UNIVERSITEIT VAN PRETORIA
UNIVERSITY OF PRETORIA
YUNIBESITHI YA PRETORIA

Exploring the lived experiences of parents raising a child diagnosed with Duchenne Muscular Dystrophy (DMD) in South Africa: Challenges and coping strategies

Casey-Garnett Forman

12289681

Supervisor: Dr. Linda M Eskell Blokland

Dissertation submitted in partial fulfilment of the requirements for the degree of Master of Arts in Counselling Psychology in the Faculty of Humanities at the University of Pretoria, South Africa, in August 2020.

Declaration

I, Casey-Garnett Forman, declare that the dissertation *'Exploring the lived experiences of parents raising a child diagnosed with Duchenne Muscular Dystrophy (DMD) in South Africa: Challenges and Coping Strategies'* is my own academic work and has no previous submissions for a degree by me at any other institution. All citations have been acknowledged and are deemed to be complete and reliable.

Casey-Garnett Forman

Date: 26/08/2020

Acknowledgements

To the participants in this study, thank you for your genuine warmth and honesty. You have contributed immeasurably to the findings in this study and you have ignited a flame in me to continue raising awareness.

To my supervisor, Dr. Linda Blokland, thank you for sharing your knowledge and insights with me.

To my editor, Noleen Loubser, thank you for going the extra mile for me and for all your encouragement, insight and knowledge.

To my mother, thank you for always having my best interests at heart and for giving me the time and space to write this. Your constant support has been invaluable.

Dedication

I dedicate this research, with deep gratitude, admiration and love, to my mother, who was the inspiration for this study, and without whom, this dream of mine would not have been possible.

Abstract

This study set out to explore the lived experiences of parents raising a child with Duchenne Muscular Dystrophy (DMD) in South Africa. It addressed the challenges and coping strategies faced by three married couples of children, specifically male children, who are affected by this degenerative and fatal genetic condition. Purposive sampling was used and themes were highlighted through semi-structured interviews, employing interpretative phenomenological analysis (IPA).

Major themes found included the reactions to the diagnosis, facing many losses and experiencing anticipatory grief, and learning to adapt. It was found that parents tended to vacillate between hope, grief, avoidance and presence, depending on their perceived level of support, contextual factors, and social and economic challenges. It was important for the parents to plan in advance and gain practical information about the illness in order to implement the necessary changes. The adjustment process was shown to be complicated. The final theme entailed what it meant for each family to create a meaningful life beyond the loss, and to remain hopeful about what lies ahead.

It was concluded that amidst the unique and far-ranging challenges experienced on a daily basis, the parents started making changes that have had a positive impact on the lives of their sons and the entire family. These parents have found ways of cultivating meaning and hope in their everyday lives, doing everything in their power to grant their sons fulfilling lives.

The findings support the need to incorporate strategies into existing services and health promotion programmes to build on the parents' support structures so that they are able to adapt to the challenges they face on a regular basis. Further

research into the disease impact in a South African context is necessary to improve care provision and inform policy.

Keywords: Duchenne Muscular Dystrophy (DMD); degenerative; fatal; genetic condition; male children; parents; lived experiences; challenges; coping strategies.

Abbreviations

BiPAP	Bi-level Positive Airway Pressure
DMD	Duchenne Muscular Dystrophy
HMV	Home Mechanical Ventilation
IPA	Interpretative Phenomenological Analysis
MDFSA	Muscular Dystrophy Foundation of South Africa
NMD	Neuromuscular Disease
SA	South Africa
UK	United Kingdom
USA	United States of America

Contents

List of Tables

Chapter One: Background and Rationale

1.1 Introduction

Over the past few decades, there has been a significant increase in the number of studies conducted on the challenges experienced by parents raising a child with a chronic or rare disease (Alexander et al., 2018; Cornelio, Nayak, & George, 2016; Nicholas et al., 2009; Pourghaznein, Heydari, Manzari, & ValizadehZare, 2018; Ringnér, Jansson, & Graneheim, 2011). While there is a developing body of literature surrounding the challenges faced by these parents within a global context, Samson et al (2009) found that the coping mechanisms of parents have scarcely been explored. Additionally, significant gaps continue to exist in rare disease discourses in South Africa (Ross, 2016).

There is currently no universal definition of a rare disease or a chronic disease. The use of inconsistent definitions and the variation in public information often results in inconsistencies in access to treatment and exclusion from mainstream society and poses serious implications for parents managing a chronic or rare disease (Richter et al., 2015; World Health Organisation, 2012).

A rare disease is generally referred to as a chronic health condition that affects a small number of people compared with other prevalent diseases in the general population, and may be life-threatening (Richter et al., 2015, p. 907; WHO, 2012). The more commonly diagnosed chronic diseases include cardiovascular diseases, cancer, chronic respiratory diseases, and diabetes. Genetic disorders may or may not be considered to be rare, depending on their prevalence, but they are considered to be chronic diseases, contributing significantly 'to the burden of disease on individuals, families, societies and countries' (World Health Organization, & Public Health Agency of Canada, 2005, p. 35).

As will be discussed in the literature review, a wealth of information exists on the undeniable impact of DMD on the parents, other family members and family functioning as a whole – both positive and negative. It has been suggested in the literature that a reciprocal relationship exists between the son's illness and parental functioning. Furthermore, stressors include the treatment and management of the child's illness through requirements for care assistance, consistent supervision or guidance and an increase in day-to-day caregiving requirements, the complexities of disease management activities, long-term treatments and periods of exacerbation that often lead to prolonged hospitalisations, as well as the associated financial, emotional, physical and psychological adjustments (Brown et al., 2008). Additionally, parents may experience role strains, marital conflict and separation, interruptions in daily routines and plans for the future, and uncertainty with regards to the child's prognosis – often leading to feeling overwhelmed, anxious or depressed (Youngblut, Brady, Brooten, & Thomas, 2000). These challenges are likely to affect family dynamics and may influence long-term health outcomes for the parents, the affected child, and the healthy siblings (Glover, Hendron, Taylor, & Long, 2020).

According to Richter et al. (2015), an increased focus on chronic and rare diseases has been spurred by legislation intended to facilitate patient access to effective treatments in the United States of America (USA) by 'incentivising pharmaceutical and biotechnology companies to develop new medicines that would otherwise not be profitable' (p. 907). The current surge in research studies could also be attributed to the growing concern about assisting parents in a more practical and supportive manner, due to the myriad of stressors that they experience on a daily basis (Alexander et al., 2018; Chen & Clark, 2007; Emery, 2003; Gilgoff, 1983;

Gravelle, 1997; Magliano, 2014; Parker, Maddocks, & Stern, 1999; Polita et al., 2018; Samson et al., 2009).

According to the World Health Organisation (WHO, 2012, p. 1),

> Rare diseases present fundamentally different challenges from those of more common diseases, such as asthma. This is most apparent during the clinical development stage when rarity significantly complicates the task. Problems include the small number of patients, the logistics involved in reaching widely dispersed patients, the lack of validated biomarkers and surrogate end-points, and limited clinical expertise and expert centres.

Additionally, medical expertise for a rare disease such as Duchenne Muscular Dystrophy is a scarce resource. It is therefore critical to address fragmented and inconsistent disease knowledge by investing in dedicated infrastructure and international networks such as registries and networks of expertise (WHO, 2012).

Muscular dystrophies are a heterogeneous group of genetic disorders that are characterised by progressive muscle weakness that affects mobility (Emery, 2003; WHO, 2012). There are differences in the various types of muscular dystrophy in the rate at which muscle weakness progresses and the muscle groups which are most severely affected (Emery, 2003). Duchenne Muscular Dystrophy is the most severe form of muscular dystrophy with a prevalence of one in 3500 male births regardless of race or ethnic origin (Emery, 2003; WHO, 2012). It is a degenerative disease, inherited as an X-linked recessive disorder caused by a mutation or a lack of the protein dystrophin. DMD causes progressive weakness by affecting all body muscles and leads to early death (Erby, Rushton, & Geller, 2006). Boys with DMD manifest initial signs and symptoms in preschool and early school years and boys are usually wheelchair-dependent by the age of two to twelve years (Erby et al.,

2006). Despite there being no cure, survival into adulthood has become more common due to improvements in interventions such as ventilation and cardiac care (Hoskin, 2017).

While external associations such as The Muscular Dystrophy Foundation of South Africa (MDFSA) exist to alleviate and assist with some of the emotional and practical challenges parents experience, Esterhuizen, Greenberg, Ballo, Goliath, and Wilmshurst (2016) have acknowledged that health professionals providing treatment in a hospital setting in South Africa often find it impossible to provide the parents and their affected children with the necessary emotional support due to resource constraints. In keeping with international protocols, the diagnosis in clinically suspected cases is confirmed by DNA analysis or a muscle biopsy, and clinic visits are scheduled every three to six months, where the families have access to genetic counselling and support from the MDFSA. Corticosteroid intervention is implemented at around four years old, or as soon as the boy's motor function is affected (Esterhuizen et al., 2016). Associate Professor Jo Wilmshurst, Head of Paediatric Neurology at the Red Cross Children's Hospital, acknowledges that while she oversees a few support groups at the clinic, there is often "…very little space to ask, how are you feeling?" due to the enormous pressure each health professional is under in the South African health care system (personal communication, January 9, 2020). Furthermore, the lived experiences of parents raising a child with Duchenne Muscular Dystrophy (DMD) within a South African context have been virtually unexplored.

In an annual report on rare diseases by the WHO (2012, p. 3), it has also been stated that further research needs to be conducted on:

- Networks of excellence that focus on research infrastructure as well as provision of disease-related information at EU level. These networks can be utilised to train health professionals on rare diseases;
- Patient and caregiver experiences, and that assessment methods are 'adapted to small and very small patient populations' (WHO, 2012, p. 3);
- Initiatives that focus on rare disease classification; and
- Fundamental research into the disease process to increase understanding of rare diseases.

To address these challenges, public funding of fundamental research into the disease process is necessary, both at the national and global level (WHO, 2012).

1.2 Problem Statement

Research has shown that during the course of DMD there will inevitably be many inherent stages that predispose an atmosphere of emotional turmoil and family strife as parents become consumed by their child's illness.

In response to the dearth of research in a South African setting around the complexity and diversity of the challenges and stressors affecting the adjustment process that parents face while raising a child with DMD, it seemed fitting to begin to address this gap of knowledge on their lived experiences within our South African context. Not only does this study have theoretical implications, providing descriptive experiences through the use of IPA and adding to the body of knowledge on this topic in South Africa as this is the first study on the lived experiences in SA, it also has practical implications. An additional perspective may be gained as to how parents cope with the challenges with which they are faced in a South African context. This perspective will appeal to individuals with DMD and their families, as well as a wide range of therapists and other professionals interested in

understanding the lived experiences of parents of children with DMD within a South African context. The current study could additionally have a more profound impact by benefiting other families who are raising a child with DMD in South Africa, and internationally. Furthermore, this information may benefit families raising a child with various other chronic or rare diseases. This may help them to manage the challenges they face more effectively.

1.3 Rationale

Various international researchers have explored some of the strategies parents employ to cope with raising children with a rare disease or chronic illness. Far fewer studies have been conducted in South Africa on how to manage DMD in a marginalised society. Much of the research conducted on DMD in South Africa to date has been quantitative (Esterhuizen et al., 2016) and has not explored the vast challenges parents are likely to encounter in a third-world country with limited resources to assist their children.

This research study was informed by an interpretive, phenomenological perspective (Smith & Osborn, 2007). The aim of this study was to explore the lived experiences of parents of children, diagnosed with Duchenne Muscular Dystrophy (DMD). It aimed to explore how these parents make sense of and give meaning to their personal world through their experiences that undoubtedly affect their lives, and the strategies they adopt to deal with these challenges. Themes were highlighted through semi-structured interviews with these parents, employing interpretative phenomenological analysis (IPA).

I believe that carrying out a qualitative study on the challenges and coping strategies of parents in South Africa is likely to benefit South African families dealing with this very demanding condition.

1.4 Research Questions

The primary research question was:

- How do parents experience raising a child with DMD?

The secondary research questions were:

- How did the parents perceive and react to the initial diagnosis of DMD?
- What are some of the challenges the parents have faced?
- What are some of their coping strategies?
- What kind of support have they received from their community?

1.5 Objectives

The objectives of this study were to:

- Explore the lived experiences of parents raising a child with Duchenne Muscular Dystrophy (DMD); and
- Identify the needs of parents who are raising a child within the South African context.

1.6 Organisation of the Dissertation

This dissertation consists of five chapters organised as follows:

Chapter One – Introduction: This chapter introduces the study by stating what the study is about and providing a context within which it exists. In addition, it provides the background for the study, the purpose statement, the problem statement, research questions and aims; as well as a brief explanation of the methodology used.

Chapter Two – Literature Review: This chapter offers a review of some of the relevant literature, situating the study within the South African context and discussing rare and chronic illness, the impact of the initial diagnosis, adjusting to a new way of

life, the different appraisals of DMD, challenges encountered and coping strategies adopted, depression, treatments used, palliative care, as well as a discussion on clinical practice, recent developments and socio-economic concerns in the South African context.

Chapter Three – Research Methodologies: This chapter engages with the methods used in the study, including the research design, selection of participants, procedure, and means of data analysis. Ethical issues and reflexivity are also discussed in this chapter.

Chapter Four – Results and Discussion: This chapter provides an in-depth analysis of the results and findings from the data collected in the study, and is concluded with a general discussion of the results and discussion. It is organized around four themes, including the 'reactions to the diagnosis' (encompassing the associated frustrations with the medical fraternity); 'grief and anticipatory loss' (the consistent losses faced by the parents); 'learning to adapt' (the appraisal of the illness, the importance of planning in advance, support versus isolation); and finally 'seeing beyond the illness'.

Chapter 5 – Conclusions and Recommendations: This chapter encompasses my reflections of the study and presents the findings, limitations, and conclusions, and makes recommendations for family assistance, clinical and therapeutic practice, and further research.

Chapter Two: Literature Review

2.1 Introduction

Muscular dystrophy (MD) is the name given to a group of more than seventy different genetic neuromuscular disorders causing progressive wasting and weakness of the muscles. There are differences in the various types of muscular dystrophy in the rate at which the muscle weakness progresses and the muscle groups which are most severely affected. Duchenne Muscular Dystrophy (DMD) is the most severe of the inherited dystrophies, predominantly affecting males, and necessitating specialised and time-consuming care from the child's birth, even though the terminal condition lies years ahead (Esterhuizen et al., 2016; Gravelle, 1997). Parents of children with Duchenne Muscular Dystrophy encounter numerous challenges in supporting their children throughout their lives. Some daily challenges include structural barriers to care, such as coordinating care across school, health and family settings. Parents are often concomitantly striving to manage their child's condition as well as their own health care needs, work pressures and family demands. Research has shown that attaining a work-life balance is an elusive concept for parents raising a child with a chronic illness such as DMD (Guite, Russell, Homan, Tepe, & Williams, 2018).

2.2 Duchenne Muscular Dystrophy

Muscular dystrophies are degenerative rare muscle diseases, and skeletal muscle is predominantly affected in DMD (Emery, 1987; 2003). With an estimated incidence of 1 in 3500 male births, DMD is considered a rare disease (WHO, 2012). DMD is an X-linked disease of the muscle. This means that it is a genetic condition passed from a mother to her son. It is caused by a mutation of the Xp21 gene, and

this gene encodes a rod-like cytoskeletal protein called dystrophin that affects boys who inherit the disease from their mothers (Emery, 1987; 2003). Females with a mutant X-chromosome will not be afflicted by DMD, but they will be carriers of the disorder. If the mother's mutant X-chromosome pairs with a Y-chromosome, thus producing a male child, the boy will have DMD (Chen & Clark, 2007). Research has shown that this accounts for 70 percent of DMD inheritances while another 30 percent is due to a genetic mutation in the child (Emery, 2003; Magliano, 2014).

DMD manifests during infancy but it is usually recognised between three and six years of age, and its symptoms include general muscle weakness and wasting affecting the pelvis, upper arms, upper legs, and eventually all voluntary muscles (Emery, 2003; Magliano, 2014). The majority of the research reveals that a child with DMD is usually non-ambulatory by ten to twelve years of age, and life expectancy does not usually exceed twenty years (Pascual, et al., 2019; Yamaguchi & Suzuki, 2015). Serious life-threatening complications may develop including disease of the heart muscle (cardiomyopathy); and breathing (respiratory) difficulties. Despite the fact that there is no cure for DMD, significant advances have been made in terms of DMD management over the years. Treatment includes the use of corticosteroids and the use of intermittent positive pressure ventilation, which improves quality of life, life expectancy and ambulation (Yiu & Kornberg, 2008). Well-documented side effects of corticosteroid treatment include behavioural and emotional problems (Biggar, Harris, Eliasoph, & Alman, 2006; Dubowitz, 2005). Additionally, providing care to a child with DMD on a daily basis includes assisting him with wheelchair transportation, and ongoing physical therapy (Nereo, Fee, & Hinton, 2003).

The progressive muscle wasting usually results in the boy becoming severely debilitated. To date there is no cure. Initial symptoms include a waddling gait, toe walking, overdeveloped calves, and difficulty rising from the floor once they have started walking (Emery, 2003). Death often occurs due to respiratory or cardiac failure in the boy's late teens (Magliano, 2014; Webb, 2005). Other complications include scoliosis and fractures of long bones and vertebrae due to osteoporosis (Pascual et al., 2019). Over the past 30 years or so, the life expectancy of boys has risen due to proactive care and more aggressive treatments (Emery, 2003; Webb, 2005). Yamaguchi and Suzuki (2015) reported that mechanical ventilation has improved the quality of life for adolescents and young adults. A survival analysis depicted that the mean age of death of DMD boys in the 1960s was 14.4 years while in the 1990s it was 25.3 years. A recent study showed that the estimated probability of survival up to age 30 was 85 percent due to the use of mechanical ventilation and having access to medical support (Kohler et al., 2009).

Research has also shown that boys with DMD are likely to have some specific psychological or learning disorders, including problems with learning potential, and general psychosocial adjustment (Hendriksen et al., 2007). Hendriksen et al.'s (2007) study included the administration of psychosocial and emotional distress questionnaires to parents, and compared the psychosocial adjustment of 287 Dutch and American boys with DMD to findings reported in boys with other chronic illnesses. They found that boys with DMD did not appear to be at greater risk for adjustment problems than boys with other types of chronic conditions. Additionally, overall psychosocial adjustment in affected boys appeared to be rated higher as age increased, despite the progression of the disorder. This is an encouraging finding, as one would expect more problems to arise as the disorder progresses. Cotton,

Voudouris, and Greenwood (2005) suggested that the development of effective coping strategies or the improvement in cognitive and problem-solving skills is likely to aid psychosocial adjustment.

2.3 Raising a Child with a Chronic or Rare Disease

Duchenne Muscular Dystrophy (DMD) is a complex chronic and rare disease that poses significant challenges to the physical and psychological well-being of the affected child and his parents. Research has shown that during the course of DMD there will inevitably be many inherent stages that predispose an atmosphere of emotional turmoil and family strife as parents become consumed by their child's illness and the necessary specialised and time-consuming care (Alexander et al., 2018; Emery, 1987; Emery, 2003; Gilgoff, 1983; Gravelle, 1997; Parker et al., 1999; Magliano, 2014; Samson et al., 2009).

The current literature reveals that the lived experiences of parents who care for their children diagnosed with a terminal illness can be likened to a never-ending struggle (Chen & Clark, 2007; Erby, Rushton, & Geller, 2006; Folkman, 1997; Gottlieb, 1997; Meiring, 2011). Regardless of the substantial progress in the treatment of the illness, it is generally presented as emotionally, physically and psychologically challenging for parents and other caregivers. For the parents and relatives, long-term assistance of the child has been described as devastating and demanding, as well as rewarding (Magliano et al., 2014). Despite possessing the knowledge of the inevitability of the child's final days, the ultimate end often leaves the parents totally unprepared as they have poured so much energy and emotion into providing a nurturing environment for this child (Dawson & Kristjanson, 2003; Gilgoff, 1983; Gottlieb, 1997).

It is therefore imperative for parents to acquire knowledge on the signs and symptoms of DMD, and on ways to manage and care for their child effectively (MDFSA, 2018). Equally important is acknowledging their own needs in order to cope with the anxieties, fears and frustrations they face through interacting with the child, and integrating the child's needs into established family routines in addition to managing the strain this diagnosis places on family life (Dawson & Kristjanson, 2003; Emery, 2003; Meiring, 2011; Peay, Meiser, Kinnett, Furlong, & Tibben, 2015).

Several researchers have commented on various psychological aspects of the affected child, including care and management, intellectual changes and personality or emotional reactions (Donald, Mathema, Thomas, & Wilmshurst, 2011; Soutter et al., 2004; Webb, 2005). However, the emotional strain on the family is often overlooked. Far more qualitative studies on the lived experiences of parents have been conducted on more commonly diagnosed illnesses such as cancer (see, for example, Alexander et al., 2018; Cornelio et al., 2016; Leite, Garcia-Vivar, Neris, Alvarenga, & Nascimento, 2019; Nicholas et al., 2009; Pourghaznein et al., 2018; Ringnér et al., 2011). Alexander et al.'s study explored the challenges and coping strategies employed by parents of a child diagnosed with cancer in Trinidad. Parents reported that since their child's diagnosis, they had no time for a social life, and often isolated themselves due to complete exhaustion. Looking back, they acknowledged that allowing themselves to open up more might have helped them cope better in the future.

Mothers are likely to have the added burden of knowing they are the carriers of the mutation responsible for DMD, and may develop an attitude of self-blame as a result (Chen & Instone, 2008; Rubin, 1987). Additionally, a recent study revealed that most mothers tended to assume the role of the primary caregiver, providing

more emotional support, whereas the fathers tended to act as providers and distance themselves emotionally from the situation (Peay et al., 2015). Other research suggests that paternal involvement often provides a coping resource that supports both the mother's and the child's adaptive capacity (Wysocki & Gavin, 2006). Youngblut et al. (2000) described the role changes in marital functioning while raising a child with a chronic illness and the negative impact this had on marital satisfaction, while Lavee and Mey-Dan (2003) described a greater sense of cohesion in the marital relationship due to the need for more open communication.

2.4 The Impact of the Initial Diagnosis

A DMD diagnosis occurs in early childhood through genetic testing, a muscle biopsy and biochemical tests (Pascual et al., 2019). Studies investigating the psychosocial outcomes of DMD seem to support the obvious, that DMD has a major impact on the lives of stricken individuals and families, and important psychosocial issues of the child arise at a time when the family is dealing with severe psychological trauma (Charash, 1983; Samson et al., 2009).

Gilgoff (1983) described DMD as an ironically tragic disease that typically occurs at a time in a boy's life when he should be gaining in strength and body control, but instead he finds himself losing control. It occurs at a time when other children are beginning to experience a greater sense of autonomy, while he is losing independence. This creates severe emotional stress for the entire family. According to studies conducted on distress in paediatric chronic illnesses, uncertainty is one of the greatest psychological stressors faced by families, often pervading various aspects of the parents' lived experience, including disease course ambiguity and the persistent and consistent losses that the parents and children face as the disease progresses (Bell, Biesecker, Bodurtha, & Peay, 2019; Han, Klein, & Arora, 2011).

Despite the fact that uncertainty is likely to increase feelings of helplessness, anxiety and depression in parents (Abi Daoud, Dooley, & Gordon, 2004), studies have conveyed that a parent's appraisal of uncertainty affects his or her ability to cope, and is not always perceived as a danger or threat (Bally et al., 2018). In contrast, parents have even alluded to the fact that uncertainty allows for a sense of hopefulness and possibility for positive outcomes for their affected child (De Graves & Aranda, 2008; Magliano, 2014).

DMD usually presents itself unexpectedly, giving parents little opportunity to prepare for the demands of caring for their newly diagnosed child. One study conducted in the United Kingdom (UK) sought to describe the lived experiences of hope of twelve parents raising a child with DMD (Samson et al., 2009). Major themes that emerged included the profound sense of loss experienced by each parent. Some parents tended to suppress their grief while others expressed this loss openly. Both ways of confronting the loss led to exacerbated anxiety. All of the parents indicated that they felt clinically depressed within the first few months after the diagnosis. Other major themes included seeing beyond the illness and its physical implications, hope for normality for the child, and hope for a cure or medical breakthrough to DMD through research in gene and stem cell therapies (Samson et al., 2009).

Parents may react to the diagnosis in many different ways. Some may react with feelings of disbelief, denial, anger, anguish, anxiety, guilt, fear, confusion, powerlessness, rejection, sorrow, or parent-child related stress (Gilgoff, 1983; Webb, 2005).

The chronic stress and never-ending responsibilities increase the likelihood of parents experiencing financial strain, anxiety, depression, post-traumatic stress

symptoms, sleep disorders, adjustment disorders, changes in marital relationships, and generally feeling physically, financially and emotionally overwhelmed and overburdened (Peay et al., 2015).

2.5 Learning to Live with DMD: Adjusting to a New Way of Life

Although the child with DMD is the identified patient, the parents and healthy siblings are also likely to endure chronic stress and prolonged periods of grieving. In a family unit it is impossible to look at wellness unless one looks at the function of the unit or system holistically (Becvar & Becvar, 2013). In order to function in a family in which an individual has a chronic or life-limiting disorder that requires extensive care it is short-sighted to focus solely on the individual, as all lives are intertwined. The progressive chronic illness of one family member often places pressure on the healthy family members to adjust quickly and adapt to the new challenges with which they are faced. Family functioning and roles are altered and the family members, including the healthy siblings, are forced to examine their beliefs about the illness, the existing behavioural patterns in the family and the effects it is likely to have on the family unit, in order to collaborate in creating new roles within the family. This is important to co-develop ways of managing new situations (Årestedt, Persson, & Benzein, 2014; Becvar & Becvar, 2013; Kazak, Sims, & Rourke, 2002).

