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The Value of Life

An Ethnography of Duchenne Muscular Dystrophy in

Aotearoa New Zealand

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Abstract

How is life valued for people living with Duchenne muscular dystrophy (DMD) in Aotearoa New Zealand? I examine this question through a study of the community of people in New Zealand living with DMD, the Muscular Dystrophy Association, and the state systems of health and social services in which they are embedded. DMD is a serious genetic condition in which boys and young men with the affected gene experience degenerative muscle wasting. It is characterised by a predictable decline in all the muscles of the body, including the heart and the thoracic muscles. It is a life shortening condition. My detailed ethnographic analysis of situations as diverse as enthusiastic computer coding, paid caregiver support and power chair soccer has called for a range of theoretical approaches, including local biologies, assemblages, governmentality, and recent literature in the anthropology of policy and the anthropology of hope.

In my analysis of people's day to day lives, I learned that pain and suffering are unavoidable but pain and hope are intertwined. Boys and men with DMD and their families focus on living purposeful lives. The interests and passions that they have are indicative of a distinctive immediacy in which the now and the near future is paramount. These young men often have exceptionally devoted families who highly value their lives.

Using the analytic of biopower, I explain how the bureaucratic entanglements of young men and their families have the effect of devaluing their lives. As Foucault proposed, biopower, exercised through New Zealand's neoliberal state health and social systems is focussed on the needs of the majority. Because DMD is such an extreme condition, people living with it often find themselves outside the limits of state care. The ensuing frustration creates social suffering, compounding the condition itself. Consequently, I conclude that inclusive provision and effective care for people with DMD necessitates a specific national system of comprehensive, multidisciplinary care. International and local models for such provision already exist.

Keywords:

Duchenne muscular dystrophy, medical anthropology, genetic conditions, hope, policy.

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And finally to Kerry Hills, thank you for opening my eyes to some of the realities of living with a neuromuscular disease. I dedicate this thesis to you.

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List of Abbreviations

ACART	Advisory Committee on Assisted Reproductive Technology
ACC	Accident Compensation Corporation
ADHB	Auckland District Health Board
AGM	Annual General Meeting
ANN	Australasian Neuromuscular Network
ANT	Actor Network Theory
Bi-pap	Bilevel Positive Airway Pressure
BMD	Becker Muscular Dystrophy
CEO	Chief Executive Officer
CiCL	Choices in Community Living
CK or CPK	Creatine Phosphokinase
CO2	Carbon dioxide
C-pap	Continuous positive airway pressure
CVS	Chorionic Villus Sampling
D&E	Dilation and Evacuation
DHB	District Health Board
DMD	Duchenne Muscular Dystrophy
DNA	Deoxyribonucleic acid
ECART	Ethics Committee on Assisted Reproductive Technology
ED	Emergency Department
EMA	European Medicines Agency
EMA	Early Medical Abortion
EMS	Equipment and Modification Services
FDA	Food and Drug Administration
FFC	Family, Friends and Community
FINZ	Fund Raising Institute of New Zealand
GP	General Practioner
GST	Goods and Services Tax
HDC	Health and Disability Commissioner
IEA	Institute of Economic Affairs
IEP	Individual Education Plan
IF	Individualised Funding
IVF	In Vitro Fertilisation
MD Prev	Muscular Dystrophy Prevalence
MDA	Muscular Dystrophy Association of New Zealand
NASC	Needs Assessment Service Co-ordinator
NFP	Not for Profit
NMD	Neuromuscular Disease
NRFT	Neuromuscular Research Foundation Trust
NZORD	New Zealand Organisation for Rare Disorders
ORS	Ongoing Resourcing Scheme
OT	Occupational Therapy

PBF	Population Based Funding
PGD	Pre-implantation Genetic Diagnosis
PHARMAC	Pharmaceutical Management Agency
PPMD	Parent Project Muscular Dystrophy
RNA	Ribonucleic acid
SRT	Selective Reproductive Technology
TGA	Therapeutic Goods Administration
UNCRPD	United Nations Convention on the Rights of Persons with Disabilities

Frontispiece

> The disease is the most interesting and at the same time the most sad, of all those with which we have to deal: interesting on account of its peculiar features and mysterious nature; sad on account of our powerlessness to influence its course, except in a very slight degree, and on account of the conditions in which it occurs. It is a disease of early life and early growth. Manifesting itself commonly at the transition from infancy to childhood, it develops with the child's development, grows with his growth – so that every increase in stature means an increase in weakness, and each year takes him a step further on the road to helpless infirmity, and in most cases to an early and inevitable death. (Gowers, 1879:1)

This classic description of Duchenne muscular dystrophy (DMD) was written by the 19th century physician, Sir William Gowers. It summarises both the clinical and emotional aspects of Duchenne muscular dystrophy. Despite advances in treatment, DMD remains a particularly devastating neuromuscular condition. As children with this condition grow their muscles weaken. Gradually they lose the ability to move independently and continue to face "an early and inevitable death".

SECTION 1

CONTEXTUALISING DUCHENNE MUSCULAR DYSTROPHY IN AOTEAROA NEW ZEALAND

Chapter 1: How are Lives with DMD Valued in Aotearoa New Zealand?

Duchenne muscular dystrophy (DMD) is a genetic condition which causes muscle wastage in all muscles of the body. It is a condition that primarily affects males, although there are a small number of manifesting female carriers. DMD is a family disease. It impacts not only those individuals who have the condition but also other family members. It is a condition that causes serious disabilities for those directly affected. This thesis is a study of Aotearoa New Zealand's Duchenne muscular dystrophy community. It examines the concept and practice of 'value of life' in the context of lives lived with this genetic condition in New Zealand society by asking, how are the lives of people with DMD valued?

I explore two types of response to this question. The first response addresses the way individuals and families deal with and reconcile the condition. The second addresses the way systems of care and governance respond to the condition. These are not discrete responses; they intertwine in complex ways. I examine multiple, contradictory and overlapping factors that define the experience of living with Duchenne muscular dystrophy. I explore how the physical, embodied experience of living with DMD occurs within a complex web of cultural values and social structures which shape that experience. The experiences of individuals and families living with DMD support a distinctive analysis of some of New Zealand's fundamental social services (health, education and community care). Central to this analysis is an exploration of the rationales that underpin these services. These offer critical insights into the way people with disabilities are valued in New Zealand society. These experiences of individuals and families as they attempt to establish fulfilling lives, also offers a lens for understanding culturally constructed notions of the value of life in Aotearoa New Zealand. This chapter outlines key ideas that emerge from my research and I discuss relevant literature that exposes the social mechanisms and cultural values that co-create these findings.

Thesis Question and Overview

Initially this research project intended to examine how reproductive decisions were made by couples with a family history of DMD; I envisioned focusing on the moral decision-making, the everyday ethics, of couples as they planned their families. I asked broad, general questions which I felt would facilitate my understanding of the experiences of families living with DMD as this information would be central to an analysis of the decisions that couples

made. However, as I collected this general information regarding the experiences of families living with DMD, it became apparent that an academic focus on reproductive decision making did not align with the issues that were of overwhelming significance for the community. Although reproductive choices were obviously a concern, it became increasingly evident that the issue that was fundamental to participants was the ways in which individuals and families attempted to create meaningful and purposeful lives. Those who were involved in tertiary education talked about their studies and future ambitions, others discussed the work they did designing websites and coding, going to church, engaging in sports both as fans and players, following the news and planning accessible journeys. This positive information contrasted with the stories about the repeated difficulties people experienced with bureaucracies: schools, healthcare and community services. These two distinctive discourses both address ideas about the value of life lived with this significant disability.

