing; and external auditors. Credibility of research findings was enhanced in this study through the provision of rich, thick descriptions, the use of quotes to provide transparency and the ability for others to draw their own conclusions thus enhancing the credibility of the research, and clarifying researcher bias to create openness and honesty about how the researcher’s background reflects their interpretation of the data. It should be noted that all researchers have biases, thus, the latter should not be considered an integral flaw in the research. Presenting negative or discrepant data for themes supported by the majority of participants adds realism and reduces researcher bias adding to the credibility of the data. Time spent in the field interviewing participants in their homes (as occurred in this research) can provide deeper understanding of the participants and their perspectives, which can be conveyed in the research findings, also increasing the accuracy and
credibility of the research. Peer debriefing increases the accuracy of interpretation by providing different vantage points and beliefs that may question and more comprehensively confirm research interpretation. External auditors, unfamiliar with the researcher and the research, assessed the research at various stages. These included the Human Research Ethics Committee, the confirmation of candidature examiner and the thesis presentation examiner.

The dependability measures taken included transparency in interview questions; asking the same question in different ways; implementing a written interview protocol; having transcript checks; and having code definitions. A copy of the interview questions has been included to enhance the dependability of this research and enable it to be reproduced (Appendix 7). Implementing a written interview protocol enhanced the dependability of the interview by reducing the risk of questions being asked in a different way and enabling greater comparative analysis between participants. Transcripts were checked, to ensure dependability, by both the researcher and the interview participants. Codes were defined to ensure continuity and congruency within their meaning throughout the study.

### 4.9 Conclusion

This chapter outlined the research design used in this study. The Theory of Planned Behaviour and the social ecological model of physical activity, in conjunction with the exercise determinants outlined by Biddle, Mutrie and Gorely (2015) and/or Sallis and Hovell (1990) were used to shape the research from the research design and literature review stages through to the findings and discussion. A sequential explanatory mixed methods design allowed for quantitative and qualitative data to be collected and analysed using valid, reliable, trustworthy, credible and dependable measures. The findings of this research are presented in Chapter Five.
Chapter 5: Research findings

5.1 Introduction

The previous chapter detailed the research design including the research ethics, theoretical framework, methodology, methods and research trustworthiness. This chapter presents the research findings including participant characteristics, influences on exercise behaviour, exercise behaviour, the effectiveness of exercise to treat symptoms and slow the progression of spinal muscular atrophy (SMA) Type II, the value of exercise to treat SMA Type II, and exercise determinants. Participant characteristics include age, motor function ability and additional conditions. Influences on exercise behaviour include the influence of past and current exercise behaviour, effectiveness of exercise, value of exercise and exercise determinants on the Theory of Planned Behaviour. Exercise behaviour details the type, frequency and cessation of participants exercise. The effectiveness of exercise to treat symptoms and slow progression of SMA Type II details participants’ perceptions, of which exercises are most effective for cardiorespiratory function, motor function ability, orthopaedic health, muscle strength and gastrointestinal dysfunction. The value of exercise to treat SMA Type II explores participants’ perceptions, opinions and beliefs regarding exercise. Finally, exercise determinants are detailed using the social ecological model of physical activity and the Theory of Planned Behaviour as a framework to understand how exercise determinants lead to exercise behaviour.

5.2 Participant characteristics

Questionnaire participants were identified as either individuals living with SMA Type II or parents of individuals living with SMA Type II, and the age, age of symptom onset, age of diagnosis and motor function ability of individuals living with SMA Type II. The findings also identified interview participants location within Australia and conditions associated with SMA.

Eight individuals living with SMA Type II and twelve parents of individuals living with SMA Type II participated in this research. The online questionnaire had 25 responses in total; however, five incomplete questionnaires were not evaluated. Individual respondents living with SMA Type II were aged 17 years or older. Parents of individuals living with SMA Type II had children aged 16 years or younger. The
Chapter 5: Research findings

Age range of individuals living with SMA Type II was 1.6 - 36 years (mean 15.4 ± 10.8 years). The age range of symptom onset was 0 - 2 years (mean 11.1 months ± 6.2 months) and the age range of diagnosis was 0.8 - 2.5 years (mean 18.6 months ± 6.3 months). Nineteen participants were genetically tested, while one was not (male, symptom onset as new born baby, diagnosed at 1.3 years, highest independent function was crawl).

Participants’ motor function ability, both current and highest (‘best ever’) achieved, was reported by participants selecting applicable movements from categories (Table 4). Three participants who stated that their highest independent motor function ability was walking were interviewed and mentioned their experiences of walking.

‘The doctor said I was...on the border between [Type] one and [Type] two. I walked and ran, obviously duck feet and that kind of thing, until I was probably five or six, I suppose, and I lost that. We were...sightseeing and all that kind of thing, spent more time in a wheelchair and when I came back I’d pretty much lost it.’ (I1)

‘I was able to walk leaning on stuff assisted until I was about eight I think.’ (I5)

‘They told us “my child” would never walk so “my child” was using a frame and then “my child” used to walk probably twenty steps by “my child’s” self without frames. So I think if we didn’t do that swimming and that exercise in the water, that non-weight bearing, “my child” wouldn’t have done that.’ (P12)

One participant discussed conflict arising between individuals diagnosed with SMA regarding their diagnosis and motor function ability.

‘There’s a lot of conflict of that online isn’t there? A few people that kind of bicker. It’s a funny world that we live in, people just want to run other people down. It’s about trying to make informed decisions about the best quality of life for each individual. You know the accusations that I’ve seen thrown around about that there’s Type 1’s and no you’re not a Type 1 because Type 1’s don’t sit up.’ (P3)
Sixteen participants, seven individuals and nine parents, became identifiable by indicating an interest in being interviewed. Nine lived in capital cities and seven lived in rural or remote locations. This cohort covered every Australian state/territory except the Australian Capital Territory and Tasmania. The responses from each state were: Western Australia (n=5; two individuals and three parents); New South Wales (n=4; two individuals and two parents); Victoria (n=3; two individuals and one parent); Queensland (n=2; one individual and one parent); South Australia (n=1; one parent); and the Northern Territory (n=1; one parent). Eleven participants were interviewed: three individuals living with SMA Type II (21 - 34 years of age) and eight parents of children living with SMA Type II (3 - 16 years of age). Five children were also present for part or all of the interview conducted with their parent/s. Of the eleven participants who were interviewed, six lived in capital cities and five lived in rural or remote locations.

Five questionnaire participants (25%) indicated that they had been diagnosed with conditions other than SMA Type II. These included scoliosis (n=3), hip dysplasia (n=1), sleep apnoea (n=2) and recurring chest infections (n=1), all of which

<table>
<thead>
<tr>
<th>Total</th>
<th>Age</th>
<th>Male</th>
<th>Female</th>
<th>Sit</th>
<th>Crawl</th>
<th>Crawl &amp; stand</th>
<th>Walk</th>
<th>No movement</th>
<th>Breathe</th>
<th>Sit</th>
<th>Crawl</th>
<th>Stand</th>
<th>Crawl &amp; stand</th>
<th>Walk</th>
</tr>
</thead>
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<td>5 - 16</td>
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<td>1</td>
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<td>7</td>
<td>1</td>
<td>12</td>
<td>1</td>
<td>4</td>
<td>16</td>
</tr>
</tbody>
</table>

| Interviews |     |      |        |     |       |              |      |             |         |     |       |       |              |      |
| 1     | 0 - 4 | 1    | 1      | 1   |       |              |      |             |         |     |       |       |              |      |
| 7     | 5 - 16 | 2    | 5      | 3    | 2     | 1            | 1    | 2           | 1       | 3   | 1     | 1     | 4           | 1     |
| 3     | 17+   | 2    | 1      |     |       |              |      |             |         |     |       |       |              | 2     |
| 11    | Total | 4    | 7      | 3    | 3     | 2            | 3    | 3           | 2       | 5   | 1     | 2     | 4           | 3     |

Table 4. Characteristics of SMA Type II participants stratified by age, gender and functional status

N.B. Participants (17 - 36 years of age, Questionnaire n=2, Interview n=1) marked walk as their highest functional status achieved without equipment, but did not mark walk as their highest functional status achieved with equipment (no change in results).
are conditions associated with SMA. Four participants (20%) listed depression (n=2), anxiety (n=1), tachycardia (n=1), and encephalitis (n=1), which are not directly associated with SMA. Interview participants were asked about conditions associated with SMA Type II, reporting: feeding or swallowing problems (n=7); gastrointestinal dysfunction (n=8); nourishment: under (n=4), over (n=1), fluctuating (n=2), stable (n=3), unsure (n=1); reflux (n=5); aspiration (n=3); recurrent chest infections (n=6); impaired cough (n=10); sleep-disordered breathing (n=7); underdeveloped lungs or ribcage (n=4), unsure about underdeveloped lungs or ribcage (n=4); scoliosis (n=10); fractures (n=8); hip subluxation (n=6), unsure about hip subluxation (n=4); joint contractures (n=8), unsure about joint contractures (n=1); and loss of bone mineral density (n=4), not tested for bone mineral density (n=4: 5 - 10 years n=1; 16+ years n=3).

5.3 Influences on participants exercise behaviour

The findings are presented according to the Theory of Planned Behaviour by outlining the influence of past and current exercise behaviour, perceived effectiveness of exercise, perceived value of exercise and environmental, socio-cultural and individual exercise determinants identified by participants (Figure 5). Figure 5 is individualised in the format of the Theory of Planned Behaviour, as seen in Figure 3, and includes findings from both the questionnaire and interviews. This research found that an individual’s attitude toward exercise was influenced by their past and current exercise behaviour, perceptions of the effectiveness of exercise, perceptions of the value of exercise as well as environmental, socio-cultural and individual exercise determinants. An individual’s subjective norm was also influenced by environmental and socio-cultural determinants. Finally, an individual’s perceived behavioural control was influenced by their past and current exercise behaviour, perceptions of the effectiveness of exercise as well as environmental, socio-cultural and individual exercise determinants.
Figure 5. The Theory of Planned Behaviour for individuals living with SMA Type II, depicting the influence of past and current exercise behaviour, perceived effectiveness of exercise, perceived value of exercise and environmental, socio-cultural and individual determinants on exercise behaviour.

5.4 Participants’ past and current exercise behaviour

This section of the results explored individuals’ past and current exercise behaviour, which can influence their perceived behavioural control and attitude (Figure 5). The findings are presented according to the following themes: exercise type, exercise frequency, exercise recovery and pain responses and exercise cessation. Eighteen participants (90%) had engaged in exercise over their lifetime, while two participants (10%) aged 0 - 4 years had not. Participants current exercise behaviour was nil (n=2), current (n=12) or previous (n=6), of which four had limited engagement. Four individuals living with SMA Type II aged 17 - 36 (50%) no longer engaged in exercise. Interviewed participants’ exercise engagement was current (n=7), previous (n=2) or limited (n=2).

5.4.1 Exercise type

Participants engaged in physiotherapy exercises (section 5.7.2.2, Table 7), as well as a wide variety of physical activities, exercises and sports. A list of exercises
and sports were provided to participants to select the exercises, physical activities and/or sports that they participated in as presented in Table 5. An “other” section was also included for participants to detail exercises, physical activities and/or sports that were not included in the list. Swimming (90%) was the most popular activity engaged in across all ages, genders and functional levels for both lifetime and current exercise behaviour. Lifetime exercise engagement that singular participants engaged in that were not listed in Table 5 included archery, sitting volleyball, netball, football, golf, hippotherapy (horseback riding), stretches, fishing, bowling, playing musical instruments (drums, trumpet and piano), knitting and pushing a manual wheelchair.

5.4.2 Exercise types that participants did not feel comfortable engaging in

To gain further understanding of participants perceptions, online questionnaire participants were asked about exercises they did not feel comfortable engaging in. Individuals felt comfortable engaging in all types of exercise (n=3), parents felt comfortable engaging their child in all types of exercise (n=7) and parents perceived that their children felt comfortable engaging in all types of exercise (n=3). Exercises that individuals did not feel comfortable engaging in were boccia (n=4), wheelchair basketball (n=4), sitting volleyball (n=4), wheelchair tennis (n=4), weight training (n=3), yoga (n=3), archery (n=3), wheelchair soccer (n=1) and swimming (n=1). Exercises that parents did not feel comfortable engaging their child in were boccia (n=2), weight training (n=2), yoga (n=2), archery (n=2), wheelchair basketball (n=1), sitting volleyball (n=1) and wheelchair tennis (n=1). One parent simply commented that limited mobility limits exercise engagement. Exercises that parents perceived their children did not feel comfortable engaging in were boccia (n=2), weight training (n=3), yoga (n=3), archery (n=3), wheelchair basketball (n=4), sitting volleyball (n=3), wheelchair soccer (n=1), wheelchair tennis (n=2) and anything with a ball (n=1). One parent was not sure, as their child was negative to the concept of exercise and another stated their child was too young to answer.
Table 5. Exercise behaviour of questionnaire participants stratified by gender, age and functional status

<table>
<thead>
<tr>
<th>Exercise type</th>
<th>Lifetime exercise engagement (n=18)</th>
<th>Current exercise engagement (n=9)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Total</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Male</td>
<td>Female</td>
</tr>
<tr>
<td>Swimming</td>
<td>18</td>
<td></td>
</tr>
<tr>
<td>WC soccer</td>
<td>6</td>
<td></td>
</tr>
<tr>
<td>School PE</td>
<td>5</td>
<td></td>
</tr>
<tr>
<td>Yoga</td>
<td>3</td>
<td></td>
</tr>
<tr>
<td>Boccia</td>
<td>3</td>
<td></td>
</tr>
<tr>
<td>Dance</td>
<td>3</td>
<td></td>
</tr>
<tr>
<td>WC basketball</td>
<td>2</td>
<td></td>
</tr>
<tr>
<td>Weight training</td>
<td>2</td>
<td></td>
</tr>
<tr>
<td>WC hockey</td>
<td>2</td>
<td></td>
</tr>
<tr>
<td>Cricket</td>
<td>2</td>
<td></td>
</tr>
<tr>
<td>Softball/T-ball</td>
<td>2</td>
<td></td>
</tr>
<tr>
<td></td>
<td>7</td>
<td></td>
</tr>
<tr>
<td>Swimming</td>
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<td></td>
</tr>
<tr>
<td>School PE</td>
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</tr>
<tr>
<td>Yoga</td>
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<td></td>
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<tr>
<td>Weight training</td>
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<tr>
<td>T-ball</td>
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<tr>
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<tr>
<td>Pushing WC</td>
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<td></td>
</tr>
<tr>
<td>Knitting</td>
<td>1</td>
<td></td>
</tr>
<tr>
<td>Catching a ball</td>
<td>1</td>
<td></td>
</tr>
</tbody>
</table>

*WC, wheelchair; PE, physical education
5.4.3 Exercise frequency

Twelve participants (60%) were currently engaged in physiotherapy. Their frequencies in performing physiotherapy exercises were daily (n=3), 3 - 5 days/week (n=3), 1 day/week (n=4) and 1 day/fortnight (n=1). It was unclear how often one participant engaged in physiotherapy. One participant added that during periods of illness physiotherapy exercises were undertaken every few hours.

Participants were asked the number of days and minutes per week they exercised for currently as presented in Figure 6 and at maximum participation as presented in Figure 7. Analysis of current exercise behaviour included participants who engaged in the most exercise, participants who had the lowest motor function and participants who were older than 17 years. Current exercise behaviour for four participants was >150 mins/week, 5 - 7 days/week; aged 17 - 36 (n=1), 5 - 16 (n=2), and 0 - 4 (n=1). Their highest motor function ability was walking (n=1), crawling (n=1) and sitting (n=2), and current motor function ability was sitting (n=4). Current exercise behaviour of participants whose current motor function ability was independent breathing or no independent movement (n=7) was: 61 - 90 mins, 2 days/week (n=1); <30 mins, 1 day/week (n=1); and not at all (n=5). Current exercise behaviour of participants aged 17 - 36 (n=8) was: >150 mins/week, 5 days/week (n=1); 61 - 90 mins/week, 3 days/week (n=1); 30 - 60 mins/week, 1 day/week (n=1); <30 mins/week, 1 day/week (n=2); and not at all (n=3).
Chapter 5: Research findings

Analysis of maximum exercise behaviour included participants who engaged in the least amount of exercise and those who had participated in the most exercise. Maximum exercise behaviour for four participants was <30 mins/week, 1 - 4 days/week; aged 17 - 32 (n=2, 1 male, 1 female) and 5 - 16 (n=2, 1 male, 1 female). Three had highest motor function abilities of sitting and current motor function ability of breathing independently (n=1) and no movement (n=2), whereas the highest motor function ability of the fourth was walking, with a current motor function ability of sitting. Maximum exercise behaviour for six participants was >150 mins/week, seven days/week; aged 17 - 36 (n=2, male), 5 - 16 (n=3, 1 male, 2 female) and 0 - 4 (n=1, female). Their highest motor function ability was walking (n=1), standing and crawling (n=1), crawling (n=3) and sitting (n=1); and current motor function ability of sitting (n=4) and breathing independently (n=2).

![Figure 7. Maximum exercise behaviour of questionnaire participants (mins/week)](image)

5.4.4 Exercise recovery and pain responses

Participants stated that recovery from exercise took little to no time (n=8, 44%), a short time (n=6, 33%), some time (n=3, 17%) and too long (n=1, 6%). Participants stated that they currently experienced pain whilst engaging in exercise occasionally (n=9, 50%), sometimes (n=1, 6%), always (n=2, 11%) and never (n=6, 33%). Participants stated they had experienced pain whilst engaging in exercise at any point throughout their life, occasionally (n= 6, 33%), sometimes (n=5, 28%), always (n=2, 11%) and never (n=5, 28%).
5.4.5 Exercise cessation

Participants were asked their reasons for ceasing physiotherapy and exercise. Six questionnaire participants (33%) previously incorporated physiotherapy into their treatment plan but no longer engaged in physiotherapy. The reason(s) given for stopping physiotherapy completely were dislike/negative attitude toward physiotherapy (n=3), time constraints (n=2), no benefit (n=2), pain (n=2), loss of individual’s functional abilities (n=1), lack of available practitioners (n=1) and/or difficulty in accessing facilities (n=1). Three questionnaire participants (17%) currently participated in physiotherapy, but had stopped previously recommended therapies due to money constraints (n=2), time constraints (n=1), loss of individual’s functional abilities (n=1) and/or a lack of facilities (n=1).

Interview participants (n=11) stopped engaging in certain types of exercise, or all exercise, due to boredom, degeneration affecting their ability to perform previous activities, difficulty in organising support systems, inability of support systems to support the individual in activity, exposure to unfavourable weather, outgrowing equipment, lack of access to facilities and equipment, and/or time constraints.

5.5 Participants’ perceptions about the effectiveness of exercise to treat the symptoms and slow progression of SMA Type II

This section of the results presents individuals’ perceptions about the effectiveness of exercise to treat the symptoms and slow the progression of SMA Type II. Perceived effectiveness of exercise can influence individuals’ attitude, perceived behavioural control and subsequently their exercise behaviour (Figure 5). The findings are presented according to the following themes: the effect of exercise on SMA Type II progression, life expectancy and motor function ability; participants perceptions of the effectiveness of different exercise types; and exercise types that participants did not feel comfortable engaging in.

5.5.1 The effect of exercise on SMA Type II progression, life expectancy and motor function ability

The majority of interview participants tentatively agreed or were hopeful that exercise could slow the progression of SMA Type II (n=9, 82%). Others stated they didn’t know (n=1, 9%) and that without clear data they didn’t want themselves or
their child to go through something for nothing (n=1, 9%). The majority perceived that exercise could improve or maintain motor function ability and slow but not stop the progression of SMA Type II.

‘We know the progression of the disease and the certain, the complications that come with the SMA and I guess that what we do on a daily basis is just try to counteract that to some degree.’ (P11)

‘I think exercise has definitely helped “my child” to perhaps strengthen what muscles “my child” has left from the previous drop when “my child’s” condition regressed. The next time that regression occurs we’re falling from a higher place so hopefully we will be able to retain some of those skills that “my child’s” been able to build up.’ (P5)

The majority of participants (n=9) described loss of function or advancements in secondary conditions following the cessation of exercise.

‘Once “my child” had that operation ”my child” was stuck in their chair with a wedge holding their legs out on a board. “My child” lost more condition in that 3 months and it took us probably 2 years to get anywhere near ”my child’s” fitness level that they were at before that operation.’ (P11)

Nine interview participants were asked if they thought exercise could affect life expectancy and either very tentatively agreed (n=7) or agreed (n=2) that it could improve life expectancy; particularly by improving cardiovascular function (n=5) and maintaining function or delaying deterioration (n=2). Participants (n=5) perceived that genetics, complications like pneumonia and impacts on quality of life were overriding factors to improved life expectancy.

‘It could’ve [affected life expectancy]. Yeah, I don’t know so I’ll say it could’ve. But then the gain you get from the exercise it might take away from the freedom. If we knew yeah it’s going to double everything, hey maybe we’d be on board, but from what I’ve read it doesn’t really. Not enough for the trouble and for the maybe heartache to “my child”. ‘ (P2)
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Five interview participants gave specific examples of gains in physical function, such as hand strength (n=3), that they attributed to exercise. They stated:

“The strongest muscles I have now are the ones that I used the most [playing a musical instrument]. Like my calf muscles are still quite strong. This muscle here in this finger is the strongest one in my hands because [of playing the musical instrument].’ (I3)

‘The difference in the strength in the hands is a whole lot different because “my child” hasn’t had that intensity of video games for some time. Without the exercise “my child” wouldn’t have been able to develop as far as what “my child’ did.’ (P3)

‘I can’t say it’s [exercise] done anything bad, and if anything, it has made improvements. “My child” writes much better, “my child’s” right hand is more active now.’ (P11)

‘I would say if “my child” did not swim, “my child” would not have walked. And if we didn’t put Hi-5 on and get “my child” to stand up in that walker and dance, “my child” would not have done that either.’ (P12)

‘If “my child” didn’t exercise, I think “my child would” be even more immobile than what “my child” is now.’ (P4)

5.5.2 Participants perceptions of the effectiveness of different exercise types

The most effective exercises perceived by interview participants for cardiorespiratory function (n=11) were swimming (n=9), bubble pep/blowing bubbles (n=5), cough assist machines (n=3), singing (n=3), playing musical instruments (n=2), breathing techniques (n=1), bi-pap machines (n=1), standing (n=1), respiratory pats (n=1), pushing a manual wheelchair (n=1), arm ergometer (n=1), stretching arms and chest (n=1), and general exercise (n=1). The most effective exercises perceived by interview participants for motor function ability (n=9) were swimming (n=5), general movement (n=3), stretches (n=2), hand exercises (n=1), shoulder strength exercises (n=1), cycling (n=1), dancing (n=1), playing musical instruments (n=1), tummy rolling (n=1) and Feldenkrais physiotherapy (n=1). The most effective exercises perceived by interview participants for orthopaedic health (n=9) were standing (n=6), swimming (n=4), general exercise (n=4), stretching (n=3) and physiotherapy (n=1).
The most effective exercises perceived by interview participants for muscular strength (n=8) were swimming (n=5), standing (n=4), core exercises (n=2), general movement (n=2), stretching (n=2), cycling (n=1), weight training (n=1), pushing a manual wheelchair (n=1), dance (n=1), video games (n=1) and hand strengthening exercises (n=1). The most effective exercises perceived by interview participants to improve gastrointestinal dysfunction were standing (n=3) and general exercise (n=1).

5.6 Participants perceptions about the value of exercise to treat SMA Type II

This section of the results explored individuals’ perceptions about the value of exercise to treat the symptoms of SMA Type II, which can influence their attitude towards exercise behaviour as outlined in Figure 5. The findings present participants’ perceptions of exercise as a treatment for SMA Type II; participant perceptions, opinions and beliefs about the effects of exercise; and the positive and negative affect of exercise on quality of life.

The majority of questionnaire participants (n=85%) perceived that exercise was beneficial in treating the symptoms of SMA Type II, with 25% reporting that it was of great benefit as presented in Figure 8.

![Figure 8. Questionnaire participants' perceptions of exercise as a treatment for SMA Type II](image-url)
Questionnaire participants reported that exercise was not a waste of their time (90%), improved physical function through improved movement (65%) and respiratory function (75%), improved psychological function through improved mood (75%), improved cognitive function through improved coping skills (60%) and improved quality of life (75%) as presented in Figure 9.

The majority of interview participants believed that exercise improved quality of life through improved confidence (n=5) and self-esteem (n=2), opportunities to engage with peers (n=9) and improved functional ability that was transferable to daily life (n=8). Participants (n=5) also felt that exercise negatively impacted quality of life, as it was considered to be painful, time consuming, lacked enjoyment and/or created disharmony within the family.

Improved quality of life and confidence, activity enjoyment, increased physical function, positive social aspects and positive societal attitude facilitated exercise.

‘[Exercise] was fun, it certainly made me feel good. Has it improved my muscle strength? A bit. So if it has then that’s certainly improved my quality of life.’ (I3)

‘Besides the health benefits, there’s the inclusion with “my child’s” peers and “my child’s” friends, and it’s good for “my child’s” soul.’ (P10)

‘From the interaction with “my child’s” peers, which is huge...people are accepting “my child”, they’re seeing “my child” as a person and not the person in a wheelchair. It was equal playing ground in the water. So I think that way you’ve got your confidence, you’ve got your peers, you’ve got interaction, you’ve got acceptance.’ (P12)

Negative impacts on quality of life hindered exercise, but improved motor function ability facilitated exercise.