Parental stress is evidently a pervasive challenge for couples who are raising a child with DMD, and studies have revealed that stress levels are exacerbated in the transitional periods due to challenges in social interactions more than the increased physical demands of DMD (Buchanan, LaBarbera, Roelofs, & Olson, 1979; Nereo et al., 2003; Yamaguchi & Suzuki, 2015). Nevertheless, studies have also conveyed the immense resilience and hope of parents and the likelihood of

finding enrichment and fulfilment beyond the stress by developing effective coping strategies arising from these challenging experiences (Carnevale, Alexander, Davis, Rennick, & Troini, 2006; Samson et al., 2009; Webb, 2005).

Chen and Clark (2010) conducted a correlative study in Taiwan with 126 parents of children with DMD (46 couples, 26 single mothers, and eight single fathers) recruited from the Taiwan Muscular Dystrophy Association, to determine which family resources affected parental health. The results showed that the constructs 'family hardiness' and 'family support' influenced parental health directly, and supported the need to develop strategies to increase family hardiness and family support. 'Education' and 'employment' played mediating roles in the influence of caregiving on parental health (Chen & Clark, 2010, p. 246).

2.6 Different Appraisals of DMD

Cohen and Lazarus (1979) noted that there is no standard or uniform way of adapting to life after the diagnosis of a chronic illness has been made. The cognitive appraisal of a situation or illness is likely to play a role in the process of adaptation. A primary appraisal refers to the process of gaining a deeper understanding of the consequences DMD is likely to have, while a secondary appraisal would entail the consideration and evaluation of potential coping resources in order to cope with the demands of DMD. Using these resources would ultimately aid the process of adaptation (Cohen & Lazarus, 1979).

It is necessary to highlight that as the experience of muscular dystrophy influences the emotional response of parents in relation to their child's needs, the ability of the child to deal with adverse situations is related to the parents' ability to cope (Chen & Clark, 2007). The child's self-concept and appraisal of the disorder is thus influenced by each parent's perception of the disorder (Alexander et al., 2018).

Raina et al. (2005) found self-perception (or higher caregiver self-esteem) and stress management to be important predictors of caregiver psychological health.

While the above may be true in some family contexts, a study in the Netherlands by Opstal, Jansen, Van Alfen, and De Groot (2014) has shown that 40 boys with DMD (19 ambulant and 21 wheelchair-dependent) rated their health-related quality of life as similar to their healthy peers, while their parents who completed the same health-related quality of life assessment underestimated the perception of their son's happiness, self-perception and social acceptance. One could assume that these parents might be hypersensitive to negative comments or criticism of their environment or have a tendency to project their negative frame of mind onto their son by interpreting the limitations of their son from their perspective (Opstal et al., 2014). This study vividly demonstrated the importance of using self-report questionnaires to measure health-related quality of life in boys with DMD rather than making assumptions based on their parents' pessimistic perceptions. It is therefore necessary for parents to pay close attention to their sons' realities as the benefits are likely to be twofold. Firstly, involving their sons in therapy decisions is likely to be empowering for the sons as they exercise autonomy and responsibility in their own lives. Secondly, parents are also likely to feel some relief to discover that their son is managing far better than they had imagined.

2.7 Challenges Encountered and Coping Strategies Adopted

Buchanan et al. (1979) conducted a study at a clinic in America in which 25 families were interviewed to examine their adjustments to having a child with Duchenne Muscular Dystrophy (DMD). One of the most prominent psychological effects on parents included the parents' anticipation of a future stressor due to the unpredictability of the disease course. When the parents were asked to identify what

they experienced as the major problem of DMD, and the majority of the families identified a 'psychological problem' as a major stressor, rather than "physical issues such as lifting, carrying, or turning" as their primary difficulty (Buchanan et al., 1979, p. 263). Recent studies have confirmed similar findings, indicative of the negative psychological impact of DMD on parents (Abi Daoud et al., 2004; Bell et al., 2019; Chen & Clark, 2010).

Peay, Hollin, and Bridges' (2016) recent study led by 'Parent Project Muscular Dystrophy' in the United States of America (USA) documented parents' prioritisation of short-term DMD-related worries, and identified whether worry prioritisation varied based on their child's ambulation status. It is relevant to note that this was an online survey administered over one year. While recruitment occurred using newsletter notices, social media, word-of-mouth and emails from 'Parent Project Muscular Dystrophy', the participants were predominately married, biological mothers. This study, as well as many others, suggests that mothers are more likely than fathers to get involved in understanding their son's condition and that their reactions to it may be better (Arias et al., 2011; Peay et al., 2016). Fathers tend to experience intense sadness as a result of the loss of expectations for their sons and seem to have more difficulty accepting the diagnosis (Abi Daoud et al., 2004). The results revealed that the participants' gravest concern related to their 'child getting weaker'; and 'getting the right care over time' for their child. Participants also highlighted their worry about prioritisation of 'managing uncertainty about my child's future' as well as 'my child feeling happy'. Interestingly, this study revealed that, in comparison to the other 'worry items', parents did not prioritise 'having time for myself' or 'the well-being of my other children' or 'feeling isolated from other families' as highly (Peay at al., 2016, p. 309).

This implies that the impacts of DMD on the family and parents' well-being are prioritised less than child disease management and also points to the selflessness of the parents. It could also reveal a vulnerability to dysfunction in the family system, where parents seemed likely to neglect the psychological well-being of their healthy children and themselves. Interventions that aim to address and reduce the negative emotional impact of caring for a child with a progressive disease should thus be implemented.

2.8 Depression

Abi Daoud et al.'s (2004) Canadian study explored depression in parents of 33 males with DMD, and confirmed that parents of males with DMD "had a higher probability of experiencing a major depressive episode" (Abi Daoud et al., 2004, p. 18). The parents of older males reported having more distress in their lives which was likely due to the son's greater physical dependence and increased awareness of the inevitable prognosis. The majority of the parents reported experiencing all the major symptoms of depression, such as feelings of hopelessness or worthlessness. Single parents harboured a lower self-esteem and felt less in control of their lives, revealing a sense of isolation and desperation (Abi Daoud et al., 2004).

2.9 Treatments Used and Psychological Impact on Parents

Recent studies by O'Brien (2001) and Yamaguchi and Suzuki (2015) have focused on parental care for a child with DMD on mechanical ventilation. Despite the advantages of ventilation prolonging the life of an individual with DMD, it is often overwhelming for parents to adjust to this change psychologically, emotionally and physically. The parents indicated feeling isolated and frustrated due to the perceived lack of support. Doctors working at clinics have acknowledged the need to provide anticipatory guidance to families related to options for managing respiratory

complications during routine clinic visits, but that it is not always possible during very busy times (Carnevale et al., 2006; Mah, Thannhauser, McNeil, & Dewey, 2008).

Mah et al., (2008) conducted a study in Canada which sought to describe the experience of fifteen mothers and four fathers caring for their child with neuromuscular disease (NMD) on home mechanical ventilation (HMV). Parents conveyed their experience of being the 'lifeline' for their child's quality of life. Most parents relayed a sense of ambivalence around the initiation of assisted ventilation. On the one hand, it provided them with a sense of relief and security knowing that the assisted ventilation provided respiratory support for their child, while on the other hand, the use of assisted ventilation required increased parental support in an already very demanding day-to-day life. Parents had to grapple with new complications such as power outages and potential machine malfunctioning, not to mention the stress of additional expenses. O'Brien's (2001) similar American study interviewed fifteen families to explore the experience of providing long-term home care for a child who was technology-dependent from the family's point of view. Prominent themes that emerged from the study included 'living in a house of cards' which essentially referred to the utter unpredictability of daily life with a child with DMD. A major challenge in the lives of these families included their consistent struggle to maintain a functional family when their son's condition was constantly changing, forcing the family to develop a new sense of routine amidst the chaos (O'Brien, 2001, p. 15). Although the machine for bi-level positive airway pressure (BiPAP) or tracheostomy ventilation was funded by government programmes, parents had other out-of-pocket expenses such as facemasks, backup batteries, pulse oximeters, and cough assist devices (O'Brien, 2001).

These studies have shown that advances in health care and life expectancy do not necessarily translate into increased support at home and in the community.

2.10 Coping Strategies

Coping refers to the cognitive and behavioural efforts to overcome, reduce or endure both the internal and external demands of an adverse event or experience (Rajandram, Jenewein, McGrath, & Zwahlen, 2011). Coping may also be described as the diverse responses to distressing experiences that attempt to stabilise the demands of the environment and the available resources of the individual. Furthermore, coping is not only associated with the demands of a particular situation but is also affected by the resources available to an individual (Pat-Horenczyk & Brom, 2007). Austin (2012) described three coping styles as attempts to alter negative emotions, attempts to alter the stress inducing situation, and avoidance of the stressor. Researchers have found that parents often adopt unhealthy ways of dealing with their child's illness, including overprotection, denial, and magical thinking where they often believe that their child will get better (Buchanan et al. 1979).

Peay et al (2015) acknowledged the importance of social support to parents of children with DMD. Their findings revealed that from the initial diagnosis, therapists should assess the parents' psychological needs, particularly their ability to cope with uncertainty and fear. Interventions should then help the parents to build on existing support structures, and facilitate the use of and comfort with respite care (Samson et al., 2009).

Some studies have revealed that parents tend to rely on spirituality, as well as a strong support structure (Alexander et al., 2018; Samson et al., 2009; Webb, 2005). Magliano (2014) conducted a study based on the psychological benefits of

caregiving in 502 key relatives of patients with DMD, and the results suggested that, despite the practical difficulties of caring for patients with DMD, 86 percent reported at least one positive aspect of caregiving, including personal growth, resilience, altruism, and sharing of the experience with people in similar situations.

A considerable amount of family support, including support groups, provides comfort, reassurance and hope, and spirituality aids in the relief of social, emotional and spiritual distress to enhance quality of life (Peay et al., 2015; Samson et al., 2009). Hope and spirituality assisted most of the families in the study "…to let go of what they cannot control while holding onto what they can control", thereby turning pain into hope (Alexander et al., 2018, p. 7). Similarly, Bregman (1980) noted that some families coped by focusing on the present, living as normal a life as possible, adopting a proactive attitude regarding the care and services for the child's condition, and by becoming trained disability advocates, thereby developing coping resources based on personal strength.

2.11 Developments in South Africa

2.11.1 Muscular Dystrophy Foundation of South Africa.

Various symposia have been held and associations have been formed internationally, and also in South Africa, as the need to support parents raising a child with a chronic condition has gained recognition as a cornerstone of effective treatment. These platforms have sought to outline the key challenges and opportunities to assist parental coping (Guite et al., 2018).

In South Africa specifically, various organisations exist to alleviate and assist with some of the challenges parents experience on a daily basis. The Muscular Dystrophy Foundation of South Africa (MDFSA) was founded in 1974 by parents who had a son with DMD due to their frustrations with the level of support available

to them. The foundation provides valuable and comprehensive information to parents, ranging from awareness campaigns and parent projects, to support services and psycho-education on muscular dystrophy (MDFSA, 2018).

2.11.2 Clinical Practice.

Genetic testing for DMD in the South African public sector is performed within the National Health Laboratory Service at the Groote Schuur Hospital in Cape Town, and at various neuromuscular clinics in Johannesburg and Pretoria. It has been extended to the state and includes private patients inside and outside of the country (Esterhuizen et al., 2016).

A neuromuscular service has been operating at The Red Cross War Memorial Children's Hospital for over twenty years and referrals include individuals from all provinces in South Africa and from other African countries. It is the only certified Duchenne Muscular Dystrophy Centre in South Africa. In keeping with international protocols, the diagnosis in clinically suspected cases is confirmed by DNA analysis or a muscle biopsy. Clinic visits are scheduled every three to six months, where the families have access to genetic counselling and support from the MDFSA (Esterhuizen et al., 2016). The children undergo regular sessions with a physiotherapist and regular cardiac assessments. Corticosteroid intervention is implemented at around four years old, or as soon as the boy's motor function is affected. As steroid side-effects include immunosuppression, children are vaccinated against varicella and undergo bone mineral density assessments. Boys with scoliosis are referred to an orthopaedic specialist (Esterhuizen et al., 2016).

Additionally, the Red Cross clinic also assists with the placement of DMD boys in schools with suitable facilities. The clinic acknowledges that families and their affected children often need emotional support, which is only partially

addressed at the clinic due to resource constraints (Esterhuizen et al., 2016). External associations such as the MDSFA offer a great deal of emotional support to families, and counsellors from the association offer sessions with parents and their children (Associate Professor Jo Wilmshurst, Head of Paediatric Neurology at the Red Cross Children's Hospital, personal communication, January 9, 2020; Esterhuizen et al., 2016).

Wilmshurst mentioned that, although she does oversee a few support groups for boys and their families at the clinic, there is often "…very little space to ask, how are you feeling?" due to the enormous pressure each health professional is under to best serve the community (personal communication, January 9, 2020).

2.11.3 Socio-economic concerns in South Africa.

Families with higher education levels may have greater access to financial or other resources that may serve to reduce stressors on a day-to-day basis. Wilmshurst expressed her concern for affected boys living in rural areas as their parents often do not bring them into the clinic until the later stages of the disease as they have not realised that their child has a progressive disease (personal communication, January 9, 2020). It is currently necessary to develop educational programmes and training initiatives for resource-poor communities in order to reduce stressors, and to support and educate the parents.

Wilmshurst also indicated that while direct disease modification has been implemented in clinics in Oxford in the UK, the treatment strategies employed in South Africa are comparatively conservative (personal communication, January 9, 2020). Despite the fact that she and her colleagues are working towards implementing gene replacement and gene modification strategies in South Africa, "We simply do not have the financial resources to implement these treatments as it

would cost roughly $300 000 per year for each affected boy and South African medical aids will not cover these exorbitant costs" (personal communication, January 9, 2020).

Donald et al.'s (2011) South African study described the cognitive and behavioural profile of boys with DMD in South Africa. Their results revealed that boys with DMD experience mild cognitive dysfunction, predominately affecting visual memory, verbal and nonverbal executive functioning. They found that environmental factors are likely to contribute to the performance of children with DMD, and risk factors included violence in the household, social isolation, poverty and maternal depression (Donald et al., 2011). Furthermore, emotional and behavioural problems were reported by the parents who participated in the study. The parents reported comparatively higher rates of behavioural problems, including social problems, somatic complaints, thought problems and depressed and withdrawn behaviour), possibly due to "…the exposure of most of our children to significant socioeconomic stressors" (Donald et al., 2011, p. 967). While parents might be more concerned about their son's physical well-being, it is clear that boys with DMD are likely to experience cognitive and behavioural problems too, which are often under-recognised, especially in resource-limited settings (Donald, et al., 2011).

2.11.4 Registries in South Africa.

The Division of Human Genetics at the University of Cape Town obtained approval for a formal biological material repository and database in 2010 (Esterhuizen et al., 2016). This was a massive feat, as the repository holds all DNA material which will be used for further DMD research. The Red Cross Clinic, in collaboration with the Division of Human Genetics at UCT, has created a database listing each patient with a diagnosis of DMD currently attending the clinic

(Esterhuizen et al., 2016). The purpose of this is to identify DMD patients who could be candidates for a mutation-based intervention, as well as providing valuable information for future research in South Africa (Esterhuizen et al., 2016).

2.12 Sons with Duchenne Muscular Dystrophy Transitioning into Adulthood

Although a curative treatment is not available, survival into adulthood has become more likely over the past twenty years due to improvements in interventions such as ventilation and cardiac care (Hoskin, 2017). With treatment, the mean age of death was reported to be 27 years of age in 2007 (Eagle et al., 2007), and without treatment the mean age of death was reported to be 19 years of age (Bushby et al., 2010). This comes with its own challenges and concerns about well-being in this seemingly unexpected population of adolescents and adults where information and guidance for parents and individuals with DMD seems to be scarce (Yamaguchi & Suzuki, 2015).

Several studies have been conducted on the transition to adulthood, with the vast majority showing that the transition to adulthood for adults with DMD and their families "…has not been a positive one" (Hoskin, 2017, p. 166). Abbott, Carpenter, and Bushby (2012) conducted their study in the UK, where young men with DMD and their families were interviewed about the transition to adulthood. Their findings showed that, for parents, getting their son's needs met while encouraging their independence was often a challenging task, often involving fights and battles with their sons. Some families in this study also showed that the young adults with DMD tried to protect their parents by not expressing their difficult emotions or talking about their concerns. Parents are likely to find it difficult to know what to tell their sons, given the changing medical technology and increases in life expectancy, and results from the 'Hospital Anxiety and Depression Scale' revealed that 80 percent of parents

reported clinical depression and anxiety (Abbott et al., 2012). Additionally, families conveyed their distress at the prospect of having to figure everything out themselves with very little support from key professionals on how to plan for this transition to adulthood, both practically and psychologically (Abbott et al., 2012).

2.13 The Importance of a Shared Community

Many young adults with DMD in the UK have experienced the provision through transition to adulthood to be inadequate (Abbott et al., 2012; Hoskin, 2017). They often struggle with high levels of anxiety in a world that does not expect much of them or consider their long-term futures, "…because no one expected them to be alive" (Hoskin, 2017, p. 166).

Hoskin (2017) reported on the parent evaluation of a lottery funded 'Transition to Adulthood' project which sought to empower boys with DMD and their parents by preparing them for the future with a focus on employment, independence, health and social inclusion, and hosted 'Letting Go' meetings for the twenty parents where information, strategies and support systems were shared. All parents recommended the project and said how beneficial it was for their sons to speak to adult role models within the DMD community as it gave them a sense of hope for their futures. Most families expressed a strong need to be part of a shared community (Hendriksen et al., 2007), which is an important reason to initiate new projects of this nature in South Africa.

2.14 Palliative Care

Palliative care refers to the care of terminally ill individuals and includes physical, psychological, emotional and spiritual support for the parents and also includes bereavement care. Effective palliative treatments have become increasingly necessary for boys with DMD who are now living longer, to address

psychosocial problems encountered by parents and their affected sons (Arias et al., 2011; Dawson & Kristjanson, 2003; Weidner, 2005).

According to Arias et al. (2011, p. 93),

> Palliative care now encompasses an array of services that extends beyond the care that is usually associated with the end stages of diseases. These services may range, for example, from respiratory care to improve functioning and maintain quality of life, to case management services, and may also include counselling of families about decision-making they will face as the disease progresses and legal planning such as advance directives.

Additionally, Arias et al. (2011) vividly highlighted the importance of implementing palliative care services throughout the life of an individual with a chronic and progressive disease rather than only focusing on end-of-life-concerns. In their study, Arias et al. (2011) described the palliative care services that parents of males with DMD felt they received. Thirty-four families completed a comprehensive interview. The vast majority, 82 percent of respondents, were mothers. Their findings revealed that most parents did not know what palliative care entailed or how to access the services, and they felt inexperienced in communicating about advanced care planning (Arias et al., 2011). Erby, Rushton, and Geller (2006) reported similar findings. Although respiratory care was a service received by the majority of young men with DMD, other pertinent services such as pain management, hospice services and respite care were reported to be received by less than twenty percent of the respondents. Additionally, very few participants reported having a living will or advanced directives in place (Arias et al., 2011).

A study conducted on palliative care among parents of individuals with DMD showed that while some families were aware of what palliative care entailed, they

were reluctant to use hospice for respite care as this simply signified the grim reality of their son being "…on their way out – and you don't need that" (Parker et al., 1999, p. 247). This conveys how difficult it is for parents to acknowledge and discuss issues of emergency treatment or advanced directives, or to consider that palliative care could be useful.

Parents also conveyed their loss of confidence in the medical fraternity because sometimes professionals had been unable to assist them and some parents recalled the clear discomfort of health professionals with advanced directives (Parker et al., 1999). Due to this, parents preferred to engage in a "parent support network" (Parker et al., 1999, p. 247). Most families expressed their life philosophy of living for the moment and their reluctance to discuss end-of-life care (Arias et al., 2011; Parker et al., 1999). Reid and Renwick's (2001) study indicated the intense feelings of guilt some parents experienced when discussing death issues with their children. Furthermore, the level of discomfort expressed by professionals and families in facing these issues is likely to make it even more challenging for parents to ask for and receive the help they may need.

These studies also suggest a need for improved awareness of palliative care among families and the lack of coordination of care and access to competent carers. Parents exposed to more consistent and reliable information would encourage them to make use of the available services.

2.15 Conclusion

The studies reviewed in this chapter have indicated that raising a child with DMD poses significant challenges to the physical and psychological well-being of the parents and the entire family unit. The initial diagnosis of DMD marks a turning point in the lives of the families, forcing them to adapt to a new way of life. Several studies

revealed the vast challenges parents face on a daily basis, pervading every aspect of their lives. A few studies in this literature review acknowledged the importance of social support to parents of children with DMD. Parents expressed feeling overwhelmed, fearful and uncertain and in need of comfort, reassurance, practical and psychological support due to a very unpredictable future (Alexander et al., 2018; Samson et al., 2009; Webb, 2005). The literature review was concluded by outlining the current developments and clinical practice in South Africa, and the importance of palliative care, highlighting the need to develop educational and training initiatives for resource-poor communities in order to reduce stressors and provide much needed support. This provides the motivation for the present study. The next chapter presents the research methodology used in the current study.

Chapter Three: Methodology

3.1 Introduction

This chapter describes the research approach and how it was actualised in this study. It provides a brief discussion of the paradigm and research design, followed by an explanation of the participants. Each procedure and technique utilised in the data collection and data analysis is described in detail. The chapter concludes with a discussion of various issues of quality assurance and ethics that informed the study.

3.2 Research Aims

The aim of this study was to explore the lived experiences of parents of how a diagnosis of DMD affects their lives. This study endeavoured to describe the participants' challenges and the strategies they adopt to deal with these challenges. Themes were highlighted through semi-structured interviews, employing interpretative phenomenological analysis.

Samson et al. (2009) found that while the challenges experienced by parents have been investigated to a greater extent, the coping mechanisms of parents have scarcely been explored. In addition, few qualitative research studies have been conducted on the challenges and coping strategies of parents living with a child diagnosed with DMD in South Africa.

Results from the current study may lead to advocating for policies to improve support for parents with DMD, and provide practical information to assist parents with their daily challenges. Furthermore, an additional perspective may be gained as to how parents cope with the challenges with which they are faced, which is yet to be explored in a South African context. This perspective will be helpful to families, therapists, clinicians and other professionals interested in understanding the lived

experiences of parents of children with DMD in a South Africa, in order to implement positive change.

3.3 Research Questions

This study aimed to explore the experiences of parents raising a child with DMD. It thus aimed to answer the following questions:

- How did the parents perceive and react to the initial diagnosis of DMD?
- What are some of the challenges the parents have faced?
- What are some of their coping strategies?
- What kind of support have they received from their community?

3.4 Research Design

An exploratory qualitative approach was used for this small-scale study to gather and convey an in-depth understanding of the research questions using semi-structured interviews. A qualitative research paradigm aims at gathering a thorough understanding of the processes that underlie an individual's unique experiences, beliefs and perspectives by examining the social and cultural contexts which shape behavioural patterns (Mistry, 2012; Wagner, Kawulich, Garner, & Botha, 2012). My qualitative study adopted an interpretivist stance which focused on people's subjective experiences. Interpretivists assume that reality is not objectively determined, and accept that there are multiple realities of phenomena. I therefore sought to examine the meanings that individuals associated with their experiences and perspectives. The interpretivist stance also acknowledges that the researcher's values, beliefs and prior knowledge will inevitably inform and direct his or her research (Wagner et al., 2012).

3.5 Interpretative Phenomenological Analysis (IPA)

The theoretical framework adopted in the current study is interpretivist phenomenological analysis (IPA), which aims to give a voice to the parents' experiences and accounts of raising a child with DMD. It also aims to contextualise and derive value and meaning from these reported accounts from a psychological perspective (McLeod, 2011; Smith, 2012; Willig, 2013). Phenomenology is the study of phenomena or the structure of experiences that appear in one's consciousness from a subjective point of view (Smith & Shinebourne, 2012). In addition to describing the phenomenon in question, IPA methodology draws attention to how participants make sense of their experiences, thereby enabling first-person research that can also be refined into superordinate and sub-themes (Shaw, Burton, Xuereb, Gibson, & Lane, 2014; Smith, Flowers, & Larkin, 2009). These themes enable the researcher to arrive at a deeper understanding of the connections evident between the research participants' experiences through the analysis of detailed, reflexive, first-person accounts. Willig (2013) noted that phenomenology relies on the representational validity of language and the reflections that individuals make regarding their experiences.

Husserl (Husserl & Gibson, 2002) and Heidegger & Dahlstrom (2005) are considered to be the dominant pioneers of phenomenology, and both hold views that rejected a dualist approach to reality (Larkin, Watts, & Clifton, 2006; Reiners, 2012). Phenomenology refers to the method of inquiry developed by Edmund Husserl, who introduced the concept of 'lived experience' and defined phenomenology as an interest in those things that can be directly understood through one's senses (Koch, 1996; McLeod, 2011; Wallace & Wolf, 2006). This concept underpins the belief that individuals and the world around them are intrinsically linked – essentially revealing

that it would be impossible to dismiss the subjective influence of notions like culture, language, and ideology (McLeod, 2011; Willig, 2013).