When research participants were describing their own and others' actions and decisions, what they seemed to be talking about was the way in which various people attributed meaning to the value of life lived with DMD. Some research participants felt that other people acted as if their lives were devalued or less important than others. This thesis is an anthropological analysis of the many overlapping and sometimes conflicting processes that inform these understandings about value of life. Central is the predictive, degenerative nature of the condition itself. DMD facilitates the determination of those most immediately affected to make the most of each good day, to engage in fulfilling and purposeful activities. This positive approach is balanced by the physical and psychosocial suffering which is an inevitable part of the condition. These experiences of hope and pain occur within a wider social context in which bureaucratic hurdles often compound suffering but medical care may alleviate it.

Woven through out my analysis is a synthesis of Bruno Latour's actor network theory (ANT) (2005), Gillian Tett's work on alternative epistemologies (2015) and Margaret Lock and Vinh-Kim Nguyen's concept of local biologies (2010). Latour's actor-network theory is pertinent because families are closely connected to objects (powerchairs, cough assist machines and ventilators). Living with DMD involves being embroiled in complex networks of technology, biomedical expertise and bureaucratic structures. ANT facilitated my analysis of the interactions between key stakeholders and technologies, it enabled a detailed investigation of the different strategies and tactics that the participants in various networks adopted. Tett's ideas about silos of expert knowledge (2015) were also helpful in clarifying

the way the actors in various networks talked past each other. Biomedicine dominates the medical discourse within which DMD is framed. Applying Lock (1995 and 2001) and Lock and Nguyen's (2010) ideas about local biologies was helpful in distinguishing the ways in which New Zealand's health and social systems created a distinctive experience of DMD for people living in Aotearoa New Zealand. The way in which DMD has a local biological manifestation is not in changes to the predictable pattern of degeneration but rather in the way distinctive local factors impact on life experience and expectancy.

During my research I learned that it is almost impossible to conduct research in the field of disability without being confronted by the marginalisation of, and discrimination against, people with disabilities. Some of this discrimination results from barriers created by policies and criteria produced by the state or agents thereof. This type of discrimination can be confronted by addressing the relevant policies and criteria. When conducting research in the field of disability one is faced with the dilemma of the role of the researcher versus the role of activist / agent for social change. This dilemma has a long history in anthropological research and other identity-based research areas such as women's studies, indigenous/postcolonial studies and queer studies (see McClaurin 2001, Gardiner 2006, Naisargi 2012). Fassin (2012:115) notes that this quandary also exists in the field of medical anthropology. His argument, that the crucial role for anthropologists is the critical thinking we bring to understanding discrimination rather than simply the "denunciation of injustice" is useful. Here I attempt to uncover the often hidden or concealed social mechanisms and political issues that create the marginalisation and discrimination that people with DMD experience. In order to do this I use Foucault's ideas about governmentality and biopower and the anthropology of policy. Before addressing these anthropological theories in detail I note that this thesis also addresses an inter-disciplinary debate.

Debates in Disability Studies

Here I consider the work of Tom Shakespeare, a sociologist and disability activist who has contributed significantly to the field of Disability Studies. Shakespeare's challenge to the idea that the marginalisation and discrimination of people with disabilities can be addressed primarily by changes to structural exclusions fits with the accounts that form the basis of this research. My analysis of the hope with which people with DMD live considers the important role of biomedicine as a way of overcoming the social exclusion that the weakening Duchenne body creates. The cultural dominance of the biomedical model of disability has

been a topic of heated debated within Disability Studies particularly following the publication of Shakespeare's controversial book Disability Rights and Wrongs (2006). This challenged a widely accepted research methodology which was particularly prevalent in Britain. Disability studies had emerged from disability politics and focused on structural issues rather than personal issues. Claims made by disability studies traditionally included arguments that disability is a social and political issue, not simply a medical or rehabilitation issue; that disability is an issue of identity: "nothing about us without us"; and that people are disabled by society, not by their bodies (Oliver 1996).

This led to dichotomised models in which research fell into either a medical or social model. The medical model has a focused primarily on a clinical diagnosis and individual treatment. This model sees the disabled person as having a problem that needs to be fixed or cured and treatment of the individual as the answer. Boxall (2007:228) characterises the medical model as referring to ideas of "individual deficit and defective personhood". The social model focuses on the wider context. People are disabled by society rather than by their bodies. In this model discrimination and prejudice are identified as the problem and the removal of barriers is the answer. Disability studies has traditionally rejected the medical model in favour of the more liberating and empowering social model. The focus of disability studies has been to address barriers that limit the social participation of people with disabilities.

Shakespeare (2006, 2012 and 2013) however, sees this dichotomy as flawed. Others (Sheldon 2007:209, Boxall 2007:228, Oliver 2007:231 [in Sheldon et al. 2007]) argue that his analysis is overly simplistic and the social model is not as dogmatic or inflexible as Shakespeare (2006) suggests. Shakespeare argues that disability is not similar to other identity political movements such as the women's movement (feminism), religious identity, sexuality identity or ethnic identity. He argues that it is not sufficient to aim for the removal of barriers (be those physical or attitudinal). Removing barriers is a vital part of creating a fully inclusive society but acknowledging the intrinsic nature of an individual's impairment is fundamental to ensuring that all people with disabilities are able to fully participate in society. Many factors need to be taken into account as societies attempt to become fully inclusive; health concerns, psychological and emotional experiences, the physical environment, social and cultural factors, as well as economic, legal and political factors. Disability needs to be understood as a complex phenomenon with interventions possible at many sites. For many disabled people their physical or mental impairments are central to their experiences and these must be addressed. Shakespeare advocates for an approach to disability studies that,

"understand(s) disability as the dynamic interrelationship of an individual with a health condition and the environment in which they find themselves" (2012:129). Proponents of the "strong" or "British" social model do not agree with Shakespeare's representation of it and argue that his interactionist approach risks depoliticising disability studies (Oliver 2007: 232-233 and Goodley 2014:660).

In my own research I found that because of the nature of the DMD – the continual loss of muscle strength – health and medical issues were central to the way life is lived. It is not possible to research in this field without considering medical issues. Some members of the community are very interested in medical research, in knowing what the latest research means for them, how effective various treatments are and what pharmaceuticals are in development. Some families are well versed in the effect of alternative therapies and nutritional supplements. Others have been instrumental in the establishment of a New Zealand Neuromuscular Disease Registry which facilitates the participation of people with neuromuscular conditions living in New Zealand in international drug trials. This aligns with the French experience of disability activism (Callon and Rabeharisoa 2008) where activism diverges from the undertakings usually associated with disability activists. From the point of view of individuals and families directly affected by this condition, whom I spoke to during the course of my research, there was no rejection of research that focused on the individual's disability, no criticism of this sort of research. In fact, quite the opposite, currently fund-raising efforts are underway within the community to assist some families to travel to Australia to participate in drug trials. This indicates a widespread appreciation for, and desire to participate in, medical research.