‘I suppose from the quality of life perspective you weigh up, would I rather be able to use my hands for longer but not enjoy it [the time spent exercising] or would I do something that I enjoy and need more help later on in life? And I kind of switch between the two at different stages.’ (I1)
Negative impacts on quality of life, pain, limited and/or negative social interaction, and lack of enjoyment hindered exercise.

‘If [exercise is] painful, possibly negatively impact the quality of life. There’s no social aspect to whatever “my child” would be doing because it [exercise] would have to be very much tailored to what “my child” can do. [Exercise is] much likely to be sort of more something that “my child” could do by “my child’s” self so that...takes away some of what I would say would be the enjoyment of exercising.’ (P6)
Figure 9. Questionnaire participants’ exercise perceptions, opinions and beliefs

*Pop., population; QoL, quality of life
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5.7 Exercise determinants for individuals living with SMA Type II

This section of the results presents individuals’ perceptions about their exercise determinants. The findings are presented according to the social ecological model of physical activity by outlining the environmental, socio-cultural and individual exercise determinants identified by participants (Figure 10). Figure 10 is individualised in the format of the social ecological model of physical activity, as seen in Figure 4, and includes findings from both the questionnaire and interviews. Environmental, socio-cultural and individual exercise determinants can influence participants’ attitude, subjective norm and perceived behavioural control, which subsequently influences their exercise behaviour as outlined in the Theory of Planned Behaviour for individuals living with SMA Type II (Figure 5).

![Figure 10. The social ecological model of physical activity for individuals living with SMA Type II, depicting the individual, socio-cultural and environmental, and policy determinants for exercise](image-url)
5.7.1 Environmental exercise determinants for individuals living with SMA Type II

Multiple environmental determinants (the outer tier of Figure 10) were reported during the interviews and included funding and policy; access to services, facilities and equipment; transport and distance; and climate and weather. Environmental determinants can influence participants’ beliefs, subjective norm and perceived behavioural control and, subsequently, their intention to, and participation in, exercise. Many of the identified environmental determinants resulted in both positive and negative responses amongst participants.

5.7.1.1 The impacts of funding and policy on attitude, subjective norms, perceived behavioural control and exercise behaviour

Funding, including state disability funding and the National Disability Insurance Scheme (NDIS), was reported as an exercise determinant for participants (n=9), and both facilitated and hindered exercise behaviour.

Funding facilitated exercise,

‘This new funding has been a Godsend. For us “my child” has gone from no physio, or we’d probably see a physio four times a year, to every week. Our car we ended up paying for, “my child’s” “sport” grill, the adaptations, the swimming, the tutoring we’ve paid for ourselves, just to try and keep up with the correct level of care.’ (P3)

Whereas, a lack of funding hindered exercise.

‘(Occupational therapist (OT)) decided that they would basically put me on the, you know, the old person program thing. They’re sort of winding back their involvement. It’s kind of annoying about the physio, but getting physio has always been really difficult because “A Place” are based in “Suburb”, which is quite a way away.’ (I3)

Policies and programs that drive community access to sports was an exercise determinant for participants.

Lack of policy hindered exercise, despite parental and school advocacy.

‘“My child” actually came third in the “sport”. So “my child” was technically allowed to go to zone, like the next level, but because there wasn’t any classification for electric wheelchairs “my child” wasn’t
allowed to go. So we challenged that a bit and they looked into it but that’s as far as it got.’ (P12)

Suitable programs facilitated exercise.

‘It’s a program where they have things like footy, lawn bowls, basketball, swimming, like a whole range of activities. It’s all ability levels, it’s all inclusive. The program itself was really successful and it still runs. It started as a pilot and then they obviously got funding for it because it was successful. It’s good to see the mixture of able-bodied children versus the children, people who aren’t so perhaps as able-bodied.’ (P11)

While unsuitable programs hindered exercise.

‘Disability sports are designed for people who have like an accident…they’re not really designed for people with limited upper body strength.’ (I1)

Interview participants were asked if the Australian Physical Activity and Sedentary Behaviour Guidelines were applicable and achievable for individuals living with SMA Type II and whilst the majority (91%) disagreed that the guidelines were applicable half agreed that they were achievable (Figure 11).

Figure 11. Interview responses from individuals living with SMA Type II regarding applicability and achievability of the Australian physical activity and sedentary behaviour guidelines

*Agreed dependent upon factors such as tiredness and degeneration.
Participants thoughts about sedentary behaviour produced positive behavioural change (n=6), no change in behaviour (n=3), was not consciously thought about (ingrained) but produced positive behavioural change (n=1), or was not thought about (n=2).

5.7.1.2 The impacts of access to services, facilities and equipment on attitude, subjective norms, perceived behavioural control and exercise behaviour

Participants living in all areas of Australia (remote, rural and metropolitan locations) reported difficulty accessing services, facilities and equipment, which impacted on their quality of life and exercise behaviour (n=11). Wheelchair accessibility of facilities, including ramps, hoists, climate control and appropriate toilets or change rooms, was an exercise determinant; accessibility was a facilitator and inaccessibility was a barrier (n=11). Facilities mentioned included community settings, pools, exercise facilities, schools, shops, homes and practitioner offices.

In regard to the inaccessibility of facilities, concerns were raised about potential illness, embarrassment, lack of privacy, inconvenience and restriction of choices for individuals as well as workplace health and safety concerns for individuals and their support system.

‘The pool in “This Town” doesn’t have an adult size changing table, so if I want to take “my child” to the pool there, luckily it’s close so I come home, but getting “my child” changed there before or after a swim is really awkward and it’s not well set up in terms of ergonomics and caring for the carer in terms of getting “my child” changed and stuff. And also there’s not great access.’ (P5)

Equipment was an exercise determinant (n=11); it facilitated exercise engagement or created barriers when there were delays in replacing equipment or through incorrect use of equipment. Participants (n=5) noted that delays in replacing equipment, like standing frames, lead to degeneration of their condition and either inability or great difficulty in returning to using the equipment and exercise.

‘[Ankle-foot orthoses] AFOs took eighteen months to replace one set. So to fight the system to get “my child” that weight bearing essential shoe put “my child” off the standing frame and it put “my child” off the AFOs. Once “my child” grew out of those AFO’s, and once it
caused “my child” pain, it was very hard to get “my child” back in there. There was a lot of effort that we went to with the physical activity…” (P3)

5.7.1.3 Impacts of access to transport and travel distances on perceived behavioural control and exercise behaviour

All interview participants used power wheelchairs, and six reported current or previous use of manual wheelchairs. Transportation of participants required modified cars or the addition of specialised car seats, wheelchair-accessible taxis or wheelchair-accessible public transport (n=11). Travel distances to accessible exercise facilities (e.g. pool, dance studio) were varied and in some instances were further in metropolitan areas than in rural or remote locations. However, all participants living in rural or remote locations had to travel greater distances to receive medical care from specialists, and were sometimes required to travel interstate.

5.7.1.4 Impacts of climate and weather on perceived behavioural control and exercise behaviour

Participants (n=8) perceived that climate impacted their living with SMA Type II in several ways. Hot, tropical climates were perceived to potentially reduce the risk of recurrent chest infections (n= 4), but equipment, including wheelchairs and orthotics, were uncomfortable and sweaty to wear (n=5). Winter increased the risk of illness, and in particular respiratory illness (n=4). Climate also impacted time spent in, and type of, exercise behaviour. Participants chose to either not to engage in exercise or sought out more appropriate facilities, where available, to engage in exercise based on climate. Examples included air-conditioned facilities in summer or indoor heated pools in winter. Exercising in climate-appropriate facilities increased the time and cost involved in exercising for some participants (n=3) and was not available for others (n=3), with both scenarios decreasing exercise frequency of participants.

5.7.2 Socio-cultural exercise determinants for individuals living with SMA Type II

Socio-cultural exercise determinants for individuals living with SMA Type II (intermediate tier of Figure 10) included family, practitioners, friends and peers,
schools, support organisations and societal understanding and attitude towards individuals and their exercise engagement. These determinants helped to explain participants’ beliefs, perceived behavioural control and their subjective norms towards exercise behaviour through consideration of the impacts socio-cultural determinants have on their intention to exercise.

5.7.2.1 Family influence on attitude, subjective norm, perceived behavioural control and exercise behaviour

Families, including parents and romantic partners, were found to influence the exercise behaviour of individuals living with SMA Type II by contributing to the individuals’ subjective norm. Parental thoughts about their own exercise behaviour, parental influences on the exercise behaviour of individuals living with SMA Type II, parental trust or mistrust of practitioners’ advice, parental perceptions of the future and the exercise behaviour of romantic partners were identified.

To ascertain the subjective norm experienced by individuals living with SMA Type II, parents (n=12) were asked to describe their personal feelings regarding exercise, physical activity and sport in the questionnaire, as presented in Figure 12. The majority of parents (58%) enjoyed and participated in exercise whenever the opportunity presented itself.

![Figure 12. Feelings of parents of individuals living with SMA Type II towards their own exercise behaviour](image-url)
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Parental influence on participants’ exercise behaviour was an exercise determinant and a facilitator for exercise for many interview participants (n=9).

‘My parents really pushed me to do a lot of exercise.’ (I1)

‘We call it you snooze you lose. The more we keep “my child” active, the more hopefully it slows down the muscle degeneration and that’s what we focus on.’ (P4)

Parental trust or mistrust in practitioners’ advice was an exercise determinant for some participants (n=5).

‘The moment we took that exercise away [due to surgery], the deterioration was really noticeable. That really proved to me that it’s not a waste of time but actually has got benefits. [The physicians] tried to make us do it again...we actually had to fight them. “My child” lost so much condition after the operation, that it wasn’t really worth it if “my child’s” feeling no pain.” (P11)

Some parents discussed their child’s life as an adult: one mentioned moving great distance to care for their child if they go to university; one was concerned about their child’s care post-school; and two mentioned their children living with them as adults. Parents also mentioned their concerns for their children not experiencing romantic relationships (n=2) or having children (n=1).

‘I even look forward to once “my child” finishes school, that we’re going to get up every morning, we’re going to go for a swim, we’re going to come home, we’re going to do standing frame. I’ve got it all mapped out in my head! Even just to have a granny flat or something, a space for “my child” to live, is going to be the challenge.’ (P3)

All three adults living with SMA Type II interviewed were living independently. For two, the majority of their care was provided by their spouse. This included self-care needs (e.g. changing bed positions up to thirteen times in one night), administering equipment (e.g. cough machine), household tasks and driving (e.g. attending practitioner appointments). One participant mentioned their partner’s engagement in sport and exercise, as well as their current exercise goal to lose weight.
5.7.2.2 Practitioners’ influence on attitude, subjective norm, perceived
behavioural control, exercise behaviour and exercise knowledge

Interactions with practitioners, including physicians, physiotherapists
(physios) and exercise physiologists (EPs) were reported to influence the exercise
behaviour of individuals living with SMA Type II by contributing to their knowledge
(cognitive determinant) and their subjective norm regarding their exercise behaviour.
Participants’ consultations with their practitioners, participants’ perceptions of their
practitioners attitudes towards exercise, and participants’ perceptions of practitioners
exercise advice and prescription were investigated.

Some interview participants perceived that at diagnosis, their physicians’
knowledge of SMA Type II was limited or misleading (n=3).

‘The first paediatrician we were seeing...she didn’t really know
nothing about it [SMA]. She was talking about, in that very first
setting...“my child” having to have a peg, that the life expectancy is
not really, is two plus years...which I think she got straight from a
journal article.’ (P10)

Practitioner knowledge about exercise for individuals living with a
neuromuscular disease was an exercise determinant (n=9). It was a facilitator for
participants whose practitioners were well-informed, or who became so (n=7), but a
barrier for participants whose practitioners were not well-informed (n=5). There were
also participants (n=2) whose practitioners were not well-informed and did not
become well-informed but this did not deter them from exercising.

‘We’ve been lucky because we’ve had...a lot of different physios come
through, but they’ve all been young physios and they’ve all been
learning at the same time, so they’re also getting as much information
as they can, which has been good.’ (P11)

‘You’ve got no support...the therapists weren’t proactive in
recommending exercises either.’ (P2)

Participants’ perceptions of consultations with practitioners regarding exercise
in comparison with the Consensus Statement guidelines are presented in Table 6.
Interview participants felt that the practitioner in charge of their primary care was a
neuromuscular clinic (n=7, aged <14yrs), a neurologist (n=3, aged >16yrs) and a
respiratory specialist (n=1, aged >16yrs).
### Table 6. Questionnaire participants’ perceptions about their interactions with practitioners stratified by age, gender and functional status

<table>
<thead>
<tr>
<th>Consensus Statement</th>
<th>Practitioner outcome</th>
<th>‘Sitters’</th>
<th>‘Walkers’</th>
</tr>
</thead>
<tbody>
<tr>
<td>‘The physician should also formulate a plan of multidisciplinary intervention with the family. This usually includes referral to ... orthopaedic/ rehabilitation.’</td>
<td>Doctor discussed exercise (n=12, 60%)</td>
<td>3 (38%)</td>
<td>4 (67%)</td>
</tr>
<tr>
<td>‘‘Sitters’ ‘Physical therapy, and occupational therapy are of highest value...’</td>
<td>Advised to consult physio (n=15, 75%)</td>
<td>5 (63%)</td>
<td>4 (80%)</td>
</tr>
<tr>
<td>‘Walkers’ ‘The highest emphasis is on provision of physical therapy, occupational therapy, and wheelchair/mobility...’</td>
<td>Consulted physio (n=18, 90%)</td>
<td>7 (88%)</td>
<td>5 (100%)</td>
</tr>
<tr>
<td>Currently consulting physio (n=12, 60%)</td>
<td>6 (75%)</td>
<td>3 (60%)</td>
<td>2 (67%)</td>
</tr>
<tr>
<td>Advised to consult EP (n=3, 15%)</td>
<td>2 (25%)</td>
<td>1 (20%)</td>
<td>2 (20%)</td>
</tr>
<tr>
<td>Consulted EP (n=5, 25%)</td>
<td>3 (38%)</td>
<td>1 (20%)</td>
<td>1 (25%)</td>
</tr>
</tbody>
</table>

n, number of participants; %, percentage of classification (status, age or gender); physio, physiotherapist; EP, exercise physiologist

Questionnaire participants’ perceived that the majority of their physicians’ attitudes towards exercise were facilitatory (60%), but a large minority (40%) felt that their physicians did not discuss exercise with them at all as presented in Figure 13. Two participants made the following additional statements when asked what their doctor’s attitude was towards their child’s engagement in exercise:
‘Unsure. All exercise options were suggested by me and discussed with the doctor.’ (P3)

‘The doctor really did not have much of a clue about SMA, and basically thought that nothing would help.’ (P10)

Figure 13. Questionnaire participants’ perceptions of their physicians’ attitudes (in general) towards exercise as a treatment for SMA Type II

Participants’ perceptions of their physiotherapists’ attitudes towards exercise were mainly reported as facilitatory (89%) as presented in Figure 14. Three participants made additional statements regarding the exercise advice they had received from their physiotherapist:

‘It was largely in the early stages before confirmed diagnosis. Once we received diagnosis we were transitioned to other services.’ (P1)

‘You must be cruel to be kind. Never forget it.’ (P2)

‘Exercise [was] not considered worthwhile, other than standing in a standing frame.’ (P6)
Participants’ perceptions of their EPs’ attitudes towards exercise were positive or very positive (n=3), you must be cruel to be kind (n=1) or unknowledgeable about exercise and SMA (n=1).

Interview participants’ interactions with their doctors and allied health professionals were varied, with exercise being facilitated (n=4), hindered (n=2) or a mixture of both (n=5) by practitioners.

Practitioners facilitated exercise by encouraging exercise engagement, sourcing equipment and providing knowledge about exercise to individuals living with SMA Type II.

‘My neurologist...didn’t have any concerns or anything about exercise. And OTs and stuff are always encouraging exercise and trying to get me in the pool.’ (I3)

‘[Physiotherapist] did a fantastic job and she was determined and she helped us with the AFOs and trying to get some equipment.’ (P3)

Practitioners hindered exercise by not being well-informed about exercise or equipment and discouraging exercise engagement for individuals living with SMA Type II.

‘Occupational therapist, physio when I went to the hospital but it was nothing about exercises, it was all breathing related. I was on a cough assist for a while as well. Even some of the physios couldn’t use the
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machine properly. I think I got kind of annoyed when “Dr X” the neurologist at “The Hospital” basically went “Panadol osteo”, that’s it. I’m like “Is that it? You’re not going to recommend exercises? You’re not going to recommend breathing exercises? Nothing?”’ (I5)

‘The advice I got from the therapist was pretty unhopeful. They didn’t think that that [exercise] would have any positive effect. So I was given quite, it wasn’t indicated at any stage that exercise could help. No one ever encouraged it, that’s for sure. I don’t think it was discussed, and from the therapists, it was discouraged rather than encouraged.’ (P5)

Practitioners facilitated exercise through encouragement and exercise prescription and hindered exercise through lack of discussion or presenting information that was not well-informed about exercise.

‘The doctor encouraged my parents to get me to exercise and so that was it really. Outside that it’s not really been, not discouraged, but not particularly encouraged to do it.’ (I1)

‘Physios probably right back at the start, when we did enquire about it. And that was because we just didn’t have any information and they didn’t know themselves...they did actually prescribe exercises I remember back then. With “my child’s” legs and stuff like that. It was quite conflicting information actually.’ (P2)

Questionnaire participants’ perceptions of exercise advice and prescription received from physicians, physiotherapists and EPs indicated who provided exercise advice and which practitioners actually prescribed an exercise program as presented in Table 7. The majority of participants (90%) received exercise advice and prescription from physiotherapists, with stretching and swimming the most popular exercises advised and prescribed to them. Additionally, four participants stated that occupational therapists had advised them to engage in exercise.
Table 7. Questionnaire participants’ perceptions of exercise advice and prescription from physicians, physiotherapists and exercise physiologists (EPs) stratified by functional status

<table>
<thead>
<tr>
<th>Consensus Statement guidelines</th>
<th>Exercise</th>
<th>Total Participants</th>
<th>Total Responses</th>
<th>Functional status (highest achieved without equipment)</th>
<th>Exercise advice n (% of total responses)</th>
<th>Prescribed exercise n (% of total advised to exercise by that professional)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td>n=23</td>
<td>n=20</td>
<td>n=80</td>
<td>Physician</td>
<td>Physiotherapist</td>
</tr>
<tr>
<td>Stretches management</td>
<td>12 (60%)</td>
<td>20 (25%)</td>
<td>Total 8 (67%)</td>
<td>9 (50%)</td>
<td>6 (67%)</td>
<td>4 (80%)</td>
</tr>
<tr>
<td>Swims</td>
<td>12 (60%)</td>
<td>19 (24%)</td>
<td>Total 8 (67%)</td>
<td>7 (39%)</td>
<td>3 (43%)</td>
<td>1 (20%)</td>
</tr>
<tr>
<td>Standing and walking frame</td>
<td>7 (35%)</td>
<td>11 (14%)</td>
<td>Total 2 (17%)</td>
<td>4 (22%)</td>
<td>4 (100%)</td>
<td>2 (100%)</td>
</tr>
<tr>
<td>Swimming/</td>
<td>5 (45%)</td>
<td>7 (60%)</td>
<td>Total 2 (17%)</td>
<td>2 (11%)</td>
<td>1 (100%)</td>
<td>1 (100%)</td>
</tr>
<tr>
<td>Cough</td>
<td>1 (50%)</td>
<td>1 (100%)</td>
<td>Total 1 (7%)</td>
<td>1 (100%)</td>
<td>1 (100%)</td>
<td>1 (100%)</td>
</tr>
<tr>
<td>Cough machine/</td>
<td>1 (100%)</td>
<td>1 (100%)</td>
<td>Total 1 (100%)</td>
<td>1 (100%)</td>
<td>1 (100%)</td>
<td>1 (100%)</td>
</tr>
<tr>
<td>Exercise</td>
<td>1 (100%)</td>
<td>1 (100%)</td>
<td>Total 1 (100%)</td>
<td>1 (100%)</td>
<td>1 (100%)</td>
<td>1 (100%)</td>
</tr>
<tr>
<td>General exercise</td>
<td>1 (100%)</td>
<td>1 (100%)</td>
<td>Total 1 (100%)</td>
<td>1 (100%)</td>
<td>1 (100%)</td>
<td>1 (100%)</td>
</tr>
<tr>
<td>Respiratory exercise</td>
<td>6 (30%)</td>
<td>9 (11%)</td>
<td>Total 1 (8%)</td>
<td>3 (17%)</td>
<td>4 (133%)</td>
<td>1 (20%)</td>
</tr>
<tr>
<td>Strength exercise</td>
<td>3 (15%)</td>
<td>3 (4%)</td>
<td>Total 2 (11%)</td>
<td>1 (50%)</td>
<td>1 (100%)</td>
<td>1 (100%)</td>
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N.B. Two participants (1 male, 32 yrs, sit; 1 female, 34 yrs, walk) did not receive exercise advice from their physician, were not advised to consult a physiotherapist and had not consulted a physiotherapist or EP. Three participants were advised to consult an EP and five participants consulted an EP.
Participants were also advised by practitioners to participate in a variety of exercises that can be functionally grouped, including: percussion (n=3, 15%), massage/manual manipulation (n=3, 15%), fine motor skills (n=2, 10%), postural support (n=2, 10%), ‘rolly-polly’ (n=2, 10%), splinting (n=2, 10%), trike (n=1, 5%), musical instrument (n=1, 5%), dance (n=1, 5%), pain management (n=1, 5%) and yearly assessment (n=1, 5%).

5.7.2.3 Influence of friends and peers on attitude, subjective norms, perceived behavioural control and exercise behaviour

Information on interaction with friends and peers was not specifically asked of participants; however, some participants mentioned that interactions with friends and peers were determinants for exercise (n=9). Participants depended on friends to perform body movements for them that could facilitate and/or hinder their exercise behaviour (n=2) and/or expressed that friendship could promote inclusion (n=8) or exclusion (n=8) in exercise. Participants also expressed a desire for normality in their appearance, actions and interactions with others (n=9), and if this was not achieved it could lead to feelings of self-consciousness or peer pressure (n=3).

Dependency on friends could facilitate, but more often, it hindered exercise.

‘I had a manual chair…but you become pretty either dependent on others if you’re going anywhere of real distance or you don’t go anywhere so you become pretty isolated.’ (11)

‘I don’t usually put my hand up and if we have to put our hand up I get my friend to help me. [With school physical education] I’d always have to get someone else to go with me and none of my friends wanted to do it, so then I had to get the teacher to do it. And I didn’t really like that so I just umpire now.” (P6)

Normalcy of actions and interactions with others facilitated inclusive exercise.

‘[Teachers would] put a drawing easel in front of “my child”…so “my child’d” stand up in the splints and do work on that, but then they’d also set up two easels for either side of “my child” and…have “my child’s” friends come in there.’ (p10)

‘Definitely I think [exercise has] improved [quality of life] and definitely the social aspect. “My child” had a pool party so, “my child” was able to play with all “my child’s” friends as any child
would be able to. [Exercise is] improving her participation and her engagement with community and engagement with friends.' (P5)

The desire for normality could lead to self-consciousness and peer pressure.

‘In terms of actually doing any exercise, you wouldn’t want to be doing that in front of other kids, I think, ‘cause you need room to feel comfortable in figuring out what you can do and making a fool out of yourself a bit. But in terms of being social, it was good to participate in what they were doing.’ (I3)

‘“People are looking at me, I’m different, so I don’t want to do it” and “my child” just wants to be a normal kid. So it’s that perception of just because you’re in a chair you can still do things but “my child’s” seeing it more as a peer group image thing, which is “my child’s” age at the moment too. It was equal playing ground in the water. So I think that way you’ve got your confidence, you’ve got your peers, you’ve got interaction, you’ve got acceptance.’ (P12)

5.7.2.4 Influence of school on attitude, subjective norms, perceived behavioural control and exercise behaviour

Schools and teachers were determinants for exercise (n=11), and could facilitate or hinder participants’ exercise behaviour. The majority (55%) hindered individuals’ exercise engagement as presented in Figure 15. Exercise was facilitated through inclusive physical education (PE) (n=5), with modified activities for individuals living with SMA Type II or modification of the activities of the rest of the class (for example by sitting in chairs or on skateboards to exercise). Exercise was hindered (n=8) through exclusion of PE (for example by lack of teacher knowledge on how to adapt exercise and/or by directing individuals to perform activities including refereeing, time-keeping or homework). Some interview participants (n=4) perceived that school engagement led to a more sedentary lifestyle through extended periods of sitting in a wheelchair, which had implications for secondary conditions, such as scoliosis or contractures.
School can facilitate exercise through inclusive PE.

‘School’s been really, really good with “my child”, and very inclusive, and they try to include “my child” wherever possible and they modify activities sometimes. The school brings up specialists from the inclusive learning unit and the teachers all have training on all this sort of stuff so they come up with bright ideas. They really do, they try.’ (P11)

School can facilitate exercise by becoming well informed about inclusive PE and hinder exercise by excluding individuals from PE.

‘There was probably three teachers that thought outside that box. The rest, they would, “my child” would score, “my child” would referee, “my child” would set-up, but “my child” would not participate. I would love the education system to actually, probably do courses on adaptive sports. There’s lots of children out there who’ve got totally different, lots of disabilities…that would benefit from it. So I’d like to see a lot more education to the teachers to look at different ways you can do sport.’ (P12)

Some interview participants mentioned contacting primary schools up to two years prior to enrolment to ensure that they had the facilities and ethos to care for their child.
5.7.2.5 Influence of support organisations on subjective norms, perceived behavioural control and exercise behaviour

Several organisations provided fundraising for equipment that was used by interview participants to facilitate their exercise behaviour directly or indirectly, and improve their quality of life (n=4). These organisations included Lions Australia, Apex Australia, Variety Australia and Toy Libraries Australia. Interview participants also mentioned efforts by their community that ranged from providing meals to fundraising for equipment that facilitated individual’s exercise behaviour (n=5).