Philosophers and scientists were largely responsible for the transition into modernity and thus sought to create new ways of thinking about the world (McLeod, 2011). Husserl sought to 'enlighten people' in Europe who had been dominated by a Christian world-view, often relying on prejudice to justify their beliefs, until the seventeenth and eighteenth centuries (Wagner et al., 2012).

Husserl devoted most of his life to creating a phenomenological understanding of the world, holding the basic assumption that knowledge is acquired through direct experiences that are created by sensory stimuli and phenomena. Spinelli (1989, as cited in Willig, 2013, p. 85) conveyed that phenomenological psychology is concerned with vivid descriptions of the diversity of human experience, whereas Husserl's phenomenology was more concerned with the 'identification of essences'.

Two approaches to phenomenological analysis are prominent in social science research. Hermeneutic or interpretative phenomenology is derived from the work of educator Van Manen (1990, as cited in Wagner et al., 2012, p. 238), who referred to his research as interpreting the 'texts' of life, varying from interviews, observations, journals, art and poetry. Following this approach would entail a written description of the phenomenon being studied, where the researcher's interpretation is mediated between the lived experiences of the participants (Wagner et al., 2012)

A second form of phenomenological analysis is often referred to as empirical or psychological, tending to be less focused on the researcher's interpretative analysis. This second form can be likened to Husserl's notion of 'bracketing' or

setting aside one's assumptions by adopting a transcendental attitude in order to gain a new perspective on the phenomenon being studied (Wagner et al., 2012).

An IPA seeks to understand the social world by exploring the richness, depth and complexity of phenomena by engaging actively with the research participants in order to identify some shared phenomenon or experience (Wagner et al., 2012). Phenomenological research, specifically IPA, has thus been utilised in the current study as it is inductive, interpretative and interrogative, with the primary goal of exploring the nuances, commonalties and differences evident in the parents' lived experiences, and determining how they make sense of their experiences by often asking open-ended questions such as, "What is it like to raise a child with DMD?"

Engaging with an IPA research study entails the description of phenomena by analysing themes and patterns that arise, and incorporates previous literature and theory to interpret them (Smith & Osborn, 2003). In using IPA, I also recognise that such a study is likely to implicate my unique worldview in the form of meanings that I inherently bring to the interaction between myself and the participants (Smith et al., 2009). An IPA can therefore be likened to Van Manen's hermeneutic phenomenology, in which an understanding of reality is derived through reflexive analysis of perceptions and reflections, attempting to describe an individual's consciousness in context (Willig, 2013).

This study aimed to describe vividly the reflected experiences of parents raising a child with DMD. As internal states cannot be fully understood or comprehended, a reflected version of the parents' personal experiences is presented. As a qualitative research approach, IPA allowed for an opportunity to understand the innermost deliberation and perceptions of the 'lived experiences' of the research participants. Furthermore, the number of participants can vary from two

to twenty-five (Alase, 2017; Creswell, 2012). In the current study, three couples (six people) were interviewed.

An important and constant consideration in qualitative research is reflexivity, which refers to the researcher's acknowledging their influence and how they shape the research process as a person and as a theorist (Madill et al., 2000; Willig, 2013). Additionally, IPA researchers are required to maintain a realistic and reflexive approach to knowledge production by acknowledging the researcher's influence on it, and still striving to give voice to the participants by contextualising and interpreting their experiences using existing psychological knowledge (Larkin, 2006).

In this study, I kept a reflexive journal, to understand my responses and experiences to the research (Ortlipp, 2008). The process of journaling may have increased the level of transparency in the study. Many of the reflections made in the journal were taken to research supervision. In this way, my emotional and occasionally subjective responses were acknowledged and separated. I considered my position as a student hearing confidential information about the experiences of these parents and was mindful of the effect this had on me, and on the writing of this research report. I experienced a strong advocacy response to many of the participants when hearing some of their negative experiences. These experiences were acknowledged in my reflections and balanced with the production of quality research.

Much of the effect of the reflexivity of the study is discussed in the results and discussion chapter, and I tried to restrict my subjective experiences from entering into the discussion of the participants' experiences when it was not helpful, to ensure that the data used to answer the research question was the reflected experience that

had been co-constructed. It is hoped that, through this process, the data reflects the experiences of the participants from the interview process.

3.6 Participants

Purposive sampling was used to select participants. Participants were chosen for their relevance to the research questions (Willig, 2013) and inclusion criteria (Guest, Bunce & Johnson, 2006). DMD is a genetic disorder that occurs primarily in boys (Chen & Clark, 2007; Emery, 2003). Therefore, to be eligible for the study, parents were required to be currently raising a son with DMD. Inclusion criteria therefore included a recognised diagnosis by a specialist physician.

According to Peay et al. (2015), mothers tend to assume the role of the primary caregiver, whereas fathers tend to act as providers. Additionally, both parents are likely to be heavily involved in caring for their son with DMD due to the demanding nature and wide-ranging responsibilities of the illness. Despite the nature of their marriage or relationship, both parents play an important role (Peay et al., 2015; Wysocki & Gavin, 2006). This was the primary reason for interviewing both parents in one sitting, as I sought to gain a holistic view of the parents' experiences.

Parents were recruited via an online support group, 'The Muscular Dystrophy Foundation of South Africa (MDFSA)'. This platform serves to reach out to parents and families in addition to supporting research into this disease to find a cure (MDFSA, 2018). Support groups in the Johannesburg and Pretoria region were contacted to find out which members would be willing to take part in the study. The rationale for sourcing these participants was due to the need for face-to-face interviews and thus finding parents in the Johannesburg or Pretoria region was ideal.

Participants needed to be able to communicate in my spoken language (English). The online discussion group platform was used to assist me in determining the participants' language competency.

The duration of diagnosis of DMD needed to be longer than one year, as the study sought to report on the parents' lived experiences thus far. Additionally, parents of children who had been newly diagnosed were likely to find it more challenging to disclose their feelings openly or to report on their lived experiences due to the terminal nature of DMD. This was evident when four couples with sons who were recently diagnosed initially volunteered to participate in the study by contacting me, but withdrew from the study before it commenced. Their primary reasons for withdrawing included not finding the time to be interviewed in between all the specialist appointments, and feeling too overwhelmed to speak openly about their experiences thus far.

Three sets of parents of children with DMD were interviewed as a larger sample size could not be attained after ethical clearance was obtained in January 2019 (Certificate Reference Number 12289681 (GW2018115H). Please see Appendix A.) There was no restriction on the age of the child as an exploration of a range of experiences was considered to be beneficial to parents and professionals alike. The parents' experiences often echoed those of other parents in similar studies, and at times enabled vivid comparisons of their personal coping strategies and challenges, thereby enriching this research study. Wagner et al. (2012) referred to this process as a 'cross-analysis' that 'identifies convergent and divergent themes' (p. 239).

The details of the parents who participated in this research, their sons, and other family members are presented in Table 3.1.

Table 3.1: Relevant characteristics of participants

Name of parents* (age)	Sons* with DMD	Current Age	Date of initial diagnosis	Age when diagnosed	Other children* (age)
Sarah (51) and Chris (54)	Michael Jeff	18 14	2006 2006	6 2	Grace (13) Kevin (10)
Lesley (46) and Mark (59)	Eric	9	2014	4	Stuart (18 Months)
Michelle (25) and Scott (26)	Paul	8	2015	4	Leanne (4)

*Pseudonyms used

3.7 Research Instrument

IPA is characterised as inductive whilst allowing a sense of flexibility, thereby enabling an intensive qualitative analysis of detailed personal accounts derived from participants, and is generally conducted on relatively small sample sizes which are sufficient for the potential of IPA to be realised (Smith & Shinebourne, 2012).

The information sheet and informed consent introduced each participant to the study (Wagner et al., 2012). These may be seen in Appendix B and Appendix C.

Semi-structured interviews were used for data collection. The motivation for semi-structured interviews was that although I was guided by what the interviewee said during the interview, there was an element of flexibility where key questions were asked (Wagner et al., 2012). Additionally, the semi-structured nature of the interview schedule allowed for increased flexibility by adding follow-up questions, a feature which is lacking in structured interviews. Semi-structured interviews are generally used to answer more complicated research questions (Fylan, 2005) and this enabled each couple to describe freely what they felt strongly about, rather than me dictating the agenda of the interview.

The interview schedule (Appendix D) included open-ended questions which aimed to elicit information needed to answer the research questions of the current study. This helped to facilitate the conversation between the participants and myself.

The final set of questions was formed after reviewing the pertinent literature on the topic and ascertaining the relevance of the questions. The interview schedule was designed based on the broader topic of parental perceptions of raising a child with DMD and started with the more general experiences of each couple and developed to each parent's personal experiences and more specific questions regarding the initial diagnosis, interpersonal factors, challenges and coping strategies. The questions were piloted by two academics to determine their suitability for the study.

3.8 Data Collection

The parents of the children with DMD were interviewed at their homes or another venue of preference in Johannesburg and Pretoria. The reasoning behind this was to accommodate the parents who all had busy schedules. The interviews were approximately sixty minutes long and were audio-recorded.

Once the interviews were completed, they were transcribed verbatim before being subjected to analysis. All the transcriptions have been kept in my and my supervisor's possession and have been safeguarded in electronic and paper format.

3.9 Data Analysis

After a thorough analysis of the text, emergent themes that characterised each section were identified and integrated into meaningful clusters, according to Willig's model of IPA. This was done in order to capture the experiential quality of what was described. An integration of themes described the quality of the

participants' shared experience of the phenomenon under investigation. This revealed something about the essence of the phenomenon itself (Willig, 2013).

I did not wish to test hypotheses, but rather to vividly explore the rich meanings associated with the parents' lived experiences of raising a child with DMD. This exhaustive description was generated from the analysis of the interviews, by discarding preconceived notions about the possible outcomes of the study to allow the meaning to be derived solely from the data collected, through "… the detailed examination of one case; then other cases are analysed, one by one, until they are all included in the analysis…" (Wagner et al., 2012, p. 239).

The procedure of IPA involves specific steps that should be followed with an attitude of reflexivity and a focus on data that is grounded in the transcriptions (Willig, 2013). The intention behind this process is to avoid imposing my views or beliefs on the participants, in an attempt to "…unravel the meanings contained in […] accounts through a process of interpretative engagement with the texts and transcripts" (Smith, 1997, as cited in Willig, 2013, p.87).

Willig (2013) proposed the following stages of IPA, which were followed:

Stage one involved the reading and re-reading of the text. In the process I was able to reflect on my initial thoughts and observations. I made notes regarding important questions, experiences, descriptive comments and summary statements of each interview. Jotting down the significance of the tone of voice and vocabulary used by each couple used throughout the interview by way of linguistic comments appeared to be particularly useful to me, as I was able to analyse each couple's raw and uncensored subjective experiences of raising a child with DMD.

Stage two of the data analysis required me to identify and label emergent themes that characterised each section of the text. Thematic analysis is described

as a method of identifying and describing patterns or themes (Braun & Clarke, 2006). It was important to be cognisant of the fact that phenomenological research is interested in the nature, quality and meaning of experience by constantly referring to the original data, highlighting "…theme labels [that] ought to capture the experiential quality of what is being described" (Willig, 2013, p. 88). In other words, I sought to elucidate the essence of the phenomenon as it existed in the participants' concrete experience. Throughout this process, an integrative approach was utilised where I incorporated theory to interpret the data in a holistic way. In addition to this, I endeavoured to make a contribution to the field of mainstream psychology through enhancing existing knowledge (Smith, 2003; Smith & Shinebourne, 2012).

Stage three involved listing the identified themes and engaging in critical thinking and analysis around how the themes related to one another. The themes were grouped into clusters, along with relevant quotes, labels or keywords placed next to each theme. It was useful to record the page number and line number of each quote, sentence or paragraph that alluded to a shared concept or meaning. I ensured that I followed this process for each transcript, giving each cluster of themes a label that captured their essence. Some were significant quotations or descriptive labels taken from the participants that captured their lived experiences vividly (Willig, 2013, p. 88). The interviews could be regarded as a co-construction between the participants and myself, albeit that the themes emerged from an in-depth engagement with the participants' expressions alone. Through this process, emergent or master themes were revealed, and some emergent themes subsumed others (Willig, 2013).

The final stage of IPA involves the production of a summary table of the structured themes, including relevant quotations and keywords. The interpretation

phase of IPA can be done at various levels. Initially the transcriptions were interpreted from an empathic and meaning-making level. I then engaged in a critical interrogation of the hermeneutics behind the initial meaning (Smith, 2003; Smith & Shinebourne, 2012). The rationale behind interpretation was to understand the data within existing literature and to answer the research questions by contextualising the findings by incorporating existing literature and theory.

3.10 Ethical Considerations

Ethical concerns are at the centre of the research procedures. In the field of psychology specifically, various boards and institutions are in place to ensure that the correct ethical procedures are adhered to, such as the American Psychological Association (APA); the Health Professions Council of South Africa (HPCSA); and the Research Ethics Committee at each university (Willig, 2013). The proposal for the current research was presented to a panel of supervisors and submitted to the University of Pretoria Research Ethics Committee. I was granted ethical clearance (Certificate Reference Number 12289681 (GW2018115H) in January 2019. Please see Appendix A.). Due to the voluntary nature of the study, parents were invited to respond to my online post. My details were posted on one of the support groups created by the Muscular Dystrophy Research Foundation of South Africa (MDSA).

After ethical approval, I contacted the parents who showed a keen interest in participating in the study by responding to the online request to participate in the study, and a time and day to conduct the interview was arranged. Each couple received an information sheet with all the pertinent information of the study (Appendix B) and they were asked to read this carefully before the interview. This included the research questions, the aims of the research, the procedures regarding

the interview and the contact details of myself and my supervisor. The letter also detailed the importance of participant confidentiality, voluntary participation, which is unpaid, as well as each individual's right to withdraw from participating in the study at any time. Participants were given the opportunity to ask questions about any aspect of the research before and after signing the consent form (Appendix C). Debriefing took place after the interview.

Discussing the challenges and coping mechanisms of living with a child with DMD is likely to be a sensitive topic due to the terminal nature of the disease, with the potential to open emotional wounds. Additionally, interviewing both parents is likely to stimulate issues around differing responses to their situations. This was acknowledged from the start. All participants were informed that should the interviews raise any difficult emotions, they could withdraw from the study or choose to not answer certain questions, with no consequences. This did not happen though, as all of the parents were comfortable with completing the interview. They were also provided with the contact details of a reliable counsellor or psychologist, free of charge, and should they feel they would benefit from counselling at any future time, free psychological services in the Johannesburg and Pretoria areas would be arranged and provided. The contact details of Itsoseng Clinic (Pretoria) and FAMSA (Johannesburg) were given to the parents. In addition, I provided a debriefing session for the parents at the end of each interview. A debriefing session allowed for the closure of each participant's experience (Qu & Dumay, 2011). Debriefing is worthwhile as it creates a safe, non-threatening space to discuss any sensitive thoughts or feelings that the interview process may have elicited. Some of the debriefing questions asked included:

1. How did you experience the interview?

2. Is there anything else you would like to raise or discuss?
3. Did the interview raise any difficult emotions for you that you would like to discuss further?

The parents seemed to appreciate this considerate approach, but did not raise any other personal concerns. Instead, they reiterated how crucially important they believed it to be to raise awareness of the challenges faced whilst raising a child with DMD and the importance of ongoing research in this area. Their primary hope was to establish a larger support system in South Africa, similar to the effective support structures available to parents in America. Furthermore, they seemed grateful to have the opportunity to inform as many people as possible of the gaps in the DMD medical fraternity in SA.

One father, Mark, decided directly before the interview that although he would participate, he did not 'plan on saying much'. This interview (between Lesley and Mark) was deemed by me to have perhaps been a personally painful process for both participants in that the discussion stimulated issues around their conflictual marriage and their tendency to withdraw from each other during stressful times. He explained that it would be difficult to summarise their experiences, and let his wife do most of the talking. When asked if he would like to withdraw from the interview altogether, he noted that he would rather stay. I did not coerce him to participate at any point during the interview and respected his wish to 'sit in' but not participate fully. These participants were debriefed at the end of the interview and advised to seek counselling. The possible reasons for this are interpreted in the next chapter.

Willig (2013) encourages a non-invasive form of recording interviews, and thus a cell-phone was used to create an environment that was relatively comfortable for self-disclosure of experiences. The participants were also informed of their right

to request a document of the results of the study and were provided with my and my supervisor's details if they wished to contact us at a later stage (Willig, 2013).

In line with the ethical considerations stipulated by the American Psychological Association and the relevant ethical boards, pseudonyms have been used to protect the participants' identities and confidential information (Willig, 2013). While full anonymity could not be assured in the study due to the personal nature of the interview process and the use of extracts in the analysis, participants were assured that all identifying information would be eliminated. Additionally, I was the only individual who listened to the interview recordings and the data were kept on a password-encrypted computer.

3.11 Credibility and Quality Assurance

All qualitative methodologies acknowledge that the researcher is always implicated in the research process as the researcher is both a participant and an observer (Speer, 2002). However, there are still certain criteria that need to be adhered to and standards by which the quality of data can be evaluated (Willig, 2013). Willig (2013) noted that if it is evident that the researcher has attempted as best as possible to achieve his or her research goal by answering the research question and he or she has adhered to the assumptions and directions of its epistemological and theoretical position, it is considered quality research. The research questions and design were succinctly and coherently stated in order for the reader to be aware of the goals, assumptions and methods underpinning the research. The data collection and data analysis were aligned with the research question and design. When interpretations were made, they were grounded in examples from the participants' own words, and these examples were situated according to their context and situation (Madill et al., 2000; Willig, 2013).

It was my responsibility not to alter the data to serve my own personal subjectivity, but rather to ensure that the data analysis was led by the participants' experiences. In this study, the data were considered against already published studies. Speer (2002) noted that the researcher's perspective is valued and incorporated in so far as it enriches and validates the data. I used non-directive techniques in the interview to ensure that the participants' voices were prioritised. Additionally, participants were aware that they were in an interview, making it necessary for me to consider that the direct words of the participants might reflect some interactional resistance, which is alluded to in the data analysis (Speer, 2002).

This study sought to explore the lived experiences of parents raising a child with DMD by capturing the subjective 'feel' of their experiences and identifying recurring patterns of experience among the parents. A relativist epistemological position was adopted in this study, implying that obtaining and describing a 'pure experience' is virtually impossible. Therefore, my aim was to explore multiple versions and narratives of the parents' experiences, largely dependent on cultural and discursive resources (Willig, 2013).

The interpretative phenomenological researcher aspires to capture, above all, the participants' feelings, thoughts or perceptions, but not in a way that takes account of experiences entirely at face value; description of experience always involves a certain amount of interpretation (Willig, 2013). Larkin et al. (2006) said that the researcher's interpretations arise from reflecting on the participants' descriptions in relation to a wider social, cultural and theoretical context. My reflexive experiences have been included in this study to bring the reader into an awareness of the meaning-making made between myself and the participants.

3.12 Conclusion

This chapter discussed the implications of the methodology and described how the procedure within this study was conducted. The primary research questions were presented and used to guide both the methodology and the data analysis. Following this, the research approach that laid the foundation for this study was discussed and justified. In addition, the participants and the data collection method were described, and a table of the participants in this study was provided. The data analysis and all the processes underlying it were discussed and the ideas that underpinned the method were included. As qualitative research involves subjective perceptions, issues of credibility and quality were presented. Ethical considerations were further discussed. This chapter also included an exposition of the reflexivity that was maintained by the researcher to increase the quality of this study. The next chapter provides and discusses the results.

Chapter Four: Results and Discussion

4.1 Introduction

This chapter presents an overview of the themes that emerged as well as an analysis of the data. I remained cognisant of the impact my own social, cultural and historical influences and how they were likely to affect my interpretations. I acknowledged these influences through thorough reflexive engagement throughout the results and discussion chapter.

4.2 Sample Characteristics

Six parents (three couples) were interviewed. Participants were aged between 25 and 54 years old. All of the participants were married. The length of membership of the Muscular Dystrophy Association of South Africa ranged from two to fifteen years. The parents' level of involvement within the support group varied considerably.

Before starting the discussion of themes, information about the participants as outlined in the methodology chapter is briefly reiterated.

Sarah (51) and Chris (54) are married with four children. They have one healthy son named Kevin (10) and one healthy daughter named Grace (13). They have two sons with DMD. Michael is 18 years old, and Jeff is 16 years old. Michael was diagnosed with DMD at the age of six, while Jeff was four years old.

Lesley (46) and Mark (59) are married with two sons. Stuart (18 months) is healthy and Eric is nine years old and was diagnosed with DMD at the age of four years old.

Michelle (25) and Scott (26) are married with two children, one healthy daughter named Leanne (4) and one son named Paul (8). Paul was diagnosed at four years old.

For easiness of reference, the sample characteristics are summarised in Table 4.1.

Table 4.1: Summary of sample characteristics

Parents	Boys with DMD	Other Siblings
Sarah and Chris	Michael (18) and Jeff (14)	Kevin (10) and Grace (13)
Lesley and Mark	Eric (9)	Stuart (18 months)
Michelle and Scott	Paul (8)	Leanne (4)

4.3 Themes Identified

Analysis of the interviews allowed for the emergence of four major themes within the parents' lived experiences of raising a child with DMD. The first major theme identified was reactions to the diagnosis. This included frustrations with the medical fraternity as a sub-theme. The second major theme was facing the loss in terms of grief and anticipatory loss. The third major theme was learning to adapt, or 'living with it'. This theme had a number of subthemes, namely, the appraisal of the illness in terms of daily challenges and responsibilities, vacillating between hope, grief, avoidance and presence, the importance of planning in advance and gaining practical information, and support versus isolation. The fourth major theme was looking beyond the illness. These themes are summarised in Table 4.2.

Table 4.2: Themes

Number	Theme	Subthemes
1	Reactions to the diagnosis	Frustrations with medical fraternity
2	Facing the loss	Grief, anticipatory loss
3	Learning to adapt (Living with it)	Appraisal of illness, daily challenges, hope vs grief, avoidance vs presence, planning, support vs isolation
4	Looking beyond the illness	Finding meaning

4.4 Reactions to the Diagnosis

The parents indicated that their sons were all diagnosed at around the age of four years old. Furthermore, it was the parents who were initially the most affected and devastated by the initial diagnosis.

In accordance with the current literature on the reactions to the initial diagnosis of a chronic condition or terminal illness, the parents revealed that the period following the disclosure of the diagnosis was characterised by shock and a profound sense of loss (Alexander et al., 2018; Bell et al., 2019; Han et al., 2011; Samson et al., 2009). Processing the loss and the period of grieving following the diagnosis was different for each parent, and the parents reiterated throughout the interviews that, to a large extent, they still felt as though they were grieving the loss of their sons.

The parents described their initial state of shock and panic vividly, and how they truly believed that they would never adjust to their new reality. All of the couples described how little they knew about DMD, and how they knew something was not quite right with their sons from early on. Initial signs included that their sons were slow to reach their milestones, tended to walk on their tip toes, were clumsy and had overdeveloped calves (Emery, 2003). One of the mothers jokingly recalled that she always used to say that if her son was not so clumsy, he could be a gymnast because he had such well-developed muscles, but he always had balance issues. One mother recalled the day her eldest son was diagnosed:

> *I had never even heard of the word Duchenne until that day. Michael was struggling at school and Michael's teachers thought he might be ADD and the school suggested that we take him to a psychologist. He was slow to reach his milestones too. The psychologist noticed signs of ADD but felt that there*

was something more going on. She recommended that we take him to a physio, who picked up that Michael might have muscular dystrophy, but suggested that we take him to a paediatrician [Sarah].

Her husband also vividly recalled his initial reaction of being 'horrified' by the diagnosis of both of their boys:

Yes, and I remember being in a board meeting when Sarah called to tell me what the physio had said. Of course, I approached 'Dr Google' to find out what Duchenne muscular dystrophy was, and was horrified at what came up. The very next day, we took him to a paediatrician, who confirmed that 'all the signs' of DMD were evident. All that needed to be done now was a blood test, as the creatine kinase (CK) levels needed to be checked. At this time, Michael was 6, Jeff was two years old, and Grace was five months old. We decided to check Jeff and Michael at the same time. It was quite traumatic for them as they took blood from their necks (it is the best way for kids). A few days later, the results came back positive for both of our boys (elevated CK levels). We were both shocked.

One mother shared her state of shock and disbelief on the day of the diagnosis, when she exclaimed to the doctor:

Are you going to tell me that my son won't be able to speak eventually and he's going to be in a wheelchair? That is not happening! [Michelle]

Lesley expressed similar concerns, noting her son's developmental delays. In her initial state of shock and denial, she believed that her experience with children with intellectual disabilities would assist her, and there almost seemed to be a glimmer of hope that her son might 'get better'. This knowledge seemed to reassure her at the time:

I always knew there was something going on with Eric, I mean I didn't expect this kind of diagnosis. He was always slow with all his milestones though and before he started Grade R, I think I convinced Mark to have one final check. The paediatrician immediately picked it up. It was his first case ever, and he didn't handle it very well. He basically wrote 'Duchenne' on a piece of paper and told me to google it! [Lesley]

At this point I commented: "Obviously you didn't know what that was at that time."