Current medical research is raising a whole new gamut of discriminatory issues that will require ongoing social research and activism. The increasing Influence of Science and Technology Studies (STS) allows those involved in disability studies to move away from unhelpful, polarising research positions and to view all research as part of a wider social construction. As Shakespeare revisits his 2006 text (Shakespeare 2013), the controversy between the strong or 'British' social model and the emerging focus on an interactionist approach has diminished. As Goodley (2014) notes, "disability studies has matured" (2014:660). In his revised text Shakespeare clearly demonstrates a continued allegiance to political progress and the social movement of disabled people. "It is vital to state that in rejecting the 'strong' social model of disability, I am not rejecting the human rights approach to disability (UN 2006)" (Shakespeare 2013:1). This interactionist idea that disability is best

understood as a dynamic relationship between a person with a health condition, in this case DMD, and their environment is central to my research findings.

The structure of my thesis aligns with the conceptual schema described in this introduction. I begin with a consideration of the way the concepts of resilience and hope have been adopted in anthropological literature and their application to my own research. I then address the way Foucault's ideas of governmentality and biopower and the anthropology of policy have been helpful in illuminating the structural nature of social exclusion. Finally, I address the integrative literatures that run throughout the thesis particularly Latour's ANT (2005), Lock and Nguyen's (2010) work on local biologies and Fassin's (2012) discussion on the value and worth of lives.

Concepts of Resilience, Hope and Value

Participants were eager to discuss the aspects of their lives that were creative and positive. They demonstrated the way they made meaning of lives which were severely impacted by impairments. This recurrent aspect of the research data was not about dealing with external barriers. Rather it was about finding novel ways of living meaningful lives, of adjusting to the intrinsic difficulties that their bodies presented and finding activities and social events they could do. Initially I anticipated that the literature on resilience would be useful here. Panter-Brick (2014) situates resilience as a useful (and increasingly popular) concept within social and biomedical sciences. She acknowledges that resilience is a polysemous term, ambiguous, with "multiple but related meanings" (2014:432). I came to realise that the concept had several shortcomings. It would be easy to frame the stories people shared with me about the way they created meaning in their lives as a way of resisting common experiences of being under-valued, dismissed as being useless or of little value. However, I think this is too simplistic. The passion with which some people shared their accounts of making meaning in their lives suggests more than just resistance to social marginalisation. Resisting marginalisation was certainly part of these stories but people were also enthusiastic to talk about their activities and to show me what they do because the activities had intrinsic value. Like many people engaged in activities that they were passionate about, research participants were eager to discuss their interests at length and in great detail. Furthermore, despite Panter-Bricks's comments that resilience is a useful concept in the social sciences for overcoming generalisations that suggest victimhood of particular communities (2014:439), others argue that resilience can be used to further individualise suffering and push the burden of

responsibility onto individual sufferers and their families (Neocleous 2013, Gill and Donaghue 2016). I found that the anthropology of hope similarly moves beyond a focus on suffering and victimhood and enables a consideration of the techniques and strategies that individuals and communities adopt to improve their situations, but the concept of hope has not been co-opted by neoliberalism in the same way that resilience has. (There are, for example, no workshops to promote hopefulness in the under-staffed academic workplace as there are for resilience [Gill and Donaghue 2016]). My analysis suggests that the desire of participants to be engaged in meaningful occupation goes beyond neoliberal values of autonomy and productivity and addresses the quality of human life at a more spiritual level.

I use hope, rather than resilience, as a concept to explain some of the seeming contradictions of my research. I talked to young men who were excited, animated and passionate about their particular occupation but were also despondent and depressed about their decreasing muscle strength, and their social invisibility. Hage's (2003) description of the contingent nature of hope and the important role that the state has in providing citizens with hope is fundamental to my analysis. I do not use the concept of hope to contribute to a discourse that places increasing responsibilities onto individuals and families, rather I use the concept of hope to acknowledge the responsibilities of the state in structuring a society in such a way that all lives are valued. My analysis of the hopeful nature of the DMD community builds on the work of Mattingly (2010) who discusses some of the complex ideas embedded in this concept. Mattingly (2014:15) acknowledges that while hope supports the "rampant individualism" that characterises aspects of American culture, such as the ethos of 'The American Dream' it also underpins the struggles of marginalised communities for social inclusion. The detailed example I give in Chapter Five about the establishment of a powerchair soccer team is an example of the communal application of hope by those in a local community of powerchair users. This idea that hope is a concept related to community endeavor can be found in the work of Novas (2006). He notes that some patient organisations' lobbying and support of science and research are based on hope, hope that such efforts will facilitate effective treatments and eliminate suffering, "the hope invested in science is not only an aspiration, but can also be thought of as having a political and economic materiality that seeks to bring to fruition the many future possibilities inherent in the science of the present" (2006:289). Such an analysis has direct application to parts of this thesis as I examine the establishment of New Zealand Neuromuscular Disease Registry and consider it as an example of a complex community enacting hope.

The anthropology of hope has been central to clarifying various ways in which lives lived with DMD are valued in Aotearoa New Zealand. By using hope as a conceptual tool I have been able to address issues of immediacy for this particular community, I have centred people's hopes within communal frames and acknowledge the important role of the state in ensuring that marginalised people can access hope. In 1993 Coleridge, writing about disability and international development, noted

> … there are other needs which are just as basic: the need to be creative, to make choices, to exercise judgement, to love others, to have friendships, to contribute something of oneself to the world, to have social function and purpose. These are active needs; if they are not met the result is impoverishment of the human spirit, because without them life itself has no meaning. The most basic need of all is the need for meaning. (Coleridge 1993:213)

It is this essential and most basic need that the young men I interviewed were addressing when they shared their passions and enthusiasms with me. Hope entwines with the fulfillment of this need –living a purposeful life that has meaning.

This thesis adds to a developing body of literature in anthropology that indicates the possible universality of hope. It is a contribution to an anthropological discourse that demonstrates the importance of hope for the human condition (following Appadurai, 2004). Hope is about laughter and resilience and bravery and believing that there is purpose and meaning in life. This thesis and the work of Ghassan Hage (for example) show that although the propensity to hope may be a universal phenomenon the conditions of hope are socially constructed and that governments have an important role to play in that construction. The examples of hope that I discuss, powerchair soccer and pharmaceutical developments are not only about hope. Powerchair soccer is also a fun activity whilst those engaged in pharmaceutical research may well have other agendas such as career advancement or simply holding down a steady job with a reliable salary. However these things are not mutually exclusive to hope. The examples I cite demonstrate the importance of hope for a community that may not generally be associated with hope. In much of the literature about DMD the focus is on the degeneration of the muscles and the inevitability of an early death. However my research uncovered a community keen to discuss the things that gave their life value and meaning.

Value is a term that is repeated throughout the thesis. It is in the title and the concluding paragraph. It is an integral part of this thesis. I have used it both as a noun and a verb. I use it

in the manner one might commonly find it used in everyday communication. Value is about personhood, visibility and recognition. The term the value of life describes the importance or significance of a life. The discussion of research findings in Chapter 11 clarifies this. The distinction that Fassin (2012) makes between the biological valuing of lives and the political valuing of lives is crucial to my analysis and to clarify, Fassin argues that it is much easier to generate an emotional consensus around the value of one individual life rather than around the value of masses of lives – "we can give a human face to dying patient or the suffering child while the concreteness of disparities remains elusive". I use this point to explain the huge investments being made in pharmaceutical developments. Focussing on treatments that respond to the individual, biological body, responds to the individuals right to life. It affirms life. Pharmaceutical research is well funded and valued by those with DMD, their families and the medical community. It has emotional consensus. My findings about the social exclusions that characterise some aspects of service provision are described by Fassin as evaluations of the worth of lives (plural) where statistical significance rather than individual significance dominates. This is an ethnographic account – an emic account of the perceptions and experiences of people with DMD so it is subjective. I am not trying to judge if one group is valued over another. Rather I make the point that if value is about personhood, visibility and recognition then the way that services are provided makes people with DMD feel undervalued, invisible and unrecognised. To be valued is a political issue. This is not to do with the decisions of assessors but rather the way the systems are constructed. I am not suggesting that people working within the current system are purposefully diminishing or undermining those with DMD by not providing sufficient services rather I argue that the system itself creates these circumstances. The funding of service is about prioritisation and this process of prioritising is a concrete reflection of value. Those services within the disability sector that receive funding benefit those who are valued. Conversely if an individual does not receive essential services they are not socially valued.