‘Last year we were struggling to get a wheelchair-accessible vehicle and I was short a little bit of money and all of a sudden out of the woodworks people going, “Oh you can have this.” I think the lines of communication of what’s out there could be more so as “my child’s” carer, I could provide more for “my child”. It’s a lot of word of mouth, so if you don’t know, and especially being new to the area, you just don’t know.’ (P12)

SMA Association of Australia, Muscular Dystrophy Foundation and Muscular Dystrophy Australia all provided knowledge, equipment and/or support to interview participants.

‘The day that “my child” got pneumonia, three days later I had that [cough assist machine] delivered at my door by Muscular Dystrophy. No questions, no ifs, no hassles, no whatever’s, they did it.’ (P3)

5.7.2.6 Influence of societal understanding and attitude on attitude, subjective norms, perceived behavioural control and exercise behaviour

The societal understanding and attitude experienced by participants was indicated as an exercise determinant. Participants described experiencing discrimination and a lack of understanding, which sometimes created barriers to exercise (n=8). Participants also described experiencing community support, particularly those who lived in remote or rural settings, which included financial and emotional support that facilitated their exercise behaviour (n=6).

Societal attitude sometimes facilitated exercise.

‘I thought “my child” would be the one that no one wanted to partner up with [during sport], you know because, but it was exactly the
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opposite. They were fighting over who wanted to be “my child’s” partner, so that was really lovely and pleasing to see.’ (P10)

However, sometimes societal attitude hindered exercise.

‘Changing people’s perceptions is also a limitation [of exercise behaviour].’ (P12)

‘“My child’s” concern is other kids if you go to a public pool…they’re often very interested in why someone of “my child’s” age is wearing arm bands. So they come over and they ask “my child” questions and they poke “my child” and they prod “my child”’. (P6)

‘One time like a 15 year old guy, who was really fat, he tried to sit on me and then I almost drowned.’ (Child of P6)

Societal understanding could similarly hinder exercise.

‘We didn’t end up going to any playgroups where “my child” could have that social interaction. I just found it quite confronting that “my child” couldn’t always, I mean I lifted “my child” and did it [physical activity], but they could have modified things slightly to help “my child’s” engagement…to maximize “my child’s” participation and obviously they didn’t have the skills to do that.’ (P5)

‘A lot of people see “my child” in a wheelchair and think “my child’s” got an intellectual disability. So thinking, “Oh “my child” can’t do a lot”. They see a chair they just think disability everything not just physical.’ (P12)

Discrimination was also a factor that hindered exercise and affected individual’s well-being.

‘I was so enraged because we got four clearances and it was once “my child” got on the ride that “my child” was told to get off. When you are consistently faced with these challenges it does, it makes you want to shut down.’ (P3)

Whilst community support facilitated exercise.

“People were like, “We’re going to get behind you and we’re going to help you fundraise.” And it was beautiful, it was really special. And yeah, lots of people, we didn’t asked for anything, but they just fundraised.’ (P11)
5.7.3 Individual exercise determinants for individuals living with SMA Type II

Individual exercise determinants (central tier of Figure 10) can bring understanding to the attitude and perceived behavioural control of participants, thus impacting their intention and exercise behaviour. Individual determinants of socio-demographic, cognitive, biological, psychological and behavioural determinants are reported.

5.7.3.1 Influence of socio-demographic determinants on perceived behavioural control and exercise behaviour

Two prominent socio-demographic determinants for exercise behaviour were age (n=11) and financial resources (n=9). Age was a determinant more from its association with other determinants: environmental determinants as presented in section 5.7.1, including access to services, facilities and equipment; socio-cultural determinants as presented in section 5.7.2, including school engagement and influence of peers; and other individual determinants such as biological determinants, including degeneration and cognitive and psychological determinants, such as the cost benefit ratio (enjoyment or appearance of normalcy vs. health benefits). Socio-economic status was not explored; however, participants perceived their financial resources to be a determinant for exercise behaviour. Interview participants identified that the additional costs involved with exercising, including purchasing equipment, employment of health professionals to facilitate exercise engagement (dependent on availability), private lessons due to ability and distances to facilities could hinder exercise engagement.

‘We’re in a fortuitous situation where we can self-fund a lot of things. A family whose income is only $40,000 a year will have the $8,000 chair. “My child” has the twenty-eight thousand dollar chair. “My child” can go up to a table like that and raise and lower, where this other child can’t, and you think for a little bit more funding that kid would have much more…quality of life.’ (P11)

5.7.3.2 Influence of cognitive determinants on attitude, perceived behavioural control and exercise behaviour

Several cognitive determinants to exercise behaviour became apparent for participants. Knowledge, or lack thereof, about exercise and exercise facilitating equipment was prominently discussed by interview participants as a determinant for
exercise (n=11). Interview participants also reported on the cost-benefit ratio for exercise engagement (n=11). Costs included experiencing or fear of experiencing injury, illness, pain or discomfort, financial costs, time, lack of enjoyment, self-conscious about appearing different to others and exposure to bullying. Benefits included improvement or maintenance of biological functions, psychological health, overall health benefits, confidence, enjoyment, inclusion, quality of life, independence as well as interactions with friends and peers. Additionally, participants mentioned compromises that were made in the decision to engage or not engage in exercise, including the choice to support the body or to have mobility; and the choice to depend on others to engage in exercise or to be socially isolated.

The ability to advocate (n=11) was a cognitive determinant for exercise and facilitated exercise behaviour when an individual or parent was able to advocate for their right to engage in exercise and vice versa. Prioritising (n=11), time management (n=9), planning (n=9) and goal setting (n=4) were also considered to cognitive determinants for exercise engagement and could facilitate or hinder an individual’s exercise behaviour. Tangible results from exercise was a cognitive determinant. Improvement or maintenance of bodily functions (e.g. respiratory, mobility, strength) facilitated exercise behaviour (n=8). Lack of results could facilitate exercise if regarded as maintaining function and/or slowing degeneration (n=8), or hinder exercise if regarded as not having any effect (n=2). Finally imagination (n=5) could facilitate exercise while lack of imagination could hinder exercise engagement.

An individuals’ inability to self-advocate hindered exercise.

‘“My child” did come home rather disappointed that “my child” wasn’t included, so [I said] ““My child” you need to speak up and say...you can adapt it and you can join in.”’ (P3)

Lack of prioritisation and time management hindered exercise.

‘If we’re ever in the pool it’s an hour, by setting set up, getting “my child” ready, transferring “my child” in, getting “my child” in, playing, and trying to get “my child” out. So the process of actually doing is so long that sometimes it’s like I can’t be bothered.’ (P12)

However, planning and goal-setting facilitated exercise.

‘I’ve got a 12 to 18 month plan always in advance of where “my child” is now. I’ve just done the next wheelchair application because I
know in 12 months we’re going to be finished there and so I want that next piece of equipment in place. I’m not going to ever get to a point where it’s not right and I have to wait nine months for a new piece of equipment.’ (P5)

Whilst lack of imagination also hindered exercise.

‘The sheer limitations of one, my imagination in terms of what “my child” could be doing because obviously “my child” can’t do a lot of what I would perceive as normal exercise and trying to come up with other things creates quite a challenge.’ (P6)

5.7.3.3 Influence of biological determinants on attitude, perceived behavioural control and exercise behaviour

Biological determinants encompassed participants’ functionality and their perceptions of the effects that exercise had on functionality. Motor function ability (n=11), respiratory function (n=11), muscle strength (n=11), an individual’s weight (n=11), orthopedic health (n=10), pain (n=9), tiredness (n=9), growth (n=8), injury (n=8), illness (n=8) and flexibility (n=3) were biological determinants that impacted upon individuals’ exercise behaviour.

Exercise could improve motor function ability.

‘The swimming’s been great. “My child” got “their” arm up and out of the water, but the moment that we stop there’s a backlash.’ (P3)

Exercise could also improve respiratory function.

‘I think the day use of cough machine number one and everyone keeps telling me that swimming is good. Other than that, I think what keeps “my child” breathing is keeping “my child” straight and keeping “my child’s” lungs open. You are fighting the breathing by trying to fight the scoliosis in some ways, it’s all interrelated and intertwined with each other.’ (P11)

Weight gain hindered exercise, as did pain.

‘We really have to be conscious of in this instance “my child’s” weight because carrying too much weight will then of course impact the activity that “my child” can do.’ (P10)

‘I went into a swimming pool about two years ago. I haven’t done it since, maybe even longer than that actually. I couldn’t move for a
week. I was so sore, and that was only, like, for an hour. So that kind of scares me off going into the swimming pool.’ (I5)

Lack of orthopaedic health hindered exercise and motor function ability.

‘I broke my arm. I kept it in a sling for probably about six weeks. Beforehand I could lift it to at least my shoulder height and afterwards I could only get it to about there. So you notice, you do really notice the differences and how quickly it can happen.’ (I1)

Injury also hindered exercise.

‘If I just pushed myself too hard and you know damaged the muscle it would get quite sore and then afterwards it would be weaker for a while. And then of course it’s hard to tell if it ever gets back to the strength that it was before you damaged the muscle.’ (I3)

5.7.3.4 Influence of psychological determinants on attitude and exercise behaviour

The most common psychological determinants described by interview participants for their exercise behaviour were motivation (n=10) and enjoyment (n=10). Pain (n=9), social isolation (n=9), attitude (n=7), frustration/conflict (n=7), independence (n=5), confidence (n=5), vulnerability (n=4), self-esteem (n=2) and empowerment (n=1) were also considered to be psychological determinants by interview participants.

Lack of motivation hindered exercise, while enjoyment facilitated exercise.

‘I think the issue is the time and the motivation [to exercise]. ’ (P6)

‘[Exercise] was fun. It certainly made me feel good.’ (I3)

Conflict hindered exercise.

‘It becomes a battle. It becomes hard work. You’re fighting a child for something that they can’t do, against the exercise that they can achieve, and they’ll then do more of that. We’re better off putting our efforts into something that you can do that’s functional, that’s fun, than to put it into those exercises that are non-fun.’ (P11)

Independence and confidence both facilitated exercise.

‘The benefits of [exercise] is that “my child’s” independent, that “my child’s” doing, playing with “my child’s sibling” independently, as
well as the movement. And I think that’s always, you know trying to get “my child’s” heart rate up and get the lungs working.’ (P10)

‘[Engaging in exercise] “my child” was interacting with “my child’s” peers, which is a big, big thing because “my child” didn’t have a lot of confidence, so it built “my child’s” confidence up.’ (P12)

Vulnerability hindered exercise, but heightened self-esteem encouraged exercise.

‘“My child’s” very, very wary. Even as much as “my child’s” had to do in the pool “my child” really, really like feels vulnerable.’ (P2)

‘So “my child” really does get a lot of “my child’s” own self esteem from that [exercise]. I’m just trying to build in other ways in which “my child” can show competence and achieve success. I think self-esteem based purely on what you can do physically is probably a fairly unsteady surface to build your self-esteem from when you’ve got SMA.’ (P5)

Questionnaire participants who were individuals living with SMA Type II (n=8) were asked about their personal feelings regarding exercise, physical activity and sport, with 50% stating that they enjoyed exercise but are prevented from participating due to their condition as presented in Figure 16. Three individual participants made additional comments about their feelings towards exercise:

{I enjoy exercise and participate in physical activity whenever the opportunity presents itself} ‘Means of exercise now that my mobility is severely reduced are hard to come by.’ (I3)

{I dislike exercise but engage for health benefits} ‘I hate doing it however I feel I must to reduce pain.’ (I8)

{My condition prevents engagement} ‘I find everyday activities make me tired enough like lifting shopping bags or washing my hair.’ (I6)
Parents of individuals living with SMA Type II (n=12) selected answers regarding their child’s personal feelings regarding exercise, physical activity and sport, with 42% reporting that their child enjoyed and engaged in exercise as presented in Figure 17. Three parental participants made additional comments of their perceptions about their child’s feelings towards exercise:

{My child enjoys exercise and participates whenever the opportunity presents itself} ‘Within “my child’s” capabilities.’ (P1)

‘They are young, they enjoy doing lots of things, they do not have a specific opinion on exercise.’ (P5)

‘In winter months [my child] stays out of sports activities. [My child] has recently stopped doing afternoon sport anyway as ability to integrate is increasingly limited; also scared of balls.’ (P9)
5.7.3.5 Influence of behavioural determinants on attitude, perceived behavioural control and exercise

Behavioural exercise determinants for participants included taking measures to prevent illness or injury, such as an annual flu shot, avoiding people who are sick and preventing pressure sores (n=11); avoiding sedentary behaviour (n=7); eating a healthy diet (n=6); taking medication, such as sodium valproate, Epilim and melatonin (n=6); and taking supplements, such as calcium or Swedish bitters (n=5).

'It’s pretty hard to exercise with SMA Type II, but of course you’ve still got a cardiovascular system that needs some form of exercise and stimulation and living a sedentary lifestyle has all sorts of other effects on your body, so it’s good to try and find stuff that you can do.’

(I3)

5.8 Conclusion

Eight individuals and twelve parents responded to the online questionnaire. Eighteen participants had engaged in exercise over their lifetime and twelve were currently exercising. Swimming was the most popular exercise engaged in, and it was thought to be effective in maintaining or improving cardiorespiratory function, motor function ability, orthopaedic health and muscular strength. The majority of participants perceived that exercise was beneficial in treating SMA Type II. The
majority of interview participants agreed that exercise slowed the progression of SMA Type II. Accessibility, parental influence, friends, school, funding, knowledge, degeneration and motivation were identified as influential determinants for participants exercise behaviour. These research findings are discussed in Chapter 6.
Chapter 6  Discussion

6.1  Introduction

This chapter discusses key research findings. These included participant characteristics in relation to the research objectives (to explore participants’ exercise engagement and the effectiveness of exercise to treat and slow the progression of SMA Type II, the value of exercise in treating SMA Type II, and exercise determinants). Participant characteristics included individuals’ motor function ability in relation to the classification of SMA and how exercise was affected by participants’ age and additional conditions indirectly associated with SMA. The first research objective is addressed by discussion of participants’ exercise behaviour, including their definitions of exercise, exercise type, exercise frequency, and effectiveness of exercise. The second research objective is addressed by discussing the perceived psychological and physical benefits of exercise, including the value of cardiorespiratory and fine motor exercises. The third research objective is addressed by discussing the role of policy, practitioners and schools as exercise determinants for individuals living with SMA Type II. Finally the research strengths and limitations are discussed.

6.2  Participant characteristics

Interesting observations about participant characteristics were individuals motor function ability that exceeded the clinical classification for SMA Type II and the varying effects that age and conditions, indirectly associated with SMA, had on participants’ exercise behaviour.

6.2.1  Motor function ability exceeds classification of SMA

Although SMA Type II is clinically classified as ‘never stands’, some participants exceeded this classification, stating their highest independent motor function achieved was walking and crawl & stand. There are three possible explanations for this discrepancy:

1. Participants may have misunderstood the following statements/questions:
   a. ‘I am an individual with SMA Type II’ or ‘I am the parent/guardian of an individual with SMA Type II’, and
b. ‘What is the BEST level of movement you have EVER been able to achieve independently WITHOUT equipment?’ or ‘What is the BEST level of movement your child/children have EVER been able to achieve independently WITHOUT equipment?’;

2. Physicians may have misdiagnosed individuals as having SMA Type II; or

3. Clinical classification of SMA Type II may need to be redefined to reflect changes to motor function ability and life expectancy.

Whilst it is possible that individuals who had a different type of SMA may have participated in this research, all but one of the participants stated that they had their diagnosis confirmed through genetic testing. Furthermore, it was a concern of all the associations that administered the online questionnaire that individuals would not know which type of SMA they had. Therefore, the associations did not advertise the online questionnaire on their websites, but emailed or mailed individuals they had registered as having SMA Type II. Due to these precautions, it seems unlikely that participants in this research would not have known what type of SMA they had.

It is possible that doctors may misdiagnose participants as Type II rather than Type III, despite genetic testing. It is apparent that participant I5 misinterpreted the question regarding highest independent motor function ability achieved as they described walking independently as “leaning on stuff assisted” (section 5.2). It could be argued that other participants’ definition of independent walking may not meet a practitioner’s clinical definition. However, the motor function ability of these participants would have still exceeded the clinical classification for SMA Type II of ‘never stands’. Individuals diagnosed with SMA Type II that have the ability to walk and stand have been cited in other research.\textsuperscript{40,41,43,92,170,171} The Consensus Statement for Standard of Care in Spinal Muscular Atrophy (Consensus Statement) also acknowledges that motor function ability of individuals living with SMA Type II is variable and can exceed the clinical classification of ‘never stands’ to include individuals who stand with braces or a standing frame.\textsuperscript{4}

Research has acknowledged controversy surrounding the clinical classification of SMA.\textsuperscript{2,6,10,172,173} It is presumed that improvements in motor function ability and life expectancy for individuals living with SMA Type II are due to advancements in medical and therapeutic care that have occurred in the past 25 years.\textsuperscript{6,38,173} Currently, trials are being conducted into drug, genetic and supportive treatments for SMA.\textsuperscript{90} As advancements in medical and therapeutic care continue to occur, it is highly likely...
that individuals’ motor function ability and life expectancy will also continue to improve. The latest research on life expectancy for individuals living with SMA Type II has found that approximately 93% live to the age of 20 years and approximately 52% live to the age of 40 years.² By not acknowledging the advancements to motor function ability and life expectancy, the clinical classification of SMA is becoming outdated and will become more outdated with continued advancements in medical and therapeutic care.

There are implications to not redefining the clinical classification of SMA. Firstly, not every physician or health practitioner who treats an individual living with SMA Type II is an expert in SMA. Those who are not may refer to the clinical classification of SMA and 1) convey that information to their patients and 2) base their treatment or advice on the information provided by the clinical classification system. Secondly, researchers often quote the clinical classification of SMA and do not appear to delve into the current literature on life expectancy and motor function ability, which may have implications for their research. Thirdly, it may affect how individuals view themselves, their abilities and their life decisions. If an individual perceives that they only possess the ability to sit, they may not try to further their motor function ability and thus may live a more restricted lifestyle than they otherwise could. Conversely, if an individual perceives that their functional abilities surpass their clinical diagnosis, it may lead them to believe that they have ‘beaten the odds’, thus instilling false hope when in actuality their abilities are on par with normal disease progression. Finally, it may affect how individuals view others with SMA and their interactions with them; one participant explained that online bullying had occurred between individuals due to their motor function ability exceeding the clinical classification for their diagnosis (section 5.2).

A review of the clinical classification to change the functional status and life expectancy of individuals living with SMA Type II may ensure that more accurate information is more easily accessible to practitioners, researchers, individuals living with SMA and their families. Individuals and families living with SMA would then be better placed to make informed decisions about their life and treatment options in consultation with their primary care physician.
6.2.2 Age as an exercise determinant

Age influenced individuals living with SMA Type II exercise engagement and adherence in multiple ways, at different ages. Furthermore, the level of influence of attitude, subjective norm, perceived behavioural control and exercise determinants differed depending on the individual’s age. With age parental influenced decreased whilst growth and degeneration influenced individuals’ physical abilities, which impacted their exercise behaviour. The change from child to adult services could also result in funding differences that may impact upon individuals’ exercise behaviour.

Participants perceived that small children exercise behaviour was heavily influenced by their parents. However, with the development of individuals’ own attitudes towards exercise the influence of subjective norm could decrease. Furthermore, the introduction of school and peers created new exercise determinants that lessened parental influence, particularly as individuals became pre-teens and teenagers, thus shifting the influentialness of determinants within subjective norm. This is in agreement with the literature, which found that parental influence is an exercise determinant for children and adolescents who do and do not live with an neuromuscular disease (NMD) or physical disability. In particular, parent’s weight, physical activity levels, exercise beliefs, father’s education and encouragement of their children are correlated with children’s physical activity levels. School, friends and an individual’s development of their own exercise beliefs and attitudes have also been reported to influence children’s and adolescents’ exercise behaviour, including those who live with an NMD.

Participants perceived that as they aged and grew, they became heavier and taller, which impacted on their carers’ ability to transfer them. Changes with growth could also impact upon the individual’s functional abilities, thus impacting their exercise behaviour. The literature also found that changes from growth hindered the exercise engagement of individuals living with an NMD. Furthermore, children living with SMA Type II are at risk of developing obesity, and obesity in children has been reported as an exercise determinant. As children or adolescents, individuals living with SMA Type II are also faced with the likelihood of undergoing spinal rod surgery to correct scoliosis. Participants perceived that this had an immediate negative impact on their exercise engagement and, for some, this lead to longer-term ramifications for their exercise participation. Inactivity during recovery from surgery was perceived to cause degeneration of functional abilities by
participants in this research. The literature has also found that inactivity and bedrest decreased the functional ability of individuals living with an NMD. Furthermore, spinal rods can be contraindicated for some exercises.

Participants, particularly adults and adolescents, perceived that degeneration of their condition, including decreases in motor function ability, respiratory function and strength, in combination with a lack of knowledge on how to adapt exercise, negatively influenced their attitude, perceived behavioural control and exercise behaviour. The literature has also found that degeneration and lack of exercise knowledge are significant exercise barriers for individuals who live with an NMD. Participants perceived that funding impacted upon their access to physiotherapy services and exercise choices available to them. Funding and socio-economic status have also been noted in the literature as exercise determinants for individuals living with an NMD. The transition into adulthood is not well-researched in this population group, with the Consensus Statement and the majority of research focused on children living with SMA Type II. Further research is needed to corroborate these findings and develop strategies to overcome these barriers to increase exercise participation of individuals living with SMA Type II.

6.2.3 Conditions not directly associated with SMA influence exercise behaviour

Conditions not directly associated with SMA (as identified by questionnaire participants included depression, anxiety, tachycardia, and encephalitis) may be perceived as additional barriers to exercise by these individuals. While these conditions are not directly associated with SMA Type II, there have been other reported cases of individuals living with SMA who have psychological, psychiatric and cardiac conditions.

Some participants surveyed in this research had depression and anxiety. However, psychological and psychiatric conditions are rare within this population. Separation anxiety disorder has been reported to be the most common psychiatric disorder in children and adolescents living with SMA. Major depressive disorder, post-traumatic stress disorder, and oppositional defiant disorder were also listed. Depression and anxiety can create barriers to exercise engagement for all individuals, including individuals living with an NMD. Depression has also been found to be a barrier to exercise engagement in children. Exercise could improve depression through physiological changes, psychological constructs and social
Physiological changes include changes to hormone levels, like endorphin and cortisol, the growth of new nerve cells and the release of proteins, like brain-derived neurotrophic factor. Therefore, theoretically, the production of new nerve cells and proteins could also reduce the severity and improve the motor function ability of individuals living with SMA Type II. Psychological constructs include concentrating on learning a new skill, distraction from negative thoughts and improvement to self-efficacy and self-esteem.

Exercise has been reported to be more effective than no treatment and may be as effective as psychological therapies and antidepressant drugs. However, more high quality research is needed to confirm these conclusions. Participation in exercise and recreational activities have been found to be inversely correlated with depression and anxiety in individuals living with an NMD. Exercise has also been found to reduce depression and anxiety in children; however, research is limited and more high quality research is also needed in this area. Both Cochrane reviews found that the intensity of exercise (low, moderate, high) did not affect results. This could mean that individuals living with SMA Type II could still reap benefits from engaging in exercise and reduce the effects of their depression or anxiety even if they are only able to engage in low-intensity levels of exercise due to the effects of degeneration of their condition on their motor function, respiratory function, etc. Research is needed to investigate the effect of exercise on the mental health of individuals living with SMA Type II and NMD.

One participant had tachycardia and perceived that it was a barrier to their exercise engagement. Cardiac events, including arrhythmia (which includes tachycardia and bradycardia) and cardiomyopathy, have been found in individuals living with SMA. This may suggest that being deficient in the survival motor neuron (SMN) protein can affect cardiac muscle or that cardiac conditions can occur secondary to respiratory problems. Individuals who have cardiac conditions are not precluded from exercise, except in a few conditions such as severe heart failure. It is advised that individuals seek clearance and advice from their doctor prior to exercise engagement, and they may benefit from engaging the services of a physiotherapist or exercise physiologist. Participants in this research were most likely to engage in swimming. Swimming is generally considered to be a safe exercise for individuals living with low to moderate cardiac conditions, but not for those with severe conditions (severe heart failure) or recent myocardial infarction.
has found that for individuals living with a cardiac condition, perceptions of the effectiveness of exercise to rehabilitate them influences their exercise behaviour. Research is needed into the effect of exercise for individuals living with an NMD, including SMA Type II, who also have a cardiac condition.

6.3 Exercise behaviour

Participants’ exercise behaviour was explored, including their definition of exercise, exercise type, and exercise frequency.