The first thing I said to him was that 'this isn't a death sentence' because I have the experience with physically and mentally retarded children and my mom used to work with physically and mentally retarded children so I have more exposure to that kind of thing than most people have. Obviously, it was devastating and I spent the whole night reading up on it and I knew without a doubt that that was it! In the meantime, we had blood tests done, but we were both expecting the worst. So yes, it was a terrible, terrible time and very emotional and you have bouts of crying, but you become a lot more hardy because you have to. It is still terrible. [Lesley}

4.4.1 Frustrations with the medical fraternity.

I am very disappointed with the level of support in South Africa – it is pathetic! Ah, I don't know, some people are just handling this the only way they know how. [Lesley]

After the initial diagnosis has been made, parents often experience debilitating anxiety, panic and uncertainty triggered by this completely 'foreign disease' as one participant described, that they somehow have to come to terms with, 'with no answers and a handful of wrong diagnoses' as one mother noted.

Other studies have also reported similar findings (Bally et al., 2018; Gilgoff, 1983; Webb, 2005).

During this tumultuous period in their lives, they turned to medical professionals for reassurance and practical advice. It appears that they initially felt disillusioned by the health care system as they encountered physicians who made incorrect diagnoses or prescribed 'the wrong medicines' or doses. All of the participants in the study described their consistent disappointments and frustrations with the medical fraternity in South Africa vividly. In the following extracts, participants relayed these sentiments:

> *The signs were there but they were never linked to the condition! I don't understand how the medical fraternity didn't pick it up earlier. When we began reading up on Duchenne muscular dystrophy, all his symptoms were there! [Chris]*
>
> *We took him to different paediatricians because we had our doubts; he simply checked his throat, ears and chest and prescribed an antibiotic for flu and sent him off. It was ridiculous. [Sarah]*

Additionally, Michelle and Scott argued that if it was not for meeting other knowledgeable and supportive parents via support groups, they would have had no idea which specialists to turn to or rely on for the best medical care for their son. Michelle conveyed the alarming reality of it all, that some doctors have made mistakes, leading to the rapid deterioration of a boy's muscle mass and overall quality of life:

> *We hear a lot of stories in Pretoria with the doctors prescribing the wrong medicines, too high dose, then taking the kids off resulting in scoliosis and lordosis, then their hearts and lungs fail. [Michelle]*

I then asked: “So that’s why some boys die at such a young age?”

Yes, I can show you a photo of a boy at the doctor who was initially diagnosed, when that boy turned ten she took him off steroids because he was in a wheelchair, now she feels they don’t need steroids when they are in a wheelchair, but the steroids still keep their heart and lungs strong. Now his heart and lungs are taking strain and he’s very thin, no muscle mass at all, all the muscle mass deteriorates, so he’s very weak, he can’t pick up a glass. So, it makes a huge difference how you take care. [Michelle]

I added: “Making those informed decisions, that seems really important…”

Yes – and quality of life is what you are trying to give them [Michelle]

Although the parents suspected that there might have been something wrong with their son early on, inconsistent information and a few incorrect diagnoses from specialists left them feeling confused, frustrated, desperate for some sort of miracle, and disheartened. They were led to believe that:

No one [of the doctors] could be trusted because they all said something different! [Chris].

These frustrations with the medical fraternity were echoed throughout the interviews as the parents sought support, accurate answers and concrete advice, and received very little practical and emotional support at the start of this journey. Having said this, the parents did eventually find ‘the right doctors’, specialists and other medical professionals who continued to provide valuable guidance and support.

As the parents relayed their narratives in vivid detail of how they felt so discouraged and disillusioned by the health care system due to being given little direction by professionals on the basic steps to take, who to see and how to go

about adjusting to a very different way of living, their immense fear of the unknown emerged. A completely warranted fear, too, as this was a life-changing diagnosis for the entire family and the doctors they first encountered seemed to have little experience with this genetic disorder. Research has shown that uncertainty is one of the greatest psychological stressors faced by families, along with disease course ambiguity and the consistent losses that the parents face as the disease progresses (Bell et al., 2019; Han et al., 2011).

Despite these frustrations, nothing could have truly prepared them for the diagnosis of DMD or what this would mean for their family in the long-term. They would learn over time that the impairment effects include muscle degeneration, leading to loss of ambulation, weakness in heart and breathing muscles, the likelihood of behavioural, cognitive and adjustment problems, and early death. (Hoskin, 2017).

In summary, the parents perceived this illness as an intrusive, foreign and destructive force that threatened to destroy their lives and their child. At this time, they demanded answers and solutions. Their perception was that a miracle cure was needed, and as Lesley stated, she believed that the diagnosis was not 'a death sentence' as she compared her experience and exposure to physically and mentally disabled children to this diagnosis. Of course, there is no cure and her son will eventually die, but her initial reaction of shock and denial was similar to that of many other parents, that this cannot be a death sentence, and it is necessary to find a solution or cure.

4.5 Grief and Anticipatory Loss

The thing with any degenerative condition such as Duchenne is you know what is going to happen but you just don't know when it will happen. It is just

learning to live with that knowledge every day. Although it is the most aggressive form of MD, it still takes time. [Lesley]

Throughout the interviews, parents tended to vacillate between hope, intermittent periods of grieving, avoidance of the fact that their son will die, and presence – living each day as if it were their last and finding meaning in their everyday lives. These emotional states are interwoven and often exist simultaneously, and at other times the parents were able to identify periods in their lives where they were clearly grieving more than feeling hopeful, for example.

As the parents relayed their lived experiences to me, an overarching theme emerged, one of grief and anticipatory loss. Additionally, a sense of urgency was revealed in their diction, tone and fast pace at which they spoke, alluding to their desire to grant their sons a short, albeit fulfilling life, and almost being under constant pressure to achieve this. For all of the couples this involved putting plans into action by 'doing things that most kids want to do' in a responsible way:

Well, we are trying to cram a lifetime (70+ years) into eighteen to twenty years. So, we've travelled a lot. We've calmed down a bit now. We just want to make sure they are doing things that most kids want to do. [Sarah]

In the end, we realise that there is nothing we can do to stop our kids from dying. Yes, we can prolong their life and ease their suffering, but they will die, whether they receive the best medical care or not. Boys with DMD usually die from cardiomyopathy. We're lucky because we're spoilt. We get the physio to help us. [Chris]

The biggest thing for us is that people don't understand the daily struggles that we experience to keep our child alive. Catching the simple flu can kill him [Michelle]

At this point, we are managing what he has because we started steroids early at age four, he's stronger than boys that didn't have steroids and weren't strict. We let him jump on a trampoline, but not that much, you know. We limit him and try and save his muscles as much as possible, going down the stairs and things like that, so to keep him walking longer and saving the muscles. [Michelle]

The three couples alluded to the fact that some health care professionals continued to 'prescribe' stages of grief to their behaviour, which was disconcerting rather than comforting at the time. The mothers all expressed that they felt that 'there was something wrong' with the way they were reacting to the diagnosis if they did not experience a set pattern of specific reactions in order to adapt to the many losses they were facing:

I felt that there was something wrong with me or like I was processing it all wrong, the loss, I mean. It took me forever to actually start grieving, and the other mothers I had spoken to, said that they had already accepted the loss and that this had helped them. How did they just move forward so quickly? I didn't understand how some of them seemed to be okay and I just wasn't. [Michelle]

I hated the expectations associated with the diagnosis. I was in shock and felt that I wasn't as proactive as other mothers, seeing psychologists, planning ahead and dealing with it. I grieved, but not in a normal way. I saw someone eventually, a therapist, but it was honestly disappointing. It is hard to explain. You can't just deal with the loss and move on, it's not that simple. I don't think it ever has been. [Lesley]

I couldn't even say the word 'Duchenne' without becoming emotional. I had to basically bury them [Jeff and Michael] back then. For a year and a half, I mourned. It's as simple as that. [Sarah]

The above quotes illustrate that grief is often experienced on a continuum. At the one end of the continuum, Michelle suppressed her grief, and at the other extreme, the grief Sarah experienced was clearly observable by others. These mothers expressed their insecurities about the future, and how incompetent they felt as they attempted to tackle this gargantuan task. The parents felt uncertain and insecure, doubting whether they possessed the necessary skills to cope with the demands of the illness:

I didn't know how to react. We were told that he will die at a young age but he could live to be an adult too. So obviously it is a strange time. In the beginning I felt that there was something wrong with the way I handled things. I was very, very depressed for many months and felt disconnected from everything. I didn't like that the first psychologist I went to made me feel that there was a clear-cut formula to follow. That was rubbish. Maybe it was just because she was young and didn't know how to handle our situation, but it was annoying to me that she kept 'matching' my reactions to a theoretical model of grief! My husband didn't go back to her, and I ended up finding a great psychologist who really seemed to understand what I was going through. I still see her from time to time. [Lesley]

In the above excerpt, this mother refers to Kübler-Ross's very influential and well-known stage theory of grief. The five stages of grief, according to Kübler-Ross, include denial, anger, bargaining, depression, and acceptance (Kübler-Ross & Kessler, 2005). In my training thus far I have rarely questioned the legitimacy,

reliability and usefulness of the stage theory due to its widespread teachings. After this particular interview though, and the ones that followed, I did some more research into theories of grief. I was intrigued by how seemingly unhelpful this psychologist had been by utilising the stages of grief to try to help this mother process her grief in their therapy sessions. Of course, I acknowledged the many other factors that may have been involved in truly connecting with a psychologist or therapist, but it was interesting to note that this mother found the prescription of stages of grief to be annoying rather than comforting or informative. It appears that this mother felt pressured to proceed from one identifiable reaction to another in an almost orderly fashion, which was not how she experienced her grieving process.

Stage theories have remained largely influential in psychology, offering different ways of understanding complex systems of human behaviour and guiding intervention (Kübler-Ross & Kessler, 2005; McGorry, 2007). While the stage theory of grief might be useful to some, the above quotes illustrate that this is not always the case. Many researchers have criticised Kübler-Ross's theory and expectation of recovery in the acceptance stage of grieving to be simplistic, noting that this theory lacks empirical evidence, leading to a prescriptive use by practitioners (Corr, 2015; Friedman & James, 2008; Parkes, 2013).

Therefore it can be seen that while it is useful to consider the existing theoretical models of grieving and Kübler-Ross's enormous impact on the care of dying patients, her well-known stages of grief – denial, anger, bargaining, depression, and acceptance – describe a somewhat oversimplified model that does not account for the diversity in grief reactions. It does not consider the complexities of coping with loss, "[taking] no account of [the] recuperative purpose of avoiding reality of death at times of doing other things to regain strength to cope" (Stroebe,

Schut, & Boerner, 2017, p. 462; see also Parkes, 2013). Many contemporary studies have refuted the use of stages due to their prescriptive statements, implication of a smooth progression, failure to account for secondary stressors (new roles and identities formed and ongoing life changes) and neglect of the social and cultural context of grieving (Bonanno, 2009; Holland & Neimeyer, 2010; Parkes, 2013; Stroebe et al., 2017). Stroebe et al. (2017, p. 455) presented a compelling argument in their article intended to caution health-care professionals that bereaved people are misguided through the stages of grief. They noted that stages of grieving "…should be discarded by all concerned" due to the implicit expectation that a person needs to go through these stages in order to recover, implying that there is a "right way" to grieve when clearly this does not exist.

Stroebe and Schut (1999) proposed a dual process model of coping with bereavement that described the ways that people come to terms with the loss of a close person by confronting and avoiding grief in order for adaptive bereavement to occur. It is evident that this model shares similar traits to the themes of avoidance and presence, derived from the interviews conducted with the parents. This model involves the vacillation between both 'loss-oriented' and 'restoration-oriented' coping. These are referred to as "…two categories of stressor, each of which requires coping efforts during bereavement" (Stroebe & Schut, 1999, p. 212). Furthermore, this dual model refers to coping as being embedded in the parents' everyday life experience.

'Loss-orientation' alludes to the traditional theories that focus solely on the relationship or bond with the deceased person, such as the grief work concept of confronting the reality of death to adjust to life without the person, and Bowlby's (1969) seminal work on bereavement, which focuses on attachment theory and the nature of the loss relationship (Stroebe, 2008).

In the extracts below, the parents described how, at times, they were confronted by the anticipated loss and overwhelmed with fear and uncertainty, and how, at other times, they sought relief by oscillating or concentrating on other things. Oscillation, a cognitive process, is a regulatory mechanism utilised when it is too painful to confront some aspect of their reality (Erby, Rushton, & Geller, 2006; Stroebe & Schut, 1999).

> *I will go with to the specialist, but all the therapy I don't have time for because I work full time. I manage my father's business. I try not to think about Paul's illness, we keep busy on the week-ends and I don't like thinking about what is going to happen in the future. Focusing on other things – that helps me to cope. [Scott]*

While Chris coped by planning in advance and Scott coped by working long hours, Sarah's process included ruminating about her sons' eventual deaths, imagining what life might have been like if her sons could have lived a full life, and mourning the loss of them. She was filled with despair and grief and decided to go for regular therapy sessions. It appeared to me that after this repeated exposure to and confrontation with the anticipated loss, habituation or desensitisation had taken place (Stroebe & Schut, 1999):

> *Chris coped in the initial years by building a room for the boys' wheelchairs etcetera during the first few years, and got all the information overseas, and for me, I couldn't even say the word 'Duchenne' without becoming emotional. I had to basically bury them back then. For a year and a half, I mourned. It's as simple as that. I asked Chris to wait for me to catch up. For us, we knew how important it was to stand together; the kids are already in such a difficult*

place and if the parents aren't supportive of each other, it just makes the whole journey so much more difficult. [Sarah]

Sarah experienced anger initially. She recalled the incredibly painful experience of grieving the loss of both of her sons with DMD. She acknowledged that while this was a painful process, it taught her and her husband valuable lessons that have enabled them to instil kindness, gratitude and the art of living in the present in their children. Furthermore, Sarah conveyed an attitude of hopefulness and gratitude whilst ensuring that they made the most of each day:

I was angry, at God, but that was years ago. We've been through a lot of trauma over the years. I take a happy pill every day and a sleeping pill every night. This must have happened to us to teach us valuable lessons. It isn't really a religious thing as much as a spiritual one to live your life well and be kind to other people. We've instilled those values in the children. We don't go to church, but Jeff is very spiritual. Our family motto is: "Live one day at a time, and make each day count." [Sarah]

Michelle and Lesley mourned in their own way, but revealed that for the first few years their child's health became their main priority. They poured their time, energy and finances into the best treatment for their sons, often at the expense of their own mental health. This was evident when Lesley noted that she coped by delving into research on treatments and the best medical care and steering clear of personal therapy after a disappointing first encounter. Similarly, Michelle said that therapists did not really think of their psychological well-being, and that most of the families she met later on in support groups disclosed that none of them went for individual therapy because the children were the focus of attention. Consistent with these findings, a recent study conducted by 'Parent Project Muscular Dystrophy' in

the USA illustrated that the parents' gravest concern was related to their child getting weaker and finding the right care over time (Peay et al., 2016). Similarly, the parents did not prioritise their own well-being (Peay et al, 2016).

Thereafter, the parents described how they started doing new and meaningful things as a family. Chris and Sarah decided to home school their boys as it allowed them the flexibility and freedom to travel. This provided a healthy distraction from the grief and enabled the entire family to create new narratives, identities, roles and relationships amongst themselves.

It gave us flexibility; we could start travelling and stop dwelling on the loss. They were so excluded in mainstream schooling anyway. [Chris]

They described that they:

...no longer felt like victims of DMD. [Michelle].

Stroebe and Schut (1999) present findings consistent with those reflected in this study: "Whereas emotions come and go, sometimes unwilled and unexpected, in the early days negative affect tends to predominate; however, as time goes on, positive affect plays an increasingly important role in the recovery process" (p. 213). It was evident throughout these interviews, and through the large body of knowledge on this topic, that loss orientation dominates early on in bereavement whereas the restoration-oriented dimension involves accepting the reality of the changed world and taking time off from the pain of grief.

The second type of bereavement stressor is restoration-orientation, which refers to the parents' ability to cope with the stress of everyday life (Stroebe & Schut, 1999). The parents collectively described social loneliness and isolation, and how they dealt with this by joining support groups and reaching out in order to feel connected. It was important for their experience to be heard.

Confrontation with the reality of the eventual loss is the essence of adaptive grieving, but, as numerous other studies have noted, this is a messy and unstructured process that occurs in between other tasks that are concomitant with loss (Bonanno, 2009; Corr, 2015; Friedman & James, 2008; Holland & Neimeyer, 2010; Parkes, 2013; Stroebe, 2008; Stroebe & Schut, 1999).

> *Well, we haven't got to the hard part yet – this is still easy. The hard part will be when they can't move at night and you have to turn them every couple of hours. The steroids are hard – they make them very hungry. [Sarah]*
>
> *We still grieve. That's the thing most people don't understand. They thought we would grieve in the beginning, but then when Paul's calves get weaker, we know that that means he's one step closer to being in a wheelchair. They also have dreams. It's sad to see that he can't open a bottle. Before doctors' appointments I feel extremely anxious because I don't know what's waiting for me. [Michelle]*

In the above extract, Michelle relays the severe emotional stress she experiences prior to each doctor's appointment due to disease course ambiguity, which is a common reaction, according to other studies (Bell et al., 2019; Han et al., 2011). Overall, some aspects of bereavement are changeable whereas others are not, especially in the case of this progressive disease. All of the parents have realised that the damage might be irreversible, but there are certain things that each family could realistically implement and prioritise in order to cope and adapt to living with DMD.

4.6 Learning to Adapt: Living with DMD

The parents alluded to a brief period of passivity in dealing with their new circumstances, where they hoped and prayed for something that does not exist, a

cure. Thereafter, they started engaging in proactive forms of coping, such as educating themselves and seeking emotional support from professionals, family and friends. They experienced a range of emotions, some of which included feeling overwhelmed, isolated and ill-equipped to cope with the situation emotionally, financially and physically. Parents initially believed that they did not possess the resources they needed, as evidenced in the following excerpt from a father:

> *Sarah and I felt like idiots. We knew nothing and were forced to teach ourselves most of what we know today. We felt that we couldn't handle it and prayed for some miracle – well this was wishful thinking! [Chris]*

One mother, Lesley, expressed wishful or magical thinking when she noted that the very first thing she said to the doctor was that 'this isn't a death sentence' because she felt that her exposure to mentally and physically retarded children had prepared her for this diagnosis.

Additionally, the mothers experienced guilt when they realised they were the carriers of the mutation responsible for DMD. This attitude of self-blame has also been well-established in other studies (Chen & Instone, 2008; Rubin, 1987). DMD is an X-linked disease of the muscle, meaning that if the mother's mutant X-chromosome pairs with a Y-chromosome producing a male, the boy will have DMD (Chen & Clark, 2007).

Two of the mothers alluded to the guilt they experienced soon after discovering that they were carriers:

> *I am a carrier – I got tested. Some moms aren't carriers. In the beginning, I experienced a lot of guilt knowing I gave this to my children. [Sarah]*
>
> *I felt guilty. If it weren't for me, Eric wouldn't have this terrible disease [Lesley]*

As a general observation, the mothers processed the diagnosis quite differently to the fathers. Peay et al. (2015) showed that most mothers tended to assume the role of the primary caregiver, expressing themselves more openly and providing more emotional support to their sons, whereas fathers tended to act as providers and distance themselves emotionally from the situation.

In the current study, similar, subtle nuances were observed among the mothers' reactions to the diagnosis, and their experiences of the subsequent challenges they encountered in the first few years shared many commonalities. Although each parent's experience was unique and thus cannot be generalised, the mothers clearly expressed their tendency and preference to express their emotions more openly while the fathers expressed their need to work harder in order to finance all the new exorbitant costs involved in treatment. The fathers focused their time and energy on developing practical coping strategies. While this has inevitably caused some friction in their relationships to varying degrees, Wysocki and Gavin (2006) indicated that paternal involvement often provides a coping resource, supporting both the mother's and the child's adaptive capacity. Overall, the mothers concurred with this statement, stating that it would have been extremely difficult to manage everything as a single parent:

> *For us, we knew how important it was to stand together. The kids are already in such a difficult place and if the parents aren't supportive of each other, it just makes the whole journey so much more difficult. This has actually brought us closer together more than anything else. It has strengthened our connection and marriage. It could have easily destroyed our marriage but we realised that this isn't a 'single parent' journey. It is so important for both of us to be on the same page! [Sarah]*

Scott and I work as a team and he is my biggest support structure. [Michelle] For one parent to do that would be very difficult. And, of course, the complication with Stuart. Getting a divorce would mean splitting the brothers. [Lesley]

Sarah and Chris, whose sons who were diagnosed around twelve years ago, have accumulated a wealth of knowledge and experience over the years in comparison to the other two couples with sons who were diagnosed around five years ago. Furthermore, Chris and Sarah relayed that the intensity of the trauma they experienced has faded over time, and that while some memories of the early days will be forever etched into their memory, they felt much more in control of their lives now compared to parents with sons who have been diagnosed more recently. They appeared calm and confident, and I commented on the fact that they both seemed to be equally informed and accepting of their reality. Chris intercepted:

'No, no, no…I mean, sure, we seem calm now. That's because time heals and our old age helps us to forget the intensity of the trauma (laughs). Sure, we remember certain highlights or lowlights, but if you interviewed us ten years ago you'd probably have a whole other perception of us! Yes, I've always been the more grounded, logical parent, and I suppose I am more practical in general. I don't just let it all out like Sarah does, but then, most men wouldn't spend days sobbing (laughs), but then she was probably there for them, emotionally, more than I was. I had to get out and do something. It hit me at certain times, but generally I prefer to keep my feelings to myself, it's how I'm wired. I do remember working longer hours and taking my mind off things by just working more. [Chris]

Scott appeared to have a similar mentality to Chris in some respects. In contrast to Chris, though, he alluded to how difficult it *still* is for him to accept this reality:

I don't know, I haven't really taken it in. [Scott]

I asked: "Okay, so it's been hard for you to come to terms with everything?"

No, I have accepted it, but I don't want to think about it the whole day. [Scott]

I said: "Okay…"

So eventually when he goes into a wheelchair it's going to hit me bad.

I added: "Of course, it's very upsetting."

I try to work the whole day because if I do nothing I start thinking about it all. [Scott]

Scott expressed that while he has 'accepted it [the diagnosis]', he 'doesn't want to think about it the whole day'. He made it clear that he was not in denial and insisted on the best possible treatment for his son, but he found it easier to cope by distracting himself by working long hours and not looking too far into the future.

Yes, I just want to add, the day Paul was diagnosed, I accepted the diagnosis. I didn't want him to suffer and go without the best treatment possible. We have seen families in denial and the children are suffering as a result. [Scott]

Michelle also acknowledged that as a married couple in their twenties, they would have liked to study towards a degree and lead a more career-driven life, but that circumstances had not allowed for this. Michelle reiterated that seeking psychological support was not deemed essential, or even considered at the time, 'because the children were the focus of attention'.

The biggest thing for us is that when Paul was diagnosed we spent all our money on doctors' appointments. We didn't really think of our psychological

well-being, and with all the families I have spoken to in our support groups, none of them went for individual therapy because the children were the focus of attention. We took Paul to a state hospital for two years, but you don't get much help or any answers and that's when we decided we would rather sell things and start seeing private doctors and now we have a better medical aid. It is better than trying to pay for it all on your own. The MDFSA also just doesn't have the funds anymore, and there wasn't a proper structure in place to see who really needs help the most – for example, the one family we know received two wheelchairs because they were friends with the manager. We have made lots of sacrifices and people don't understand that. At our age we would like to be further in life, but we are dealing with this now and we don't really have a choice. [Michelle]

All of the parents noted that they had not received much financial assistance from the government or from the MDFSA over the years. As a result, they had made many sacrifices to their standard of living in order to put their sons on a reliable medical aid, as the physiotherapy and occupational therapy work out to R8000-00 a month; and all the tests and hospital visits are covered. Their medical aid covers these expenses in addition to any equipment needed (such as the BiPAP, night splints or wheelchair).

The other parents shared similar sentiments, revealing their frustrations with the mismanagement of funds within the association, in addition to always feeling misunderstood by others. Moreover, their focus has largely been on the well-being of their children. Although Michelle has insisted that her son goes for therapy as he has 'become more moody on the steroids', she has not sought personal therapy since the initial diagnosis. At some point, the other parents went for counselling to

learn how to understand and cope better with their sons with DMD, but Lesley noted that she did not feel that they received much practical guidance:

> *I would like to understand where Eric is at. He has been to psychologists and play therapists but we have really struggled. The psychologist only did play therapy with Eric. He did play therapy with Eric for six months and seemed to forget about us. We didn't know how to articulate our needs and we didn't know how to answer his questions. We didn't get enough practical coping techniques to aid Eric in coping.*

These findings are consistent with other studies, suggesting that parental responsibility and commitment to their child's well-being supersedes the focus on their own well-being. Additionally, an over-protective style of parenting suggests a sense of emergency, which serves to ease the parents' feelings of anxiety and guilt (Carnevale et al., 2006; Koufaki, Kedraka, Ierodiakonou-Benou, & Samakouri, 2019).

4.6.1 Appraisal of the illness in terms of daily challenges and responsibilities: Vacillating between hope, grief, avoidance and presence.

Studies have shown that a parent's appraisal of uncertainty undeniably affects his or her ability to cope. Moreover, uncertainty is usually, but not always, perceived as a danger or threat (Bally et al., 2018). In some instances, parents have even alluded to the fact that uncertainty allows for a sense of hopefulness and possibility for positive outcomes for their affected child (De Graves & Aranda, 2008; Magliano, 2014).