Governmentality, Biopower and the Anthropology of Policy

Complementing my analysis of the value of life within the DMD community is my analysis of the state systems of care and governance and my use of the concepts of governmentality and biopower. These ideas have proved useful in exposing the concealed social mechanisms and political concerns that create some of the marginalisation and discrimination that people with DMD experience. People with DMD often do not receive health, education and other

services that appropriately meet their needs and engaging with bureaucratic hurdles causes additional suffering. During my research I heard about a family that entered into a complex appeals process to get a subsidy for the steroid deflazacort, which suited the particular needs of their son better than the routinely subsidized prednisone. This was a long and difficult process that involved an assemblage that included a paediatric neurologist, pharmaceutical manufacturers, bureaucrats from PHARMAC (the government's drug purchasing agency), the family and deflazacort. Other parents paid to provide this non-subsidised pharmaceutical for their sons. Another family repeatedly applied to a charitable trust that funded mobility vans so they could transport their son in his powerchair. Each time they had to provide all the required documentation, reiterating their financial circumstances and the needs of the family. Several times the trust was over-subscribed and they missed out. So for over a year their son spent long periods of time at home. He was able to attend school because transport for school was provided but apart from trips to the local shop there were few accessible venues available to him. These two examples and others I detail in Section Three of this thesis give a message about the way New Zealand society, as represented by these agents, values lives lived with DMD. The message is that these lives are not highly valued. Foucault's ideas about governmentality and biopower are useful tools for explaining these anomalies in service provision. Despite a shift away from the welfare state the New Zealand government continues to accept a duty of care for citizens. A key analysis of this thesis comes from Foucault's explanation of biopower, namely, that governments provide some services to meet the needs of the population as a whole, rather than the needs of the individual.

While Foucault did not analyse contemporary networks of power, like the one that exists within the DMD community, his accounts of systems of control and modes of resistance can be applied to my data. Foucault had a lot to say about power; central to his analysis is the rejection of the idea that power is simply repressive. Foucault argues that power is productive and constitutive. In Foucault's publications and lectures there is a chronological development of his ideas. Initially he discusses his ideas about disciplinary power (Foucault 1979) his thinking and language here facilitates later ideas about biopower and governmentality. Foucault explains the development of disciplinary power partly as a response to the limits of traditional (European) sovereign power following the industrial revolution and the increasing numbers of people living in industrial, capitalist cities (Foucault 1997:249). He adopts a broad view of historical changes across centuries noting how disciplinary power had gradually come to surpass sovereign power. He considers the significance of "mechanisms,

techniques and technologies of power" that were focussed on disciplining the individual through surveillance and training (1997:242). Foucault says that through disciplinary power the body is individualised. Social mechanisms manipulate the behaviour of the individual.

Many of the individuals and families who participated in my research expressed frustrations regarding their interactions with these mechanisms, particularly schools and hospitals. This frustration could be explained in Foucauldian terms. Even those families that Foucault might describe as docile (but others might describe as compliant) who appreciated the observation and expert knowledge that schools and hospitals offer, were unable to benefit from these institutional forms of disciplinary power because they were unable to conform to institutional ideas of productivity. The families' own distinctive knowledge was not valued and did not align with these technologies.

In the second half of the eighteenth century, Foucault argues, a new technology of power emerged. It was not about disciplining or manipulating the individual, rather this technology was developed to apply to the population more generally. Foucault defines biopower as "an explosion of numerous and diverse techniques for achieving the subjugations of bodies and the control of populations" (Foucault 1990:40). The focus of government strategies was not only the control and organisation of individuals but also the control and organisation of the population as a whole. Biopower developed in the context of major social changes when industrialisation caused populations to shift into large cities. With the rise of masses of people living together in cities new ways of managing these masses of people were also needed. Biopower developed to address these changes. Policies and procedures that manage births, deaths, reproduction, and health and illness work together to influence the population as a whole. The role of PHARMAC, mentioned in the example above, fits into this analysis. PHARMAC is the government's drug buying agency and has an important role to play in managing the health of the overall population. PHARMAC provided one standard subsidised steroid treatment but its side effects for this particular child (and others) were unmanageable. The very complicated bureaucratic process the family had to go through to get an alternative steroid subsidised reiterates that the role of the state was to meet general need and not specific need.

As well as addressing universal issues relating to rates of births and deaths, things that affect the entire population, biopolitics (the particular strategies of biopower) began to intervene in

areas where large groups, but not everyone, were affected. Strategies to deal with accidental incapacitation and old age were established. Key for those with DMD is Foucault's point that,

> The field of biopolitics also includes accidents, infirmities, and various anomalies. And it is also to deal with these phenomena that biopolitics will establish not only charitable institutions … but also much more subtle mechanisms which were much more economically rational than an indiscriminate charity … insurance, individual and collective savings, safety measures and so on (1997:244).

He goes on to say that these subtle regulatory mechanisms are designed to compensate for variations within the general population to create an average balance and acknowledge the chance or random element at a population level. The aim of such mechanisms is to "optimize the state of life" (1997:246). Foucault is clear that biopower does not address individual concerns rather it will "achieve overall states of equilibrium or regularity", which perhaps helps to clarify why those with DMD repeatedly feel marginalised by the mechanisms of biopower. According to Foucault these mechanisms are not designed to meet their individual needs but rather to enable the internal conditions of society to remain stable and constant despite a level of risk, or chance, within a population. The security measures of biopower are designed to optimise an average state of life not an individual state of life. Foucault also points out that while under the earlier model of governance, "Sovereignty took life and let live", biopower, on the contrary, "consists in making live and letting die' (1997:247) which is highly apt for the DMD community. This is an analysis which matches much of the DMD community's experience with the way health services operate in New Zealand. The problem for people with DMD is that their condition is so serious, so catastrophic, that the technologies of government that are meant to offer a level of security for protecting average states of life are not sufficiently extensive to always protect their lives.

Foucault explains that disciplinary power and biopower work together, for example in medicine. "Medicine is a power-knowledge that can be applied to both the body and the population, both the organism and biological processes, and it will therefore have both disciplinary effects and regulatory effects" (1997:252). Thus a Foucauldian analysis of DMD is not straightforward. The combination of disciplinary power and biopower is discussed further by Foucault. He suggests that together these powers cover the entire surface of society, although not always evident, they are inescapable, (1997:253). He goes on to develop this suggestion into an analysis of governmentality.