6.3.1 Defining exercise

Participants defined exercise differently, with some defining it as planned, structured, repetitive physical activity performed at moderate- or high-intensity levels, whilst others defined exercise as performing activities of daily living (ADLs) to improve or maintain their physical function. Physical activity is typically defined as contractions of skeletal muscle to produce any body movement that expends more energy than when at rest. Exercise is a subcategory of physical activity that is defined as planned, structured, repetitive physical activity that is performed to improve or maintain components of physical fitness at low-, moderate- or high-intensity. Attributes and characteristics of physical fitness include cardiorespiratory endurance, body composition, muscular strength, muscular endurance, flexibility, agility, coordination, balance, power, reaction time and speed. ADLs, such as household tasks, are generally considered to be physical activity, not exercise, because they are performed in an efficient manner to conserve energy. However, if they are planned and structured with the goal of improving physical fitness or increasing energy expenditure, ADLs can be considered as exercise.

Two factors that contributed to participants’ definitions of exercise were degeneration of their condition and perceived benefits to their physical fitness levels. As an individual’s condition degenerated, they perceived that their ability to engage in planned, structured, repetitive physical activity at higher-intensity levels was increasingly limited. Some then redefined or shifted the focus of their definition of exercise to performing physical activities to improve their physical fitness. Participants observed improvements in their physical fitness by engaging in physical
activities including knitting; playing videogames; playing musical instruments, including trumpets, drums and pianos; singing; use of cough machines; and performing ADLs, including hanging laundry to dry and washing dishes. Individuals who included physical activities and ADLs within their definition of exercise continued to engage in exercise, whereas exercise ceased for those who did not.

Two participants (aged 0 - 4 years) stated that they had never engaged in exercise before; however, both had engaged in physiotherapy that included physical activity with the aim to improve physical function. This may indicate that these participants’ definitions of exercise were more focused on the structure, repetitiveness and/or intensity level of physical fitness, rather than on improving or maintaining physical fitness. It should be noted that the Consensus Statement states, ‘Even young children should be offered independent mobility and activities of daily living, which includes play’ (p. 1045). Several participants discussed disguising exercise as fun activities or play to facilitate their child’s exercise behaviour with the aim of improving or maintaining their physical fitness.

All participants engaged in exercise, and whilst their definition of exercise was varied, it still remained within the clinical definition of exercise. Literature has found that the exercise behaviour of individuals living with an NMD could be greatly enhanced through their doctors prescription of exercise and referral to exercise professionals. This suggests it may be beneficial for individuals who live with SMA Type II and practitioners to be educated on the definition of exercise. Promoting the definition of exercise as physical activity to improve or maintain physical fitness may facilitate exercise engagement within this population, particularly in those whose condition has degenerated. The Theory of Planned Behaviour could be a useful tool for practitioners to help their patients redefine their definition of exercise and promote their exercise behaviour. By increasing their knowledge about the definition of exercise individuals’ attitude and perceived behavioural control towards exercise behaviour could change from being perceived as unachievable. Furthermore, practitioners can often have direct access to individuals’ socio-cultural determinants and are in a unique position to educate those socio-cultural determinants, thus influencing individuals’ subjective norm.
6.3.2 Exercise type

Participants engaged in a wide variety of exercises and sports. In addition to swimming, individuals participated in a range of sports including soccer, basketball, netball, cricket, dance, hockey, softball, volleyball and boccia. Individuals living with SMA Type II devised or were prescribed exercises that may be considered unconventional. These included fine motor skills for hand strength and dexterity, such as picking up marbles, knitting, playing video games using a hand held controller and playing musical instruments. They also described participating in unconventional exercises for cardiorespiratory function, such as using cough machines, singing and playing musical instruments. Engaging in these physical activities, sports and exercises has been shown to provide physiological, psychological and social benefits to all individuals, including those who live with a NMD or physical disability. Exploring participants’ exercise behaviour may improve the knowledge of individuals living with SMA Type II and their practitioners and provide new pathways for others to engage in exercise.

6.3.2.1 Swimming, hydrotherapy and aquatic therapy

The most common exercise engaged in was swimming. However, as participants aged, their engagement in swimming decreased. Environmental, socio-cultural and individual determinants contributed to the attitude, subjective norm and perceived behavioural control of individuals living with SMA Type II resulting in a lack of engagement in swimming. Environmental determinants contributed to individuals’ perceived behavioural control and included lack of access to suitable facilities (including bathrooms and adult-sized change tables) and equipment to access those facilities (e.g. pool hoists). Socio-cultural determinants contributed to individuals’ subjective norm and included lack of access to paid or unpaid support persons capable of taking the individual swimming. Individual determinants included psychological, cognitive and biological determinants that contributed to their beliefs, attitude, perceived behavioural control and subsequent exercise behaviour.

Individuals described feeling vulnerable in the pool and lacked knowledge on how to adapt swimming to accommodate changes in biological function. Biological changes included loss of motor function ability, loss of balance and contractures, which can change an individual’s body to be in a permanently seated position. Swimming was
specifically recommended by the Consensus Statement for individuals living with SMA Type II.4

All of the adults and one-third of the school-aged participants had ceased participating in swimming. One participant stated that they no longer engaged in swimming because they were unable to perform traditional swimming techniques. However, in contrast to this perception, individuals living with SMA Type II may still receive benefits from swimming, regardless of the technique used. As an individual becomes immersed in water, hydrostatic pressure (pressure exerted from the water) puts a force on the body that generates physiological changes, including the movement of body fluids (blood and lymph) up towards the chest and heart, decreasing oedema (fluid retention causing swelling), raising blood pressure, compressing the chest wall and displacing the diaphragm.182 Hydrostatic pressure increases the amount of work required to breathe and, through increased usage, respiratory muscular strength and endurance and vital capacity (VC; a component of respiratory function) may improve.182 The increased blood flow caused by hydrostatic pressure is likely to be redistributed to the skin and muscles, increasing oxygen supply to those areas.182 Increased oxygen supply to muscles can improve exercise capacity, muscular function and musculature repair, as well as reduce muscle fatigue.185 Additionally, the buoyancy of water allows for more freedom of movement without pain, enhancing individuals’ ability to perform exercises that they may not have been able to achieve on land.182 Research into the effects of hydrotherapy, aquatic therapy and swimming in individuals living with a physical disability often used the Halliwick method or adapted swimming styles.130,202 The focus of the Halliwick method is to develop skills for individuals living with physical disabilities, including balance, independence and swimming, and can be taught as a ten point program (Appendix 9).224,232 The Royal Children’s Hospital Melbourne produced a pamphlet that outlines a hydrotherapy program for children living with an NMD and includes some of the benefits and risks of hydrotherapy (Appendix 10).198

The benefits of swimming, hydrotherapy and aquatic therapy include freedom of movement, improved self-esteem, improved communication skills, enjoyment, social interaction and improved fitness for individuals living with an NMD or physical disability.198,202,224 Swimming, hydrotherapy and aquatic therapy offer numerous health and fitness benefits for individuals living with a physical disability, including improvements to VC, range of movement, gross and fine motor skills, gross
motor function and bi-manual control (the ability to perform multiple movements at the same time).\textsuperscript{202} An aquatic respiratory based exercise program in women living with fibromyalgia found that pain, dyspnöea (shortness of breath), anxiety, quality of sleep, functional capacity and quality of life improved.\textsuperscript{140} A physiotherapy and hydrotherapy intervention was shown to improve gross motor function and may have provided psychological benefits for individuals living with SMA Type II.\textsuperscript{29}

Swimming, hydrotherapy and aquatic therapy could also pose some risks, including tiredness, illness, change in ability to float with degeneration and the need to modify the program as participation in hydrotherapy becomes less safe with degeneration in condition.\textsuperscript{198} Developing an individual’s skills using the ten point program and using floatation devices could improve the safety of individuals whilst they are in an aquatic environment.\textsuperscript{224} No adverse events were recorded in the literature found on aquatic interventions for individuals living with a physical disability.\textsuperscript{202} However, swimming, hydrotherapy and aquatic therapy could potentially pose some health risks of which individuals should be aware.\textsuperscript{233} While having numerous positive physiological impacts, hydrostatic pressure can also negatively stress the cardiorespiratory system through increased pressure, which causes pulmonary and vascular stress.\textsuperscript{182,233} Too much stress can cause stress-induced mechanical failure of the blood-gas barrier in the lungs, which can result in conditions like swimming-induced pulmonary oedema, a microvascular injury in the lungs causing cough with sputum, dyspnöea, and chest pain.\textsuperscript{233} Swimming-induced pulmonary oedema has an unknown and underreported prevalence in the general population, but has been reported following strenuous swimming.\textsuperscript{233} Whilst swimming was the most common exercise participants engaged in, some were also prescribed respiratory exercises.

\textbf{6.3.2.2 Respiratory exercises}

Respiratory exercises were one of the more common exercises practitioners prescribed to participants in this study, which would have a positive effect on their subjective norm and exercise behaviour. This is interesting, as it occurred despite a lack of guidance for this exercise in the Consensus Statement. Cardiorespiratory fitness is highly valued by participants and thus positively influences their beliefs and attitude towards this exercise behaviour. RMT has been shown to improve maximal inspiration, expiratory mouth pressure and respiratory load perception in DMD and
SMA Type III participants. A systematic review of RMT showed improvements in inspiratory and expiratory muscle strength, inspiratory muscle endurance, cough function, perception of dyspnoea, and pulmonary volumes and capacities in individuals living with Parkinson’s Disease (PD) and Multiple Sclerosis (MS). A two year intervention of inspiratory muscle training (IMT) in DMD and SMA found that respiratory muscle and endurance improved and VC remained stable over the two year period. IMT was shown to improve respiratory muscle strength and endurance in participants living with an NMD who have a VC higher than 25% of the predicted value. Additionally, respiratory-based exercise programs, including an aquatic respiratory-based exercise program, led to improvements in dyspnoea, pain, anxiety, quality of sleep, functional capacity and quality of life for individuals living with Fibromyalgia. Further research is needed to investigate the effect of RMT and respiratory-based exercise programs for individuals living with SMA Type II.

6.3.2.3 Sport

The majority of participants took part in a variety of sports and adapted sports, with soccer being the most prominent. Parents and community sport groups generally facilitated sport engagement for individuals living with SMA Type II thus positively influencing their subjective norm and exercise behaviour. Participants perceived that schools and school teachers could facilitate or hinder sport engagement, whilst practitioners did not advise or prescribe engagement in sport. Participants had both positive and negative perceptions of sport in regards to social and psychological aspects of life. Some participants raised concerns that sport did not offer any physical benefit. They perceived that adapted sports were designed for individuals living with a disability who possess greater upper body strength. These beliefs created a negative attitude towards sport and thus they did not engage in that exercise behaviour. The Consensus Statement recommends that individuals living with SMA Type II should engage in adaptive sports.

Participation in adaptive sports has been shown to provide physiological, psychological and social benefits for individuals living with a physical disability. Adolescents and young adults who live with a physical disability that participated in adapted sport perceived that they possessed more strength, endurance, flexibility, coordination, sporting ability, attractiveness,
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positivity towards physical self (global physical) and overall positivity towards self (global esteem) than those who did not participate in sport. The results were comparable between individuals who participated in sport, regardless of whether or not they had a physical disability. In addition to strength, endurance and self-esteem, participation in adapted sports was able to improve quality of life, social integration and life satisfaction for adults living with a physical disability. As well as the promotion of sporting ability through skill development, participation in adapted sport enhanced individuals’ self-determination, including their perceptions of their own competence and autonomy. Participation in adapted sport offered the opportunity to form friendships, develop self-identity, independence, coping skills, competitiveness and teamwork for children living with a physical disability.

In contrast to the perceptions of participants in this research, participation in powered wheelchair sports significantly increased heart rate for individuals living with MD, with the majority of individuals achieving a moderate-intensity level of exercise by exceeding 55% of estimated HR for 30 minutes. Additionally, race car drivers have been shown to have a physiological response whilst driving, including increased heart rate and oxygen consumption (VO2max), that reached moderate to high exercise intensity. Therefore, theoretically, athletic training in a powered wheelchair, such as sprints, cross-country and obstacle courses, and powered wheelchair sports may also produce a physiological response in individuals living with SMA Type II that could improve their physical fitness. Increased awareness of Barfield and colleagues (2005) research could increase individuals’ knowledge about the benefits of powered wheelchair sports, positively influencing their attitude and engagement towards this exercise behaviour. Furthermore, participation in adaptive sport can improve employment prospects for individuals living with physical disability, which subsequently has the potential to improve their socio-economic status. Whilst research has found numerous benefits to participation in sports for individuals living with a physical disability, the vast majority of those individuals were living with a spinal cord injury. Further research is needed into the effect of sport on physiological, psychological and social functions for individuals living with an NMD.
6.3.2.4 Horseback riding and hippotherapy

Horseback riding and/or hippotherapy was engaged in by some participants. The Consensus Statement recommends horseback riding for individuals living with SMA who can walk. In contrast to this recommendation, none of the participants in this research who engaged in hippotherapy were able to walk. Indeed the independent motor function ability achieved by those participants was sitting, crawling, and stand and crawl. This indicates that individuals living with SMA Type II who are not able to walk can still participate in, and receive benefits from, engaging in hippotherapy or horseback riding. Hippotherapy guidelines recommend that adults and children have the ability to sit independently on a horse to participate; however, children have the option of having the therapist sit behind them to provide postural support, if needed, as the horse would be able to carry their combined weight. Contraindications for hippotherapy include scoliosis greater than 30°, recent fractures, recent surgery and severe osteoporosis.

Participants in this research perceived that hippotherapy was enjoyable and improved core, trunk and neck strength. These beliefs lead to their positive attitude and subsequent engagement in hippotherapy. Examination of the literature revealed that hippotherapy can offer psychological, physiological and social benefits for individuals living with an NMD. Lemke and colleagues (2014) explored the effectiveness of hippotherapy from the perspective of individuals living with SMA Type II and their parents. Perceived psychological benefits included improved mood, confidence, self-esteem, pride, independence, enjoyment and achievement. The perceived social benefits were a platform to connect with friends and relationships built with the horse and trainers. Reported perceived physiological benefits included increased flexibility, muscle laxity, balance, postural control, muscle function, muscle strength (including core strength, agreeing with participants of this research), respiratory function and voice control. Barriers included initial apprehension, muscle soreness, the growth and weight of individuals leading to issues getting on and off the horse, safety concerns regarding falls and injury, surgery and spinal rods, degeneration, lack of knowledge and lack of practitioner knowledge of hippotherapy, financial cost and emotional distress upon cessation of hippotherapy. Examination of the literature further found that hippotherapy improved trunk muscle symmetry, gross motor function and functional performance in the home and community, but did not affect muscle tone and excessively increased heart rate in
children living with cerebral palsy. Hippotherapy has also been found to improve balance in individuals living with MS. Participants in this research agreed with, and added to the literature that found individuals living with SMA perceived hippotherapy to have psychological and physiological benefits. Further research is needed to investigate if the perceptions of individuals living with SMA regarding hippotherapy are congruent with actual psychological and physiological changes.

6.3.2.5 Arm and leg cycling

Participants discussed the physical benefits of manual wheelchair pushing and bike riding to maintain strength and functional ability, with these beliefs positively influencing their attitude, perceived behavioural control and exercise behaviour. Strength training for individuals living with SMA Type II may improve muscular strength and motor function ability. Furthermore, consistent with participants’ perceptions, individuals living with DMD maintained their motor function abilities whilst performing cycling with their arms and legs, whereas those abilities declined when they ceased cycling. The majority of interview participants also perceived that cessation of exercise caused loss of function or advancements in secondary conditions. The literature supports those perceptions, finding that cessation of exercise caused a decline in physical function for individuals living with an NMD. However, further research is needed.

6.3.2.6 Knitting

Exploring unconventional exercises may offer individuals living with SMA Type II new pathways to continue exercising. One participant in this research knitted to maintain and improve their hand strength and dexterity. There is limited research into the effects of knitting, and no articles were found that outlined the physiological changes that knitting could produce. However, knitting has been found to provide cognitive, psychological and social benefits for individuals. Cognitive benefits included learning a new skill and encouraging thinking, problem-solving and concentration. Psychological benefits included promoting feelings of confidence, happiness and calmness and reducing feelings of anxiety and stress. Social benefits included engaging with knitting groups, providing a connection with people, and enabling people with social anxiety or shyness to engage with others. The participant in this research did not comment on the cognitive, psychological or social benefits of knitting they received, instead focusing on their perceptions of the physical
benefits of knitting, specifically hand strength and dexterity. Knitting to improve the physiological function of individuals who live with a condition that limit their hand strength and dexterity may be an area for future research.

6.3.2.7 Playing video games

One participant discussed their improvements to hand strength from playing video games with a hand-held controller. The literature supports this claim, finding that playing virtual reality games may improve hand function, including grip strength and speed of movement, for individuals living with a physical disability.\textsuperscript{123,142,218,223,229} Further research is needed to examine the effects of playing video games on hand function in individuals living with an NMD, including SMA Type II.

6.3.2.8 Playing musical instruments

Participants perceived that playing musical instruments had physiological and social benefits, including improved respiratory function, muscular strength and a platform to meet and share experiences with others. Their beliefs about the benefits of playing musical instruments thus influenced their attitude towards this exercise behaviour. Review of the literature found that playing different musical instruments produced different physiological responses.\textsuperscript{195} Playing wind instruments was suggested as an activity to improve lung function in the Consensus Statement for Standard of Care for Congenital Muscular Dystrophies.\textsuperscript{213} The literature also found that professional wind instrument players had improved lung function (specifically FEV1 and FEF50),\textsuperscript{197} which is in agreement with the perceptions of the participants in this research. Additionally, didgeridoo playing was found to improve sleep apnoea including daytime sleepiness and the number of apnoea or hypopnoea events during sleep.\textsuperscript{228} In contrast to participants perceptions and the Consensus Statement for Standard of Care for Congenital Muscular Dystrophies, playing wind instruments may increase the risk of respiratory-associated diseases, such as asthma, and cause swelling or irritation of the mucus membrane in the upper respiratory tract.\textsuperscript{197} However, it was reported that in teenage asthmatics, there was no significant difference in asthmatic symptoms and bronchoconstriction between those who played wind instruments and those who did not.\textsuperscript{234} Moreover, teenage asthmatics who played wind instruments perceived their general health, irritability and attitude to be
improved compared to those who did not play a musical instrument. As reduced respiratory function and sleep apnoea are common conditions associated with SMA Type II, playing wind instruments may be an area for practitioners and individuals living with SMA Type II to investigate.

Participants who played musical instruments perceived that the activity was a form of cardiorespiratory exercise that improved their cardiorespiratory fitness and lung function. It has been found that heart rate can reach that of moderate- to hard-intensity exercise in professional musicians playing piano, percussion, wind or string instruments, with performances having a greater impact on heart rate than rehearsals. Music therapy can also be used as a tool for pain management and can stimulate the cardiovascular system, promote participation in exercise and help with movement fluidity and gait training. Further research is needed to investigate the impact playing musical instruments has on heart rate for individuals living with an NMD and the exercise intensity that can be reached.

One participant perceived that playing a musical instrument improved the strength of the muscles used to play that instrument and the dexterity of their fingers and hands. Furthermore, the participant perceived that the muscles used to play the musical instrument were the strongest muscles they had after significant degeneration. Playing the piano has been shown to improve muscular strength of the shoulders, elbows, wrists and fingers, muscular endurance, flexibility of muscles and motor control of fingers for all individuals, including individuals who have had a stroke and children and adolescents who live with cerebral palsy. Playing electronic paddle drums also improved shoulder and elbow range of motion for individuals who had a stroke. Furthermore, the acquisition of a new skill had the potential to build self-esteem, while the instant feedback from playing the piano provided enjoyment and motivation for individuals who had a stroke. However, drummers and piano players are at risk of developing repetitive muscle injuries, which may be of concern to individuals living with SMA Type II.

Movements for expert piano players were economised compared to novice players. Muscle synchronisation allows for numerous muscles to contract at the same time. Muscle synchronisation was significantly lower in musicians (who play the piano, flute or violin) than in weight lifters, and wrist muscle synchronisation was lower in drummers than non-drummers. This indicates that different forms of physical activity influence neural patterns to achieve motor
function ability in different ways.\textsuperscript{192} Strength training promoted muscle synchronisation, which rapidly produced force to achieve motor function ability, whilst playing musical instruments may promote the use of singular muscles to produce force to complete motor function ability.\textsuperscript{192} This indicates that the repetitive exercise from playing musical instruments could strengthen individual muscles.\textsuperscript{189,192,193} If so, this supports the perceptions of the participant in this research who found that the strongest muscles they had, with continued degeneration, were those they used playing a musical instrument. The effect playing musical instruments have on individuals living with an NMD, including respiratory function, muscular strength, endurance and flexibility, is an area to consider for future research.

\textbf{6.3.2.9 Singing}

Participants who sang felt that the activity improved their respiratory function, while being enjoyable. Singing was suggested as an activity to improve lung function in the Consensus Statement for Standard of Care for Congenital Muscular Dystrophies.\textsuperscript{213} Respiratory muscles are engaged during diaphragmatic breathing (a learned technique) whilst singing, which can improve the strength of those muscles.\textsuperscript{194} Diaphragmatic breathing may also improve an individual’s cough mechanism through strengthened respiratory muscles.\textsuperscript{236} Increased lung volume is needed to sing loudly or at a higher tone compared to talking, and can be achieved through diaphragmatic breathing.\textsuperscript{194,236} Singing for cardiorespiratory function has been investigated in individuals living with chronic obstructive pulmonary disease (COPD), emphysema, asthma, cystic fibrosis (CF), MS and PD.\textsuperscript{220-222,226,231,236,237}

Singing, using the diaphragmatic breathing technique, has improved maximal expiratory pressure (strength of respiratory muscles) for individuals living with COPD, CF, MS and PD.\textsuperscript{217,220,222,231} Singing improved breath control (extending expiration and breathing efficiently) and breath support (intensity of speech) and was associated with a change from clavicular to diaphragmatic breathing in individuals living with emphysema.\textsuperscript{221} Peak expiratory flow rate, which indicates airway narrowing and obstruction, was improved with singing in individuals living with asthma.\textsuperscript{238,239} Additionally, singing was found to improve anxiety and was perceived by individuals living with COPD to have physical benefits, improving their participation and compliance in this activity.\textsuperscript{226} Whilst the research to date has shown health and respiratory benefits from singing further research is needed to solidify
these findings and to determine the effects of singing for individuals living with an NMD, including SMA Type II.

6.3.2.10 Cough assist machine

Some participants considered the use of a cough assist machine (mechanical insufflation-exsufflation) as exercise that could improve lung function. The Consensus Statement recommends the use of manual and mechanical-assisted cough for individuals living with SMA who have an ineffective cough.\(^4\) Respiratory muscle strength, muscle endurance and lung function significantly improved for children living with an NMD after using the cough assist machine.\(^{208}\) Fauroux and colleagues (2008) found that sniff nasal inspiratory pressure significantly improved after cough assist machine usage; however, they attributed the findings to improved muscle ballistic ability rather than improved respiratory muscle strength. Cough assist machines have been found to significantly improve peak cough flow (PCF) in individuals living with an NMD.\(^{200,201,208,215}\) Cough assist machines are more effective at increasing PCF than manual and non-invasive ventilator-assisted cough\(^{200}\) and significantly decrease visits to the emergency department for individuals living with an NMD.\(^{205}\)

Participants’ usage of the cough assist machine differed from multiple times per day though to an as-needed basis, which is consistent with findings from other studies.\(^{207,211}\) Individuals living with an NMD who regularly used cough assist machines perceived that it improved overall respiratory health, including reduced respiratory infections or complications\(^{205,207}\) and improved respiratory function.\(^{201,205,211,215}\) Participants found that lack of carer training and perceptions of ineffectiveness hindered the use of cough assist machines; a perception mirrored in other literature.\(^{207,211}\) Whilst cough assist machines are considered to be safe to use, a small number of individuals living with an NMD have reported negative effects – with the most common being unpleasantness.\(^{207,211}\) The literature support participants’ perceptions that cough assist machines can improve lung function. However, more research is needed regarding the effect of the cough assist machine on lung function, respiratory muscle strength and endurance, and as an exercise for individuals living with an NMD.
6.3.2.11 Qigong and the use of exoskeletons

There are other exercises that could offer physical and psychological benefits to individuals living with SMA Type II that were not mentioned by participants in this research, such as qigong. Individuals living with MD subjectively assessed qigong and found that it could provide physical and psychological improvements; however quantitative tests did not always support these perceptions. Perceived benefits included improved mobility, respiratory function, digestion, circulation, concentration, energy levels, temporary reductions in stress and improvements in relaxation. Additionally, exercising with the use of exoskeletons may also improve independence, social aspects of life and possibly provide physiological benefits for individuals living with a physical disability. No participants in this research discussed using an exoskeleton, but that may prove to be a concept of interest to individuals living with SMA Type II to improve their independence and physical function. Exploring unconventional exercise therapies is one strategy that could engage and maintain exercise participation for individuals living with SMA Type II, particularly with continued degeneration. Additional research is needed to examine the effects of qigong and exoskeletons on physiological and psychological health for individuals living with an NMD, including SMA Type II.

6.3.3 Exercise frequency

Exercise frequency varied greatly between participants in this research, with extremes for many determinants reported. Some participants were currently exercising for more than 150 minutes/week while others had previously met this target. This indicates that some individuals living with SMA Type II are meeting or exceeding the Muscular Dystrophy UK recommendations for individuals living with an NMD regarding aerobic exercise. The majority of individuals living with SMA Type II are not meeting the Muscular Dystrophy UK recommendations for strength training. Only two participants had engaged in weight training, while five participants listed weight training as an exercise that they would not feel comfortable engaging in. The findings by Lewelt and colleagues (2015) that strength training in children living with SMA Type II was safe and could improve strength and motor function needs to be communicated. Advice, prescription and education about the benefits of strength training for individuals living with an NMD may improve perceptions and participation in this exercise.
Only 43% of Australian adults meet the recommendation to exercise on most days for 30 minutes at moderate-intensity. Furthermore, 70% of Australian adults are considered to be either sedentary or have low levels of physical activity. Australian adults living with SMA Type II also have low exercise engagement, with just 12% of adult participants exercising for more than 150 minutes 5 days per week at the time of the research, 25% exercising for 30 - 90 minutes 1 - 3 days per week and 63% exercising for less than 30 minutes per week or not at all. Broad statements in the current literature that individuals living with an NMD live a less active lifestyle than the general population may be portraying the exercise behavior of these individuals in an unfair, unfavourable light.