Cohen and Lazarus (1979) noted that there is no standard or uniform way of adapting to life after the diagnosis of a chronic illness has been made, and the

cognitive appraisal of a situation or illness is likely to play a role in the process of adaptation.

A primary appraisal refers to the process of gaining a deeper understanding of the consequences DMD is likely to have, while a secondary appraisal would entail the consideration and evaluation of potential coping resources in order to cope with the demands of DMD. Using these resources would ultimately aid the process of adaptation (Cohen & Lazarus, 1979).

The parents in the current study perceived this illness as an outside but intrusive force. The illness was perceived as a loss in the context of the crisis following the initial diagnosis. They were experiencing a crisis and their primary goal was to find a solution or a way of restoring their lives to normality. Furthermore, coping was mainly cognitive in nature and their hope was that through research, an effective cure for the illness would be developed (Alexander et al., 2018; Bell et al., 2019; Han et al., 2011; Samson et al., 2009). Therefore, the parents' hopefulness tended to be concrete and specific. The parents also experienced intermittent periods of grieving as they tried to grapple with the complexities of coping with loss. Each parent experienced grief differently, and the parents tended to suppress their emotions at times by avoiding the reality of the impending death. These findings are consistent with the experiences of other parents raising a child with DMD (Koufaki et al., 2019; Samson et al, 2009).

One father, Mark, clearly was not as comfortable speaking about his challenges or coping strategies as much, which became elucidated when he succinctly stated:

I don't plan on saying much today. I hope that is okay. It really is hard to summarise what we've been through. I prefer to not speak about this, but I don't mind listening or commenting here and there. [Mark]

Lesley also noted that she had grown apart from her son and her husband, possibly due to her clinical or avoidant approach to her relationships.

Although Lesley spoke throughout the interview while her husband remained silent, it became evident that he, in fact, spent a lot more time with Eric than she did as she is often away at work conferences. Furthermore, it could be harder for him to speak about his challenges or grieving process as he appeared to be the primary or consistent caregiver. Speaking about his challenges might be too close to home. In psychodynamic terms, he could have developed this defense or protective strategy in order to cope. Perhaps his reluctance to communicate with me was how he coped on a day to day basis, by repressing painful memories or thoughts, compartmentalising or intellectualising (Lemma, Target, & Fonagy, 2011; McWilliams, 2011). Similarly, Lesley alluded to becoming clinical in her approach. This could imply that she tended to employ intellectualisation to distance herself from the painful emotions she might otherwise experience:

When it comes to Eric, I have become quite clinical and scientific about everything…when it comes to Eric I seem to have grown a thick skin. I don't know why, but that is my coping mechanism now. I think it must be a lot more difficult for Mark because he spends a lot more time with Eric. I travel a lot, internationally as well. So, it must be hard if it is in your face all the time. [Lesley]

In diametric opposition to this, Sarah and Chris were only too happy to share their experiences openly, both good and bad, with me. To me it seemed that they

were managing their everyday challenges and seemed hopeful about the future at times. At other times, they expressed some ambivalence, vacillating between bouts of hopefulness, grief and presence.

Sarah's ambivalence around her ability to cope was elucidated when she said that their family motto was to live one day at a time, and make each day count; she then highlighted the entirely different concept of the importance of planning in advance. In the excerpt below, she highlighted additional stressors and expressed feelings of guilt about her daughter's well-being:

> *While it might look okay now, it isn't always okay. I feel it is important to plan in advance so we can relax when the time arrives. I haven't been to my psychologist in a while, but the Grace thing [healthy daughter] has knocked me quite a bit though. She has been very depressed lately and of course that gets me down. I feel like sometimes I neglect her, unknowingly. [Sarah]*

Once the parents realised that they might have had unrealistic expectations for their child's full recovery and finding an elusive cure, gaining knowledge and practical information became crucially important to them. These findings are consistent with other studies, revealing that a sense of hopefulness for a cure or medical breakthrough propelled the parents to engage in in-depth research in order to find the best possible medical care for their child (De Graves & Aranda, 2008; Magliano, 2014; Samson et al., 2009).

Thereafter, their new goal became facing this new reality and meeting their child's physical needs, whilst keeping up to date with the latest medical and technological advancements. This became more complicated to achieve due to the negative influence of external factors and pressures such as bullying and being excluded from mainstream schools, and personal challenges such as depression,

withdrawing from their spouse, struggling financially, having little to no support from family members, and experiencing personal health challenges. Other factors included the inevitable progression of the illness and the associated challenges like reaching puberty and adulthood, becoming non-ambulatory, and constantly having to re-adjust to the progression of the illness. This learning process required dedication from both parents, and an active involvement in the development of their own personal resources and abilities.

The implementation of new coping strategies seemingly becomes a lifelong process of learning as the parents are faced with two realms of reality. One involves their relationship with their son along with their growing and ever-changing understanding of their son's physical, emotional, behavioural and intellectual challenges and the associated treatments and day-to-day responsibilities. The other realm includes each parent's ability to adapt to a new family dynamic and their relationship with the outside world. In their own unique way, each parent experienced difficult emotions, and through their different grieving processes they developed their own sense of normality. Moreover, parents also experienced increased confidence in their abilities to manage their son's condition as DMD became incorporated into their daily lives (Samson et al., 2009). One father, Chris, and the other mothers related how they felt it was their responsibility to raise awareness and inform newly diagnosed parents of all they had learned through educating themselves through trial and error.

The gargantuan responsibilities and adapting to the challenges of their day-to-day lives often became all-encompassing and consuming for both the child and the parents. They were left feeling drained, isolated and worried about the uncertainty of the future. Similar findings have been reported in families of children and

adolescents living with a chronic illness (Amador, Reichert, Lima, & Collet, 2013; Cornelio, Nayak, & George, 2016; Hinton & Kirk, 2017). Some common challenges included feeling overwhelmed by the loss of mobility, increased dependence, intellectual and behavioural changes or challenges, and the consistent worries about their sons being bullied at school or excluded from mainstream society. This was revealed in the excerpts below:

> *Thirty percent of Duchenne boys are likely to have learning difficulties, um, but researchers haven't found out what the exact effects of DMD are on the brain. It does affect the way the brain develops though, especially with short term memory and reading. So, they tend to be a year or two behind their peers and mainstream schools just don't accommodate them. [Chris]*
>
> *The biggest thing for us is that people don't understand the daily struggles that we experience to keep our child alive. Catching the simple flu can kill him. [Michelle]*

Most of the parents expressed their tendency to worry constantly about their son's health and the progression of the illness. Worry involves thoughts and images that induce negative emotions and are relatively uncontrollable (Peay et al., 2016). Although pathological worry is likely to have dire long-term effects on parents' health, non-pathological worry could serve to motivate proactive and positive health behaviours by taking action sooner rather than later (Magnan, Köblitz, Zielke & McCaul, 2009).

The parents shared similar narratives, pointing to experiences of discrimination and exclusion of their sons from mainstream society due to their disabilities. Disability is defined by the United Nations Convention on the Rights of Persons with Disabilities as a "long-term physical, mental, intellectual or sensory

impairment which may hinder full and effective participation in the society on an equal basis with others" (UN, 2006, p. 5). Research has shown that boys with DMD are likely to have learning disabilities or behavioural problems (Hendriksen et al., 2009; Soutter et al., 2004) and that boys are likely to be non-ambulatory by 10 to 12 years old (Morena et al., 2019; Yamaguchi & Suzuki, 2015). Furthermore, DMD is considered a form of disability. It became clear through the narratives of the parents that all of the boys were systematically disadvantaged and limited in their capacity to navigate their learning and social environments at a mainstream school, leading to their experiencing heightened anxiety levels, bullying, and negative thoughts about death and dying. As a result of this, the parents had decided to remove their sons from a mainstream school. Wu, Chin, Haase, and Chen (2009) showed that children and adolescents with a chronic illness often harbour negative thoughts about death and dying, which is revealed in the excerpt below:

> *Eric is repeating grade two now. At a mainstream school, Eric kind of shut down on an executive level. He just needs more time, and the pace at a mainstream school was just too fast. He is in a special-needs school now, but unfortunately, for the first time ever, he has been bullied at this school. It has been very tough. This bully is quite devious and bullies him behind the scenes. The bully is on the spectrum so we are dealing with it. We've tried to get his stress and anxiety levels down. He is fixated on death. Apparently all children go through this, although I don't remember going through this! He asks existential questions like, 'Am I going to die from this disease?' [Lesley] Paul is also affected by the stares and nasty comments which is very upsetting for him – and for us. [Michelle]*

I am most worried about how DMD will affect him socially and at school as he loves playing and running around' his friends, and the outdoors. Arrhh…that is what's killing me is what it's going to do to that. In South Africa, unfortunately, it is a lot worse than in the UK or USA where there is so much more inclusion. [Lesley]

And we didn't see the point of putting them through so much stress at school and then not being able to walk up the stairs, as they did go to a mainstream school for a period of time. Also, they were often excluded from activities at school and we all decided that it just wasn't worth it. Sometimes some of the teachers also had a problem with Michael's guide dog (Vuvuzela). [Chris]

The above excerpts have shown that children with disabilities face many challenges. In addition to facing a rare and life-threatening disease, these sons with DMD and their families continue to experience social exclusion with their social networks and support systems limited to their families and other youth with disabilities (Schneider & Hattie, 2016).

4.6.2 The importance of planning in advance and gaining practical information.

All of the parents that volunteered to take part in this study reiterated just how crucially important they believe it is to go to every effort to 'raise awareness' of the implications of raising a child with DMD. This was their greatest motivation for participating in the study:

We hope that by talking to you we will educate more people about it. Many people mistake DMD for Multiple Sclerosis, for example, and they're nothing alike. The one is nerve-related and the other is muscle-related! No one really

knows about it or what we go through as parents, and we really need to educate people about this! [Sarah]

When information is lacking or contradictory information is given to the parents, their sense of desperation intensifies as they feel increasingly isolated and overwhelmed at this crucial time (Hinton & Kirk, 2017; Ringnér et al., 2011).

The three couples experienced similar frustrations with the medical fraternity in South Africa, and as a result they decided to reach out to other parents (Ringnér et al., 2011), charity centres, the internet, associations and health professionals locally and abroad (Samson et al., 2009), for the latest information and advice.

Michelle noted that she created her own content based on her experiences, and facilitated a training workshop for the social workers at the Muscular Dystrophy Foundation, shedding some much-needed light on the grieving process she has been through, as well as the emotional strain caused by DMD. She elaborated:

Even the professionals stop caring after a while and they don't realise what it takes to care for someone with a rare disease. [Michelle]

Similarly, Lesley expressed her concern at the general level of ignorance of people, and the inconsistencies of the health care system in South Africa. She was also surprised that I did not know that the Red Cross has the only certified DMD centre, alluding to the general level of ignorance surrounding muscular dystrophy. She suggested that I contact Professor Jo Wilmshurst, the Head of Paediatric Neurology and Head of the muscle clinic for further information. I followed up on this, as reported in Chapter Two, Section 2.13. Furthermore, Lesley endeavoured to get involved in assisting professionals at the Red Cross Clinic to create a patient registry of all DMD patients, which has proven to be a real challenge:

So, this lady started doing the registry, and started engaging with universities across SA. She hit a brick wall, hey, they just didn't want to share information! So, there seems to be a lot of academic competition as well, so she just left it. But she did manage to do a patient registry for the Western Cape. UCT and Red Cross are the only institutions that are doing active work in DMD in South Africa at the moment. There are a few doctors in SA that are trying, though. [Lesley]

Clearly Lesley is a committed parent, trying everything in her power to raise awareness. It was a privilege for me to speak to Professor Wilmshurst, as she shared some possible reasons for the parents' frustrations with the level of emotional support offered at the clinics, which is only partially addressed due to resource constraints (Personal Communication, January 9, 2020). Lesley works at a company which offers short courses. She revealed her interest in enrolling for a CRISPR course (an acronym for genetic modification). Despite the course being offered, implementing genetic modification strategies in South Africa is not yet a reality, due to a lack of financial resources.

So, there's only one thing that will help Eric, and even if it does, it might be too late! And that is called CRISPR. It is an acronym for genetic modification. I mean, you know the science behind DMD? Well Eric has a duplication which is relatively uncommon as well. A lot of people in Johannesburg have the more common form. But with all these treatments, they work on the existing muscle and once that has deteriorated, that's it! [Lesley]

I work at a company called [...] which offers short courses. They are quite well known by Harvard and all the big universities and there is a CRISPR

course offered by them, and I'm going to do that course soon. We would love to get something like this going in SA. [Lesley]

Sarah and Chris have two boys with DMD that are older (Michael is 18 and Jeff is 16). This couple has travelled to America several times over the years to attend DMD conferences. The biggest non-profit organisation in America conducts extensive research studies into finding a cure for DMD. These platforms restored their much-needed sense of hope, as Chris conveyed:

Well, as humans, you want to go to the places that give you the best story. First it was coming to terms with what it all meant for us, and then wanting to believe the best story. I went to a conference in Philadelphia, USA, as a new parent. It was a four-day conference, usually attended by 500+ families, and all the specialists and researchers in their fields were there, along with other families going through the same. We met families whose sons had been recently diagnosed like ours at the time, and others who have known for years. The foundation is the biggest non-profit organisation in America, raises millions of dollars, and was started by a mother who believed that more could be done for her son, and other children living with DMD. So, I learnt a lot about how to raise our children – and then you come back to SA – and the doctors used to say there is nothing they could do for our boys!

Gaining information on the latest treatments and meeting up with dedicated and like-minded people from around the globe was of utmost importance to this couple, specifically after the initial diagnosis had been made. Chris and Sarah did everything in their power to ensure that their boys received the most up-to-date treatment, and Chris soon realised that he knew more about the disease than some of the doctors in SA did, as so little research had been conducted in SA at the time.

As Chris spoke, he revealed his complete dedication to advocating for the latest treatments and I noticed how emotionally charged he became, as this conversation seemed to bring back distressing memories. He confirmed this assumption:

> *Yes – the thought that my boys would not be getting the treatment hundreds of other boys were getting abroad was almost too much to bear. [Chris]*

This is a testament to his love and commitment towards his children. It would seem that he coped by educating himself as much as possible in order to plan in advance, thereby avoiding unnecessary stress and panic.

Despite the fact that there is no cure, the parents turned to research and sought support from other families with more experience and employed practical strategies to cope with their everyday life. Sarah described how she grieved while Chris suppressed his grief and focused on preparing for their new reality:

> *I anticipated and prepared for things that weren't even close to happening yet. So, I bought a wheelchair in 2012 so it was ready and waiting. [Chris]*
>
> *Yes, the wheelchair was always there. We did this so we didn't have to say, 'Oh God, now he has stopped walking, we have to quickly find a wheelchair!'…and the boys enjoyed riding in the wheelchair too before Michael became wheelchair-bound. We did this so we could deal with it better when it happened. [Sarah]*

Several treatment options are available to help slow the progression, including the use of steroids, regular physiotherapy sessions and intermittent positive pressure ventilation. Treatment improves quality of life, life expectancy and ambulation (Yiu & Kornberg, 2008). Chris noted that:

Steroids help preserve muscle function and quality of life [and that] without steroids, children usually stop walking between eight and ten years old and are dead by twenty. [Chris]

The dosage of steroids is dependent on their body mass. Chris indicated that they took 0.75 mg per kg of body mass, up to 30mg of steroids, basically six tablets daily. The side effects include behavioural and emotional problems (Biggar et al., 2006; Dubowitz, 2005). Chris revealed that their sons had to take heart medication along with the steroids and Sarah noted the side-effects that her boys have experienced included stunted growth, weight gain, 'moon face', depression, mood swings and aggression. She added that their sons took calcium because steroids deplete the bones of calcium and vitamin D, which helps the body absorb vitamin C.

Chris and Sarah described the stressful period they endured as they encountered resistance from doctors who refused to prescribe steroids, and insisted on and advocated for the same treatment used in the USA at the time. Chris was infuriated by this, as he had gathered so much information on the latest treatments available at the time after his trip to America in 2006. Lesley also conveyed her immense irritation with the fact that South African doctors still seemed to be very conservative with treatments and that there was no consistency in the health care system. Similarly, Michelle also contended that even now she disagreed with the treatment and would be trying new things. Some doctors refused to prescribe steroids due to the side-effects, and, until relatively recently, steroids were not a recognised form of treatment according to the Lancet Journal.

Chris described this as a frustrating process:

We went to a doctor here and he refused to give us steroids. I went with a huge file of notes from the US. I told him I am not leaving until I have steroids

for my boys. The doctor kept saying that steroids weren't a recognised method of treatment at the time. I thought this was ridiculous because he didn't have an alternative form of treatment either! I concluded that we'd have to agree to disagree, and that we needed steroids! He eventually did agree with us, and later on he became quite a good ally of ours. He always used to tell the new doctors at Baragwanath hospital about me, that I knew everything about DMD. It was a frustrating process, but now it is an accepted treatment in SA. [Chris]

Michelle shared similar sentiments, revealing vividly how challenging, albeit important, it was for her and Scott to find the 'right' assistance. This was a costly process and the family had to make huge financial sacrifices for little Paul. Michelle and Scott are a much younger couple in comparison to the other two couples, and therefore they revealed just how grateful they were to have met Chris and Sarah through a support group, as this couple has been a huge source of support, selflessly and freely sharing their knowledge with Michelle and Scott over the years:

The biggest thing for us was finding the right doctors. Everybody says they know everything about it, but actually they don't know anything. Um… and finding the right therapists, then financially as well. [Michelle]

We paid R 22 000 and our medical aid didn't pay for anything. We had to pay everything cash. We had to find out which doctors to see. No information was given to us that tells you what doctors to see, and if it wasn't for Colin we wouldn't know what we know now. [Michelle]

So most of the people don't have a cardiologist, they [the other doctors] don't refer people to the right doctors. The neurologist did refer us to a cardiologist, but she knows nothing about Duchenne, so she can tell you nothing, so it's

hard. So that is why we drive to Sunninghill to see the specialist there as this group specialises in Duchenne. [Michelle]

I asked: "Did Chris tell you about this [specialist]?"

Yes, he did, thank goodness. Without him we would have been lost. *[Michelle]*

Chris and Sarah have selflessly dedicated their lives not only to engaging with professionals for the benefit of their own sons, but are also passionate about sharing their knowledge and connections with other South African families with newly diagnosed sons:

We have also brought many professors out here to educate our medical fraternity, from pulmonologist to cardiologist, you name them! SA is so behind so we have been trying to bring them to the twenty-first century! [Chris]

If you don't have the funding and medical aid etcetera, there is a lot you can do by educating yourselves as parents. A lot of parents do their own physio and take care of their kids. [Chris]

4.6.3 Support versus isolation.

This sub-theme vividly demonstrates the wide range of support needed and utilised by the parents throughout their journey thus far. A sense of belonging and inclusion was contrasted with feelings of isolation and exclusion.

4.6.3.1 Practical and emotional support.

A myriad of emotional reactions were shown be involved in coping or learning to cope, from relief and pride that one has mastered a new skill or taken the courage to reach out, to exacerbated anxiety, fear and despair at the prospect of often feeling out of one's depth. Parents expressed feeling extremely isolated or lonely at times,

even in the presence of others. The importance of being part of a shared community became increasingly valued by the parents, where they joined support groups and found the 'right' doctors' and other health care professionals who have provided invaluable support and assistance over the years. Furthermore, coping is not only associated with the demands of a particular situation but is also affected by the resources available to the parents (Pat-Horenczyk & Brom, 2007). This is revealed in the excerpts below:

> *Paul's physios are so much more than physiotherapists to us! We enjoy our bi-weekly physio sessions so much. They have made the last four years so much lighter with all their assistance, care and love. They have become such an important part of our lives! [Michelle]*
>
> *I also see a psychologist. She is great. Yes, I'm on anti-depressants and sleeping pills too. Two physiotherapists come to the house on a weekly basis, and have also provided emotional support to all of us. They are amazing. I have two very close friends, but the one doesn't really get it. We always talk about that. It's hard for anyone to understand what we're going through unless they experience it themselves. That's why going to the conference in America is so amazing, because you walk into a room and everyone gets it! I've been three times. Chris went twice on his own, and we went together with the kids in 2010. We went to the conference in 2010 and took the kids to Disneyworld. 'Give kids the world', a holiday resort. All decked out for disabled children, who are terminally ill, and their families. We went to Disneyworld in June and we were ushered to the front of the queue. They provided refreshments to us and the boys. [Sarah]*

We are lucky that we are on a good medical aid. The boys generally have to go for physio two to five times a week. This would be impossible to do if we weren't on medical aid. We are grateful for them. We also take them to the pulmonologist and cardiologist for regular check-ups. [Chris]

Sarah and Chris's faces beamed with joy as they recalled their unforgettable holiday to Disneyworld, which certainly did not entail standing in long queues and the regular treatment! They were astounded by the hospitality and support they received at the holiday resort that catered for families and their children with terminal illnesses. Throughout the interview, they highlighted how grateful they were to be able to attend the DMD conferences in America, as, "Everyone gets it!" Thousands of families just like their own flock to America once a year to attend this world-renowned conference. The sense of belonging, support and inclusion they experienced in America, in addition to the invaluable knowledge they gained, has really changed their perception of what it means to be truly supported.

Michelle and Scott revealed that their faith in God, as well as relying on each other and their close friends, fosters hope and courage in their daily lives.

Lesley acknowledged that while she has an amazing family that served as her support structure, Mark received little support due to their conflictual relationship and his uninvolved family. She coped by involving herself in community projects, attending support groups and got involved in the organisation where she could as she firmly believed that South African parents need this sort of emotional support.

These findings are consistent with the current literature, revealing that most families express a strong need to be part of a shared community, and the importance of social support in its many forms (Hendriksen, et al., 2007; Peay et al., 2015).

4.6.3.2 Social comparison within support groups: positive and negative affective reactions.

An important theme that was woven through all of the conversations was the tendency of parents to compare their current situation to that of other parents in a similar situation to their own, in order to evaluate their own successes, failures and coping strategies. Other recent studies shared similar observations of the tendency of parents to engage in social comparison (Dibb & Yardley, 2006; Hodges & Dibb, 2010). Social comparison is usually implemented during the times of great uncertainty or fear with which parents are inevitably faced at various stages of the disease. There could be both positive and negative effects of social comparison, depending on the interpretation or appraisal of the comparison, and this is likely to, in turn, influence the adjustment process of the family (Hodges & Dibb, 2010).

The parents who volunteered to take part in this study were part of a support group at the time. Despite not always feeling satisfied by the level of support offered in SA, they seemed to find solace in attending a support group or self-help group.

> *In America there are so many different platforms available – support groups and networks. In South Africa we hardly have anything like that. We have become friends with a few different families in SA though, and we share information and get together quite often. [Sarah]*

Research has shown the many benefits of being part of a support group, including gaining practical information, a sense of shared purpose and advocacy, and social support (Law, King, Stewart, & King, 2001).

4.6.4 Positive affective reactions to downward comparisons.

Dibb and Yardley (2006) investigated the role that social comparison could play in adjusting to a chronic illness, within a self-help group. The results revealed

that positively interpreted downward comparisons led to a better quality of life, and that downward comparisons are used by people who are stressed to enhance their self-image (Dibb & Yardley, 2006).

All of the mothers had met other parents whose son's difficulties were far greater than their own, which put their own problems into perspective and increased their sense of capability and confidence:

> *At this point, we are managing what he has because we started steroids early at age four. He's stronger than boys that didn't have steroids and weren't strict. We let him jump on a trampoline, but not that much, you know. We limit him and try and save his muscles as much as possible. [Michelle]*
>
> *Between the ages seven to ten they are likely to be in a wheelchair. I am glad he is seven and still walking. He has a friend that is quite bad and that is upsetting me as he isn't properly maintained. Luckily, we do everything we can and he is still walking. [Michelle]*
>
> *We are on [a medical aid plan]. We are lucky that we can afford this. I mean you can't really blame the parents…not much help is available if you aren't on a medical aid in SA. We know of families in one of our support groups who don't have medical aid, and their son has to literally sit on the floor the whole day because they cannot afford to buy him a wheelchair! [Sarah]*
>
> *We are lucky that Eric is well looked after because many parents don't have this peace of mind. [Lesley]*

The above comparisons led to a sense of gratitude and positive affect for these parents, knowing that all the time, money and effort has not been utilized in vain, and had a positive impact on their son's health.

Sarah expressed concern for other parents who do not have access to financial or other resources, thereby experiencing heightened stress and feelings of guilt on a day-to-day basis. Michelle said that although some other parents did have the financial means to assist their son, some were not strict, or went to the first doctor they could find without doing the necessary research. She noted that some doctors do not prescribe steroids once the boy is in a wheelchair, and described the vast consequences associated with this lackadaisical approach, including the deterioration of the boy's muscle mass:

> *Yes, I can show you a photo of a boy that sees this one doctor. When that boy turned ten the doctor took him off steroids because he was in a wheelchair; she felt steroids were unnecessary. The steroids still keep their heart and lungs strong! Now his heart and lungs are taking strain and he's very thin with no muscle mass. He's very weak, he can't pick up a glass. So, it makes a huge difference how you take care... [Michelle]*

As a result, Michelle and the other parents vowed to not make the same mistakes as some other parents. She and Scott decided to make the necessary sacrifices for little Paul:

> *We took Paul to a state hospital for two years, but you don't get much help or any answers and that's when we decided we would rather sell things and start seeing private doctors and now we have a better medical aid. We have made lots of sacrifices and people don't understand that. [Michelle]*

4.6.5 Positive affective reactions to an upward comparison.

The upward comparison with a parent or family who is better off often results in a sense of hopefulness (Hodges & Dibb, 2010).