Governmentality is a term Foucault uses to describe the way the state exercises control over, or governs, citizens. Governmentality also refers to the way in which people are incentivised and mobilised to govern themselves; power is not only imposed from a central authority, like a state or institution, but is also enacted through the daily activities of people. In Society Must Be Defended, a text based on his 1976 lectures at College De Paris, Foucault reiterates that biopower comes to prominence with the rise of the nation state. The nation state replaces an earlier conceptual structure for organising society, the sovereign state. The primary aim or purpose of the sovereign state was to maintain control over territory (Foucault's Governmentality lecture 1978, in Burchell, Gordon and Miller 1991). This contrasts with the nation state, the complex, modern, capitalist entity which he argues has a plurality of aims, "for instance, government will have to ensure that the greatest possible quantity of wealth is produced, that the people are provided with sufficient means of subsistence, the population is enabled to multiply, etc" (1978 in 1991:95). Similarly, in 1976 he explains the basic function of biopower is to "improve life, to prolong its duration, to improve its chances, to avoid accidents, and to compensate for failings" (from Society Must Be Defended in Rabinow 1997:254). Clearly Foucault is not just suggesting a negative, oppressive form of social control with his ideas about governmentality. Rather he sees the governance of complex societies as requiring strategies that are both controlling and repressive AND creative. These seemingly contradictory phenomena are experienced by families living with Duchenne muscular dystrophy. Where the needs of this community align with standardised services designed for the population as a whole they (sometimes) have a positive experience of technologies or strategies that do improve and prolong life. In other cases, where the needs of this community are not aligned with the population more generally, the mantra of fiscal responsibility is used to withhold services, people experience an exclusion which can be understood through an awareness that biopower will assist some to stay alive others are allowed to die (Foucault 1997:247).

Governmentality, says Foucault, is a complex form of power. It involves self-government which reflects morality, aspects developed from the government of the family – based around economics that require the same "meticulous attention" that the head of a household pays to the family, and the family fortunes. This meticulous attention to detail is often seen in the careful allocations of capped budgets. Bureaucrats demand comprehensive documentation to ensure eligibility criteria are met and prioritisation of service can be ensured. Painstaking reporting and auditing processes are followed. Foucault examines metaphors for pastoral

care; the role of the shepherd caring for his flock, the captain governing the ship, a father's care for the family, a superior of the convent or a teacher for the school. Connections between governance of the self, the family and the state are established, that is, between morality, economics and politics.

Foucault does not argue that one type of social control replaces another but rather that social structures and institutions develop slowly over time so aspects of the medieval sovereign society and the disciplinary society exist within the rationality of the current system which he calls governmentality. There is no simple definition for this system for organising society. It is a complex form of power using "institutions, procedures, analyses and reflections, the calculations and tactics that allow the exercise of this very specific albeit complex form of power, which has as its target population, as its form of knowledge political economy, and as its essential technical means apparatuses of security (1991:102)".

So governmentality can be understood as the application of a new science – political economy – that results from an assemblage or a complex network or multiple relations, between population, territory and wealth. The state is no longer defined in terms of territory or land (although this is still important) but rather in terms of its population. This population is governed by the considerations of political economy and controlled by apparatuses of security. The equipment and support needs of people with DMD are costly and they often do not receive services that adequately meet their needs. Foucault's notions of governmentality, that the principles of economy and efficiency are applied to the governance of individuals and society, helps to explain the lack of fit between the needs of people with DMD and healthcare provided under a neoliberal system of government. This is an anomalous group within the population and the systems of government are not designed to meet exceptional or unusual needs.

In addition to using the concepts of governmentality and biopower to expose the concealed social mechanisms and political concerns that create some of the marginalisation and discrimination that people with DMD experience, I also use the anthropology of policy. Applying an anthropological study of policy to my analysis of the state systems of care and governance has enabled the often concealed power of polices to be revealed. The anthropology of policy demonstrates that polices often disguise power relations, despite being accepted as an unquestioned given (Shore and Wright 1997). My research reiterates the work of Shore and Wright (2011:2) that policies are themselves political technologies and

condensed symbols whose power is often not apparent or obvious. Policies can be powerful agents of social change – either reinforcing or challenging existing values, for example buttressing fiscal responsibility or refocussing health systems on person-centred, rather than system-centred care – and are barometers of social values and beliefs and culturally pervasive ideas. Also, policies have unexpected and unintended consequences or social lives of their own. In the case of people with DMD, this is sometimes exemplified in how the provision of funding specifically designed to support people with disabilities to participate in their communities actually compromised their care in hospital. By scrutinising policies through a Latourian lens, where the polices are considered as actants in an assemblage, where they have agency, cause change and interact with other aspects of the assemblage, their dynamic and unpredictable results are most clearly seen.

Integrative Concepts

As well as these two bodies of literature – an anthropology of hope that unpacks what the ambitions of people with DMD to live purposeful and meaningful lives says about the way life is valued; and what the application of theories of governmentality, biopower and an anthropology of policy uncover about the messages that social systems provide about the way life lived with DMD is valued – there are other theories running throughout my thesis. I rely on Tett's work on epistemologies (2015), Latour's actor network theory (2005), Lock and Nguyen's (2010) local biologies (as mentioned on page 3) and also Fassin's ideas about the politics of life (2012). These theories are applied throughout the thesis and integrate the two distinctive themes that emerged into a cohesive consideration of how lives lived with DMD are valued in Aotearoa New Zealand.

My data contained many themes which were open to a range of anthropological analyses. I had information about suffering and about living a fulfilling life in the present moment. I had information about the people's interactions with medical specialists and the health system. I had information about the meetings and appointments that structured daily life. I had information about the Neuromuscular Disease Registry and about international conferences and participation in drug trials. I needed an analytical frame that would contextualise all these diverse data. Aspects of Tett's The Silo Effect (2015) offered fruitful insights. Her ideas about different types of knowledge and expertise resonated with the various ways of understanding DMD that my data reflected. Participants were describing different sorts of expertise. There was the expertise of the people who lived with DMD and their families,

there was the biomedical expertise of doctors and specialists, there was the professional and managerial expertise of the staff at the Muscular Dystrophy Association (the MDA) and embracing all these epistemologies were the structures of government and the state, and these relied on another form of expertise. Tett's analysis of different epistemologies, different ways of knowing and understanding was useful and helped to clarify some of my data, although her discussion of silos and strategies to overcome such silos was not particularly relevant to my research.

Tett uses the term silos to describe situations where there was no value given to working collaboratively and where specialist silos developed in relative isolation. This is not the case with the epistemologies I discuss. In the DMD community no one was developing specialist knowledge or a level of expertise in isolation. People involved with DMD were connected with each other. Fragmentation occurred because people had an allegiance to their own particular epistemology. There were clear, formalised pathways for collaboration to occur – the election of people living with neuromuscular diseases on to the National Council of the MDA and the allocation of a position for a person with a neuromuscular disease on a government disability advisory panel, for example. Unlike Tett's corporate examples, fragmentation developed despite attempts at collaboration within the DMD community. For people living with DMD the negative impacts of fragmentation continue, despite attempts amongst various parts of the community to operate collaboratively.