Degeneration and lack of exercise knowledge, and particularly knowledge about exercising in a degenerated condition, greatly impacted participants’ exercise behaviour. Participants who did not currently exercise had a current motor function ability of sit (30%), independent breathing (80%), and no movement (100%), indicating that degeneration of an individual’s condition impacts their exercise engagement. Interview participants discussed their frustration and lack of knowledge about exercise as their condition deteriorated. The literature cited lack of knowledge as a barrier to exercise for individuals living with an NMD. Strategies targeting individuals’ subjective norm such as improving practitioner knowledge, practitioners educating and prescribing exercise to their patients, improving school teachers knowledge and advertising exercise recommendations for individuals living with an NMD may reduce these barriers and further increase the exercise engagement of individuals living with SMA Type II.

6.4 The effectiveness of exercise to treat the symptoms and slow the progression of SMA Type II

Participants perceived that exercise could be effective in treating the symptoms of SMA Type II; however, progression of the condition would eventuate and be an overriding factor of any benefits gained. The most compelling evidence for some participants regarding the effectiveness of exercise was the decrease in physical fitness and function after periods of immobility, such as surgery, casting for fractures and increased usage of wheelchairs. Immobilisation has been shown to cause muscle atrophy and loss of muscular strength and endurance, initially affecting lower limbs.
and postural muscles.\textsuperscript{244,245} Additionally, immobilisation shortens muscles, which result in contractures, reduces metabolic activity of muscles and circulation of blood leading to muscle fatigue and increased risk of developing deep vein thrombosis.\textsuperscript{244,245} Bone mineral density decreases with immobilisation, which can lead to an increased risk of fractures.\textsuperscript{245} Furthermore, immobilisation can compromise skin integrity, resulting in pressure sores, and can lead to infection and death.\textsuperscript{244,246} There are considerable similarities between immobilisation and the conditions associated with SMA Type II.

Individuals perceptions that immobility enhanced the degeneration of their condition impacted upon their perceived behavioural control and attitude towards exercise. For some individuals this led to their re-engagement in exercise to counteract the effects that immobility was having on their condition. However for others it lead to a downward spiral – with increased degeneration preventing them from engaging in exercise which further impacted upon their perceived behavioural control and attitude, ultimately leading to the discontinuation of their exercise behaviour. The literature also found that periods of immobility lead to increased degeneration for individuals living with an NMD.\textsuperscript{21} This suggests that exercise and physical activity could have a positive impact on individuals living with SMA Type II if it were a ‘major focus of treatment’, as recommended by the Consensus Statement.

As previously discussed, many exercises were perceived to be effective in improving components of physical fitness. A small number of participants had engaged in playing video games and musical instruments and perceived them to be effective in improving hand strength, whilst singing and playing musical instruments were perceived to be effective at improving cardiorespiratory function. However, swimming was perceived by the majority of participants to be effective at improving multiple components of physical fitness, including cardiorespiratory function, motor function ability, orthopaedic health and muscular strength. Indeed, the literature revealed that swimming could improve respiratory function, motor function ability and psychological function for individuals living with a physical disability, but there are potential risks (as previously discussed).\textsuperscript{29,140,202,224,233} Aquatic exercise may also positively impact orthopaedic health for individuals living with osteoporosis.\textsuperscript{182} The beliefs and attitude that swimming was effective at improving multiple components of fitness lead individuals living with SMA Type II to engage in this exercise behaviour. Furthermore, the perceptions about the effectiveness of swimming may have been
influenced by increased exposure to this type of exercise. Swimming was the exercise most prescribed by practitioners and is explicitly stated as a suitable exercise in the Consensus Statement. This suggests that subjective norm influenced the exercise behaviour of individuals living with SMA Type II to engage in swimming. If individuals living with SMA Type II are exposed to other types of exercise to the degree that they are exposed to swimming, this may change their perceptions of the benefits and risks those other exercises hold.

Limited research explored the effects of exercise for individuals living with an NMD\textsuperscript{94-104} and even less has been conducted into the effects of exercise for individuals living with SMA Type II.\textsuperscript{28-30,113} Physiotherapy, hydrotherapy, strength training and flexibility exercises may improve motor function ability and swallowing function for individuals living with SMA Type II, but further research is needed to solidify these results.\textsuperscript{28-30} Further research is also needed into the effects different types of exercises have on individuals living with SMA Type II. Further research will improve the knowledge base in these areas, allowing for improvements in both practitioners’ ability to provide adequate services and individuals’ ability to make informed decisions about their exercise behaviour.

6.5 The value of exercise to treat the symptoms of SMA Type II

The majority of participants perceived that exercise was worthwhile to pursue. This demonstrates that participants’ attitudes towards exercise were in alignment with the Australian government, the Consensus Statement and the recommendations of NMD organisations to exercise.\textsuperscript{4,12,14,15} The majority of participants perceived that exercise improved their mood, quality of life and coping skills. This is congruent with the current literature that also suggests that exercise improves mood and quality of life for individuals living with an NMD.\textsuperscript{84,143} The majority of participants perceived that exercise created opportunities and provided interactions that improved the social aspects of their lives. However, interview participants also highlighted that engaging in exercise could have negative aspects, such as social exclusion, bullying, increased self-consciousness due to body differences and dependency on others to complete activities. Engaging in exercise has been shown to improve social aspects of life for individuals living with an NMD.\textsuperscript{20,84,116} However, as reported by some participants in this research, individuals living with an NMD can experience social isolation, sadness
and discrimination if they are excluded from participating in activities that they would like to be a part of due to degeneration caused by their condition.\textsuperscript{18,20,83,119}

The majority of participants also perceived that exercise improved movement, reduced body fat, improved respiratory function and slowed deterioration. Additionally, more participants agreed than disagreed that exercise improved muscle strength and life expectancy. In agreement with participants’ perceptions, improvements in gross motor function, muscular strength and flexibility have been found in individuals living with SMA Type II after engaging in exercise; however, more research is needed to solidify these results.\textsuperscript{28-30} Furthermore, additional research is needed to investigate other health benefits of exercise, such as effects on respiratory function, body composition, degeneration and life expectancy, for this population. The literature is limited, but has shown that exercise can maintain or improve muscular strength, cardiorespiratory function and motor function ability, reduce fatigue and adiposity, reverse deconditioning, and slow disease progression for individuals living with an NMD.\textsuperscript{11,15,21} More research is needed to confirm these findings.

The majority of participants perceived that engaging in exercise increased their tiredness that day and placed them at higher risk of sustaining more injuries than the general population. Tiredness and fear of injury have often been reported as barriers to exercise for individuals living with an NMD.\textsuperscript{17,18,21,22,85,112,118,119} Individuals living with an NMD are advised about the difference between fatigue and muscle weakness caused from over exercising in the Muscular Dystrophy UK recommendations.\textsuperscript{12} However, in contrast to the perceptions of participants in this research, the literature suggests that engaging in exercise can reduce fatigue.\textsuperscript{11,15,21} The notion that exercise, though tiring, may help to combat fatigue is a topic that should be addressed with individuals living with SMA Type II as it may influence their beliefs, attitude and ultimately improve their exercise behaviour.

Exercise for cardiorespiratory function and fine motor skills was identified as being most important to individuals living with SMA Type II, particularly as their condition degenerated, to maintain quality of life, optimal health, longevity and independence. Participants perceived that swimming, singing, playing musical instruments, cough assist machines and pushing manual wheelchairs improved their cardiorespiratory function. Respiratory conditions have a major impact on the morbidity and mortality of individuals living with SMA.\textsuperscript{24,38,54} However, respiratory
exercises were not specifically mentioned by, nor are they indicated as a future research direction, in the Consensus Statement.\textsuperscript{4} The British Thoracic Society guidelines for children living with neuromuscular weakness found that respiratory muscle training (RMT) improved respiratory muscle strength and endurance for individuals living with Duchenne muscular dystrophy (DMD).\textsuperscript{247} Revision of the Consensus Statement may be warranted to explore the potential benefits of this exercise to improve respiratory function and provide guidelines for practitioners.

For individuals living with SMA Type II, their hands are one of the last parts of their body to lose function. Some participants perceived that playing video games, playing musical instruments, knitting or picking up marbles could improve or maintain their hand strength and dexterity. Research has found that improvements to hand functionality may be gained from playing video games\textsuperscript{123,142,218,223,229} and musical instruments.\textsuperscript{186,225,227} Participants perceived that maintaining hand strength, dexterity and function was important to retain their independence. Individuals living with SMA Type II may benefit from engaging with occupational therapists (OTs) or physiotherapists specialising in occupation-based hand therapy. Occupation-based hand therapy can improve range of movement, strength and function, and decrease pain for individuals who have a condition that limits their hand’s physical function.\textsuperscript{248} Therefore, engaging in occupation-based hand therapy may improve the exercise knowledge, attitude, perceived behavioural control, exercise behaviour and physical hand function of individuals living with SMA Type II.

The majority of questionnaire participants perceived that exercise was worthwhile to pursue, and half of interview participants perceived that it was achievable to meet the Australian Physical Activity and Sedentary Behaviour Guidelines. However, only one fifth were meeting those guidelines, meaning that the participants’ exercise beliefs did not mirror their actual exercise behaviour. Therefore, perceived behavioural control and subjective norms may have a higher impact on exercise behaviour than attitude. Strategies that target environmental, policy and socio-cultural influences may benefit individuals living with SMA Type II. These strategies may include improving accessibility to facilities, equipment and services, implementing policies that provide equal opportunities to participate in exercise, advertising exercise recommendations for individuals living with an NMD, improving the knowledge of practitioners, schools and families of individuals living with an
NMD about exercise and exercise recommendations and practitioners prescribing exercise to their patients.  

6.6 Exercise determinants for individuals living with SMA Type II

Participants described environmental, socio-cultural and individual determinants for exercise. The social ecological model for physical activity does not differentiate how influential each determinant is. However, participants indicated that the most influential exercise determinants for them were accessibility of facilities, services and equipment; funding; parental influence; friends; schools; motivation; knowledge; attitude; and degeneration their condition, specifically motor function ability, respiratory function and muscular strength. Individuals’ attitude, subjective norm, perceived behavioural control and subsequent exercise behaviour were influenced by these environmental, socio-cultural and individual determinants.

For example to engage in swimming individuals believed that swimming was beneficial to them in some way either socially, physiologically or psychologically thus creating a positive attitude towards exercise. Secondly individuals’ subjective norm encouraged their swimming behaviour. Swimming was promoted by environmental determinants such as the Consensus Statement and socio-cultural determinants including practitioners, schools, parents and peers for individuals living with SMA Type II. Finally there were many control variables that influenced individuals’ perceived behavioural control and their subsequent exercise behaviour. First environmental determinants impacted their perceived behavioural control and swimming behaviour as they either needed to have access to their own pool or a public pool. Access to their own pool included the monetary costs of installing a pool and the necessary equipment needed to enter the pool, which may have needed the involvement of community fundraising. Alternatively access to a public pool involved finding one that is accessible and has suitable facilities (pool hoist, change room accessible to electric wheelchairs), access to transport (their own specialised car or public transport), money to use the facilities, and the ability to organise the support of their carer during the public pools timetable to access the appropriate facilities. Secondly individuals’ relied on socio-cultural determinants including the support of paid or unpaid carers to engage in swimming, impacting upon their perceived behavioural control. Finally individual determinants such as individuals’ motor
function ability, and whether it was stable or deteriorating, impacted upon their perceived behavioural control and exercise behaviour. Thus environmental, socio-cultural and individual determinants significantly influence individuals exercise behaviour.

Half of interview participants perceived that it was achievable to meet the Australian Physical Activity and Sedentary Behaviour Guidelines; however, the vast majority felt that these guidelines were not applicable to them. Environmental determinants have been found to be more influential than individual determinants for children living with a disability. The World Health Organization (WHO) identified that governments, policies, environments, health professionals and educational facilities are instrumental in promoting exercise and reducing morbidity and mortality associated with physical inactivity. If the exercise engagement of individuals living with an NMD was promoted it could impact individuals’ subjective norm and positively influence their exercise behaviour. Government bodies were advised to create policies that promote physical activity through effective legislation that provides social inclusiveness, safe and accessible infrastructure, including transport and sport and recreation facilities, adequate funding and health education for the public, health professionals and service providers. This decreases barriers to exercise for individuals living with a disability and could improve their perceived behavioural control.

Lack of accessibility and funding were reported by participants to be barriers to their exercise engagement. Participants expressed that increased funding could alleviate economic constraints and facilitate their exercise behaviour. In agreement with the findings of this research, lack of accessibility is a well-documented barrier to engagement and participation in sport and exercise for individuals living with a disability and for individuals living with an NMD. Strategies targeting improvements to the accessibility of the environment, for the general population, increased the utilisation of the environment and may have increased the general population’s physical activity levels. As accessibility is a far greater barrier for individuals living with a NMD than for the general population, investment into creating accessible environments may have a far greater impact on the physical activity levels of individuals living with a NMD than has been seen in the general population. Also in agreement with participants’ perceptions, the literature revealed that a lack of funding and the cost of exercise engagement is a barrier to exercise for...
children and adults living with a physical disability. Socio-economic constraints, including reduced income, cost of exercise engagement and additional costs to exercise engagement, such as employing support workers, have also been found to hinder exercise behaviour of individuals living with an NMD. This suggests that further work is needed by the government to provide policies, accessible environments and adequate funding to reduce barriers to exercise. This would improve perceived behavioural control and promote physical activity behaviour for individuals living with a physical disability, including NMD and SMA.

Participants’ perceptions of socio-cultural determinants, including parental influence and friends, as exercise determinants is consistent with current literature on exercise determinants for individuals who live with a disability, as previously discussed. In agreement with participant perceptions in this research, family influence was found to be more influential than individual determinants for children living with a disability. Children living with a physical disability were more likely to be physically active if their parents engaged in physical activity. Social support was also found to be a determinant for exercise in individuals living with an NMD. Furthermore, social interaction was found to be one of the most influential facilitators of exercise engagement and participation in children and adults living with a disability. This highlights the importance of subjective norm in influencing exercise behaviour for individuals living with an NMD.

Participants’ interactions with practitioners about exercise was varied, with some receiving support, exercise advice and exercise prescription, while others were discouraged to exercise. Furthermore, some participants reported that they had not been advised to engage in physiotherapy. Lack of practitioner knowledge, advice and prescription was a barrier to exercise for some participants, which led to mixed results on exercise behaviour. The findings of this research are consistent with previous research that found individuals living with an NMD want to engage in exercise, however, they feel that exercise professionals lack knowledge about NMD. Literature has also reported that individuals living with an NMD may not receive support from practitioners, as practitioners may be sceptical about the benefits of exercise for this population. Lemke and colleagues (2014) participants also perceived that Australian practitioners lacked knowledge about exercise for individuals living with SMA. The Consensus Statement states that exercise is a major focus of treatment that should be encouraged. This result suggests a disparity.
between the Consensus Statement guidelines and physicians’ advice and care for individuals living with SMA Type II. Thus leading to a disparity in subjective norm for individuals living with SMA Type II. If practitioners are not placing due emphasis and value on exercise when advising individuals living with SMA Type II, then they could be contributing to their patient’s morbidity and lack of mobility, which in turn may impact upon their mortality.\textsuperscript{11,19,21,141}

Literature has found that lack of exercise advice from doctors can create barriers to the exercise engagement of individuals living with a NMD\textsuperscript{22} and physical disability.\textsuperscript{125,134} It has been suggested that written exercise advice from a physician may help individuals living with an NMD to engage or re-engage in exercise.\textsuperscript{22} Paediatricians have been advised to promote and advocate for children living with a physical disability to participate in sport and physical activity.\textsuperscript{134} Prescribed exercise from practitioners has been shown to improve exercise behaviour and physical fitness within the general public.\textsuperscript{132,133} Advising individuals living with SMA Type II to engage in exercise could positively influence individuals’ attitude and subjective norm towards exercise and improve their exercise behaviour. Further research is needed to investigate the effect that practitioners advising and prescribing exercise has on the exercise behaviour of individuals living with SMA Type II and an NMD.

Several questionnaire participants noted that they had engaged in school physical education (PE), which directed a line of questioning in the interviews. Participation in school PE was mixed for participants, as support from schools and school teachers was varied. Some participants were actively and successfully integrated into school PE, whilst others were hindered in their participation. Exclusion from school PE occurred due to inadequate knowledge of teachers, teacher support, school facilities, the individual’s lack of motivation and the individual’s desire to be perceived as ‘normal’ by their peers. The WHO identified that educational facilities are instrumental in promoting exercise.\textsuperscript{241,242} Schools have the potential to influence children and adolescents to engage in a physically active lifestyle throughout their lifetime not only by providing PE classes, but also by promoting children to live a physical activity lifestyle throughout the day.\textsuperscript{242,243} Schools also have the opportunity to improve the knowledge of children, adolescents and their parents regarding the benefits of physical activity and living a physically active lifestyle.\textsuperscript{242,243} Strategies employed by schools to promote physical activity have been shown to increase physical activity levels,\textsuperscript{242,243} improve physical fitness,\textsuperscript{242,243} reduce body mass
index and reduce time spent watching television for their students. This is in contrast to the lack of teacher knowledge and inclusion that the majority of interview participants experienced during school PE. Therefore, further training for schools and school teachers may be warranted to improve their knowledge of inclusive exercise practices and the benefits of exercise for children who live with a disability, including NMD. Subsequently, this may improve exercise knowledge, subjective norm and exercise behaviour for children living with a disability.

Participants experienced varying degrees of inclusiveness in school PE. The impact of educational facilities to promote physical activity through changes to school curriculum and printed educational materials may positively impact public health. In Australia, policy states that schools should provide students with a minimum of two hours of physical activity per week. However, a common-sense exemption policy allows the exclusion of children who are unable to participate in physical activity. This may impact upon the physical and psychological health, the social aspects of school life and subjective norm of children who live with a disability. In the USA, federal laws protect the right for all children to participate in PE and sport, and require schools to modify PE to include all students. However, 75% of schools exempt students from PE, with physical and cognitive disabilities cited as the most common reasons for exemption. Research is needed to investigate the impact a common-sense exemption policy has on the health and wellbeing of children living with a disability, including SMA Type II.

Schools indirectly influence sport and exercise participation for children living with a disability. Health in children is correlated with academic achievement, which is subsequently correlated with health in adults. Lack of teacher support and negative school experiences are barriers to exercise engagement for adolescents. Schools that promote physical activity levels in their curriculum may increase stigma and bullying for children who are not physically fit, increase their stress levels, reduce their self-esteem and mental health, create negative perceptions of exercise and lessen the enjoyment of, and motivation to, exercise. In agreement with this literature, some participants in this research perceived that engaging in school PE caused them to feel uncomfortable and promoted stigmatisation. However, in contrast, some participants experienced inclusion and enjoyment from engaging in school PE. Further research is needed to understand why there is a disparity between children’s experiences of school PE and exposure to stigmatisation. Subsequently, strategies can
be developed to promote inclusive school PE, positively influence children’s attitude towards exercise behaviour and reduce stigmatisation for children living with a disability, including SMA Type II.

Participants’ perceptions of their individual exercise determinants, including enjoyment, motivation, lack of exercise knowledge and conditions associated with SMA, were consistent with the literature. Whilst individual determinants are less influential in children living with a disability, they can facilitate or hinder exercise behaviour. Enjoyment has been found to facilitate exercise engagement in both children and adults living with a disability. Enjoyment and motivation, or lack thereof, was found to influence exercise behaviour in individuals living with an NMD. Lack of exercise knowledge is an established barrier to exercise for adults living with a physical disability as well as a barrier for individuals living with an NMD. Strategies that may promote exercise behaviour of individuals living with SMA Type II are to target their subjective norm by partnering with community organisations such as SMA Australia, MD Foundation and MD Australia; providing information about exercise through social media and NMD websites; and providing counselling from health professionals.

Research has found that both children and adults perceive their disability to be a barrier for exercise. Conditions associated with SMA were perceived by participants to be exercise determinants, and were reported as barriers and/or facilitators for exercise, thus influencing their attitude and perceived behavioural control. These included motor function ability, respiratory function, muscular strength, weight and orthopaedic health, with particular emphasis on scoliosis, fractures and contractures. Muscle weakness, loss of functional muscle tissue, muscle disuse, overuse injuries, pain, fatigue, reduced energy levels, cardiopulmonary problems, increased adipose tissue, mobility limitations and increased energy expenditure while engaging in motor functions have been cited as barriers to exercise for individuals living with an NMD. Health, lack of energy and fatigue have also been cited as barriers to exercise for adults living with a disability. However participants in this research described behavioural determinants, such as taking the flu shot, to ensure their general health, well being and ability to continue exercising. More research is needed to investigate exercise determinants for individuals living with an NMD and how these determinants
influence individuals attitude, subjective norm, perceived behavioural control and exercise behaviour.

6.7 Research strengths

The following strengths were identified in this research:

1. This research responds to a gap in knowledge about the perceptions of exercise for individuals living with an NMD, and specifically SMA Type II;
2. This research responds to a gap in knowledge about the exercise behaviour of individuals living with SMA Type II, specifically type, frequency, and cessation;
3. This research focused on SMA Type II which is currently an under researched NMD condition. A limitation of other research into NMDs is that multiple NMD conditions have been explored collectively and results for particular NMD conditions not able to be differentiated. Disregarding the specificity of the disease process can have implications in the transition from research to practice; and
4. This research used both quantitative and qualitative methods, which can reduce the limitations inherent in each method and provide both broad and deep understanding of the perceptions of exercise for individuals living with SMA Type II.

6.8 Research limitations

The following limitations were identified in this research:

1. Only individuals who were associated with the Spinal Muscular Atrophy Association of Australia, Muscular Dystrophy Foundation and Muscular Dystrophy Australia were invited to participate in the research. This would have excluded an unknown number of individuals not associated with those organisations. The unknown individuals may have had a differing opinions to participants in this research, particularly if they were poorly supported;
2. The pilot groups for the online questionnaire and interview questions did not include any individuals or parents of individuals living with SMA Type II, so as to not exclude any potential participants from the actual research.
Chapter 6: Discussion

However, feedback from individuals and parents of individuals living with SMA Type II about the online questionnaire and interview questions prior to recruitment may have provided valuable information that improved this research.

3. Due to the design of the research, people who were interested in the research subject matter were more likely to participate; therefore, the viewpoints reported may not fully represent the viewpoints of the wider Australian SMA community;

4. Two surveyed individuals perceived that exercise was harmful and their perspectives may have provided a deeper understanding and more balanced view of exercise had they opted to partake in the interviews;

5. This was an Australian study, therefore, may not be as applicable to individuals who reside in other countries, particularly those from developing countries; and

6. The assumption was made that as the Australian Physical Activity and Sedentary Behaviour Guidelines were made for individuals of all abilities, they would be applicable to individuals living with SMA Type II. 

Justification for this assumption included the similar exercise advice published for adults living with a muscle-wasting condition on the Muscular Dystrophy UK website.

6.9 Conclusion

This research highlighted that the majority of individuals living with SMA Type II perceived exercise to be valuable and that it was beneficial to their physical and psychological functions and the social aspects of their life. A review of the clinical classification of SMA could be beneficial for practitioners, individuals living with SMA and their parents. Individuals living with SMA Type II are not precluded from having other medical conditions, both physical and psychological, and while the risk for this is low, it could create additional barriers to exercise. Degeneration impacted on individuals’ definition of exercise and the frequency and types of exercises they participated in. Some individuals were creative in the exercises they participated in, demonstrating that physical fitness could be achieved in a variety of ways. Individuals were most exposed to swimming and perceived it to offer benefits
in numerous components of physical fitness. However, greater exposure to different types of exercises may broaden individuals’ perceptions on the effects of exercise. Cardiorespiratory and fine motor exercises were considered to be most valuable to individuals living with SMA Type II to maintain their quality of life, health, longevity and independence. Individuals living with SMA Type II faced similar exercise determinants to individuals living with other degenerative NMDs. The majority of research participants perceived that the Australian physical activity guidelines were not applicable to them; however, 30% had still exercised for more than 150 minutes/week at some point in their lifetime. Individuals living with SMA Type II had mixed interactions with their practitioners, indicating that they did not receive standardised care. Further research is needed in the area of exercise perceptions and participation of individuals living with SMA Type II.
Chapter 7 Conclusion and recommendations

The previous chapter discussed the research findings. This chapter concludes the thesis and provides recommendations for future research, practice and policy.

7.1 Conclusion

The aim of this research was to explore the exercise perceptions of individuals and parents of individuals, living with spinal muscular atrophy (SMA) Type II. The research aims specifically sought to examine participants’ perceptions about the effectiveness of exercise to treat the symptoms and slow the progression of SMA Type II, the value of exercise and exercise determinants. Through the employment of the sequential mixed methods methodology, questionnaire and interview questions were developed to explore the exercise perceptions of individuals living with SMA Type II and their parents.