In the excerpt below, Michelle indicated that Chris was her role model. Chris coped relatively well and his thirteen-year-old son was still walking. This was something to aspire to, and certainly encouraged an optimistic and hopeful outlook for their son's future:

> *Chris is our role model and we look at his kids and hope that Paul will be the same at their age. Jeff is still walking and he is 13, you can see he has large calves and walks on his tiptoes, but other than that he is okay. [Michelle]*

Although Michelle noted that Chris and Sarah are better off than they are, their journey has inspired her to remain hopeful about their own situation. She has even become a DMD advocate, as Chris has been for several years, visiting clinics and hospitals and conducting DMD workshops to educate the nursing staff.

Additionally, Chris felt a sense of upliftment and hope for his sons when he discovered that physical limitations do not necessarily have to limit their children's goals or dreams:

> *Even though our boys have learning difficulties, it doesn't mean they aren't intelligent. At a conference we attended, we heard about DMD boys who are brilliant and have graduated with PhDs. [Chris]*

4.6.6 Negative affective reactions to an upward comparison.

Michelle described how disheartened and frustrated she felt when comparing her situation and limited resources to another family, who appeared to be in a far better situation than their own. Nevertheless, she also expressed that her difficult past has helped her to cope with her current circumstances, indicative of her resilience:

Scott is still struggling. The thing is I didn't have a good past and that's prepared me for where I am now. I am used to dealing with everything on my own. [Michelle]

Sometimes I do get frustrated and often think, "Why can't other people see things through our eyes?" If you look at Chris's support system, he has an amazing family. Sometimes I feel sad when I compare, but then I realise that that's not our reality and we just have to go on. [Michelle]

4.6.7 A daunting and isolating experience.

In the extract below, Chris alludes to the sense of isolation he and his wife experienced, as well as the often uncontrollable, recurring and ominous dark thoughts that raced through his mind. Imagining both of their sons in a wheelchair, with the need for hospitalisation, and the terrible thought of their son dying, tormented them and generated great suffering, stimulating negative thoughts:

It was so daunting – you almost live in disbelief – and everyone around you seems to disappear. You go through all the practicalities and the diagnosis is confirmed, and then everyone disappears. You wonder where all your friends are when you need them. Dark thoughts seem to rule your mind. It was the strangest time for us. [Chris]

Ja, some of our friends really didn't know how to deal with it, but at least some of our family were really supportive. [Sarah]

Having a child with DMD has been described as a daunting and isolating experience. In addition to finding the right doctors, struggling financially and having to make many sacrifices for their children, the parents described feeling disillusioned, misunderstood and excluded from mainstream society. This social isolation was evident when Sarah recounted:

It was almost as if the wind has been taken out of your sails – and all the people we thought would be around to help disappeared, and those we least expected help from actually did help. [Sarah]

As they looked at the 'ruins' around them, the couple soon realised that they were 'lucky' to have a close family to rely on for support. They would only discover later on the true value of a cohesive support group:

It is a difficult thing to deal with as a friend and so this is why most of our friends drifted from us– they never knew what to say. I realised that I needed to join a support group or something. [Sarah]

This sense of isolation became intensified when Sarah fell pregnant with another son when she already had two sons with DMD. They were grappling with feelings of guilt and confusion. Moreover, the additional judgment and criticism they experienced from family and friends was 'scary' and appears to have tormented them:

It was scary. Family and friends told us we were crazy to take the risk and we thought, "Well, if we have two with DMD, it wouldn't be bad to have another child." We knew what it was all about and really wanted a sibling for our daughter. Right in the beginning when we found out that the lifespan of a child with DMD is sixteen to eighteen, we knew that our daughter would be left on her own. Both Chris and I are very close to our siblings, and we've always believed that we wouldn't have survived without the support of our siblings. [Sarah]

Lesley described the isolating and 'devastating' effect of the diagnosis on her personal life and marriage, while Chris and Sarah openly revealed that the diagnosis could have easily destroyed their marriage, but that it actually strengthened their connection and marriage. Sarah and Chris described how they were able to stand

together in the face of adversity, relating that they are a force to be reckoned with. Michelle and Scott noted that they rely heavily on each other to 'make it work' as they did not receive much external support from their families, whereas Lesley and Sarah both felt grateful for their supportive families.

Being excluded from classroom activities due to their physical disabilities (struggling to walk on their own) and learning disabilities seemed to be another major concern, and all of the sons were bullied at school at some point. As a result, the parents decided either to home school their sons or place them in a special-needs school.

Lesley opened up about her innermost worries surrounding Eric's current struggles with bullying and her concerns about his future, as well as relaying that the diagnosis had had a 'devastating effect' on her marriage and she and Mark had grown apart. She attributed this to their diametrically opposed points of view regarding treatment and how to deal with Eric's intellectual challenges. From having four miscarriages prior to Eric's diagnosis, to harbouring very different opinions and methods of dealing with this rare disease, Lesley stated frankly that, given the appropriate circumstances, she and Mark would have split up. In addition to this, Lesley conveyed her disappointment in the level of support she and her husband received from various psychologists and teachers. Mark noted that they needed practical coping techniques to aid Eric in coping.

Both parents articulated how lost and isolated they felt, not knowing how to answer Eric's questions or help ease his anxiety:

> *There's been little guidance from the psychologists we've seen though, and nothing has materialised over the years. I know if I mention therapy to Mark again, he will freak! [Lesley]*

The psychologist only did play therapy with Eric. He did play therapy with Eric for six months, and seemed to forget about us. We didn't know how to articulate our needs and we didn't know how to answer his questions. We didn't get enough practical coping techniques to aid Eric in coping. [Mark]

Lesley noted that while she was eager to implement remedial interventions immediately, Mark was hesitant, and while she tried to accept and find a way to assist Eric when she realised he was struggling academically by finding an appropriate special-needs school, Mark was:

…in complete denial about Eric's intellectual challenges, for many, many years, and to a large extent, he still is... [Lesley]

She added:

I wonder what it is, if it is a South African thing or a reasonable thing. I actually think it is a male thing, because when I look at the support groups online (and this is all over the world) I would say that 95 percent are female and participate very actively. Not the men. Mark accepted the diagnosis, but only ever cried or sobbed, once, at the start of the diagnosis. Mark does all the physical stuff, the fetching and carrying. He actually has the closer relationship with Eric. The one thing I have noticed is that I have withdrawn from Eric. I'm not sure why. [Lesley]

The above excerpts highlight the complex circumstances in which Lesley and Mark have found themselves. They also highlight the possible cultural and gender prescriptions that often have an impact on the way that grief is manifested and the different interpersonal and intrapersonal processes of each individual and how this may either strengthen or weaken relationships. Additionally, studies have shown that the way a wife grieves, for example, is likely to affect and change the husband's

grieving process (Stroebe & Schut, 1999). Research has shown that a mother's grief tends to be more loss-oriented, while a father's grief seems to be more restoration-oriented (Stroebe & Schut, 1999). As a result, Lesley expressed feeling that Mark grieved less than she did, while he could have been grieving in a different way, but did not know how to confront, acknowledge or address these differences.

One mother, Michelle, spontaneously shared a poignant piece of writing with me during our interview. She said that she enjoyed writing about her experience as it was an outlet for her. It was an honest, heartfelt essay, entitled 'What rare disease means to me'. It depicts vividly her personal journey thus far. It amplifies the reciprocal relationship that exists between her son's illness and her daily functioning. It also highlights that medical expertise for a rare disease such as DMD is a scarce resource in South Africa, meaning that since the diagnosis, Michelle has dedicated her life to assisting her son in every way she could:

To me, rare disease means driving miles to seek medical care because not every city and state has a doctor that understands rare.

Rare is scary, it's isolating. It's getting up and wondering how my child feels today. That magnitude of worry is consuming. With every breath I take, I inhale and exhale his rare condition.

Rare is the dark circles under my eyes; it's sleepless nights and the cause of my insomnia. Rare is the lack of funds in my bank account.

Rare is what comes between my son and the world – it limits him. Rare has flipped my world upside down and forever changed me. It keeps me on my toes; it keeps me searching for answers.

Rare finds a way to be acknowledged even during the happiest of moments. Rare will keep your eyes posted to every genetic study being done

in the hope that maybe, just maybe, today might be the day that someone can help take away his pain.

Michelle's descriptive essay truly highlights the extent to which her life has become consumed by her son's illness and encompasses the constant worry, the immense sense of isolation she experiences, the insomnia, and the hope that 'someone can help take away his pain'. It also alludes to the level of exclusion she faces as the parent of a child with a rare disease, as well as the exclusion her son has faced due to his physical and learning disabilities – '*Rare is what comes between my son and the world – it limits him"*. She described what appeared to me to be a selfless and poignant depiction of dedication, as well as providing a glimpse into the life of a mother raising a child with a rare disease.

Moreover, the other parents alluded to, and identified with similar daily stressors. Some of the major stressors revealed by the parents include financial strain, worry, feeling misunderstood, and judged by close relatives during an already uncertain time when their greatest desire was to be supported unconditionally:

I just want to add that our families haven't been very supportive. Scott's parents don't accept the diagnoses and believe that Paul is fine. My sister doesn't really understand either. It was her son's birthday party and she arranged to take the kids to a water park and I immediately said that it would be too strenuous for Paul's muscles. She blocked me on whatsapp. Scott's father also criticises me by saying I don't allow my children to just live. I will do anything in my power to keep him out of a wheelchair as long as possible, even if that means missing out on a day of fun. People make ignorant comments and judgments, for example, one woman said he won't learn to

walk properly if he rides around all day and they think he is a spoilt brat because he rides around on a fancy scooter. [Michelle]

It was scary though, the criticism and judgment we experienced when Sarah fell pregnant with Kevin. I have actually written a story on how Kevin came to be. I'll send it to you. [Chris]

The challenges and varying degrees of exclusion from mainstream society reported in this study are consistent with those reported in other studies developed in the contexts of chronic, rare and terminal illnesses (Hinton & Kirk, 2017; Leite, Garcia-Vivar, Neris, Alvarenga, & Nascimento, 2019; Samson et al., 2009). The term 'inclusion' is used to refer to recognising and respecting the differences among all individuals regardless of their age, gender, sexual orientation, language, class, disability, ethnicity, culture, and race. It focuses on supporting all individuals within the education system and society as a whole, so that a full range of needs can be met and barriers can be overcome, through adaptations to the system and society (DoE, 2001). Despite the recent efforts to move toward the full inclusion of individuals with disabilities in the institutions of South African society (such as schools), Schneider and Hattie (2016) revealed that learners with disabilities continue to experience social exclusion, with their social networks and support systems often limited to their families, support groups and other youth with disabilities.

4.7 Beyond the Illness

Yeah, so basically with Duchenne life still goes on as normal. There are all the other challenges which we would face anyway, but I guess life is complicated by Duchenne. We have met so many amazing people on this journey through, so we feel blessed in many ways. [Sarah]

As the parents relayed the intricate details of their personal stories to me, it became clear too that leading a meaningful life entailed integrating their son's illness into the family dynamics in a way that encouraged rather than stifled a sense of family hope. Throughout the interviews, it became clear that a systemic perspective needed to be taken into account as families do not exist in a vacuum. Furthermore, hope is influenced and altered by the reactions, coping strategies and worldviews of the family system in its entirety. These parents have experienced the dynamism of hope by continuously striving for balance, no matter how elusive the concept.

For example, Sarah and Chris vividly relayed how thrilled they were when they decided to invest in service dogs for both of their sons, after the conference they attended in America. These four-legged friends have 'brought so much joy' to the entire family, Sarah noted. This was enlightening to hear as these Labrador retrievers are true companions to their boys, assisting them with practical activities that they would otherwise struggle to do on their own, as well as providing comfort and affection with their gentle natures. As mentioned previously, Michelle and Scott have become friends with Sarah and Chris over the years and described that as soon as they witnessed how incredibly loyal and helpful these dogs were, and the positive impact they made on their sons' lives, they immediately organised a guide dog for their son too. These dogs are an embodiment of hope and joy.

Chris and Sarah's faces lit up as they spoke about their dog, Vuvuzela:

> *Both Michael and Jeff have guide dogs, and they're now allowed to take them into a grocery store with them. [Chris]*
>
> *The Guide Dog Association trains them for about eight months. They are truly amazing. [Sarah]*

He turns on the light, opens the door, picks things up. He is a lab retriever named Vuvuzela. We found out about service dogs in America, and then looked for a litter as soon as we got back to SA. Our boys absolutely love their dogs, ya, so these little things have really made a huge difference to our everyday lives. [Chris]

Although their appraisal of the illness is often dependent on the progression of the illness and therefore changes, all of the parents described how they have never lost hope. Despite the uncertainty, bouts of depression and hopelessness and lack of support, they have all gained meaning and purpose through the adversities. This was exemplified in Chris's words towards the end of our interview:

It's been one heck of a journey, but we still feel that there is a lot to be grateful for, and a lot to still look forward to. [Chris]

Additionally, Sarah and Chris revealed that embarking on a spiritual journey has helped them to live in the moment' as much as possible. They truly believe that this must have happened to teach them valuable lessons. They conveyed their journey to be more of a spiritual journey than a religious one, instilling values of kindness in their children and showing acts of kindness by consistently raising awareness, leading support groups and helping other parents who are going through a similar experience. They have formed 'friendships for life' with some other parents in their support groups, including Michelle and Scott.

This must have happened to us to teach us valuable lessons. It isn't really a religious thing as much as a spiritual one: live your life well and be kind to other people. We've instilled those values in the children. We don't go to church, but Jeff is very spiritual. Our family motto is: "Live one day at a time, and make each day count." [Sarah]

Hope was intricately connected to finding a purpose, living by their values, a sense of spirituality, connecting with others and the search for a meaningful quality of life:

So, we started living, we really started doing meaningful things together. We travelled and gave them experiences. It was amazing; it is amazing. [Chris]

The parents expressed the importance of communicating effectively with their sons in a way that did not pathologise their behaviour, thereby empowering them to make some of their own decisions, as well as integrating their son's illness into the life of the family. Sarah articulated:

We treat them like normal children, not like they're sick or dying because we know then they'll behave like normal children. [Sarah]

While the other two couples have much younger sons, Chris and Sarah's sons are teenagers now. Michael (18) did not reach puberty as his growth was stunted due to the steroids he had taken on a daily basis. His parents respected his wish to 'feel like a man', giving him the necessary information on possible treatments. As a result, he decided to start taking testosterone supplements:

Our boys are likely to face new challenges now as there isn't much research on adults living with Duchenne muscular dystrophy as it was highly unlikely for boys to live past the age of sixteen. Due to improved treatment (steroids, heart medication) and regular physiotherapy sessions, they are living much longer. This comes with a whole new range of new problems. For example, Michael didn't reach puberty and it was his decision to take testosterone as he wants to feel 'like a man'. [Chris and Sarah]

Additionally, the parents highlighted the importance of open communication in the family system, and not neglecting their other children, even though this was not

always as simple as it sounds due to the amount of time they dedicated to their sons. While Michelle and Scott's daughter…

> *…is too young to understand what is really going on, she will have a normal childhood as she is included in [our everyday lives]. For example, she enjoys riding with her brother on his mobility scooter, and when Paul has outgrown his splints she gets them to play with, and they are stretched together. [Michelle and Scott]*

In the excerpt below, Sarah refers to the other serious challenges their family has recently faced. Grace, her daughter, has been adversely affected by her father's ill health and clearly suppresses her emotional state and she has become depressed. Nevertheless, Sarah revealed that while the siblings will inevitably suffer along the way, she feels that they lead a relatively normal life with the usual challenges that most other families face:

> *Yes, we chose to tell our children in an appropriate way, so both Kevin and Grace know what's happening with their siblings. They seem to be fine with it for the most part. We acknowledge that our DMD boys do tend to get more attention due to their condition. I think Grace is feeling it now though. I have started taking her for therapy, but I am sure a lot of it has to do with being a teenager, being hormonal. She has also had a lot to deal with, with her father. Chris has had cancer, and he is the rock in the family. And then he also had a heart attack last week, so it is a lot to deal with. She also doesn't talk very well about what is going on with her. She doesn't want to worry me, I suppose, um, I think the kids do suffer along the way, but no more than a normal family would I think. I'm not saying that they aren't affected at all by what goes on at home. [Sarah]*

4.8 General Discussion and Conclusion

Initially the parents perceived this illness as an intrusive force, and their primary goal was to find a way of restoring their lives to normality. Their appraisal of this state of shock and uncertainty meant that their sense of hopefulness was based on finding a cure and gathering as much information on DMD as possible (Alexander et al., 2018; Bell et al., 2019; Han et al., 2011; Samson et al., 2009). The treatments that are currently used to slow progression include steroids, regular physiotherapy and occupational therapy sessions, and the use of splints to prevent muscle contractures (Esterhuizen et al., 2016; Yiu & Kornberg, 2008). Michael uses a wheelchair and the other boys use electric scooters to maintain mobile independence, in addition to regular physiotherapy, occupational therapy and specialist visits.

The parents relayed their many disappointments and frustrations with the medical fraternity in South Africa. These frustrations with the medical fraternity were echoed throughout the interviews, as the parents had sought support, accurate answers and concrete advice and had received very little practical and emotional support at the start of this journey due to the lack of awareness and medical expertise around treating DMD. Information often varied between hospitals, and some doctors had very limited exposure to working with DMD patients. Similarly, schools and psychologists seemed to be relatively unaware of how to assist these boys (Bushby et al., 2010).

All of the parents noted that, despite their attempts to receive financial aid, they had not received much financial assistance from the government or from the MDFSA over the years. They all referred to the general lack of support available in South Africa in comparison to the UK and US, where so much more support and

inclusion exists. This need for greater support and inclusion in a South African context has been acknowledged in recent South African studies (Esterhuizen et al., 2016).

Hoskin (2017) argued that disability is a form of social oppression in which individuals with impairments are excluded by society:

> "According to the social model, it is not a person's impairment that disables them, but society itself which denies disabled people access to opportunities open to the non-disabled population" (Hoskin, 2017, p. 176).

This quote clarifies the misconception that disability is the burden, when lack of access is in fact the biggest burden. Hoskin (2017) explained that, while this model has been effective in the implementation of some economic, social and political rights for disabled people over the years, it is important to consider the 'impairment effects' that no level of accessibility can remove (Hoskin, 2017, p. 177). Some impairment effects include muscular degeneration leading to a weakening of the heart and breathing muscles, adjustment, and intellectual difficulties. The parents in this study also acknowledged that DMD is a demanding degenerative condition compared to other types of muscular dystrophy and some chronic illnesses.

Nevertheless, their hope is to empower their children in such a way that they are able to find purpose in their everyday lives, rather than to oppress or belittle them for what they cannot do.

The parents took matters into their own hands with regard to schooling when they noticed that their children were being excluded from everyday activities in the classroom. They believed that keeping them in a mainstream school would only lead to a lack of confidence in their abilities. It therefore seemed that the problem was not

always about their son's disability but seemed to stem from society's view or misconception of their son's abilities.

The parents fought for inclusion in their own lives and those of their children. Initially they prioritised finding a cure. However, they came to the realisation that they were 'so busy researching and worrying about every minor detail of DMD', as Lesley reiterated, that they forgot to pause and ultimately to live a meaningful life. They began to utilise a social model (Hoskin, 2017) in their everyday lives, acknowledging the importance of communication with their sons, developing close relationships and friendships and encouraging their sons to do the same, 'rather than suffocating them, as they also deserve to live and have some fun' (Sarah). They are all responsible parents, with varying levels of comfort when it comes to exposing their sons to high-impact activities due to the irreversible muscle damage that could occur, but they ensure that they are active as much as possible.

Parents revealed that they tended to move between different states: hope, intermittent periods of grieving, avoidance of the fact that their son would die, and presence – living each day as if it were their last, finding meaning in their everyday lives. They experienced the grieving process in different ways while attempting to grapple with the complexities of coping with loss (Corr, 2015; Friedman & James, 2008; Parkes, 2013; Stroebe et al., 2017; Stroebe & Schut (1999). The mothers described how 'annoying' and 'unhelpful' the health professionals were as they utilised a stage theory of grief. They expressed feeling pressured to proceed from one identifiable reaction to another, which was not how they experienced the grieving process. Stroebe and Schut (1999) proposed a dual process model of coping with bereavement that described the ways that people come to terms with the loss of a close person by confronting and avoiding grief in order for adaptive

bereavement to occur. It is evident that this model shares traits similar to the themes of avoidance and presence derived from the interviews conducted with the parents. This model involves the vacillation between 'loss-oriented' and 'restoration-oriented' coping. These are referred to as "…two categories of stressor, each of which requires coping efforts during bereavement" (Stroebe & Schut, 1999, p. 212). Furthermore, this dual model refers to coping as being embedded in the parents' everyday life experience.

Peay et al. (2015) found that most mothers tended to assume the role of the primary caregiver, expressing themselves more openly and providing more emotional support to their sons, whereas fathers tended to act as providers and distance themselves emotionally from the situation. Similar observations were noted in this study where the fathers tended to focus their energy on developing practical coping strategies while the mothers expressed their emotions more openly. In addition to this, Abi Daoud et al. (2004) revealed that fathers tended to have more difficulty accepting the diagnosis, which was evident in this study. Wysocki and Gavin (2006) indicated that paternal involvement provides a coping resource, supporting the adaptive capacity of the mother and the child. The parents in this study agreed, noting how difficult it would be to raise a child with a rare disease on one's own. Interestingly, all the mothers expressed their disappointment with some of the health care professionals they had encountered who prescribed stages of grief for their behaviour but subsequently provided little reassurance or emotional support. They felt that the stages of grief described an oversimplified model, often not accounting for the diversity in grief reactions due to the social and cultural context of grieving (Bonanno, 2009; Holland & Neimeyer, 2010; Parkes, 2013; Stroebe & Schut, 1999; Stroebe et al., 2017).

The dual process of coping with bereavement, as proposed by Stroebe and Schut (1999), was applied to the grieving experiences of the parents. This was evident when the parents relayed that, at times, they were confronted by the anticipated loss and overwhelmed with uncertainty. At other times, they sought relief by concentrating on 'other things'. Confrontation with the reality of the eventual loss is the essence of adaptive grieving, but this is a messy process that occurs in between other tasks that are concomitant with loss.

Planning in advance and gaining practical information became crucially important to the parents, and through their research into finding specialists who could help they experienced some disappointments, and later met some remarkable health care professionals who have provided invaluable support (Hinton & Kirk, 2017; Ringnér et al., 2011). A notable finding is that the parents did not prioritise their own well-being; as Michelle said, "With all the families I have spoken to in our support groups, none of them went for individual therapy because the children were the focus of attention". Similarly, a current study documenting parents' worry prioritisation revealed that child disease management was prioritised over their general well-being (Peay et al., 2016).

Adapting to their new reality and meeting their son's physical needs was shown to be complicated by external pressures such as witnessing their son being bullied and excluded at his mainstream school. Additionally, they encountered far-ranging personal challenges too, including conflict in their marriage and other relationships, financial stress, experiencing unreliable or inconsistent support from others, depression, and personal health conditions (Chris had had cancer and a heart attack). Parental employment and education seemed to play a mediating role

in the adaptation process. These findings are consistent with other studies (Chen & Clark, 2010)

Other stressors included the inevitable progression of the illness, requiring the parents to constantly re-adjust and re-examine their changing reality. One could say that they are faced with two realms of reality. The one realm involves their relationship with their son, understanding the emotional, physical and intellectual challenges and implementing the required treatments depending on the stage of the illness. The other realm includes the adaptation to a new family dynamic and its relationship with the outside world. Through each family's unique way of processing these realms of reality, they noted that they had developed their own sense of normality.

Parents adopted different ways of coping. Although they strove to live by the family motto 'live one day at a time, and make each day count', they often expressed some ambivalence regarding their ability to manage everything, often vacillating between bouts of hopefulness, grief and presence. It was, however, evident that parents had not always been as hopeful, optimistic or accepting of living with a child with DMD as they were when interviewed.

Although some studies have shown that social and economic challenges often serve to oppress disabled people and lower their drive or aspirations (Campbell, 2009), some of the parents in this study revealed that, to a large extent, their sons had not internalised all of the external 'oppression' they had experienced, such as bullying and being excluded at a mainstream school. Having said this, Lesley did express concern about Eric's fixation with death. He is still quite young and may grow out of this way of thinking with some assistance, but it is clear that he has internalised this fear of death and dying, possibly due to bullying or a lack of effective

communication between him and his parents. Lesley added that he is doing better now, since they moved him into a special needs school. Sarah and Chris also noted that they have really seen a change in their eighteen-year-old son's demeanour since they started giving him more freedom and encouragement to exercise his own autonomy and make his own choices, but that this transition has not been without its challenges. These findings are consistent with the current literature, showing that getting their sons' needs met while encouraging their independence is often a challenging task (Abbott et al., 2012).

The findings in this study have shown that different forms of peer support, support groups, and a sense of belonging and inclusion have been an essential coping strategy, particularly due to the nature of this rare genetic impairment (Hendriksen et al., 2007; Peay et al., 2015). Additionally, once parents joined a support group, they tended to compare their current situation to that of parents who were perceived to be either worse or better off than themselves. This had both positive and negative affective reactions (Dibb & Yardley, 2006; Hodges & Dibb, 2010). Overall, their perceived level of support and their ability to cope were often linked to their appraisal of DMD (Cohen & Lazarus, 1979). Furthermore, it seems that each parent's appraisal of health, rather than the son's actual health status, is a better indication of parental coping.