Examining the Relationships within DMD Networks

Latour's actor-network theory is a useful tool to examine the relational ties within the DMD community. In addition to unpacking the various epistemologies that exist (Tett 2015) and identifying the power dynamics that give some of these epistemologies more clout than others (Foucault), my analysis includes reference to the social and political effects of inanimate objects. It is not just knowledge that is important in this community but also things, often technologically sophisticated things such as cough assist machines, ventilators, powerchairs and mobility vans. This equipment is central to the community and its provision, in the context of wider New Zealand society, is inequitably funded. Actor-network theory, as advocated by Latour (2005), provides a model for detailed descriptions of relational networks. Relational networks are complex webs that include tangible and symbolic actors

that combine to create change. These networks are potentially transient; they exist as the result of an alliance that serves a purpose (Latour 2005). This means that relations need to be repeatedly performed or the network will dissolve. This idea that networks of relations are not intrinsically coherent, and may contain conflicts is relevant to the DMD assemblage. Social relations are only ever in process, and require ongoing interaction. The idea that actors (or actants as Latour [2005:75] also called them) are both human and non-human can be challenging (Sayes 2014, Brush 2015:126). The focus of ANT is the way combinations of objects and people create situations in which change occurs. Latour is not just interested in identifying different parts of the network, he is interested in tracking the flows between the different parts of the network. Latour is interested in how different phenomena in the network (assemblage) influence and change things. Phenomena in a network can be animate or inanimate; people, objects (such as cough assist machines) and policies. As Shore and Wright elaborate, policies and policy documents are classic 'actants' that are translated and inscribed and implicated in a network of relations (Shore and Wright 2011). Latour's lens is a shift from previous social analyses which focussed on either structural constraints or on the agency of participants. Latour's approach allows for a consideration of circulation and movement. Like Foucault, Latour does not see power as simply repressive. During the 1980's and 1990's as Latour and other collaborators, notably Callon and Law, developed their thinking about ANT, it offered a novel approach to Science and Technology Studies that challenged the hegemony of earlier analysis. This resulted in some controversy (Winner 1993, Bloor 1991). Mol (2010) reviews aspects of ANT. She notes that it is not a theory:

> ANT is not a theory, says Callon. This is true in various ways. For a start, ANT writings do not offer something that remotely resembles a "law of nature" – or, given that typically the social and the physical are studied together in ANT work, a "law of nature-culture". Not at all. There is no attempt to draw the findings of various studies together into an overarching explanatory framework. There is no attempt to hunt for causes: the aim is rather to trace effects. (2010:261)

Mol argues that the strength of this approach is that it sharpens the sensitivity of the research (2010:261). This is why I have found it useful. It has enabled me to explore the relationships between the various parts of networks. These networks consist of both inanimate objects and people with different forms of knowledge and expertise relating to DMD. Conceptualising the relationships between people and things as part of an assemblage, or a network in which decisions or actions or developments in one part of the network have implications for decisions or actions or developments elsewhere, helped to clarify some of the complexities and nuances of my data. ANT has been critiqued for being descriptive rather than not

analytical (Amsterdamska 1990). Mol's response counters this criticism. My analysis relies on this detailed, nuanced approach to understanding situations and circumstances.

Local Biologies and the Discourse of Biomedicine

Local biologies, a concept originally developed by Margaret Lock (1995) to explain different biological outcomes that result from the entanglements of biomedicine with social, environmental and political particularities, is central to this consideration of the values attributed to lives with DMD in New Zealand. The historic relationship between anthropology and the study of the human body is one which, among other things, involved perpetrating oppressive racial categories and engaging with eugenic philosophies (Smedley 2005). Little wonder then that for the decades prior to Lock's engagement with anthropology the discipline left the study of the human body to biology. According to a biological epistemology "bodies are everywhere biologically the same" (Brotherton and Nguyen 2013). As a result of her work on ageing and menopause in Japan, Lock came to question this doxa, this common-sense way of understanding biomedicine. Using a concept she called local biologies Lock argues against the universalising approach of biomedicine. Bodies with DMD are clearly not everywhere the same. In the case of DMD life expectancy generally increases when people receive good biomedical treatment and are able to access appropriate technology and services. Biological outcomes are different in different places because bodies are entangled with the environments in which they live, and are impacted by social, cultural and political factors which can alter biological phenomena and outcomes.

As DMD is caused by the mutation or deletion of a single identifiable gene which results in a very predictable pattern of muscle degeneration, applying the theory of local biologies to my findings may seem untenable. Lock discusses how developments in genetic and epigenetic research reinforce the findings of medical anthropology that environmental, historical and socio-political factors are important in determining biological outcomes (Lock 2013). However, the type of epigenetic research Lock refers to is in its infancy and I am not suggesting that the genetic changes that cause DMD are affected by environmental factors. Nor am I suggesting that the deterioration in strength and muscle function, the predictable pattern of losing the ability to walk, to raise the arms, to sit up, to inflate the lungs, to swallow, to empty the bowels and so forth, changes as a result of environmental factors. Some aspects of the condition may be delayed as a result of steroids, physiotherapy or other treatment regimens but the fundamental loss of function is not altered by environmental

factors. These biological outcomes remain unaffected. However, the way individuals and families experience DMD in New Zealand is affected by social and environmental factors and this is the focus of this thesis. I examine aspects of New Zealand society that contribute to this distinctive experience. It is these factors that reveal much about how the lives of those living with severe disability are socially valued. The issues I address here do not, significantly, affect the predictable degeneration associated with the condition, although the research into pharmaceutical developments which I consider in Chapter Five has the potential to do so. The biomedical approach in which much of the DMD research and treatment is centred tends to focus on the effects of muscle degeneration. Here I also consider the psychosocial, or emotional effects for those with DMD and also other family members. In addition, I examine other aspects of care, and I unpack the way New Zealand's distinctive model of service provision creates a distinctive experience of living with DMD. This experience is not the same as the experiences of other similarly resourced countries because of the particular bureaucratic nature of service provision. This is characterised by a neoliberal hybridity with internal markets and a state funded purchaser provider split and the inequitable provision of supports for people with disabilities based on the cause of the disability. This thesis considers these factors as examples of "the way in which biological and social processes are inseparably entangled over time, resulting in human biological difference" (Lock and Nguyen 2010:90). To be clear, the biological differences that my analysis of local biologies refers to are the differences that ensue from the availability of medical treatment (the reduction of contractures where effective physiotherapy is available and the increasing age at which boys require wheelchairs as a result of new regimes of steroid treatment, for example).The emotional well-being of those with DMD, whose susceptibility to depression and anxiety is well recognised, is another site of biological difference. Experiences vary and depend on various issues including social inclusion and psychological support. Similarly, the availability of equipment and services that enhance longevity are environmental phenomena that lead to a distinct local biology of Duchenne muscular dystrophy in New Zealand. It is these social and political aspects of biological outcome that this thesis addresses. My analysis of local biologies is "a product of individual lived experience in specific environmental, historical, and sociopolitical contexts" (Lock 2013:303).

Finally, I also rely on the work of Fassin (2012) in this thesis. In this chapter Fassin reflects on what is meant by the term global health, it is a philosophical evaluation of the role of anthropology in this complex, interconnected world where the various manifestations of

global health are associated with multiple possibilities. Fassin suggests rather than a focus on health, anthropologists would be better thinking in terms of life (Fassin 2012:108). He argues that this approach would encompass both a biological and a political analysis. Fassin uses the ideas of Canguilhem (1943) to explain and support his position. Canguilhem said,

> everything happens in a society as if a society had 'the mortality that suits it,' the number of dead and their distribution into different age groups expressing the importance which the society does or does not give to the protraction of life. In short, the techniques of collective hygiene which tend to prolong human life or the habits of negligence which result in shortening it, depending on the value attached to life in a given society, are in the end a value judgement expressed in the abstract number which is the human life span. (in Fassin 2012:108-109)

This quote suggests that where the services accorded to people with DMD are of the highest standard then life expectancy will be higher too. This is the outcome that the DMD community strives for. A consideration of the politics of life includes a consideration of biological phenomena of disease (in this case the degeneration of the muscles caused by the mutation of the dystrophin gene which results in an inability of the body to produce the protein dystrophin which is essential for muscle function) and political considerations (in this case the provision of life preserving equipment and services, investment and support for biomedical treatments, and the social organisation that facilitates or impedes such service delivery). Fassin distinguishes between two types of politics of life which he calls the "value of life" and the "worth of lives". The worth of lives is evaluated by "the practical ways people's lives are considered, protected and cared for, or conversely abused and sometimes eliminated" (Fassin 2012:111). While Fassin is writing about the worth of lives on a global scale, I apply his approach to those aspects of care and protection relevant to this particular case study; issues such as the provision of neuromuscular treatment centres, disability equipment and community support. When describing the value of life, Fassin says he is judging "the ideological and ethical grounds on which the very principle of life is affirmed or challenged, moralized and politicized" (2012:111) and further argues that his research indicates that the way in which life is valued has shifted from a political recognition to a biological recognition. Fassin contrasts his own theorising about the politics of life with Foucault's biopolitics and notes that while Foucault focusses on the technologies of normalisation he, Fassin, is describing the ethics of government (Fassin 2012:112).