The research findings showed that individuals living with SMA Type II have the ability to stand and walk, and are not excluded from having other conditions, including depression, anxiety, cardiac conditions and encephalitis. Individual perspectives of exercise included activities like using a cough assist machine, singing, playing musical instruments knitting and activities of daily living. At the time of the research, 20% of participants exercised for more than 150 minutes/week, while 30% achieved this milestone at some stage of their life. Swimming was the most common exercise individuals living with SMA Type II engaged in. Swimming was perceived to be beneficial for cardiorespiratory function, motor function ability, orthopaedic health and muscular strength. Individuals living with SMA Type II perceived that exercise was beneficial in treating the symptoms of SMA Type II, specifically respiratory function and motor function ability. Exercise had the potential to either positively or negatively impact quality of life; however, most perceived that exercise had a positive impact. Individuals living with SMA Type II reported that exercise for cardiorespiratory function and fine motor skills were most important, particularly as their condition degenerated, to maintain quality of life, optimal health, longevity and independence. Stretches and swimming were the most common exercises practitioners advised and prescribed to individuals living with SMA Type II. Determinants for exercise were multileveled for individuals living with SMA Type II and included environmental, socio-cultural and individual determinants.
Environmental determinants perceived to be most influential were accessibility of facilities, services and equipment, and funding. Socio-cultural determinants perceived to be most influential were parents, friends and schools and school teachers. Individual determinants perceived to be most influential were knowledge about exercise; degeneration of an individual’s condition, specifically motor function ability, respiratory function and muscular strength; motivation; and attitude.

This research found that some individuals living with SMA Type II exceeded the motor function ability stated in the clinical classification. This finding could impact physician’s knowledge and care advice, the perceptions of individuals living with SMA Type II of their own abilities and care decisions, and social interactions between individuals living with SMA. The definition of exercise varied amongst individuals living with SMA Type II; however, all still met the clinical definition of exercise. Swimming was the most common exercise prescribed and engaged in, which may indicate that practitioner advice and prescription may influence the type of exercise engagement for individuals living with SMA Type II. One-third of participants had exercised for more than 150 minutes per week, indicating that some individuals living with SMA Type II may be meeting the Muscular Dystrophy UK recommendations for aerobic exercise. Lack of practitioner knowledge and prescription could create obstacles, but did not always hinder individuals from engaging in exercise. Whilst degeneration and confinement to a wheelchair can lead to more sedentary behaviour, it did not preclude individuals living with SMA Type II from engaging in exercise. The exercise determinants outlined by individuals living with SMA Type II were supported by the current literature on exercise determinants for individuals living with an neuromuscular disease (NMD). However, the results also highlighted the role of schools and school teachers as exercise determinants for individuals living with SMA Type II.

This research contributed to the literature by addressing a few of the many research gaps in exercise perceptions for individuals living with an NMD and SMA Type II specifically. This research provided specific evidence of the exercise behaviour of individuals living with SMA Type II, including type and frequency, thus fulfilling the first research objective. The first and second objectives of this research were achieved, as specific insight was gained into the value and effectiveness of exercise to improve quality of life, physiological and psychological function and treat the symptoms of SMA Type II as perceived by individuals living with SMA Type II.
This knowledge can help to educate practitioners and individuals living with SMA Type II on the different types of exercise this population can engage in and highlight that individuals living with SMA Type II can achieve the Muscular Dystrophy UK exercise recommendations. Findings from this research suggest that optimal standardised care in regards to exercise, as outlined in the Consensus Statement for Standard of Care in Spinal Muscular Atrophy (Consensus Statement), have not yet been achieved.

The third research objective was achieved, as specific evidence of the exercise determinants for individuals living with SMA Type II was provided. Furthermore, this research provided further evidence of the exercise determinants for individuals living with an NMD. Through increased understanding of exercise determinants, the value and effectiveness of exercise strategies can be developed to improve exercise behaviour in this population. These include, but are not limited to, implementing policies to ensure that individuals living with SMA Type II have equal opportunities to engage in exercise; improving the accessibility of facilities, services and equipment; increasing funding to ensure individuals living with SMA Type II can access exercise equipment, exercise facilities and exercise professionals; advertising exercise recommendations for individuals living with an NMD; educating practitioners to improve their knowledge and ability to prescribe exercise; improving school teachers’ knowledge to enable them to provide inclusive physical education and exercise knowledge to children living with a disability; and conducting further research into the effects of exercise for individuals living with an NMD, thus providing a basis to implement exercise interventions for this population. Addressing exercise perceptions of individuals living with SMA Type II could improve their exercise participation and subsequently further improve their morbidity, mobility and quality of life.
7.2 Recommendations

Recommendations for future research, practice and policy are outlined in this section.

Research recommendations:
1. Further research into the effectiveness of exercise on physical and psychological function for children and adults living with an NMD, and specifically SMA.
2. Further research into exercise perceptions and determinants for individuals living with an NMD, and specifically SMA.
3. Further research into practitioners’ perspectives of the exercise advice and prescription they provide to individuals living with SMA Type II.

Practice recommendations:
4. Implement increased advertising of information about exercise for individuals living with an NMD, including publishing the Muscular Dystrophy UK exercise advice for adults with a muscle wasting condition on support organisation websites like SMA Trust and SMA Association of Australia.
5. Implement more comprehensive education and training for practitioners and school teachers regarding exercise for individuals living with an NMD.
6. Implement improved communication between practitioners and their patients that places higher emphasis on the exercise recommendations outlined by the Consensus Statement and Muscular Dystrophy UK.
7. Implement exercise interventions with individuals living with SMA Type II using strategies to improve their exercise intention and behaviour.

Policy recommendations:
9. Advocate for the clinical classification of SMA to be revised with regard to the motor function ability and life expectancy of individuals living with SMA Type II.

10. Advocate for the Consensus Statement to include a care topic section relating to adults who are living with SMA Type I, II and III to guide practitioners on providing the minimum level of care for adults living with SMA.

11. Advocate for practitioners to educate adult individuals living with SMA Type II about their condition, as advancements in medical knowledge, technology and treatments are continuously changing, and to ensure that individuals are well informed about their condition, can make informed decisions about their own care and are less reliant on their parents’ priorities and beliefs regarding lifestyle and treatment options.

12. Advocate for improvements to the accessibility of facilities, services and equipment and funding to increase exercise participation of individuals living with a disability.

13. Advocate for the development of policies to integrate electric wheelchairs into current sports.

14. Advocate for the development of electric wheelchair sports for individuals living with degenerative conditions that have a higher focus on improving more components of physical fitness.
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## List of Appendices

### Appendix One: Summary of Reviewed Literature

<table>
<thead>
<tr>
<th>Year</th>
<th>Title</th>
<th>Authors</th>
<th>Objective</th>
<th>Methodology</th>
<th>Sample Characteristics</th>
<th>Participants Demographics</th>
<th>Country</th>
<th>Age</th>
<th>Sex</th>
<th>Disability</th>
<th>Country</th>
<th>Number of Participants</th>
<th>Nationality</th>
<th>Sex Ratio</th>
<th>Country</th>
<th>Sex Ratio</th>
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<th>Country</th>
<th>Number of Participants</th>
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<td>2002</td>
<td>Study on physical activity and health promotion for children with neuromuscular disease</td>
<td>Hellström, K.</td>
<td>Investigating the impact of physical activity on health and well-being</td>
<td>Qualitative</td>
<td>UK</td>
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<td>UK</td>
<td>15</td>
<td>10</td>
<td>NMD</td>
<td>UK</td>
<td>28 individuals</td>
<td>Caucasian</td>
<td>1.5:1</td>
<td>UK</td>
<td>1.5:1</td>
<td>NMD</td>
<td>UK</td>
<td>28 individuals</td>
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<td>2004</td>
<td>Acceptance of pain among children with chronic disease</td>
<td>Hellström, K., Clarenbach, C. F.</td>
<td>Examining the impact of pain on the quality of life and coping strategies</td>
<td>Qualitative</td>
<td>Sweden</td>
<td>18 children</td>
<td>Sweden</td>
<td>16</td>
<td>2</td>
<td>DMD</td>
<td>Sweden</td>
<td>18 children</td>
<td>Caucasian</td>
<td>1.5:1</td>
<td>Sweden</td>
<td>1.5:1</td>
<td>DMD</td>
<td>Sweden</td>
<td>18 children</td>
<td>Caucasian</td>
<td>1.5:1</td>
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<tr>
<td>2009</td>
<td>Assessment of pain among children with spinal muscular atrophy</td>
<td>Hellström, K., McDonald, C. M.</td>
<td>Investigating the impact of pain on the quality of life and coping strategies</td>
<td>Qualitative</td>
<td>Sweden</td>
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<td>SMA</td>
<td>Sweden</td>
<td>18 children</td>
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<td>2015</td>
<td>Resilience of children with a chronic disease over ten years</td>
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<td>Assessing the impact of resilience on the quality of life and coping strategies</td>
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<td>1.5:1</td>
<td>Multiple NMD</td>
<td>Sweden</td>
<td>18 children</td>
<td>Caucasian</td>
<td>1.5:1</td>
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### Notes
- **SMA**: Spinal Muscular Atrophy
- **NMD**: Neuromuscular Disease
- **DMD**: Duchenne Muscular Dystrophy
- **PPS**: Progressive Proximal Scoliosis
- **SCI**: Spinal Cord Injury
- **JCTD**: Juvenile Cerebellar Ataxia
- **LGMD**: Late-onset Gower's Disease
- **AS**: Amyotrophic Lateral Sclerosis
- **FSHD**: Facioscapulohumeral Dystrophy
- **MMD**: Myotonic Dystrophy
- **MS**: Multiple Sclerosis
- **MD**: Myopathy and Congenital Myopathies
- **CP**: Cerebral Palsy
- **HC**: Healthy Controls
- **Caucasian**: Caucasian
- **Asian**: Asian
- **American**: American
- **Australian**: Australian
- **Swedish**: Swedish
- **French**: French
- **American**: American
- **Chinese**: Chinese
- **Japanese**: Japanese
- **Korean**: Korean
- **Caucasian**: Caucasian
- **Asian**: Asian
- **American**: American
- **Australian**: Australian
- **Swedish**: Swedish
- **French**: French
- **American**: American
- **Chinese**: Chinese
- **Japanese**: Japanese
- **Korean**: Korean
- **Caucasian**: Caucasian
- **Asian**: Asian
- **American**: American
- **Australian**: Australian
- **Swedish**: Swedish
- **French**: French
- **American**: American
- **Chinese**: Chinese
- **Japanese**: Japanese
- **Korean**: Korean

### Additional Information
- **Model of Disability**: Theoretical framework used to understand the impact of disability on quality of life and coping strategies.
- **Quantitative**: Data collected through surveys and questionnaires.
- **Qualitative**: Data collected through interviews and focus groups.

### Key Points
- Children with neuromuscular disorders experience significant pain and its impact on their quality of life and coping strategies.
- Resilience plays a crucial role in the quality of life of children with chronic diseases.
- Acceptance of pain and physical activity are significant factors in the quality of life of children with chronic diseases.
- The impact of chronic disease on the quality of life and coping strategies is significant.

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### References
Appendix Two: International classification of functioning, disability and health (ICF)

Appendix Three: Bostrom and Alstrom 2004 theory for NMD based on ICF
Appendix Four: Revised Ethics Approval

27 January 2015

Michelle Searle
Director, Office of Research
Tel: +61 7 5459 4574
Email: humanethics@usc.edu.au

F22226

Marissa Hoey
Dr Bec Mellifont
Dr Jane Taylor
Faculty of Science, Health, Education and Engineering

Dear Marissa, Bec and Jane

Expedited ethics approval for amended research project: Australian Spinal Muscular Atrophy (SMA) Type II Individuals’ Perspectives of Exercise as a Therapy (S/14/603)

This letter is to confirm that on 9 January 2015, the Chairperson of the Human Research Ethics Committee of the University of the Sunshine Coast granted expedited ethics approval for an amendment to the above project, subject to specific conditions which were satisfied on 20 January 2015.

The amendment refers to an extension of the end date from 31/12/2014 to 31/12/2016; for individual interview participants to also be invited to participate in a group interview for up to 2 hours; 3) the addition of Jane Taylor as a supervisor; and 4) the change in chief investigator from Rebecca Mellifont to Marissa Hoey.

The conditions for ethics approval for this project as outlined in our letter of 24 June 2014 continue to apply.

If you have any queries in relation to this ethics approval or if you require further information please contact a Research Ethics Officer by email at humanethics@usc.edu.au or by telephone on +61 7 5459 4574 or 5430 2823.

Yours sincerely

Michelle Searle
Director, Office of Research
Appendix Five: Online Questionnaire

Initial Survey: Australian Spinal Muscular Atrophy (SMA) Type II

Project Title and Background

Project Title:
Australian Spinal Muscular Atrophy (SMA) Type II Individuals’ Perspective of Exercise as a Therapy

Background
Exercise can improve muscular strength, endurance, cardiovascular function and motor function in healthy populations. However, limited research has been conducted into the effectiveness of exercise in treating SMA Type II. Due to the limited number of studies conducted in this area, we do not know how effective exercise is in treating SMA Type II.

It is not known how common the use of exercise is in treating SMA Type II, the extent that exercise is implemented into treatment plans of individuals with SMA Type II, the perceptions of Australian individuals with SMA Type II regarding the importance of exercise in treating the disease, or the types or combinations of exercise that individuals with SMA Type II believe are of most benefit in treating the symptoms of SMA Type II.

Research has shown that an individual’s attitudes and opinions will influence their perceptions of the benefits and risks associated with engaging in exercise. These perceptions will therefore determine whether an individual decides to include or exclude exercise as part of their lifestyle as well as their compliance whilst engaging in an exercise program.

This research project aims to evaluate the perceptions of the Australian SMA Type II community regarding the effect of exercise as a treatment for SMA Type II and to determine the type and duration of exercise individuals with SMA Type II view as the most beneficial as a treatment. This is important knowledge for the health professionals who treat individuals with SMA Type II because a person’s perceptions of exercise indicate whether they incorporate exercise into their lifestyle. Additionally, the knowledge gained from those individuals who largely focus on exercise as part of their treatment plan can be shared with the rest of the SMA Type II community.

This survey is aimed at all individuals with SMA Type II and the parent’s or guardians of individuals with SMA Type II. If a parent is completing this survey, the information must relate to their child.

Investigators:
Marissa Hoey – Masters Researcher
Dr Jane Taylor - Supervisor
Dr Rebecca Mellifont - Supervisor

This research has been approved by the Human Ethics Committee University of the Sunshine Coast – S/14/603.
# Initial Survey: Australian Spinal Muscular Atrophy (SMA) Type II

## Research Purpose

| Part 1: To ascertain your perceptions of exercise as an Australian individual with SMA Type II. |
| Part 2: To review past and current exercise programs implemented by you and to ascertain your perceptions regarding the effectiveness of each exercise program to slow the progression of SMA Type II. |
| Part 3: To evaluate the current public policy and exercise recommendations for individuals with SMA. |

### Overall Design

**Part 1:** You are invited to participate in an online survey investigating your perceptions of health professionals’ attitudes towards exercise, the amount and type of exercise you are currently engaged in, and your perceptions of the benefits or disadvantages of engaging in exercise. The online survey will take less than one hour of your time. During the online survey participants are asked if they have been genetically tested for SMA and if they are willing to share these results with the researchers of this study. The first purpose of this question is to determine how many individuals have been diagnosed with SMA Type II through genetic testing. The second purpose of this question will apply to those individuals who are willing to share their genetic test results and participate in a private interview.

**Part 2:** You are also invited to participate in a private interview and a group interview. The focus of the private interview will be to investigate the current and previous exercise programs that you have engaged in and to ascertain your perceptions regarding the benefits, risks and effectiveness of each exercise program. The private interview will take up to two hours of your time. The focus of the group interview is to reflect on and validate the range and scope of programs that are employed by the participant group. All individual data will be de-identified prior to this point. This is an opportunity to add any further information you feel applies to the study, including suggestions for appropriate future exercise programs for individuals with SMA Type II. The group interview will take up to two hours of your time.

Participants will be given a consent form prior to the interviews. The private interview will be organised at your convenience. The group interview will be organised at a time convenient to the majority of participants. It may be advisable to bring any files from your allied health professionals regarding any exercise programs you have engaged in to help you accurately recount your exercise history during the private and group interview.

To participate in the interviews, please provide phone and/or email details when prompted during the survey. Approximately 15-20 individuals who have experienced exercise or are currently undertaking exercise as a mode of therapy, will be chosen to participate in a private interview and offered the opportunity to participate in the group interview. Private interview participants who have been genetically tested and are willing to share those results with the researchers should be advised that the researchers are only interested in determining how many copies of the SMN 2 gene the individual has as it is a major determinant of SMA severity. Interviews will be recorded and a transcript of the interviews will be typed from the recording. Participants will be offered the opportunity to read the typed transcript and to make changes to any statements they have made before the data is collated.
# Initial Survey: Australian Spinal Muscular Atrophy (SMA) Type II

## Risks, Results & Confidentiality

### Risks to Participants

There may be physical discomfort felt whilst completing the online survey or during the private interview but it should not exceed your usual daily discomfort levels. If you wish, you can stop or take a break at any time. There may be questions asked during the interviews which unintentionally challenge, cause upset or the remembrance of a painful moment.

Please don’t hesitate to tell us if you feel uncomfortable during the interview process – remember you can choose, at any time, not to answer any questions or to withdraw from this study. If you engage in the group interviews you will be de-identified and given a participant letter (i.e. participant a) to enable a de-identified transcript to be produced. While every effort will be taken to ensure that the data collected will be de-identified, the Australian SMA Type II population is very small and comments published for this research may unintentionally be recognised by another member or associate of the SMA Type II community. If you are uncomfortable with these possibilities you are under no obligation to participate in the research project or you can choose, at any time, not to answer any questions or to withdraw from the study.

Should you experience any concerns from the interviews, you may wish to contact the support services at USC: USC Student Life and Learning (Ground floor, E Block, University of the Sunshine Coast, ph: 5430 1226 studentservices@usc.edu.au).

### Results

Upon completion of the study, participants will be invited via email to attend a research seminar at the University of the Sunshine Coast. A summary of the results and conclusions will also be sent to all interested participants, and will be made available on the SMA website for public viewing. The results of the study will also be prepared for submission to a peer-reviewed journal for publication.

### Confidentiality

Part 1: The initial online survey is anonymous however those individuals wishing to participate in a private interview will have the opportunity to provide their contact details after the survey’s completion which will make them identifiable to the researcher.

Part 2: Data from the private and group interviews will be entered into a simple coding system (e.g. S1, S2), with the original participant names not recorded with this code. This makes it impossible for a third party to identify any one individual from the stored data. Data will be stored on the USC network using the Chief Investigator’s password protected storage space. Data will also be stored and analysed on the student researcher’s password protected computer. Signed consent forms and any hardcopies of interviews will be stored in a lockable filing cabinet in the Chief Investigator’s office (T4.04). At the completion of this study, the private interviews will be completely de-identified, and the audio recordings from the interviews will be destroyed. Data will be retained for future research surrounding SMA, and may be released to other researchers in the future.
Appendices

**Initial Survey: Australian Spinal Muscular Atrophy (SMA) Type II**

**Consent, Complaints & Contacts**

Consent to participate:
In all research projects it is important to gain consent from participants prior to collecting data. Here, to maintain your anonymity, we will assume that if you start the survey you are giving your consent (with your parents' approval as appropriate).

Complaints
Should you have any issues with any aspect of this project (e.g. testing, procedures, security of data, etc.), and you don't feel comfortable discussing these with any of the researchers listed below, you are encouraged to lodge a formal complaint through the Secretary of the University of the Sunshine Coast Human Research Ethics Committee at the Office of Research (contact ph: (07) 5459 4574 or email: humanethics@usc.edu.au).

Participation in the following study is voluntary and participants may withdraw at any stage, without explanation and there will be no consequence as a result. It is important to note that participation in this study and the results from this research will be kept confidential and any statements made to the researchers will be de-identified.

Contacts
Marissa Hoey (Masters researcher)
Phone: 0405 113 261
Email: Marissa.Hoey@research.usc.edu.au

Dr Jane Taylor (Supervisor)
Phone: (07) 5459 4543
Email: jane.taylor@usc.edu.au

Dr Rebecca Mellifont (Supervisor)
Phone: (07) 5456 5065
Email: rebecca.mellifont@usc.edu.au

The research team and the University would like to thank you for your interest in this project and appreciate the effort involved. Your support is a key element contributing to the success of exercise science research at the University of the Sunshine Coast both now and in the future.

*1. What is your age? (note: the age of person diagnosed with SMA Type II)*

- 18 or above
- under 18

**Initial Survey: Australian Spinal Muscular Atrophy (SMA) Type II**

**Informed Consent (18 years or older)**

In all research projects it is important to gain consent from participants prior to collecting data. Here, to maintain your anonymity, we will assume that if you start the survey you are giving your consent.

I agree to participate in the research study entitled "Australian SMA Type II individual’s perspective of exercise as a therapy". I understand what is expected of me, the benefits and risks involved and that my participation in the study is voluntary. I acknowledge I have the right to question any part of the procedure and that I can withdraw at any time without this being held against me.

I consent to any data about my specific participation to be used in a confidential manner for the purposes of this research project and in future research and may be published. However, my right to privacy will be maintained and personal details will remain confidential.

The following questionnaire is an indicative survey only. If you would like the opportunity to expand upon your answers a number of private interviews will be conducted in the near future. If you are interested in participating in a private interview please provide your contact details at the end of the survey.

*2. Please select the answer that best describes you*

- I am an individual with SMA Type II
- I am the parent/guardian of an individual with SMA Type II
**Informed Consent (under 18 years)**

In all research projects it is important to gain consent from participants prior to collecting data. As you have indicated that you are under 18 years of age, or that you are a parent of a child with SMA Type II, the following information is for you.

If you are under 18 years of age, we request that you complete this survey together with your parent/guardian as legally they are responsible to give informed consent for your actions. In order to maintain your anonymity, we will assume that if you start the survey you have your parent with you giving their consent.

If you are the parent/guardian of a child under 18 years of age diagnosed with SMA Type II, we request that you answer the questions bearing in mind your child's treatment and the perspectives you have. We will assume that if you start the survey, you are giving your consent to participate.

I agree to participate in the research study entitled “Australian SMA Type II individual’s perspective of exercise as a therapy”, and give consent to provide information about me/my child. I understand what is expected of me, the benefits and risks involved and that my participation in the study is voluntary. I acknowledge I have the right to question any part of the procedure and that I can withdraw at any time without this being held against me.

I consent to any data about my specific participation to be used in a confidential manner for the purposes of this research project and in future research and may be published. However, my right to privacy will be maintained and personal details will remain confidential.

The following questionnaire is an indicative survey only. If you would like the opportunity to expand upon your answers a number of private interviews will be conducted in the near future. If you are interested in participating in a private interview please provide your contact details at the end of the survey.

**3. Please select the answer that best describes you**

- I am an individual with SMA Type II
- I am the parent/guardian of an individual with SMA Type II

---

**4% Complete Demographic Information For Individuals**

**4. How old are you?**

**5. What is your gender?**

- Female
- Male

**6. How old were you when you first showed symptoms of SMA and what symptoms did you have? i.e. At 7 months of age you were unable to sit due to muscle weakness.**

**7. How old were you when you were diagnosed with SMA Type II?**

**8. Have you been genetically tested for SMA Type II?**

- Yes
- No
- I don't know
**Initial Survey: Australian Spinal Muscular Atrophy (SMA) Type II**

**8% Complete**

9. Would you be willing to share the results of your genetic test with the researcher of this study?
- [ ] Yes
- [ ] No

10. Have you been diagnosed with any other condition/s other than Spinal Muscular Atrophy?
- [ ] Yes
- [ ] No

**12% Complete Other conditions**

11. Could you please list the other condition/s you have been diagnosed with.

<table>
<thead>
<tr>
<th>Other conditions</th>
</tr>
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<tbody>
<tr>
<td></td>
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</tbody>
</table>
### 16% Complete Movement Ability

**12. What is the BEST level of movement you have EVER been able to achieve independently WITHOUT equipment?**  
*(Please tick ALL boxes that you have been able to accomplish.)*

- [ ] There is no movement I can achieve without equipment
- [ ] Breathe independently
- [ ] Sit
- [ ] Stand
- [ ] Crawl
- [ ] Walk
- [ ] Run

**13. What is your CURRENT level of movement WITHOUT equipment?**  
*(Please tick ALL boxes that you can CURRENTLY accomplish.)*

- [ ] There is no movement I can achieve without equipment
- [ ] Breathe independently
- [ ] Sit
- [ ] Stand
- [ ] Crawl
- [ ] Walk
- [ ] Run

**14. What is the BEST level of movement you have EVER been able to achieve WITH the help of EQUIPMENT?**  
*(Please tick ALL boxes that you have been able to accomplish with equipment)*

- [ ] Breathe
- [ ] Sit
- [ ] Stand
- [ ] Crawl
- [ ] Walk
- [ ] Run

**15. What is your CURRENT level of movement WITH EQUIPMENT?**  
*(Please tick ALL boxes that you can CURRENTLY accomplish with equipment)*

- [ ] Breathe
- [ ] Sit
- [ ] Stand
- [ ] Crawl
- [ ] Walk
- [ ] Run
### Initial Survey: Australian Spinal Muscular Atrophy (SMA) Type II

#### 21% Complete Doctors

16. Have any of your doctors EVER discussed exercise as a therapy for SMA Type II?
- [ ] Yes
- [ ] No

#### 25% Complete

17. What was the exercise advice you were given from your doctor?

18. Complete the following statement: In my opinion, MOST of my doctors' had the attitude that if I exercise it would...

19. In your opinion, have you EVER been treated by a doctor who had a different attitude towards exercise as a therapy for SMA Type II compared to the majority of doctors that you have had contact with?
- [ ] Yes
- [ ] No

#### 29% Complete

20. In your opinion what was the doctors' attitude towards exercise as a therapy for SMA Type II?

21. In your opinion, of all the doctors that you have been in contact with what percentage of doctors' had this attitude towards exercise?
- [ ] Less than 10%
- [ ] 10% - 20%
- [ ] 21% - 30%
- [ ] 31% - 40%
- [ ] 41% - 50%
<table>
<thead>
<tr>
<th><strong>Initial Survey: Australian Spinal Muscular Atrophy (SMA) Type II</strong></th>
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</thead>
<tbody>
<tr>
<td><strong>33% Complete Physiotherapy</strong></td>
</tr>
<tr>
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</tr>
<tr>
<td>* 22. Have you EVER been advised to engage in physiotherapy?</td>
</tr>
<tr>
<td>○ Yes</td>
</tr>
<tr>
<td>○ No</td>
</tr>
<tr>
<td></td>
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<tr>
<td>* 23. Have you EVER consulted a physiotherapist?</td>
</tr>
<tr>
<td>○ Yes</td>
</tr>
<tr>
<td>○ No</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th><strong>Initial Survey: Australian Spinal Muscular Atrophy (SMA) Type II</strong></th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>37% Complete Physiotherapist Questions</strong></td>
</tr>
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<tr>
<td>* 24. What was the exercise advice you were given from your physiotherapist?</td>
</tr>
<tr>
<td></td>
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<tr>
<td>* 25. Complete the following statement: In my opinion, MOST of my physiotherapists’ had the attitude that if I exercise it would...</td>
</tr>
<tr>
<td></td>
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<tr>
<td>* 26. In your opinion, have you EVER been treated by a physiotherapist who had a different attitude towards exercise as a therapy for SMA Type II compared to the majority of physiotherapists that you have had contact with?</td>
</tr>
<tr>
<td>○ Yes</td>
</tr>
<tr>
<td>○ No</td>
</tr>
</tbody>
</table>
27. In your opinion what was the physiotherapists' attitude towards exercise as a therapy for SMA Type II?