Seeing beyond the illness and learning to live a meaningful life entailed integrating their son's illness into the family dynamics, which seemed to encourage family hope. Family hope has been fortified through the guide dogs that have brought so much joy into the lives of the entire family. Family hope is also evident in the constant search for a meaningful life. This could mean different things for different people. For some, it may mean bridging the gap to improve the quality of

life of their sons and other children with a rare disease through advocacy and empowerment. It could mean rediscovering one's strengths or talents, or it could mean learning to live in the moment, cherishing the special moments that will be forever etched into their memories.

Their main hope in participating in this study was to highlight some of the inconsistencies of the health care system in South Africa and, through a collaborative process, to create a more inclusive and supportive environment for other parents and families who are likely to experience similar challenges. Listening to their complete dedication as parents and their hope of creating constructive change was truly inspiring to me. They have already committed so much time to this cause, and it is exciting to think about what could still be achieved by raising awareness through a more collaborative process. By raising awareness, we can improve early diagnosis, encourage policymakers to increase access to services and fund research into rare diseases, as well as reduce the feelings of isolation or the discrimination faced by families.

4.9 Limitations

Any study has its limitations. One limitation of this study was its focus on parents in general, not specifically the parent who was the primary caretaker for the child with DMD. This focus on parents in general also did not take into account the different responses that might have been obtained had the parents been interviewed separately. In addition to this, because the parents were married and had at least some level of spousal support, studies should be conducted with single parents or other families to see if similar results are obtained.

The selection of the sample in itself was a limitation. It is likely that there was a selection bias through using a support group to recruit participants. This was possibly not inclusive of more marginalised populations.

Although this was an exploratory study involving a small sample, it shows the feasibility of recruiting parents to discuss such a sensitive topic. I had initially hoped to include interviews with a much larger sample group and from a vast range of backgrounds more indicative of our South African population. However, out of the seven couples who responded to my request to take part in the study, only three couples committed. The overall response rate could have been low because, as I discovered through the interviews with these three couples, these parents lead exceptionally busy lives. The small sample was also because some parents were not ready to speak to anyone about their challenges. They stated this as their reason for not participating in the study.

This study was limited in its inclusion of families within the Johannesburg and Pretoria regions. It focused on families in which children were not in the more severe and debilitating stages of the disease, although Michael is already in a wheelchair. Thus, the parents in this study may not be representative of parents with DMD as a whole, so these findings are possibly not more widely generalisable.

Despite these limitations, this study has provided valuable insights into the lived experiences of parents raising boys with DMD. In the next chapter I consolidate these findings, make recommendations both for families with a boy with DMD and for future research, and reflect on the process of this research.

Chapter Five: Conclusion and Recommendations

5.1 Introduction

This study set out to explore the lived experiences of the parents of three families with sons with DMD. The study put the following objectives forward in answering the research question:

- To explore the lived experiences of parents raising a child with Duchenne Muscular Dystrophy (DMD); and
- To identify the needs of parents who are raising a child with DMD within the South African context.

The above objectives were met through in-depth interviews that yielded four main themes, namely, reactions to the diagnosis, facing the loss, learning to adapt, and looking beyond the illness. Sub-themes arose within these. In this final chapter I revisit the main findings. From there I offer recommendations for the families and the professionals working with them, and then for future research. I conclude with a reflection on the research process.

5.2 Research Findings

The devastation and shock caused by the initial diagnosis of Duchenne Muscular Dystrophy created pandemonium in the parents' lives, catapulting them into a world that appeared to be diametrically opposed to the one they were familiar with. Despite the parents noticing peculiar signs and atypical symptoms in their boys, such as being slow to reach their milestones, having abnormally muscular calves, their clumsiness and 'balance issues', and their tendency to struggle at school, their suspicions were refuted by medical professionals on more than one occasion. After what had been described as a long and frustrating process, a diagnosis of DMD was confirmed by a blood test indicating the high level of creatine kinase in their son's

blood. This was a turning point for the entire family. Not only had they never before heard of DMD, but they soon realised, to their shock and dismay, that neither had some of the health care professionals, due to having had little exposure to treating a patient with this rare, genetic terminal illness.

After the diagnosis, the parents revealed that there were so many unanswered questions, so much uncertainty. They expressed feeling overwhelmed and worried of what was to come and which steps to take next. Initially they felt incapable of coping with this foreign and intrusive force and the gargantuan responsibilities. All of the parents noted their many frustrations with the medical fraternity due to the perceived lack of support. The mothers experienced guilt at the reality of having passed DMD onto their sons. They also expressed feeling that they were not provided with the necessary reassurance in these beginning stages.

Grief was, and still is, a big part of their journey. The parents have faced many losses, most stoically. They expressed grieving the loss of their 'normal healthy son' as well as their hopes for his future. They noted that they grieved the loss of their lifestyle and the many close relationships they once had. They expressed anticipatory grief, as each day serves as a reminder of the final, inevitable loss of their son. Interestingly, the parents felt that their grieving process could not always be applied to the well-established stage theory of grief by psychologists and other professionals, due to the messy and unstructured process of grief.

The process of adapting to life with DMD required many sacrifices to be made, ranging from physical and financial stress to personal health issues and marital problems. The parents endured heartache at the prospect of all the losses they would still face, but they coped by immersing themselves in their son's daily life. The two realms of reality included their growing understanding of the illness and their

relationship with their son, and adapting to a new family dynamic and developing a new narrative and relationship with the outside world.

It was vitally important for the parents to educate themselves and join a support group. This seemed to lessen the intensity of the social isolation they experienced and created a much-needed sense of support and inclusion. Slowly, with the help of their spouses or close family members, they regained confidence in their abilities and felt capable of managing the day-to-day responsibilities.

Amidst the unique and far ranging challenges experienced on a daily basis, the parents started making small changes that have had a positive impact on the lives of their sons, and the entire family. Some of these included forming close friendships with other parents in a similar situation, rediscovering their own strengths and zest for life, making their sons lives easier by valuing their concerns and making the necessary changes such as moving them out of a mainstream school. Most importantly, they have dedicated much of their time to advocacy and empowerment in the hope of raising awareness on the practicalities and emotional impact of DMD, due to the inconsistencies in the health care system.

They reiterated how important it has become to appreciate the special memories made, rather than wishing for a different reality. They have found ways of cultivating meaning and family hope in their everyday lives, doing everything in their power to grant their sons short, albeit fulfilling lives. Not only were they more than willing to divulge their personal narratives to me, but since the interviews they have spontaneously shared updates on what has been happening in their lives. Chris and Sarah welcomed me into their home, where the interview was conducted. They introduced me to their beautiful guide dogs and children. Their warmth, kindness and dedication as parents astounded me and made this process so rewarding. They have

made research and education their mission over the years, selflessly sharing their knowledge and time with other parents. Michelle reiterated throughout their interview that Chris is her 'role model'. Michelle and Scott were so grateful that I had shown an interest in their lives, and DMD in general. She shared her poetry and the contents of her training workshops with me. She gave a heartfelt depiction of the challenges she has faced and how happy she is that her son is doing really well at the moment, and that all the hours of therapy and treatment are paying off. Since the outbreak of Covid-19, she has been in touch with me. She noted that despite the chaos and daily challenges, she feels 'more alive than ever' and has started making masks and leather handbags. She feels that she is now more than a mother to a child with a terminal illness, but still looks forward to continuing with her training at the hospitals once it is safe to do so. Lesley and Mark appeared to have a strained relationship. Nevertheless, they have put their son first. Lesley has enrolled for a course in genetic modification at the company where she works.

My impression of these parents is that they will look back on the therapies, appointments, sleepless nights, tears, triumphs, milestones, ignorance, struggle, strength with few regrets, knowing that it was both heart-breaking and painful, but definitely worth it.

5.3 Reflections

The initial choice of this research topic was inspired by my mother, who lost her first daughter at the age of five due to a rare, terminal genetic disorder. She faced numerous challenges throughout her journey, including uprooting her life and moving to the Johns Hopkins Hospital in the USA, where she lived, in order to receive the treatment and support that was not available in SA at the time, while her husband worked in SA. I have often wondered how she coped under the

circumstances she was faced with, and how other families cope in similar situations. It was these ruminations that led me to want to undertake this research study. Furthermore, the connection to the study is quite personal, and my hope was to implement awareness and positive change.

My supervisor reiterated how emotionally draining or depressing this study must have been for me. While it might seem like a draining or depressing topic on the surface, I did not experience it this way. I found it to be a richly rewarding and worthwhile experience. I felt immense empathy for these parents, and although I have not experienced it myself, I felt connected to them in some way. I experienced frustration as they described some disappointing experiences with the medical fraternity, and sadness at the thought of the isolation and heartache they endured. I felt snippets of their pain and their grief, as well as their hope and fulfilment. I found the topic to be enriching and interesting and thoroughly enjoyed the learning process. I hope to conduct further research on a larger sample size one day.

In the beginning I was apprehensive at the prospect of interviewing parents who seemed to be facing unimaginable horrors and the potential wounds I might have gashed open. Nevertheless, I persisted and followed through due to my keen interest in the topic. I can honestly say that it was a privilege to speak to these parents, who truly opened up to me, unexpectedly expressing their innermost thoughts, emotions and concerns, and expressing their gratitude throughout the process. The youngest couple who are more or less my age have contacted me several times, asking if I need any more information, and subsequently sending valuable guides and personal poems to me. They noted how much they enjoyed meeting me and how they felt I was making a difference by simply raising awareness in SA. This was a heart-warming experience, as although this study provided only a

glimpse into their complex lives, they found value in it, being willing to assist me in every way. Michelle also noted that no one really shows an interest in DMD unless they have a child with it, and that it felt good to talk about it 'to someone who cares'. As a result of these interactions, I have become increasingly passionate about advocating and educating the general public as I know how important raising awareness is to these families.

I was aware of being younger than most of the participants and of being enrolled for my Masters in Psychology, and of the potential biases this may have created in not being in a position to understand due to my age and never having been through this myself and not being an expert in some way. Despite these concerns, it appeared that these factors did not come into play or influence the parents' perception of me as they shared openly and honestly, almost as if I were a friend or a trusted companion. Of course, one father, Mark, set out his boundaries at the start of the interview, stating openly that he would not be saying much. Strangely enough, the fact that he remained silent throughout most of the interview did not feel awkward. I still appreciated the fact that this might be his coping strategy, and that despite this, he agreed to participate.

Despite the diversity in beliefs, varying support structures and resources that each family portrayed through their unique narratives and amidst the uncertainty they faced, each family shared an innate belief that life is primarily a quest for meaning and hope, no matter how dire the circumstances may be, as expressed by Viktor Frankl:

> *"We must never forget that we may also find meaning in life even when confronted with a hopeless situation, when facing a fate that cannot be changed. For what then matters is to bear witness to the uniquely human*

potential at its best, which is to transform a personal tragedy into triumph, to turn one's predicament into a human achievement. When we are no longer able to change a situation – just think of an incurable disease such as inoperable cancer – we are challenged to change ourselves..." (Frankl, 1992, p.135).

This came to mind from the philosophical theory of existentialism, and, of course, the pioneer of existentialism, Viktor Frankl, and his compelling book 'Man's Search for Meaning'. Frankl wrote this book shortly after his liberation from a Nazi concentration camp, detailing the unfathomable horrors he endured, but also reiterating how his suffering enabled him to find a greater meaning and purpose in life. Through this, he developed a therapeutic process aimed at challenging individuals to find meaning through suffering, love and work (Frankl, 2010).

Similarly, the parents did not choose this path. Their circumstances have served to test the very fibre of their human character, being confronted with the last of the human freedoms, as Frankl called it. This refers to choosing one's attitude in any given set of circumstances (Frankl, 1992; 2010). This attitude does not obliterate or lessen the intensity of the suffering, grief and heartache experienced in any way, but has enabled the parents to use their experiences in a constructive way, not allowing them to dampen their hope, and reiterating that what they have become is a result of their choices.

A central existential concept is that while we might long for freedom, it is an elusive concept as we often try to escape from or avoid our freedom by defining ourselves as a fixed entity (Frankl, 2010). Adapting to this entirely new way of life, with all the associated responsibilities, daily challenges and sacrifices, was bound to stir up challenging emotions and thoughts surrounding the meaning of life. As

mentioned in the previous chapter, Michelle relayed that she 'would like to be further in life' as she performed well at school and could have studied towards a degree, as many of her friends have done. Although she has not yet studied, she remains hopeful that once day they will have the financial means to do so. Similar to the other parents in the study, she has found a way of cultivating and finding meaning in her everyday life. She has facilitated courses aimed at educating the social workers at the Muscular Dystrophy Foundation, and recently started her own business, making masks and leather bags.

Despite the diversity in beliefs, varying support structures and resources that each family portrayed through their unique narratives and amidst the uncertainty they faced, each family shared an innate belief that life is primarily a quest for meaning and hope, no matter how dire the circumstances may be.

5.4 Recommendations for Professionals and Families

Parents reported the ongoing struggles with service providers and the importance of being informed. Therefore, it is important for professionals to appreciate that parents' instincts are usually accurate and to not dismiss their concerns, whether this relates to the diagnosis, the progression of the disease or palliative care. Professionals need to work collaboratively with parents to obtain the necessary equipment and medication as the disease progresses.

My findings support the need to incorporate strategies into existing services or health promotion programmes to build on the parents' support structures so that they are able to adapt to the challenges they face on a regular basis. Chen and Clark (2010) suggested the need for government-supported out-of-home day-care for working parents. Given the resource constraints in South Africa, this may not be a feasible option.

Michelle has already conducted training on DMD with social workers, based on her experiences as a parent. It would be extremely beneficial for psychologists, nurses and social workers to work collaboratively with parents to run a programme to build on internal resources and coping strategies. It would be useful to teach effective communication and conflict management, as well as self-care (Chen & Clark, 2010). This would also encourage the parents to voice their major challenges and have them addressed in a group setting.

It is necessary to consider each family's financial position and offer personalised support to ensure that each family receives assistance. The parents indicated that the Muscular Dystrophy Association in South Africa did not seem to prioritise DMD cases over the other types of muscular dystrophy with regards to funding, sponsors and support despite the terminal nature of DMD. This needs to be further investigated.

Professionals and families alike need to be aware that the grieving process differs for everyone. The diagnosis needs to be handled with empathy. Professionals need to offer practical suggestions for working through the initial shock, and reassure the parents that there is no 'right way' to grieve.

Additional support could include organising events for parents and forming more support groups.

Health care professionals should be aware of the need for assistance and increased support during their son's transition to adulthood. Psychological support, emergency and specialised care needs and parental intervention in the self-determination of their adult sons all need to be addressed as they are challenges faced by DMD individuals and their families. Furthermore, working collaboratively

with the parents and their son could serve to empower and encourage him to fulfil his aspirations before it is too late.

5.5 Further Research

Further research is needed to understand the influence of internal resources and the difference in their influence on mothers and fathers.

Future research could be carried out on larger sample sizes of parents, discussing the disease impact in a South African context. This kind of research is necessary to improve care provision and inform policy.

The factors that affect uncertainty appraisals and predictors of depression in parents need to be investigated. This could assist healthcare professionals in the development of interventions to enhance the adaptation process. Additionally, it would be useful to explore and expand on the appraisal of a rare or chronic disability.

It is necessary to carry out further research into avenues for the management of behavioural and cognitive problems evident in DMD boys, in order for support to be mobilised in resource-limited settings (Donald et al., 2011).

It would also be useful to explore and expand on the appraisal of a rare or chronic disability.

As boys with DMD are living longer and reaching adulthood, it would be valuable to explore the experiences of growing up and living with DMD, and the particular challenges adolescent boys face.

Finally, the opinions of the healthy siblings need to be explored in conjunction with those of the parents.

5.6 Final Words

Despite the small sample size and the limitations mentioned earlier, these interviews were useful as this is the first study to explore the lived experiences of

parents raising a child with DMD in South Africa. My study has served to highlight the gaps in the medical fraternity as well as to inspire and propel further research into larger studies of this nature, to continue to raise awareness and improve various support structures in SA. In addition, it has raised awareness of the increased life expectancy of boys and therefore the necessity of educating parents, doctors and other health care professionals as to how to support these boys in finding meaning and fulfilment in their everyday lives. Overall, my research may assist other families or parents raising a child with a chronic illness to lead a deeper, more reflective and meaningful life.

References

Abbott, D., Carpenter, J., & Bushby, K. (2012). Transition to adulthood for young men with Duchenne muscular dystrophy: Research from the UK. *Neuromuscular Disorders, 22*(5), 445-446. doi:10.1016/j.nmd.2012.02.004

Abi Daoud, M., Dooley, J., & Gordon, K. (2004). Depression in parents of children with Duchenne muscular dystrophy. *Pediatric Neurology, 31*(1), 16-9.

Alase, A. (2017). The Interpretative Phenomenological Analysis (IPA): A Guide to a Good Qualitative Research Approach. *International Journal of Education and Literacy Studies*, *5*(2), 9-19.

Alexander, D., Ali-Jadoobir, D., Bedeshi, G., Mitchell, C., Lyder-George, N., Onuoha, P., Ramdeen-Mootoo, G., (2018). Lived experiences of parents with children diagnosed with cancer in Trinidad: Challenges and coping strategies. *Cancer and Clinical Oncology, 2*(7). Canadian Center of Science and Education.

Amador, D. D., Reichert, A. P. D S., Lima, R. A. G. D., & Collet, N. (2013). Conceptions of care and feelings of the caregiver of children with cancer/concepções de cuidado e sentimentos do cuidador de crianças com câncer. *Acta Paulista De Enfermagem*, *26*(6), 542–546.

Årestedt, L., Persson, C., & Benzein, E. (2014). Living as a family in the midst of chronic illness. *Scandinavian journal of caring sciences*, *28*(1), 29-37.

Arias, R., Andrews, J., Pandya, S., Pettit, K., Trout, C., Apkon, S., Meaney, F. (2011). Palliative care services in families of males with Duchenne muscular dystrophy. *Muscle & Nerve, 44*(1), 93-101. doi:10.1002/mus.22005

Austin, T. (2012). *Abnormal Psychology: A South African perspective* (2nd ed.) (A. Burke, Ed.). Cape Town: Oxford University Press.

Bally, J. M., Smith, N. R., Holtslander, L., Duncan, V., Hodgson-Viden, H., Mpofu, C., & Zimmer, M. (2018). A metasynthesis: uncovering what is known about the experiences of families with children who have life-limiting and life-threatening illnesses. *Journal of Pediatric Nursing*, *38*, 88-98.

Becvar, D. S., & Becvar, R. J. (2013). *Family therapy: A systemic integration*. Pearson Education.

Bell, M., Biesecker, B., Bodurtha, J., & Peay, H. (2019). Uncertainty, hope, and coping efficacy among mothers of children with Duchenne/Becker muscular dystrophy. *Clinical Genetics, 95*(6), 677-683. doi:10.1111/cge.13528

Bernell, S., & Howard, S. W. (2016). Use your words carefully: what is a chronic disease? *Frontiers in Public Health*, *4*, 159-159. https://doi.org/10.3389/fpubh.2016.00159

Biggar, W. D., Harris, V. A., Eliasoph, L., & Alman, B. (2006). Long-term benefits of deflazacort treatment for boys with Duchenne muscular dystrophy in their second decade. *Neuromuscular Disorders*, *16*(4), 249-255. https://doi.org/10.1016/j.nmd.2006.01.010

Bonanno, G. A. (2009). *The other side of sadness: What the new science of bereavement tells us about life after loss.* New York, NY: Basic Books.

Bowlby, J. (1969). *Attachment and loss* (Ser. The international psycho-analytical library, no. 79, 95, 109). Institute of Psycho-analysis (London: Hogarth P, England).

Braun, V., & Clark, V. (2006). Using thematic analysis in psychology. *Qualitative Research in Psychology, 3*(2), 77-101.

Bregman, A. (1980). Living with progressive childhood illness: Parental management of neuromuscular disease. *Social Work in Health Care*, 5, 387-408.

Brown, R. T., Wiener, L., Kupst, M. J., Brennan, T., Behrman, R., Compas, B. E., … Zeltzer, L. (2008). Single parents of children with chronic illness: an understudied phenomenon. *Journal of Pediatric Psychology*, *33*(4), 408-421.

Buchanan, D., LaBarbera, C., Roelofs, R., & Olson, W. (1979). Reactions of families to children with Duchenne muscular dystrophy. *General Hospital Psychiatry,* 1, 262-269.

Bushby, K., Finkel, R., Birnkrant, D., Case, L., Clemens, P., Cripe, L., DMD Care Considerations Working Group. (2010). Diagnosis and management of Duchenne muscular dystrophy, part 1: Diagnosis, and pharmacological and psychosocial management. *The Lancet. Neurology, 9*(1), 77-93. doi:10.1016/S1474-4422(09)70271-6

Campbell, F. (2009). *Contours of ableism: The production of disability and abledness*. Springer.

Carnevale, F., Alexander, E., Davis, M., Rennick, J., & Troini, R. (2006). Daily living with distress and enrichment: The moral experience of families with ventilator-assisted children at home. *Pediatrics,117*(1), 48-60.

Charash, L. I. (1983). *Psychosocial aspects of muscular dystrophy and allied diseases : commitment to life, health, and function*. Springfield: Thomas.

Chen, J., & Clark, M. (2007). Family function in families of children with Duchenne muscular dystrophy. *Family & Community Health, 30*(4), 296-304. doi:10.1097/01.FCH.0000290542.10458.f8)

Chen, J., & Clark, M. (2010). Family resources and parental health in families of children with Duchenne muscular dystrophy. *The Journal of Nursing Research : Jnr, 18*(4), 239-48. doi:10.1097/JNR.0b013e3181fbe37b

Chen, J., & Instone, S. (2008). *Family resilience and functioning in child with DMD : Functioning and resilience in families with children with Duchenne muscular dystrophy*. Saarbrücken, Germany: VDM Verlag Dr.Müller.

Coffey, J. S. (2006). Parenting a child with chronic illness: a metasynthesis. *Pediatric nursing*, *32*(1).

Cohen, F., Lazarus, RS. (1979). Coping with the stress of illness. In C.G. Stone, F. Cohen, & N.E. Adler (eds.) *Health psychology: A handbook (*pp. 217-254). San Francisco, CA: Jossey-Bass.

Cornelio, S. J., Nayak, B.S., & George, A. (2016). Experiences of mothers on parenting children with leukemia. *Indian Journal of Palliative Care*, *22*(2), 168–172. https://doi.org/10.4103/0973-1075.179608

Corr, C. (2015). Let's stop ''staging'' persons who are coping with loss. *Illness, Crisis & Loss*, *23*(3), 226–241.

Cotton, S. M., Voudouris, N. J., & Greenwood, K. M. (2005). Association between intellectual functioning and age in children and young adults with Duchenne muscular dystrophy: further results from a meta-analysis. *Developmental Medicine and Child Neurology*, *47*(4), 257–65.

Creswell, J. W. (2012). *Educational Research: Planning, Conducting and Evaluating Quantitative and Qualitative Research* (4th Ed.). Boston, MA: Pearson.

Dawson, S., & Kristjanson, L. J. (2003). Mapping the journey: Family carers' perceptions of issues related to end-stage care of individuals with muscular dystrophy or motor neurone disease. *Journal of Palliative Care,19*(1), 36-42.

De Graves, S. & Aranda, S. (2008). Living with hope and fear – the uncertainty of childhood cancer after relapse. *Cancer Nursing, 31*(4), 292-301. doi:10.1097/01.NCC.0000305745.41582.73

Department of Education. (2001). *Education white paper 6: Special needs education, building an inclusive education and training system.* Pretoria: Department of Education.

Dibb, B., & Yardley, L. (2006). How does social comparison within a self-help group influence adjustment to chronic illness? A longitudinal study. *Social Science & Medicine*, *63*(6), 1602-1613.

Donald, K. A., Mathema, H., Thomas, K. G., & Wilmshurst, J. M. (2011). Intellectual and behavioral functioning in a South African cohort of boys with Duchenne muscular dystrophy. *Journal of Child Neurology*, *26*(8), 963-9. https://doi.org/10.1177/0883073811399149

Donald, K., Wilmshurst, J., & Thomas, K. (2008). *Intellectual and behavioural functioning in boys with Duchenne muscular dystrophy: Neuropsychological testing and correlation with genotype* (Doctoral dissertation, 2008). University of Cape Town.

Dubowitz, V. (2005). Prednisone for Duchenne muscular dystrophy. *The Lancet. Neurology*, *4*(5), 264–264. https://doi-org.uplib.idm.oclc.org/10.1016/S1474-4422(05)70050-8

Eagle, M., Bourke, J., Bullock, R., Gibson, M., Mehta, J., Giddings, D., Bushby, K. (2007). Managing Duchenne muscular dystrophy – the additive effect of spinal surgery and home nocturnal ventilation in improving survival. *Neuromuscular Disorders, 17*(6), 470-475. doi:10.1016/j.nmd.2007.03.002

Emery, A. (1987). *Duchenne muscular dystrophy* (Oxford monographs on medical genetics, no. 15). Oxford: Oxford University Press.

Emery, A. (2003). *Duchenne muscular dystrophy* (3rd ed.). Oxford: Oxford University Press.