Fassin's focus on the politics of life has proved immeasurably valuable to my consideration of the ways in which lives with DMD are valued in Aotearoa/New Zealand. In explaining the

links between a concern with the way life is valued and both biological and political phenomena, Fassin has helped me draw together the theoretical approaches that I rely on in responding to my research question and the themes of my research findings.

Thesis Structure

My thesis is divided in to three parts. This first part sets the scene for the ethnographic narratives and findings discussed in Parts Two and Three. Here I provide the necessary background that informs the accounts of participants in the later sections. Having introduced my research question and the anthropological literature, Chapter Two gives an historical overview of DMD. I discuss the biomedical research and findings that define current medical knowledge regarding DMD. Chapter Three continues with an explanation of my research design and methods, including a discussion of the ethical dilemmas of researching a small community in which people are familiar with each other. And finally in this introductory section of my thesis, I describe New Zealand's health and disability system in Chapter Four. This chapter includes an analysis of the changing political ideologies and social values that have underpinned the major health reforms of the past thirty years and an explanation of New Zealand's social insurance system (ACC) that provides healthcare, equipment and support to those whose disabilities result from accidents. This system provides a different level of care to that which is provided to people whose disabilities result from medical conditions, such as people with DMD.

In the second section of this thesis I focus on situations which are illustrative of the way those who form part of the DMD community (those with the condition, their families, doctors and specialists, support workers, local and national support organisations) value lives lived with DMD. Chapter Five examines the hope that is inherent to the community. Here I apply the anthropology of hope to the activities and passtimes of people with DMD and to the activities undertaken by a wider network of actants engaged in research to find effective treatments for DMD. Discussions of the pain and suffering which are as much a part of the condition as hope, are discussed in Chapter Six. Lock's ideas about local biologies illustrate how differences in service provision create different experiences of DMD, which sometimes have different biological outcomes. The decisions that couples who know they are at risk of having a child with DMD make with regard to selective reproductive technologies provide insights into the ways families value lives lived with the condition. This is the topic of Chapter Seven; an ethnographically detailed chapter that explores the disparity of decisions that sisters,

mothers and their partners make. The love of an affected family member is enacted in very different ways.

Section Three is the section of the thesis in which I utilise the social analyses of Foucault, governmentality and biopower, and the anthropology of policy. I begin with a Latourian analysis of a cough assist machine assemblage in Chapter Eight. This assemblage operates to address a gap in service provision and provides a clear example of the different epistemologies that impinge on the DMD community. It highlights the way environmenal factors entwine with biology to create a particular local biology as well as offering insight into the operation of the politics of life. Similarly in Chapter Nine, which examines the way equipment and support workers are provided, I rely on the same literatures that reinforce the applicability of Foucault's analysis of biopower with policies that meet the needs of the masses, rather than the individual. The case study in Chapter Ten draws on the findings of the two previous chapters to reiterate the serious consequences that eventuate for those whose needs fall outside policy prescriptions designed for the general public. This chapter provides a very clear example of the way bureaucracies inform social messages about the value of lives lived with DMD.

The conclusion reflects on this research, my personal involvement with the DMD community, the theoretical frameworks of my research and the contributions of my thesis. I show how my study exemplifies the contradictory ways lives lived with DMD are valued in Aotearoa New Zealand and what lessons there are for policies that address disability.

In sum, I take the position that research knowledge is co-constructed during the research process by the interactions of the researcher and her participants, readings the researcher makes of documents and her observations. My research methodology could broadly be termed as social constructivism although I have been at pains to point out that the issues facing this community are also to do with the brute reality of maintaining bodily function, ambulation, limb movement and breathing, which are outside the parameters of the social constructivism of disability. As I began this thesis under the umbrella of the wider Marsden project I wanted to talk about reproductive technologies within the context of the whole of the participants lives. I realised that how life is valued had an enormous impact on the decisions that people made. As I went on it became clear that reproductive technologies were important at certain times but I came to realise that other issues had greater significance over the whole lifespan. These issues meant I needed a different theoretical perspective and I

found actor-network theory, discourses on hope and Lock's local biology to be additionally useful.

Chapter 2: Duchenne Muscular Dystrophy: A clinical overview

Due to its physical, emotional and social complexities there are many ways to describe DMD. This chapter offers a guide to the scientific discoveries that inform current biomedical knowledge about the genetic condition, Duchenne muscular dystrophy (DMD). The research I address here speaks, in some ways, to Fassin's careful analysis of the different measures of the value, or worth, of lives. The focus in this chapter is the biomedical understanding of DMD and is based within the frame of the medical gaze (Foucault 1973). Using this frame Foucault noted a development that occurred during the eighteenth and nineteenth centuries in which the human body came to be studied in isolation from its personality and social circumstances. While this approach is a well acknowledged phenomenon of modernity (Foucault 1973: xii-xiii), Foucault focusses on its epistemological significance and describes a shift in authority. As the medical gaze came to dominate an understanding of human health, greater authority was attributed to biomedical knowledge. This biomedical way of understanding DMD is central for the DMD community with whom I conducted my research and is one of the fundamental principles underlying the provision of services. Current biomedical knowledge about DMD is the result of the detailed observations and investigations of physicians and scientists over the past two centuries. Each discovery has added another piece to the complex puzzle that is DMD. This history informs the way DMD is treated, as knowledge is applied to treatment, and these enactments (following Mol 2002) contribute to a constant refinement of what DMD is. Furthermore they reiterate the way in which lives are valued in biological, rather than political terms (Fassin 2012:112).

Today those involved with DMD know that it is a distinct condition within a range of physiologically similar muscular dystrophies. There is a thorough understanding of molecular biology which informs the genetic nature of the muscular dystrophies and the processes whereby genetic mutations occur. The gene that is responsible for DMD and the protein this gene codes for has been discovered. Scientists and others in the DMD community know a great deal about the function of this protein and doctors know how best to treat DMD with existing therapies. The piece of the puzzle that families are waiting for is the piece that will offer patients longevity and mobility. This overview emphasises the crucial importance of this biomedical epistemology, the repeatable research outcomes that contribute to the clinical understanding and medical treatment of DMD. This particular epistemology is imbued by the

DMD community with hope. It is through biomedical research and expertise that the families maintain the hope for a future effective treatment for this devastating condition.