28. In your opinion, of all the physiotherapists that you have been in contact with what percentage of physiotherapists had this attitude towards exercise?

- Less than 10%
- 10% - 20%
- 21% - 30%
- 31% - 40%
- 41% - 50%

29. Do you CURRENTLY incorporate physiotherapy into your treatment plan?

- Yes
- No
<table>
<thead>
<tr>
<th>Question</th>
<th>Options</th>
</tr>
</thead>
<tbody>
<tr>
<td>30. How often do you engage in physiotherapy?</td>
<td></td>
</tr>
<tr>
<td>31. What type of treatment are you consulting the physiotherapist for? i.e. pain management, manual manipulation, mobility aids, exercise.</td>
<td></td>
</tr>
<tr>
<td>32. Have you PREVIOUSLY incorporated physiotherapy into your treatment plan?</td>
<td>Yes, No</td>
</tr>
<tr>
<td>33. How often did you engage in physiotherapy and why did you stop?</td>
<td></td>
</tr>
<tr>
<td>34. What type of treatment were you consulting the physiotherapist for? i.e. pain management, manual manipulation, mobility aids, exercise.</td>
<td></td>
</tr>
<tr>
<td>35. Have you EVER been advised to consult an accredited exercise physiologist?</td>
<td>Yes, No</td>
</tr>
<tr>
<td>36. Have you EVER consulted an accredited exercise physiologist?</td>
<td>Yes, No</td>
</tr>
</tbody>
</table>
37. In your opinion what was the exercise physiologist’s attitude towards exercise as a therapy for SMA Type II?

38. What type of exercise was prescribed to you by the exercise physiologist?

39. Has anyone else advised you to engage in exercise as a therapy for SMA Type II?
   - Yes
   - No
### Initial Survey: Australian Spinal Muscular Atrophy (SMA) Type II

#### 40. Who advised you to engage in exercise as a therapy for SMA Type II?

(Please tick all of the boxes that apply to you.)

- [ ] Another individual with a neuromuscular disease or a member of their family
- [ ] A relative or friend
- [ ] Information collected on the internet
- [ ] Occupational Therapist
- [ ] SMA Association of Australia
- [ ] Muscular Dystrophy Foundation
- [ ] Other

Other (please specify)

#### 41. What is your opinion on exercise as a form of treatment for SMA Type II?

- [ ] It causes great harm in treating SMA Type II.
- [ ] It causes some harm in treating SMA Type II.
- [ ] It is slightly harmful in treating SMA Type II.
- [ ] It causes harm however benefits are gained in treating SMA Type II.
- [ ] It is neither harmful nor beneficial in treating SMA Type II.
- [ ] It is only slightly beneficial in treating SMA Type II.
- [ ] It has some benefits in treating SMA Type II.
- [ ] It is of great benefit in treating SMA Type II.
- [ ] Other/I would like to expand upon my answer.

Other (please specify)

#### 42. What are your personal feelings regarding exercise, physical activity and sport?

- [ ] I don’t have any personal feelings regarding exercise.
- [ ] I don’t like exercise therefore I don’t engage in any physical activity.
- [ ] I don’t like to exercise however I participate in physical activity as I feel it will benefit my health.
- [ ] I enjoy exercise but I feel that I cannot participate in physical activity due to my condition.
- [ ] I enjoy exercise and I participate in physical activity whenever the opportunity presents itself.
- [ ] Other/I would like to expand upon my answer.

Other (please specify)
# Initial Survey: Australian Spinal Muscular Atrophy (SMA) Type II

71% Complete Your opinion questions 1/2

43. The following questions are all requesting your opinion:

<table>
<thead>
<tr>
<th>Question</th>
<th>Strongly Disagree</th>
<th>Disagree</th>
<th>Neutral</th>
<th>Agree</th>
<th>Strongly Agree</th>
</tr>
</thead>
<tbody>
<tr>
<td>Do you believe it is a waste of time to engage in exercise to treat SMA Type II?</td>
<td>☐</td>
<td>☐</td>
<td>☐</td>
<td>☑</td>
<td>☐</td>
</tr>
<tr>
<td>Do you believe that engaging in exercise will result in an injury caused by overexertion?</td>
<td>☐</td>
<td>☐</td>
<td>☐</td>
<td>☑</td>
<td>☐</td>
</tr>
<tr>
<td>Do you believe that you have a higher rate of injury when engaging in exercise compared to the general population?</td>
<td>☐</td>
<td>☐</td>
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<td>☑</td>
<td>☐</td>
</tr>
<tr>
<td>Do you believe that you should not engage in exercise if you are experiencing pain?</td>
<td>☐</td>
<td>☐</td>
<td>☐</td>
<td>☑</td>
<td>☐</td>
</tr>
<tr>
<td>Do you believe that engaging in exercise will increase the rate of deterioration in your body?</td>
<td>☐</td>
<td>☐</td>
<td>☐</td>
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<td>☐</td>
</tr>
<tr>
<td>Do you think engaging exercise will increase your level of tiredness throughout that day?</td>
<td>☐</td>
<td>☐</td>
<td>☐</td>
<td>☑</td>
<td>☐</td>
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<tr>
<td>Do you believe that you should stop exercising if you are experiencing pain?</td>
<td>☐</td>
<td>☐</td>
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<td>☑</td>
<td>☐</td>
</tr>
<tr>
<td>Do you believe that exercise can effect your mood?</td>
<td>☐</td>
<td>☐</td>
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<td>☐</td>
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<tr>
<td>Do you believe that engaging in exercise will decrease the rate of deterioration in your body?</td>
<td>☐</td>
<td>☐</td>
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<td>☑</td>
<td>☐</td>
</tr>
<tr>
<td>Do you believe that exercise will decrease the level of pain you experience?</td>
<td>☐</td>
<td>☐</td>
<td>☐</td>
<td>☑</td>
<td>☐</td>
</tr>
<tr>
<td>Do you think exercise will reduce your ability to move throughout that day?</td>
<td>☐</td>
<td>☐</td>
<td>☐</td>
<td>☑</td>
<td>☐</td>
</tr>
<tr>
<td>Do you think exercise can improve your energy levels?</td>
<td>☐</td>
<td>☐</td>
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<td>☑</td>
<td>☐</td>
</tr>
<tr>
<td>Do you believe that exercise will increase the level of pain you experience?</td>
<td>☐</td>
<td>☐</td>
<td>☐</td>
<td>☑</td>
<td>☐</td>
</tr>
</tbody>
</table>
44. The following questions are all requesting your opinion:

<table>
<thead>
<tr>
<th>Question</th>
<th>Strongly Disagree</th>
<th>Disagree</th>
<th>Neutral</th>
<th>Agree</th>
<th>Strongly Agree</th>
</tr>
</thead>
<tbody>
<tr>
<td>Do you think exercise can improve the strength of your muscles?</td>
<td>☒</td>
<td>☒</td>
<td>☒</td>
<td>☒</td>
<td>☐</td>
</tr>
<tr>
<td>Do you think exercise can increase the size of your muscles?</td>
<td>☒</td>
<td>☒</td>
<td>☒</td>
<td>☒</td>
<td>☐</td>
</tr>
<tr>
<td>Do you think exercise can reduce the amount of fat stored in your body?</td>
<td>☒</td>
<td>☒</td>
<td>☒</td>
<td>☒</td>
<td>☐</td>
</tr>
<tr>
<td>Do you think exercise can improve your ability to move your body whilst performing activities in your daily life?</td>
<td>☒</td>
<td>☒</td>
<td>☒</td>
<td>☒</td>
<td>☐</td>
</tr>
<tr>
<td>Do you think exercise can improve your ability to breathe?</td>
<td>☒</td>
<td>☒</td>
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</tr>
<tr>
<td>Do you think exercise can improve your mood?</td>
<td>☒</td>
<td>☒</td>
<td>☒</td>
<td>☒</td>
<td>☐</td>
</tr>
<tr>
<td>Do you think exercise can improve your ability to make decisions?</td>
<td>☒</td>
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</tr>
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<td>Do you think exercise can improve your ability to cope with your condition?</td>
<td>☒</td>
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</tr>
<tr>
<td>Do you think exercise can improve the overall quality of your life?</td>
<td>☒</td>
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<td>Do you think exercise can increase your lifespan?</td>
<td>☒</td>
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</tr>
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<td>Do you think engaging in exercise or sport can create the opportunity to meet new people?</td>
<td>☒</td>
<td>☒</td>
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</tr>
<tr>
<td>Do you think engaging in exercise or sport can create the opportunity to expand your social life?</td>
<td>☒</td>
<td>☒</td>
<td>☒</td>
<td>☒</td>
<td>☐</td>
</tr>
</tbody>
</table>

45. Have you EVER engaged in exercise, physical activity or sport?

- ☒ Yes
- ☐ No
**Initial Survey: Australian Spinal Muscular Atrophy (SMA) Type II**

**84% Complete**

*46. What types of exercise/physical activity/sport have you engaged in over your lifetime? (Please tick ALL of the boxes that apply to you.)*

- [ ] Swimming
- [ ] Weight Training
- [ ] Yoga
- [ ] Boccia
- [ ] Archery
- [ ] Wheelchair Basketball
- [ ] Wheelchair soccer
- [ ] Sitting Volleyball
- [ ] Wheelchair Tennis
- [ ] Other

Other (please specify)

*47. Do you CURRENTLY engage in exercise, physical activity or sport?*

- [ ] Yes
- [ ] No
48. **What type of exercise/physical activity/sport do you CURRENTLY engage in?**  
(Please tick ALL of the boxes that apply to you.)

- [ ] I do not currently engage in any physical activity
- [ ] Swimming
- [ ] Weight Training
- [ ] Yoga
- [ ] Boccia
- [ ] Archery
- [ ] Wheelchair Basketball
- [ ] Wheelchair soccer
- [ ] Sitting Volleyball
- [ ] Wheelchair Tennis
- [ ] Other

*Other (please specify)*

49. **How long does it take you to recover after engaging in exercise?**

- [ ] Little to no time
- [ ] A short time
- [ ] Some time
- [ ] A long time
- [ ] Too long

50. **Do you CURRENTLY experience pain when you engage in exercise?**

- [ ] Never
- [ ] Occasionally
- [ ] Sometimes
- [ ] Usually
- [ ] Always
Appendices

Initial Survey: Australian Spinal Muscular Atrophy (SMA) Type II

51. Have you EVER experienced pain when you engage in exercise?
- Never
- Occasionally
- Sometimes
- Usually
- Always

52. Are there any types of exercise that you WOULD NOT feel comfortable engaging in?
(Please tick ALL of the boxes that apply to you.)
- I feel comfortable engaging in all types of exercise
- Swimming
- Weight Training
- Yoga
- Boccia
- Archery
- Wheelchair Basketball
- Wheelchair soccer
- Sitting Volleyball
- Wheelchair Tennis
- Other

Other (please specify)

53. Have you EVER engaged in exercise for social reasons?
- Yes
- No

If you would like to expand upon your answer please describe

Initial Survey: Australian Spinal Muscular Atrophy (SMA) Type II

54. Have you EVER engaged in exercise using weights?
- Yes
- No
### Initial Survey: Australian Spinal Muscular Atrophy (SMA) Type II

#### 92% Complete Exercise Duration

**55. In total how many minutes do you CURRENTLY exercise for each week?**

- [ ] Not at all
- [ ] Less than 30 mins
- [ ] 30-60 mins
- [ ] 61-90 mins
- [ ] 91-120 mins
- [ ] 121-150 mins
- [ ] More than 150 mins

**56. How many days per week do you CURRENTLY exercise for?**

- [ ] Not at all
- [ ] 1 day/week
- [ ] 2 days/week
- [ ] 3 days/week
- [ ] 4 days/week
- [ ] 5 days/week
- [ ] 6 days/week
- [ ] 7 days/week

**57. What was the MAXIMUM amount of minutes you have EVER exercised for within one week?**

- [ ] Not at all
- [ ] Less than 30 mins
- [ ] 30-60 mins
- [ ] 61-90 mins
- [ ] 91-120 mins
- [ ] 121-150 mins
- [ ] More than 150 mins
### Initial Survey: Australian Spinal Muscular Atrophy (SMA) Type II

**58. What was the MAXIMUM number of days per week you have EVER exercised on?**

- [ ] Not at all
- [ ] 1 day/week
- [ ] 2 days/week
- [ ] 3 days/week
- [ ] 4 days/week
- [ ] 5 days/week
- [ ] 6 days/week
- [ ] 7 days/week

**59. How long have you incorporated exercise into your treatment plan?**

- [ ] I don't engage in any exercise
- [ ] I have previously incorporated exercise however I no longer engage in physical activity
- [ ] Since I was diagnosed with SMA Type II
- [ ] For less than 1 year
- [ ] For the past 1-2 years
- [ ] For the past 3-4 years
- [ ] For the past 5-6 years
- [ ] For more than 6 years
- [ ] Other/I would like to expand upon my answer

Other (please specify)

**60. What drives you to engage in exercise?**

Some examples are: To lose weight; To strengthen your body; To slow the progression of SMA Type II; As a social interaction with others.

**61. Are you amenable to the possibility of the researcher of this study contacting you for a private interview within the next 6 months?**

- [ ] Yes
- [ ] No
### Initial Survey: Australian Spinal Muscular Atrophy (SMA) Type II

#### 4% Complete Demographic Information for Parents/Guardians

**62. How many children do you have with SMA Type II?**

- One
- Two
- Three
- Other

Other (please specify)

### Initial Survey: Australian Spinal Muscular Atrophy (SMA) Type II

#### 8% Complete

**63. How old is your child?**

**64. What is your child’s gender?**

- Female
- Male

**65. How old was your child when they first showed symptoms of SMA?**

i.e. At 2 weeks of age the baby had trouble suckling when feeding; At 7 months of age your child was unable to sit due to muscle weakness.

**66. How old was your child when they were diagnosed with SMA Type II?**

**67. Has your child been genetically tested for SMA Type II?**

- Yes
- No
- I don't know
**Initial Survey: Australian Spinal Muscular Atrophy (SMA) Type II**

### 68. How old are your children and what are their genders? (Please describe as follows: male 5 years; female 3 years)

### 69. How old were your children when they first showed symptoms of SMA? i.e. At 2 weeks of age the baby had trouble suckling when feeding; At 7 months of age your child was unable to sit due to muscle weakness.

### 70. How old were your children when they were diagnosed with SMA Type II?

### 71. Have your children been genetically tested for SMA Type II?

- [ ] Yes, all of my children have been genetically tested for SMA Type II
- [ ] Yes, but not all of my children have been genetically tested for SMA Type II
- [ ] No
- [ ] I don't know

### 72. Would you be willing to share the results of your child's/children's genetic test with the researcher of this study?

- [ ] Yes
- [ ] No

### 73. Has your child/children been diagnosed with any other conditions?

- [ ] Yes
- [ ] No

### 74. Could you please list the other condition/s your child/children have been diagnosed with.
<table>
<thead>
<tr>
<th>75. What is the BEST level of movement your child/children have ever been able to achieve independently WITHOUT equipment?</th>
</tr>
</thead>
<tbody>
<tr>
<td>□ There is no movement my child/children can achieve without equipment</td>
</tr>
<tr>
<td>□ Breathe independently</td>
</tr>
<tr>
<td>□ Sit</td>
</tr>
<tr>
<td>□ Stand</td>
</tr>
<tr>
<td>□ Crawl</td>
</tr>
<tr>
<td>□ Walk</td>
</tr>
<tr>
<td>□ Run</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>76. What is your child’s/children’s CURRENT level of movement WITHOUT equipment?</th>
</tr>
</thead>
<tbody>
<tr>
<td>□ There is no movement my child/children can CURRENTLY accomplish.</td>
</tr>
<tr>
<td>□ Breathe independently</td>
</tr>
<tr>
<td>□ Sit</td>
</tr>
<tr>
<td>□ Stand</td>
</tr>
<tr>
<td>□ Crawl</td>
</tr>
<tr>
<td>□ Walk</td>
</tr>
<tr>
<td>□ Run</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>77. What is the BEST level of movement your child/children have EVER been able to achieve WITH the help of EQUIPMENT?</th>
</tr>
</thead>
<tbody>
<tr>
<td>□ Breathe</td>
</tr>
<tr>
<td>□ Sit</td>
</tr>
<tr>
<td>□ Stand</td>
</tr>
<tr>
<td>□ Crawl</td>
</tr>
<tr>
<td>□ Walk</td>
</tr>
<tr>
<td>□ Run</td>
</tr>
</tbody>
</table>
78. What is your child’s/children’s CURRENT level of movement WITH EQUIPMENT? (Please tick ALL boxes that your child/children can CURRENTLY accomplish with equipment.)

- Breathe
- Sit
- Stand
- Crawl
- Walk
- Run

79. Have any of your child’s/children’s doctors EVER discussed exercise as a therapy for SMA Type II?

- Yes
- No

80. What was the exercise advice you were given from your doctor?

81. Complete the following statement: In my opinion, MOST of my child’s/children’s doctors’ had the attitude that if my child/children exercise it would...

82. In your opinion, has your child/children EVER been treated by a doctor who had a different attitude towards exercise as a therapy for SMA Type II compared to the majority of doctors that you have had contact with?

- Yes
- No
Initial Survey: Australian Spinal Muscular Atrophy (SMA) Type II

32% Complete

*83. In your opinion what was the doctors' attitude towards exercise as a therapy for SMA Type II?

*84. In your opinion, of all the doctors that you have been in contact with what percentage of doctors' had this attitude towards exercise?
- Less than 10%
- 10% - 20%
- 21% - 30%
- 31% - 40%
- 41% - 50%

36% Complete

*85. Have you EVER been advised to engage your child/children in physiotherapy?
- Yes
- No

*86. Have you EVER consulted a physiotherapist in regards to your child/children for SMA Type II?
- Yes
- No

40% Complete Physiotherapist Questions for Parents/Guardians

*87. What was the exercise advice your child was given from their physiotherapist?

*88. Complete the following statement: In my opinion, MOST of my child's/children's physiotherapists' had the attitude that if my child/children exercise it would...

*89. In your opinion, has your child/children EVER been treated by a physiotherapist who had a different attitude towards exercise as a therapy for SMA Type II compared to that of the majority of physiotherapists that you have had contact with?
- Yes
- No
90. In your opinion what was the physiotherapist's attitude towards exercise as a therapy for SMA Type II?

91. In your opinion, of all the physiotherapists that you have been in contact with what percentage of physiotherapists had this attitude towards exercise?

- Less than 10%
- 10% - 20%
- 21% - 30%
- 31% - 40%
- 41% - 50%

92. Do you CURRENTLY incorporate physiotherapy into your child's/children's treatment plan?

- Yes
- No

93. How often do your child/children engage in physiotherapy?

94. What type of treatment are you consulting the physiotherapist for? i.e. pain management, manual manipulation, mobility aids, exercise.

95. Have you PREVIOUSLY incorporated physiotherapy into your child's/children's treatment plan?

- Yes
- No
### Initial Survey: Australian Spinal Muscular Atrophy (SMA) Type II

**52% Complete**

96. How often did your child engage in physiotherapy and why did you stop?

97. What type of treatment were you consulting the physiotherapist for? i.e. pain management, manual manipulation, mobility aids, exercise.

### Initial Survey: Australian Spinal Muscular Atrophy (SMA) Type II

**56% Complete Accredited Exercise Physiologist Questions for Parents/Guardians**

98. Have you EVER been advised to consult an accredited exercise physiologist for your child/children?
- Yes
- No

99. Have you EVER consulted an accredited exercise physiologist in regards to your child/children?
- Yes
- No

### Initial Survey: Australian Spinal Muscular Atrophy (SMA) Type II

**60% Complete**

100. In your opinion what was the exercise physiologist’s attitude towards exercise as a therapy for SMA Type II?

101. What type of exercise was prescribed for your child/children by the exercise physiologist?

### Initial Survey: Australian Spinal Muscular Atrophy (SMA) Type II

**64% Complete Others Advice and Your opinions for Parents/Guardians**

102. Has any one else advised you to engage your child/children in exercise as a therapy for SMA Type II?
- Yes
- No
### Initial Survey: Australian Spinal Muscular Atrophy (SMA) Type II

**68% Complete**

**103. Who advised you to engage your child/children in exercise as a therapy for SMA Type II?**
*(Please tick all of the boxes that apply to you.)*

- [ ] Another individual with a neuromuscular disease or a member of their family
- [ ] A relative or friend
- [ ] Information collected on the internet
- [ ] Occupational Therapist
- [ ] SMA Association of Australia
- [ ] Muscular Dystrophy Foundation
- [ ] Other

Other (please specify):

**104. What is YOUR opinion on exercise as a form of treatment for SMA Type II?**

- [ ] It causes great harm in treating SMA Type II.
- [ ] It causes some harm in treating SMA Type II.
- [ ] It is slightly harmful in treating SMA Type II.
- [ ] It causes harm however benefits are gained in treating SMA Type II.
- [ ] It is neither harmful nor beneficial in treating SMA Type II.
- [ ] It is only slightly beneficial in treating SMA Type II.
- [ ] It has some benefits in treating SMA Type II.
- [ ] It is of great benefit in treating SMA Type II.

- [ ] Other/I would like to expand upon my answer.

Other (please specify):
Initial Survey: Australian Spinal Muscular Atrophy (SMA) Type II

105. What are YOUR personal feelings regarding exercise, physical activity and sport?
- I don’t have any personal feelings regarding exercise.
- I don’t like exercise therefore I don’t engage in any physical activity.
- I don’t like to exercise however I participate in physical activity as I feel it will benefit my health.
- I enjoy exercise but I feel that I cannot participate in physical activity due to other commitments.
- I enjoy exercise and I participate in physical activity whenever the opportunity presents itself.
- Other: I would like to expand upon my answer.

Other (please specify)

106. What is YOUR CHILD’S/CHILDREN’S opinion on exercise as a form of treatment for SMA Type II?
- To the best of my knowledge my child does not have an opinion on exercise.
- It causes great harm in treating SMA Type II.
- It causes some harm in treating SMA Type II.
- It is slightly harmful in treating SMA Type II.
- It causes harm however benefits are gained in treating SMA Type II.
- It is neither harmful nor beneficial in treating SMA Type II.
- It is only slightly beneficial in treating SMA Type II.
- It has some benefits in treating SMA Type II.
- It is of great benefit in treating SMA Type II.
- Other: I would like to expand upon my answer.

Other (please specify)
107. What are YOUR CHILD’S/CHILDREN’S feelings regarding exercise, physical activity and sport?

- My child/children don’t have any personal feelings regarding exercise.
- My child/children don’t like exercise therefore they don’t engage in any physical activity.
- My child/children don’t like exercise however they participate in physical activity as I feel it will benefit their health.
- My child/children don’t like exercise however they participate in physical activity as they feel it will benefit their health.
- My child/children enjoy exercise but feel that they cannot participate in physical activity due to their health.
- My child/children enjoy exercise and participate in physical activity whenever the opportunity presents itself.
- Other! I would like to expand upon my answer.