Erby, L. H., Rushton, C., & Geller, G. (2006). “My son is still walking”: stages of receptivity to discussions of advance care planning among parents of sons with Duchenne muscular dystrophy. *Seminars in Pediatric Neurology*, *13*(2), 132–140. https://doi.org/10.1016/j.spen.2006.06.009

Esterhuizen, A., Greenberg, L., Ballo, R., Goliath, R., & Wilmshurst, J. (2016). Duchenne muscular dystrophy in the Western Cape, South Africa: Where do we come from and where are we going? *South African Medical Journal / Suid-Afrikaanse Tydskrif vir Geneeskunde, 106*(6), 67-71. doi:10.7196/SAMJ.2016.v106i6.10999

Folkman, S. (1997). Positive psychological states and coping with severe stress. *Soc. Sci. Med.,* 45 (8), 1207-21. doi.org./10.1016/S0277-9536(97)00040-3

Frankl, V. E. (1992). *Man's search for meaning: an introduction to logotherapy* (4th ed.). Beacon Press.

Frankl, V. E. (2010). *Feeling of meaninglessness: A challenge to psychotherapy and philosophy*. ProQuest E-book Central https://ebookcentral-proquest-com.uplib.idm.oclc.org

Friedman, R., & James, J. W. (2008). The myth of the stages of dying, death and grief. *Skeptic*, 14, 37–42.

Fylan, F. (2005). Semi-structured interviewing. In J. Miles, & P. Gilbert (Eds.), *A Handbook of Research Methods for Clinical and Health Psychology* (pp. 65-78). United States: Oxford University Press.

Gilgoff, I. (1983). The psychological effects of Duchenne muscular dystrophy on children and their families. In C. Hollingsworth (Ed.), *Coping with pediatric illness*. Lancaster: MTP.

Glover, S., Hendron, J., Taylor, B., & Long, M. (2020). Understanding carer resilience in Duchenne muscular dystrophy: a systematic narrative review. *Chronic Illness*, *16*(2), 87–103. https://doi.org/10.1177/1742395318789472

Gottlieb, B. (1997). *Coping with chronic stress* (The Plenum series on stress and coping). New York: Plenum Press.

Gravelle, A. (1997). Caring for a child with a progressive illness during the complex chronic phase: Parents' experience of facing adversity. *Journal of Advanced Nursing, 25*(4), 738-45.

Guest, G., Bunce, A., and Johnson, L. (2006). How many interviews are enough? An experiment with data saturation and variability. *Field Methods, 18*(1), 59-82.

Guite, J., Russell, B., Homan, K., Tepe, R., & Williams, S. (2018). Parenting in the context of children's chronic pain: Balancing care and burden. *Children, 5* (12). doi:10.3390/children5120161

Han, P. K., Klein, W. M., & Arora, N. K. (2011). Varieties of uncertainty in health care: a conceptual taxonomy. *Medical Decision Making*, *31*(6), 828-838. https://doi.org/10.1177%2F0272989X10393976

Heidegger, M. & Dahlstrom, D.O. (2005). *Introduction to phenomenological research*. Indiana University Press.

Hendriksen, J., Poysky, J., Schrans, D., Schouten, E., Aldenkamp, A., & Vles, J. (2007). Psychosocial adjustment in males with Duchenne muscular

dystrophy: Psychometric properties and clinical utility of a parent-report questionnaire. *Journal of Pediatric Psychology, 34*(1), 69-78. doi:10.1093/jpepsy/jsn067

Hinton, D., & Kirk, S. (2017). Living with uncertainty and hope: A qualitative study exploring parents' experiences of living with childhood multiple sclerosis. *Chronic Illness*, *13*(2), 88-99. https://doi.org/10.1177/1742395316664959

Hodges, L., & Dibb, B. (2010). Social comparison within self-help groups. *Journal of Health Psychology, 15*(4), 483-492.

Holland, J. M., & Neimeyer, R. (2010). An examination of stage theory of grief among individuals bereaved by natural and violent causes: A meaning-oriented contribution. *Omega, 61*, 103-120.

Hoskin, J. (2017). Taking charge and letting go: Exploring the ways a transition to adulthood project for teenagers with Duchenne muscular dystrophy has supported parents to prepare for the future. *British Journal of Special Education, 44*(2), 165-185. doi:10.1111/1467-8578.12173

Howitt, D. (2016). *Introduction to qualitative research methods in psychology*. Pearson UK.

Husserl, E., & Gibson, W. R. B. (2002). *Ideas: General introduction to pure phenomenology* (Ser. Muirhead library of philosophy). Routledge. https://univofpretoria.on.worldcat.org/search?sortKey=LIBRARY&databaseList=&queryString=husserl+phenomenology&changedFacet=&overrideStickyFacetDefault=&stickyFacetsChecked=on&selectSortKey=LIBRARY&overrideGroupVariant=&overrideGroupVariantValue=&scope=&format=all&year=all&yearFr

om=&yearTo=&author=all&topic=all&database=all&language=all#/oclc/881184049

Jalloh, A., Mathiassen, W., Wessels, T., Ndondo, A., & Wilmshurst, J. (2019). Development of a Duchenne muscular dystrophy registry for children in South Africa to optimize care. doi:10.17724/jicna.2018.146

Kazak, A. E., Simms, S., & Rourke, M. T. (2002). Family systems practice in pediatric psychology. *Journal of Pediatric Psychology*, *27*(2), 133-143.

Koch, T. (1996). Implementation of a hermeneutic inquiry in nursing: Philosophy, rigour and representation. *Journal of Advanced Nursing (Wiley-Blackwell), 24*(1).

Kohler, M., Clarenbach, C., Bahler, C., Brack, T., Russi, E., & Bloch, K. (2009). Disability and survival in Duchenne muscular dystrophy. *Journal of Neurology, Neurosurgery, and Psychiatry, 80*(3), 320-5. doi:10.1136/jnnp.2007.141721

Koufaki, I., Kedraka, K., Ierodiakonou-Benou, I., & Samakouri, M. (2019). Parental Experiences with Duchenne Muscular Dystrophy: Feelings of loss and Empowerment. *International Journal of Academic Research in Psychology, 6*(1), 17-31.

Kübler-Ross, E. (1969). *On death and dying*. New York, NY: Macmillan.

Kübler-Ross, E., & Kessler, D. (2005). *On grief and grieving: Finding the meaning of grief through the five stages of loss*. New York, NY: Scribner.

Larkin, M., Watts, S., & Clifton, E. (2006). Giving voice and making sense in interpretative phenomenological analysis. *Qualitative Research in Psychology*, *3*(2), 102–120.

Lavee, Y., & Mey-Dan, M. (2003). Patterns of change in marital relationships among parents of children with cancer. *Health & Social Work*, *28*(4), 255-263.

Lavoie, R. D. (2005). *It's so much work to be your friend : helping the child with learning disabilities find social success*. Simon & Schuster.

Law, M., King, S., Stewart, D., & King, G. (2001). The perceived effects of parent-led support groups for parents of children with disabilities. *Physical & Occupational Therapy in Pediatrics*, *21*(2-3), 29-48.

Leite, A. C. A. B., Garcia-Vivar, C., Neris, R. R., Alvarenga, W. de A., & Nascimento, L. C. (2019). The experience of hope in families of children and adolescents living with chronic illness: A thematic synthesis of qualitative studies. *Journal of Advanced Nursing*, *75*(12), 3246–3262. https://doi.org/10.1111/jan.14129

Lemma, A., Target, M., & Fonagy, P. (2011). *Brief dynamic interpersonal therapy: A clinician's guide*. Oxford: Oxford University Press.

Madill, A., Jordan, A. & Shirley, C. (2000). Objectivity and reliability in qualitative analysis: Realist, contextualist and radical constructionist epistemologies. *British Journal of Psychology, 91*, 1–20.

Magliano, L., (2014). Psychological and practical difficulties among parents and healthy siblings of children with Duchenne vs. Becker muscular dystrophy: An Italian comparative study. *Acta Myologica*, *33*(3), 136.

Magliano, L., Patalano, M., Sagliocchi, A., Scutifero, M., Zaccaro, A., D'Angelo, M. G., … Politano, L. (2014). "I have got something positive out of this situation": psychological benefits of caregiving in relatives of young people with muscular dystrophy. *Journal of Neurology : Official Journal of the European Neurological Society*, *261*(1), 188–195. https://doi.org/10.1007/s00415-013-7176-8

Magnan, R.E., Köblitz, A.R., Zielke, D.J., & McCaul, K.D. (2009). The effects of warning smokers on perceived risk, worry, and motivation to quit. *Annals of Behavioral Medicine*, *37*(1), 46–57. doi.org/10.1007/s12160-009-9085-8

Mah J.K., Thannhauser J.E., McNeil D.A., & Dewey, D. (2008). Being the lifeline: The parent experience of caring for a child with neuromuscular disease on home mechanical ventilation. *Neuromuscular Disorders, 18*(12), 983-88.

McGorry, P. D. (2007). Issues for DSM-V: Clinical staging: A heuristic pathway to valid nosology and safer, more effective treatment in psychiatry. *The American Journal of Psychiatry, 164*(6), 859–860.

McLeod, J. (2011). *Qualitative research in counselling and psychotherapy* (2nd ed.). SAGE.

McWilliams, N. (2011). *Psychoanalytic diagnosis: Understanding personality structure in the clinical process* (2nd ed.). New York: Guilford Press.

MDFSA. (2018). *A Guide to Our Parents*. Retrieved from: http://www.mdsa.org.za/the-guide-to-parents/

Meiring, M. (2011). Caring for children with life-limiting and life-threatening illnesses. What the GP should know. *CME: Your SA Journal of CPD, 29*(7), 286-290.

Mistry, K. B. (2012). Research and statistics: Qualitative research methods. *Pediatrics in Review, 33,* 521-523.

Nereo, N., Fee, R. J., & Hinton, V. J. (2003). Parental stress in mothers of boys with Duchenne muscular dystrophy. *Journal of Pediatric Psychology, 28*(7), 473-84. https://doi.org/10.1093/jpepsy/jsg038

Nicholas, D. B., Gearing, R. E., McNeill, T., Fung, K., Lucchetta, S., & Selkirk, E. K. (2009). Experiences and resistance strategies utilized by fathers of children

with cancer. *Social Work in Health Care*, *48*(3), 260–75. https://doi.org/10.1080/00981380802591734

O'Brien, M. (2001). Living in a house of cards: Family experiences with long-term childhood technology dependence. *Journal of Pediatric Nursing, 16*(1), 13-22.

Opstal, S., Jansen, M., Van Alfen, N., & De Groot, I. (2014). Health-related quality of life and its relation to disease severity in boys with Duchenne muscular dystrophy: Satisfied boys, worrying parents – a case-control study. *Journal of Child Neurology, 29*(11), 1486-1495. doi:10.1177/0883073813506490

Ortlipp, M. (2008). Keeping and using reflective journals in the qualitative research process. *The Qualitative Report, 13*(4), 695-705.

Parker, D., Maddocks, I., & Stern, L. (1999). The role of palliative care in advanced muscular dystrophy and spinal muscular atrophy. *Journal of Paediatrics and Child Health, 35*(3), 245-250. doi:10.1046/j.1440-1754.1999.00351.x

Parkes, C. M. (2013). Elisabeth Kübler-Ross, on death and dying: A reappraisal. *Mortality*, *18*(1), 94-97.

Pascual Morena, C., Martinez-Vizcaino, V., Álvarez-Bueno, C., Fernández Rodríguez, R., Jiménez López, E., Torres-Costoso, A. I., & Cavero-Redondo, I. (2019). Effectiveness of pharmacological treatments in Duchenne muscular dystrophy: a protocol for a systematic review and meta-analysis. *BMJ Open*, *9*(9), 029341. https://doi.org/10.1136/bmjopen-2019-029341

Pat-Horenczyk, R., & Brom, D. (2007). The multiple faces of post-traumatic growth. *Applied Psychology*, *56*(3), 379–385. https://doi.org/10.1111/j.1464-0597.2007.00297.x

Peay, H., Hollin, I., & Bridges, J. (2016). Prioritizing parental worry associated with Duchenne muscular dystrophy using best-worst scaling. *Journal of Genetic Counseling, 25*(2), 305-313. doi:10.1007/s10897-015-9872-2

Peay, H., Meiser, B., Kinnett, K., Furlong, P &, Tibben, A. (2015). Mothers' psychological adaptation to Duchenne/Becker muscular dystrophy. *European Journal of Human Genetics*, *24* (2), 2-15.

Polita, N. B., Alvarenga, W. D. A., Leite, A. C. A. B., Araujo, J. S., Santos, L. B. P. A. D., Zago, M. M. F., … Nascimento, L. C. (2018). Care provided by the father to the child with cancer under the influence of masculinities: Qualitative meta-synthesis. *Revista Brasileira de Enfermagem*, *71*(1), 185-194. https://doi.org/10.1590/0034-7167-2016-0671

Pourghaznein, T., Heydari, A., Manzari, Z., & ValizadehZare, N. (2018). "Immersion in an ocean of psychological tension:" The voices of mothers with children undergoing hemodialysis. *Iranian Journal of Nursing and Midwifery Research*, *23*(4), 253–260. https://doi.org/10.4103/ijnmr.IJNMR_156_17

Qu, S. Q., & Dumay, J. (2011). The qualitative interview. *Qualitative Research in Accounting & Management, 8,* 238-264.

Raina, P., O'Donnell, M., Rosenbaum, P., Brehaut, J., Walter, S., Russell, D., . . . Wood, E. (2005). The health and well-being of caregivers of children with cerebral palsy. *Pediatrics, 115*(6), 626-36.

Rajandram, R. K., Jenewein, J., McGrath, C., & Zwahlen, R. A. (2011). Coping processes relevant to posttraumatic growth: An evidence-based review. *Supportive Care in Cancer*, *19*(5), 583-589.

Reid, D., & Renwick, R. (2001). Relating familial stress to the psychosocial adjustment of adolescents with Duchenne muscular dystrophy. *International*

Journal of Rehabilitation Research. Internationale Zeitschrift für Rehabilitationsforschung. Revue Internationale de Recherches de Readaptation, 24(2), 83-93.

Reiners, G. M. (2012). Understanding the differences between Husserl's (descriptive) and Heidegger's (interpretive) phenomenological research. *Journal of Nursing & Care*, *1*(5), 1-3.

Richter, T., Nestler-Parr, S., Babela, R., Khan, Z. M., Tesoro, T., Molsen, E., & Hughes, D. A. (2015). Rare disease terminology and definitions—a systematic global review: report of the ISPOR rare disease special interest group. *Value in Health*, *18*(6), 906–914. https://doi.org/10.1016/j.jval.2015.05.008

Ringnér, A., Jansson, L., & Graneheim, U. H. (2011). Parental experiences of information within pediatric oncology. *Journal of Pediatric Oncology Nursing*, *28*(4), 244-251.

Ross, I.L. (2016). Exploring rare diseases in South Africa, a personal journey: Time for electronic record-keeping. *Annals of Medical & Health Sciences Research*, *6*(1).

Rubin, S. (1987) The psychological impact of genetic disease. In: L. Charash, R. Lovelace, S. Wolf, A. Kutscher, D. Roye & C. Leach (eds). *Realities in Coping with Progressive Neuromuscular Diseases.* The Charles Press, Philadelphia, PA, USA. pp. 209–215.

Samson, A., Tomiak, E., Dimillo, J., Lavigne, R., Miles, S., Choquette, M., Chakraborty, P., & Jacob, P. (2009). The lived experience of hope among parents of a child with Duchenne muscular dystrophy: Perceiving the human being beyond the illness. *Chronic Illness 5*(2):103-14.

Schneider, C., & Hattie, B. (2016). Exploring the social lives of young adults with disabilities. *ALTER, European Journal of Disability Research,* 3(10), 236-247. doi:10.1016/j.alter.2016.02.003

Shaw, R., Burton, A., Xuereb, C., Gibson, J., & Lane, D. (2014). *Interpretative phenomenological analysis in applied health research* (Sage research methods cases). Los Angeles, California: SAGE. Retrieved from https://dx-doi-org.uplib.idm.oclc.org/10.4135/97814462730501351465 6

Sheryl, J. C., Baby, S. N., & Anice, G. (2016). Experiences of mothers on parenting children with leukemia. *Indian Journal of Palliative Care*, *22*(2), 168–172. https://doi.org/10.4103/0973-1075.179608

Smith, J. A., & Osborn, M. (2003). Interpretive phenomenological analysis. In J. A. Smith (Ed.), *Qualitative psychology: A practical guide to research methods.* (pp. 51-80). London, United Kingdom: Sage Publications.

Smith, J. A., & Shinebourne, P. (2012). *Interpretative phenomenological analysis*. American Psychological Association.

Smith, J. A. (1996). Beyond the divide between cognition and discourse: Using interpretative phenomenological analysis in health psychology. *Psychology and Health*, *11*(2), 261-271.

Smith, J. A., Flowers, P., & Larkin, M. (2009). *Interpretative phenomenological analysis: theory, method and research*. SAGE.

Soutter, J., Hamilton, N., Russell, P., Russell, C., Bushby, K., Sloper, P., & Bartlett, K. (2004). The golden freeway: a preliminary evaluation of a pilot study advancing information technology as a social intervention for boys with Duchenne muscular dystrophy and their families. *Health & Social Care in the Community*, *12*(1), 25–33.

Speer, S. (2002). ‘Natural’ and ‘contrived’ data: a sustainable distinction? *Discourse Studies*, *4*(4), 511–525.

Stroebe, M. S. (2008). *Handbook of bereavement research and practice: advances in theory and intervention* (1st ed.). American Psychological Association.

Stroebe, M. S., & Schut, H. A. W. (1999). The dual process model of coping with bereavement: rationale and description. *Death Studies*, *23*(3).

Stroebe, M., Schut, H., & Boerner, K. (2017). Cautioning health-care professionals: bereaved persons are misguided through the stages of grief. *Omega: Journal of Death and Dying*, *74*(4), 455-473.

Wagner, C., Kawulich, B., Garner, M., & Botha, A. (2012). *Doing social research: A global context*. McGraw-Hill Higher Education.

Wallace, R., & Wolf, A, (2006). *Contemporary sociological theory* (6th ed.). NK: Pearson Prentice-Hall.

Webb, C. (2005). Parents’ perspectives on coping with Duchenne muscular dystrophy. *Child: Care, Health and Development, 31*(4), 385-396. doi:10.1111/j.1365-2214.2005.00518.x

Weidner, N. (2005). Developing an interdisciplinary palliative care plan for the patient with muscular dystrophy. *Pediatric Annals, 34*(7), 546-52.

Willig, C. (2013). *Introducing qualitative research in psychology*. McGraw-Hill Education (UK).

World Health Organisation. (2012). *Bulletin of the World Health Organisation, 90,* 401-476.

World Health Organization, & Public Health Agency of Canada. (2005). *Preventing chronic diseases: A vital investment*. World Health Organization.

https://univofpretoria.on.worldcat.org/search?queryString=%27preventing+chronic+diseases+is+a+vital+investment&submit2.x=0&submit2.y=0

United Nations Convention on the Rights of Persons with Disabilities (2006). *Convention of the Rights of Persons with Disabilities.* United Nations. https://www.un.org/disabilities/documents/convention/convention_accessible_pdf.pdf

Wu, L. M., Chin, C. C., Haase, J. E., & Chen, C. H. (2009). Coping experiences of adolescents with cancer: a qualitative study. *Journal of Advanced Nursing*, *65*(11), 2358-2366.

Wysocki, T. & Gavin, L. (2006). Paternal involvement in the management of pediatric chronic diseases: associations and adherence, quality of life, and health status. *Journal of Paediatric Psychology, 31*(5)*, 501-11.*

Yamaguchi, M., & Suzuki, M. (2015). Becoming a back-up carer: Parenting sons with Duchenne muscular dystrophy transitioning into adulthood. *Neuromuscular Disorders, 25*(1), 85-93. doi: 10.1016/j.nmd.2014.09.001

Yiu, E. M., & Kornberg, A. J. (2008). Duchenne muscular dystrophy. *Neurology India*, *56*(3), 236–47.

Youngblut, J. M., Brady, N. R., Brooten, D., & Thomas, D. J. (2000). Factors influencing single mother's employment status. *Health Care for Women International*, *21*(2), 125-136.

Appendix A: Ethical Clearance

UNIVERSITEIT VAN PRETORIA
UNIVERSITY OF PRETORIA
YUNIBESITHI YA PRETORIA

Faculty of Humanities
Research Ethics Committee

29 January 2019

Dear Ms Forman

Project:	**Exploring the lived experiences of parents raising a child diagnosed with Duchenne Muscular Dystrophy (DMD) in South Africa: Challenges and coping strategies**
Researcher:	**C-G Forman**
Supervisor:	**Dr L Blokland**
Department:	**Psychology**
Reference number:	**12289681 (GW20181115HS)**

Thank you for your response to the Committee's correspondence.

I have pleasure in informing you that the Research Ethics Committee formally **approved** the above study at an *ad hoc* meeting held on 28 January 2019. Data collection may therefore commence.

Please note that this approval is based on the assumption that the research will be carried out along the lines laid out in the proposal. Should your actual research depart significantly from the proposed research, it will be necessary to apply for a new research approval and ethical clearance.

We wish you success with the project.

Sincerely

Prof Maxi Schoeman
Deputy Dean: Postgraduate and Research Ethics
Faculty of Humanities
UNIVERSITY OF PRETORIA
e-mail: PGHumanities@up.ac.za

cc: Dr L Blokland (Supervisor)
Prof T Guse (HoD)

Appendix B: Participant information sheet

Invitation to participate in a research study

Dear Prospective Participant

My name is Casey Forman, and I am currently enrolled for my Psychology Master's degree in Counselling Psychology at the University of Pretoria. I am conducting a qualitative research project in the form of an individual interview which will explore your experiences of raising a child with Duchenne Muscular Dystrophy. By agreeing to participate in this study, you will be providing invaluable information on the proposed research study. I would like to invite you to take part in my research study. The interview should not take longer than an hour to complete and participation is completely voluntary. Taking part in the interview will not advantage or disadvantage you in any way; and there are no foreseen risks for taking part in this study. However, as the topic may hold emotional content for you, you can be referred to a counsellor (should this be something you want). There will be no charges for this service. I will assist you with this process.

- Itsoseng Clinic is an NGO in Pretoria. Contact: 012 842 3515 or email itsoseng.clinic@up.ac.za

- FAMSA is an NPO, with branches in Pretoria and Johannesburg.
  Contact: Pretoria branch: (012) 460 0733/8 or
  Johannesburg branch: (011) 788-4784 or email famsa@absamail.co.za

For effective analysis of the information gathered, the interview will be audio-recorded. Once the information from the interview has been transcribed verbatim, the transcripts will be kept in a secure file (on a password protected hard disk drive) and not made available to any third parties, apart from my supervisors. Should you, as the participant, request for their information to be returned or for it to be destroyed, it will be done as per request. I will assign a pseudonym to all the participants, assuring that confidentiality is maintained. It is important to note that you have the right to withdraw from this study at any time, and you are fully entitled to not answer any questions that you do not feel comfortable answering. Should you withdraw from this study, any data provided by you up to this point will be destroyed. There will be no disadvantages if you decide to do this. Once the study has been completed, you may request a summary of the findings which will be available 6 months after the data collection.

Before we commence, I will ask you to read through and sign the consent form. This form confirms that you have read and understood what is required of you; and that all information

will be kept highly confidential. If you have any questions or concerns, please do not hesitate to contact me.

Kind Regards

Casey Forman

STUDENT PSYCHOLOGIST

PS S 0144860

cgforman@gmail.com

0731751271

Dr. Linda Blokland

CLINICAL PSYCHOLOGIST

PS 0037923

linda.blokland@up.ac.za

012 420 4002

Appendix C: Informed Consent form

Research title: Exploring the lived experiences of parents raising a child diagnosed with Duchenne Muscular Dystrophy (DMD): Challenges and Coping Strategies

Thank you for expressing your interest in taking part in this research study. This form acknowledges that you understand your rights as a participant in the study.

Confirmation	Tick Boxes
I acknowledge that the researcher has explained my rights and the requirements of this study. By signing below and providing my contact information I am indicating that I consent to participate in this study, I am at least 18 years of age and I am eligible to participate in this study.	
I understand that there is no compensation for or direct benefit of participating in this study.	
I can withdraw at any stage with no disadvantages to myself.	
I understand that the information given shall be confidential and shall not be publicly attributed to me or to the company for which I work.	
I agree to the interview being recorded and that the recordings will be discarded accordingly on the completion of this research report. The data will be kept for 15 years in the Psychology Department in a secure deposit space.	

Signed name: _____

Date: _____

Printed name: _____

Phone number: _____

E-mail address: _____

Thank you for your participation.

Casey Forman
STUDENT PSYCHOLOGIST
PS S 0144860
cgforman@gmail.com
0731751271

Dr. Linda Blokland
CLINICAL PSYCHOLOGIST
PS 0037923
linda.blokland@up.ac.za
012 420 4002

Appendix D: Interview Schedule

Demographic information

1. How old are you?
2. How many children do you have?
3. How old are your other children (& gender)?
4. When did you discover that your child/son has DMD?
5. How severe is your child's/son's DMD?
6. How long ago was your child/son diagnosed with DMD? (the duration of diagnosis)

Questions

1. How did you perceive, and react to, the initial diagnosis of DMD?
2. What are some of the challenges you have faced?
3. What are some of your coping strategies?
4. What kind of support do you receive from the community in which you live?

Debriefing Questions:

1. How did you experience the interview?
2. Did the interview raise any emotional feelings for you that you would like to discuss further?
3. Is there anything else you would like to discuss about the interview?