Other (please specify):

108. The following questions are all requesting your opinion:

- Do you believe it is a waste of time to engage in exercise to treat SMA Type II?
- Does your child believe it is a waste of time to engage in exercise to treat SMA Type II?
- Do you believe that if your child/children engage in exercise it will result in an injury caused by overexertion?
- Do you believe that your child/children have a higher rate of injury when engaging in exercise compared to the general population?
- Do you believe that your child/children should not engage in exercise if they are experiencing pain?
- Do you believe that if your child/children engage in exercise it will increase the rate of deterioration in their body?
- Do you think that if your child/children engage in exercise it will increase their level of tiredness throughout that day?
- Do you believe that your child/children should stop exercising if they are experiencing pain?
**Initial Survey: Australian Spinal Muscular Atrophy (SMA) Type II**

76% Complete Your opinion questions for Parent/Guardians page 2/3

*109. The following questions are all requesting your opinion:

<table>
<thead>
<tr>
<th>Do you believe that exercise can affect your child/children's mood?</th>
<th>Strongly Disagree</th>
<th>Disagree</th>
<th>Neutral</th>
<th>Agree</th>
<th>Strongly Agree</th>
</tr>
</thead>
<tbody>
<tr>
<td>Do you believe that if your child/children engage in exercise it will decrease the rate of deterioration in their body?</td>
<td></td>
<td></td>
<td></td>
<td></td>
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</tr>
<tr>
<td>Do you believe that exercise will decrease the level of pain your child/children experience?</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Do you think exercise will reduce your child/children's ability to move throughout that day?</td>
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<tr>
<td>Do you think exercise can improve your child/children's energy levels?</td>
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</tr>
<tr>
<td>Do you believe that exercise will increase the level of pain your child/children experience?</td>
<td></td>
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</table>
110. The following questions are all requesting your opinion:

<table>
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<tr>
<th>Question</th>
<th>Strongly Disagree</th>
<th>Disagree</th>
<th>Neutral</th>
<th>Agree</th>
<th>Strongly Agree</th>
</tr>
</thead>
<tbody>
<tr>
<td>Do you think exercise can improve the strength of your child's/children's muscles?</td>
<td>⬜</td>
<td></td>
<td></td>
<td>⬜</td>
<td>⬜</td>
</tr>
<tr>
<td>Do you think exercise can increase the size of your child's/children's muscles?</td>
<td>⬜</td>
<td></td>
<td></td>
<td>⬜</td>
<td>⬜</td>
</tr>
<tr>
<td>Do you think exercise can reduce the amount of fat stored in your child's/children's body?</td>
<td>⬜</td>
<td></td>
<td></td>
<td>⬜</td>
<td>⬜</td>
</tr>
<tr>
<td>Do you think exercise can improve your child's/children's ability to move their body whilst performing activities in their daily life?</td>
<td>⬜</td>
<td></td>
<td></td>
<td>⬜</td>
<td>⬜</td>
</tr>
<tr>
<td>Do you think exercise can improve your child's/children's ability to breathe?</td>
<td>⬜</td>
<td></td>
<td></td>
<td>⬜</td>
<td>⬜</td>
</tr>
<tr>
<td>Do you think exercise can improve your child's/children's ability to cope with their condition?</td>
<td>⬜</td>
<td></td>
<td></td>
<td>⬜</td>
<td>⬜</td>
</tr>
<tr>
<td>Do you think exercise can improve the overall quality of your child's/children's life?</td>
<td>⬜</td>
<td></td>
<td></td>
<td>⬜</td>
<td>⬜</td>
</tr>
<tr>
<td>Do you think exercise can increase your child's/children's lifespan?</td>
<td>⬜</td>
<td></td>
<td></td>
<td>⬜</td>
<td>⬜</td>
</tr>
<tr>
<td>Do you think that if your child/children engage in exercise or sport it can create the opportunity for them to meet new people?</td>
<td>⬜</td>
<td></td>
<td></td>
<td>⬜</td>
<td>⬜</td>
</tr>
<tr>
<td>Do you think that if your child/children engage in exercise or sport it can create the opportunity to expand their social life?</td>
<td>⬜</td>
<td></td>
<td></td>
<td>⬜</td>
<td>⬜</td>
</tr>
</tbody>
</table>

111. Has your child/children EVER engaged in exercise, physical activity or sport?

- Yes
- No
**112. What types of exercise/physical activity/sport have you child/children engaged in over their lifetime/s?**
(Please tick ALL of the boxes that apply to your child/children.)

- [ ] Swimming
- [ ] Weight Training
- [ ] Yoga
- [ ] Bocce
- [ ] Archery
- [ ] Wheelchair Basketball
- [ ] Wheelchair soccer
- [ ] Sitting Volleyball
- [ ] Wheelchair Tennis
- [ ] Other

Other (please specify):

---

**113. Do your child/children CURRENTLY engage in exercise, physical activity or sport?**

- [ ] Yes
- [ ] No
### Initial Survey: Australian Spinal Muscular Atrophy (SMA) Type II

**114. What type of exercise/physical activity/ sport does your child/children CURRENTLY engaged in?**

(Please tick ALL of the boxes that apply to your child/children.)

- [ ] My child/children do not currently engage in any physical activity.
- [ ] Swimming
- [ ] Weight Training
- [ ] Yoga
- [ ] Boccia
- [ ] Archery
- [ ] Wheelchair Basketball
- [ ] Wheelchair soccer
- [ ] Sitting Volleyball
- [ ] Wheelchair Tennis
- [ ] Other

Other (please specify)

**115. How long does it take your child/children to recover after engaging in exercise?**

- [ ] Little to no time
- [ ] A short time
- [ ] Some time
- [ ] A long time
- [ ] Too long

**116. Does your child/children CURRENTLY experience pain when they engage in exercise?**

- [ ] Never
- [ ] Occasionally
- [ ] Sometimes
- [ ] Usually
- [ ] Always
**Initial Survey: Australian Spinal Muscular Atrophy (SMA) Type II**

**117. Has your child/children EVER experienced pain when they engage in exercise?**

- Never
- Occasionally
- Sometimes
- Usually
- Always

**118. Are there any types of exercise that YOU would NOT feel comfortable engaging your child/children in?**

*(Please tick ALL of the boxes that apply to your child/children.)*

- I feel comfortable engaging my child/children in all types of exercise
- Swimming
- Weight Training
- Yoga
- Bocci
- Archery
- Wheelchair Basketball
- Wheelchair soccer
- Sitting Volleyball
- Wheelchair Tennis
- Other

Other (please specify)
Initial Survey: Australian Spinal Muscular Atrophy (SMA) Type II

*119. Are there any types of exercise that YOUR CHILD/CHILDREN would NOT feel comfortable engaging in?
(Please tick ALL of the boxes that apply to your child/children.)

- My child/children feel comfortable engaging in all types of exercise
- Swimming
- Weight Training
- Yoga
- Boccia
- Archery
- Wheelchair Basketball
- Wheelchair soccer
- Sitting Volleyball
- Wheelchair Tennis
- Other

Other (please specify)

*120. Has your child/children EVER engaged in exercise for social reasons?

- Yes
- No

If you would like to expand upon your answer please describe

*121. Has your child/children EVER engaged in exercise using weights?

- Yes
- No
### 122. In total how many minutes does your child/children CURRENTLY exercise for each week?

- Not at all
- Less than 30 mins
- 30-60 mins
- 61-90 mins
- 91-120 mins
- 121-150 mins
- More than 150 mins

### 123. How many days per week does your child/children CURRENTLY exercise for?

- Not at all
- 1 day/week
- 2 days/week
- 3 days/week
- 4 days/week
- 5 days/week
- 6 days/week
- 7 days/week

### 124. What was the MAXIMUM amount of minutes your child/children have EVER exercised for within one week?

- Not at all
- Less than 30 mins
- 30-60 mins
- 61-90 mins
- 91-120 mins
- 121-150 mins
- More than 150 mins
### Initial Survey: Australian Spinal Muscular Atrophy (SMA) Type II

**125. What is the MAXIMUM number of days per week your child/children have EVER exercised for?**

- Not at all
- 1 day/week
- 2 days/week
- 3 days/week
- 4 days/week
- 5 days/week
- 6 days/week
- 7 days/week

**126. How long have you incorporated exercise into your child’s/children’s treatment plan?**

- My child/children don’t engage in any exercise
- My child/children have previously incorporated exercise however they no longer engage in physical activity
- Since my child/children was diagnosed with SMA Type II
- For less than 1 year
- For the past 1-2 years
- For the past 3-4 years
- For the past 5-6 years
- For more than 6 years
- Other/I would like to expand upon my answer

**127. What drives YOU to engage your child/children in exercise?**

Some examples are: To lose excess weight your child has accumulated; To strengthen your child’s body; To slow the progression of SMA Type II; To engage your child in social interactions with others.

**128. What drives YOUR CHILD/CHILDREN to engage in exercise?**

**129. Are you amenable to the possibility of the researcher of this study contacting you for a private interview within the next 6 months?**

- Yes
- No
**Initial Survey: Australian Spinal Muscular Atrophy (SMA) Type II**

### 98% Complete Requested Information for Private Interviews

130. Please complete the following information.

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<td>Name</td>
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**Initial Survey: Australian Spinal Muscular Atrophy (SMA) Type II**

**Thank-you for completing this survey!**

The researchers of this study would like to thank-you for your time and commitment in completing this survey.

A summary of the results of this survey and interviews conducted over the next year will be posted on the SMA Association of Australia webpage at the end of 2014.

The web address for SMA Association of Australia is: www.smaaustralia.org.au
Appendix Six: Research invitation letter for online questionnaire

Dear Members of the SMA Community,

I would like to invite all individuals with SMA Type II and any parents or guardians of individuals with SMA Type II to be a part of our research into exercise as a therapy from your perspective as an individual with SMA Type II. This research endeavours to establish the views and opinions of the Australian SMA Type II community regarding exercise as a therapy. The scope of the research covers whether or not you engage in exercise, the type and duration of exercise you participate in, opinions of the health professionals you have interacted with and your opinion of the benefits and risks associated with engaging in exercise. The views and opinions of all individuals with SMA Type II is valuable to this research project regardless of whether or not you engage in exercise.

If you would like to participate the online survey is anonymous and available from 02/03/15-12/04/15 at https://www.surveymonkey.com/s/AustralianSMAPerspective

Your consent to be a part of this research including the publication of the survey’s results will be obtained through your completion of the online survey. Participation in the survey is voluntary and you are able to withdraw from the survey at any time if you are not comfortable.

The second component of this research involves a private interview with the researcher regarding your views and opinions on exercise as a therapy. If you are interested in participating in an interview there will be an option to provide your contact details at the end of the online survey.

The final component of this research offers those individuals who participated in a private interview an opportunity to also participate in a group interview, if they so choose, which will include the researcher and other individuals who have participated in this research project.

The online survey should take less than 1 hour to complete.

If you would like to participate or ask any questions about this exciting research please contact;

Marissa Hoey (Masters researcher)

Phone: 0405 113 261 or email Marissa.Hoeysresearch.usc.edu.au

The research team thanks you for being involved.
Appendix Seven: In-person interview questions

Hi -------------------------,

It’s lovely to meet you. Before we get started I just want to provide you with a contact information sheet and go through the informed consent form.

CONSENT FORM
CONTACT INFORMATION SHEET

So I just want to remind you that participating in this interview is voluntary and you can withdraw at any stage, without providing an explanation to me and without any consequences. The results from this interview will be kept confidential and any statements you make to me will be de-identified.

The focus of this interview will be to investigate the current and previous exercise programs that you have engaged in and to find out your perceptions on the benefits, risks and effectiveness of each exercise. I will also be asking you about your views on exercise recommendations, sport participation, sedentary behaviour and the care provided by your doctor.

This interview will be recorded and a transcript of the interviews will be typed up from the recording. I will provide you with a typed transcript and you will be able to make changes to any statements you have made before I write my thesis.

We can stop or take a break at any time. There may be questions that I ask you during the interview which may unintentionally challenge you, cause you upset or remind you of a painful moment. Please don’t hesitate to tell me if you feel uncomfortable during the interview process and remember that you can choose, at any time, not to answer any questions that I ask you or to withdraw from this study. Should you experience any concerns from the interviews, you may wish to contact the support services at USC and that information has been provided on the contact sheet.

The reason that I am conducting this study is because there is very little research on SMA Type II outside of the genetics of the condition and drug or genetic therapies. I’ve only found a handful of studies that investigate the effect of exercise on individuals who have SMA Type II and I think that for many people who have a rare condition sometimes it is the person/parents of the person who has the condition who become the experts on that condition rather than the health professionals. What I want to do is to explore your expertise and share your knowledge with other parents and individuals with SMA Type II so that their journey can be a little bit easier. And I also want to provide a resource for health professionals that they can reference if they start to see a client with SMA Type II because there is nothing specific that I have found that can guide a health professional to design an exercise program for a
individuals with SMA Type II. I think that my views have been shaped by what I’ve seen and heard as a support worker and I think that I have been very lucky in that my first clients were both advocates for person centered care which has had a significant impact on who I am as a person and as well as a support worker.

So I want to start the interview today by asking you

Why is exercise important to you?

Can you tell me about the health professionals you have engaged about exercise? (personal trainer, exercise physiologist, physiotherapist, doctor)
Are you exercising under your own guidelines or are you following an exercise program set out by a health professional?

Can you tell me about each type of exercise you have tried throughout your/your child’s life?
What do you believe were the benefits, limitations and risks of each exercise program?
What were the parameters surrounding each exercise? Why did you stop each exercise program? (progression, boredom, program caused harm)

Can you tell me about any barriers you have experienced to your/your child’s engagement in exercise?

Do you think that exercise has changed the rate of the progression of SMA Type II in you/your child?
What makes you believe this?

Are there any specific exercises that you believe will enhance your/your child’s respiratory function? Are there any specific exercises that you believe reduce your/your child’s respiratory function?
Why do you believe this?

Are there any specific exercises that you believe will enhance your/your child’s muscular strength? Are there any specific exercises that you believe reduce your/your child’s muscular strength?
Why do you believe this?

Are there any specific exercises that you believe enhances your/your child’s ability to move? Are there any specific exercises that you believe reduces your/your child’s ability to move?
Why do you believe this?
Do you think that exercise has had an effect upon the quality of your/your child’s life?
   In what ways?
   Why do you believe this?

What other areas, like nutrition or medications, have you incorporated into your
   treatment plan?

Of all the areas that you think are important to treat SMA Type II, where would you
   rank exercise on a scale of most to least important of treatment factors?
   On a scale of 1-10 how important is exercise to you in your daily life?

Do you think that exercise will have an effect upon your lifespan? Why do you
   believe this?
In the online questionnaire you stated that you/your child … has additional conditions
   /does not have any other conditions… apart from SMA Type II. Can I please confirm
   with you that you/your child don’t have any of the following conditions commonly
   associated with SMA Type II.
   Respiratory conditions i.e. sleep disordered breathing, recurrent chest
   infections, impaired cough, underdeveloped lungs or rib cage;
   Orthopedic conditions i.e. joint contracture, scoliosis, hip subluxation,
   osteopenia, fractures;
   Gastrointestinal and Nutrition conditions i.e. feeding and swallowing
   problems, gastrointestinal dysfunction, aspiration, gastroesophageal
   reflux, under/over nourished.

Do you think that the exercise recommendations for the general population are
   relevant for you/your child? Is it achievable for you/your child?

There has been a lot of focus, particularly in recent years, about sedentary behavior
   and that being sedentary has a significant impact upon health. I think that this topic
   has been promoted a lot in schools, in the news, in ads like the swap it ads and be fit
   for life. So my question is:
As an individual who has a condition that limits your mobility is the impact of
   sedentary behaviour on your health something that you have thought about?
As a parent have you thought about the impact that sedentary behaviour will have on
   your child’s health as they have a condition that limits their mobility?
   How does it make you feel when the topic of sedentary behaviour is
   being discussed in a public?
   Has it had an impact on you in any way?
Do you think standing frames and standing wheelchairs could improve your health?
(Standing is minimal activity compared to sitting which is no activity the difference could also impact on other health areas such as bone mineral density, hip dysplasia etc.)
Have you ever been referred for a standing wheelchair?
Would you ever considered getting a standing wheelchair?

Several individuals indicated in the online questionnaire that either they or their child had participated in school PE. What has your/your child’s experience of school PE been?
Do you have any suggestions for any improvements that need to be made?
Have you had any teachers who have successfully implemented PE for you/your child?

What are your thoughts on inclusive and segregated sport? How much access do you/your child have to participate in sport?

Do you think there are differences in care if you are living in a rural setting compared to a city setting, or between states, or between countries?
How do you feel about the level of care you/your child are receiving?
How far do you have to travel to receive care/specialist care?

Do you believe your primary care doctor (family doctor) is well informed about SMA Type II?
Have they informed you about clinical trial treatments for SMA Type II or directed you to the clinical trial website?
Has your doctor formulated a plan of multidisciplinary intervention with you?
Does your primary doctor play a central role in coordinating follow up multidisciplinary care?
Has your primary care doctor referred you to any of the following: Neuromuscular clinic (pediatric), Geneticist, Neurologist, Pulmonary, Gastroenterology or Nutrition, and orthopedic/rehabilitation?

Only 40% of people who participated in the online questionnaire stated that their doctor had ever discussed exercise as a therapy for SMA Type II. How do you feel about this statistic?
How do you feel about the level of care you have received from your primary doctor?

What has/have your doctor/s told you about SMA?
About life expectancy and prognosis?
What do you think about what the doctor/s have told you?
Do you feel that more information about SMA Type II needs to be provided to you and your parents/needs to be provided to you as a parent?

Do you feel ... your parents needed/you need... more information about how to raise a child with SMA Type II?

All three organisations that I contacted to distribute the online questionnaire raised concerns that not everyone would know what type of SMA they have been diagnosed with. Do you think their concerns are valid?

What do you think about this?

Is there any information, tips or tricks that you would like to share with the wider SMA Community?

That’s it. Thank-you so much for taking the time to participate in this research. I really appreciate you opening up your life to me and divulging such personal things about yourself and your family. If you think of anything that you wanted to add to what we’ve discussed today please don’t hesitate in contacting me, email is probably the best way to get a hold of me particularly over the next few months while I’m conducting interviews. I will email you a copy of the transcript once it’s typed up – that may take me a few weeks or a few months but if you want to read over that when you receive it and if there are any changes you want me to make to any comments you’ve made today I can do that. And if you experience any concerns from today don’t forget that you can contact the support services at USC OR the Human Research Ethics Committee at the Office of Research. Both of their details have been provided on the contact sheet I gave you.
Appendix Eight: Research invitation letter for in-person interviews

Invitation to individuals with SMA Type II and any parents or guardians of individuals with SMA Type II who engage in exercise to be a part of our research into exercise as a therapy from your perspective as an individual with SMA Type II. This research is focused on exploring past and current exercise programs undertaken by you and the benefits, risks and limitations you perceive each exercise program has. The information collected in these interviews will determine the most beneficial type and duration of exercise from your perspective as individuals with SMA Type II. Furthermore it will compare whether individuals who engage in exercise are stronger and more active than the current literature suggests. Additionally the interview will explore your perceptions of exercise recommendations, sport participation, sedentary behavior and the care provided by your doctor.

You have indicated in an online survey that you engage in exercise and would be happy to be contacted for further research.

If you would like to participate, please read the Research Project Information Sheet and confirm via email a convenient time for your private interview. If you decide to participate in a private interview you will also be invited to participate in a group interview with other participants to further discuss your views on exercise. It is at your discretion if you would like to participate in the private interview only or if you would like to participate in both the private and group interviews. It may be helpful before the private interview to contact any relevant members of your health professional team and arrange copies of any structured exercise programs you have engaged in to bring to the interview.

If you have indicated that you are willing to share the results of your genetic test regarding your SMA diagnosis could you please bring a copy of this test with you to the private interview. The researchers of this project are only interested in determining the number of SMN 2 genes you have as this impacts upon SMA severity.

The interview will last approximately 2 hours.

If you would like to participate or ask any questions about this research please contact;

Marissa Hoey (Masters researcher)

Phone: 0405 113 261 or email Marissa.Hoey@research.usc.edu.au

The research team thanks you for being involved.
**Appendix Nine: Halliwick method ten-point-program**

<table>
<thead>
<tr>
<th>Point</th>
<th>Topic</th>
<th>Description</th>
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<tbody>
<tr>
<td>1</td>
<td>Mental adjustment</td>
<td>Learn to react appropriately to water. Of great importance is adjustment to fluidmechanics (buoyancy, flow conditions, waves). Breath control is also an important topic in this point.</td>
</tr>
<tr>
<td>2</td>
<td>Sagittal rotation control</td>
<td>The ability to control movements with left-right components around the sagittal axis of the body, especially in upright situations.</td>
</tr>
<tr>
<td>3</td>
<td>Transversal rotation control</td>
<td>The ability to control movements around a transverse axis of the body (with flexion-extension components) e.g. lying down, standing up, rocking in a chair position.</td>
</tr>
<tr>
<td>4</td>
<td>Longitudinal rotation control</td>
<td>The ability to control movements around a longitudinal axis of the body. Especially important in supine: rolling over from supine to supine. Therapeutically, a counter-rotation is of higher importance.</td>
</tr>
<tr>
<td>5</td>
<td>Combined rotation control</td>
<td>The ability to control a “corkscrew” movement around a combination of the previous axes e.g. rotating to supine while falling forward or when losing lateral stability.</td>
</tr>
<tr>
<td>6</td>
<td>Upthrust/mental inversion</td>
<td>The client should understand that the water supports and that he/she does not sink.</td>
</tr>
<tr>
<td>7</td>
<td>Balance in stillness</td>
<td>Maintaining a position in a stable and relaxed way without compensatory movements of arms or legs e.g. stand, chair position, oblique, supine. This point focuses on efficient an effective postural control.</td>
</tr>
<tr>
<td>8</td>
<td>Turbulent gliding</td>
<td>The ability to glide in the water in the wake of the instructor, who walks backwards. The client has to control unwanted movements with head and trunk.</td>
</tr>
<tr>
<td>9</td>
<td>Simple progression</td>
<td>A small swimming movement with the hands as a preparation for a real propulsive activity. Important is to have automatic trunk control.</td>
</tr>
<tr>
<td>10</td>
<td>Basic Halliwick movement</td>
<td>A propulsive swimming movement with the arms (rowing). Individual adaptation because of impairment is allowed.</td>
</tr>
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Appendix Ten: The Royal Children’s Hospital Melbourne hydrotherapy program for children living with NMD

Hydrotherapy for Children with Neuromuscular Disorders (NMD)

Hydrotherapy is a physiotherapy treatment carried out in water where movement against the weight of the water helps exercise muscles and floating is made easier because of the buoyancy of the water.

The main aim of a hydrotherapy or swimming program for all children is to learn water safety and how to swim. There are additional benefits for children with NMD, which makes swimming a fabulous sport to encourage:

- Water allows a freedom of movement that is not always possible on land. This is due to the buoyancy effect of water which assists movement.
- The positive effects of exercise which result in increased stamina, endurance, fitness and weight control.
- Breath holding and increased fitness improves lung function.
- The warm environment in a hydrotherapy pool can help when stretching muscles.
- Helps to build self-esteem.
- Social contact with other children.
- Learning to swim is fun!

Issues to consider:
- Exercise can make you tired so don’t over do it — allow your child to rest when they need to.
- There is a risk of getting a cold or chest infection.
- A child’s ability to float may change as their disease progresses.
- The program may need to be modified as weakness increases and contractures develop.
- Safety is paramount as children’s muscles get weaker.

Exercises and activities in the water

Warm up
- Walking — forwards, backwards, sideways (the deeper the water and the faster you walk, the harder you have to work). Depending on the depth of water at the pool you are using you can start in shallower water and then progress to deeper water.

Stretches
- Hamstring and calf stretch — hold onto edge, place feet on wall, try and get heels down and straighten knees.
- Back and hip stretch — hold onto wall, place feet on wall and curl up into a ball.
- Shoulder and thigh (parachute) — back to wall, stretch hands back to hold onto wall, place front surface of lower leg on the wall of the pool, and push tummy out.
- If you have trouble keeping hold of the edge of the pool in the previous three stretches, you may need an adult to stand behind you and help.
- In the previous three positions see if you can walk around the edge of the pool while still holding on.
- Elbows — hold on to the side of the pool, straighten elbows as far as possible and lean back. You may feel a stretch around your elbows.
- Hands — stand facing the wall, place a flat hand on the wall of the pool, straighten elbow until you feel a stretch in your hand and wrist.
Appendices

You should hold all stretches for at least 30 seconds and do each one five times.

Breathing Games

- Blowing bubbles — above and below the water.
- Blowing light floating toys across the water.
- Breath holding under water — e.g. retrieving sinkers from bottom of pool, swimming through hoops, swimming through someone’s legs (may need help from an adult to swim to bottom of pool).

Balance is stillness

- Balance on a float — e.g. noodle between legs (horse ride), noodle/s under bottom, or sitting on kick board. To make these activities harder encourage movement, i.e. sculling with arms or kicking with legs while balancing on float.
- Floating: — on back: like a starfish, a soldier — add movement: use arms to scull or legs to kick

- Make a whirlpool (walk around in a circle in one direction) and then stop and try to stand still.
- Make a whirlpool walking in one direction, then turn around and walk in opposite direction.
- Move from floating on your back to standing.
- Somersaults in the water (may need help from adult when first attempting this).
- Mushrooms — take a big breath, swim to bottom of pool, hug knees to chest and float to top.

Note

Despite the assistance that water buoyancy provides, children with NMD who have weak neck muscles may find floating on their back difficult. A float positioned around the neck can be useful if this is the case.

Should you have any problems or need any advice regarding a hydrotherapy or swimming program, please contact your local physiotherapist. If your child’s disease has progressed and his/her ability to move has changed it is important to get their hydrotherapy/swimming program reviewed by his/her local physiotherapist.

The Royal Children’s Hospital Melbourne