The following clinical history can be described as a Foucauldian archaeology of the knowledge of DMD. O'Farrell (2005:68) acknowledges that it is difficult to distinguish between the way Foucault uses ideas about an archaeology of knowledge and his ideas about genealogy. But as this chapter is a history of knowledge, rather than a consideration of the power inherent in this knowledge, it seems to be more archaeological than genealogical (O'Farrell 2005:69). Here I describe how research findings follow on from each other and how knowledge about DMD is cumulative. Foucault contends that illness is a disorder (Foucault 1973) and that science attempts to create order or understanding by imposing logical, systematic categories of knowledge into which different diseases and different aspects of disease fit. This is reflected in this account of DMD. I trace a history of scientific development and in later chapters consider some the ways this knowledge is used to condition and control people's subjectivities

What is Duchenne Muscular Dystrophy (DMD)?

Duchenne muscular dystrophy is a severe form of muscular dystrophy. It occurs as a result of mutations in the dystrophin gene, DMD locus Xp21.2. DMD generally occurs in boys and can be inherited. It is an x-linked recessive genetic condition. The DMD gene is the largest human gene and it codes for the protein dystrophin. When there is a mutation in this gene the human body is unable to produce the muscle protein dystrophin and a loss of muscle function and weakness occurs.

Duchenne muscular dystrophy affects approximately 1:3,500 male births worldwide. DMD occurs both through familial inheritance and through spontaneous mutations of the Duchenne gene. Approximately 2/3 of cases are inherited. In all hereditary cases the mother carries the faulty gene and passes it to her affected son(s). I have not heard of a case where a father has passed his faulty gene to a daughter. The degenerative nature of the disease and the premature death associated with it makes this scenario unlikely. If treatment improves it is possible men with DMD may pass the faulty gene to their daughters in the usual way X linked recessive inheritance occurs (Muscular Dystrophy Association, 2016a).

What are the Symptoms of DMD?

While the age of onset and the rate of deterioration can vary, DMD is a highly predictable condition. I have used information from the website of National Genome Institute and the website of the New Zealand Muscular Dystrophy Association to write this summary but there are many, many other similar sources that refer to this same pattern of symptoms. Initial indicators are usually noticed before the age of six and may appear in early infancy. Typically the first noticeable symptom is a delay of motor milestones, including sitting and standing independently. Usually boys do not walk until eighteen months. The muscles of the legs and pelvis waste and weaken. This can lead to a distinctive waddling gait, difficulty in climbing upstairs and a distinctive way of standing up called a Gower's manoeuvre. Frequent falls are also a feature of this condition. The muscles of the arms, neck and other areas also weaken, but not as severely or as early as those in the lower half of the body.

Boys with muscular dystrophy often appear to have large calf muscles in their early years. However the muscle tissue is replaced with fat and connective tissue. Calves may look large and strong but in fact there is no muscle strength. Associated muscle contracture causes joint problems, first in the ankles, followed by the knees, hips and joints of the upper body. Between the ages of one and six a person with DMD will in all likelihood have manifested some symptoms. Muscle strength steadily declines between the ages of six and eleven. By the age of ten braces may be required for walking and by age twelve, most boys are using wheelchairs.

Bones may be malformed and curvature of the spine can result. Curvature of the spine (scoliosis) is a serious problem in DMD. Scoliosis is a curvature to the side, accompanied by rotation of the spine. Scoliosis worsens most rapidly in the latter stages of puberty, during the growth spurt. If severe, scoliosis can be extremely uncomfortable, and limits the function of the lungs and the upper limbs. It is also disfiguring, causing the chest wall to become more prominent on one side.

In the later stages of DMD breathing difficulties occur. This can be due to both deterioration of the diaphragm and curvature of the spine. Cardiomyopathy (enlarged heart) occurs in many cases and for some this will have commenced during the early teens. Sometimes mild intellectual impairment is associated with DMD but it does not worsen over time, as the other symptoms do. Intellectual impairment occurs in 35% of DMD cases. It is uncommon for

people to live beyond 30 years, although longevity is currently increasing as treatments and management regimens improve.

History of DMD

The first case of a condition with the symptoms of muscular dystrophy, recorded in English, dates from 1830. Charles Bell, surgeon, describes a case study in his book The Nervous System of the Human Body. The symptoms Bell describes seem likely to be a case of muscular dystrophy. Bell did not undertake muscle pathology so the case is a probable, rather than definite, description of muscular dystrophy. As the patient was able to climb stairs at the age of eighteen, it is also likely that the case was Becker muscular dystrophy (BMD) not DMD (Emery and Emery 2011:18).

In 1836 an Italian physician reported brothers with probable symptoms. Professor Gaetano Conte was Head of Department at the Hospital for Incurables in Naples. He describes two brothers whose symptoms included; hypertrophy of the calf and deltoid muscles, progressive muscle weakness that began in the legs, the elder brother died at age seventeen from suspected hypertrophy of the heart, the younger brother suffered from severe joint contractures and an enlarged tongue. Again no muscle pathology was undertaken so the diagnosis cannot be certain. Nevertheless, as Emery and Emery point out "this appears to be the very first detailed clinical description of muscular dystrophy" (2011:20).

By 1847 autopsy results for a fourteen year old boy who probably had DMD were presented to the Pathological Society of London. Richard Partridge, a respected anatomist and teacher (Emery and Emery 2011:21), undertook muscle examination during an autopsy on the boy. The boy had from the age of 9 developed a progressive muscle wasting condition with the large calf muscles and contractures associated with DMD. He had died from pneumonia after catching measles. Partridge noted that the distinctive fatty nature of the calf muscles. Partridge's autopsy involved the macroscopic study of muscle tissue but not microscopic study. Microscopic study of muscle tissue was undertaken by William J Little, a senior physician at The Royal Orthopaedic Hospital, London, with a particular interest in "deformities". Little's findings (1853) almost certainly suggest that the boy suffered from DMD. Little notes, "The degenerated muscles exhibited abundance of fat cells, with few traces of muscular fibre; in some fibrillae the transverse markings were scarcely distinguishable, in others they were quite distinct" (in Emery and Emery 2011:24).

British physician Edward Meryon is, according to the account of Emery and Emery, the first doctor to distinguish the common traits of DMD, "…it was Meryon's great contribution to realize the similarity among the various cases and, most significantly, that they represented a specific and unique disease entity" (Emery and Emery 2011:35).

In 1851 he presented his findings about the similarities of a muscle wasting condition in nine boys, brothers in three separate families. His discussion included reference to the microscopic analysis of muscle tissue, and the spinal cord during an autopsy procedure. Emery and Emery (2011:36) note that at this time technological advances increased the use of microscopic study by medical investigators. Meryon also published articles about this condition in 1866 and 1870 with data that indicated the condition occurred in siblings, predominantly brothers and with recognition that it might be inherited maternally.

Although Meryon noticed a common familial pattern to the condition he was not precisely aware of the hereditary pathway of the disease. Heredity was not well understood in the mid-1800s. Mendel's work on inheritance, published in 1865, was not widely disseminated until 1900. Clues to the way this condition is transmitted did exist in the mid nineteenth century. It has the same mode of transmission as haemophilia (another X-linked recessive genetic condition) and this was well-known. In fact, the hereditary pathway of haemophilia is recognised in the Talmud, which excuses at risk male babies from circumcision.

 In 1861 Guillaume B. A. Duchenne (1806-1875), a French GP, refers in one of his publications to a patient he first treated in 1858 with clinical symptoms of DMD. In this publication the case is only briefly described and no histological (microscopic tissue study) findings are mentioned. However, Duchenne encounters other similar cases and in 1868 he published, "... an extensive series of articles" (Emery and Emery 2011:82). These articles include his original patient and twelve additional cases. Duchenne also reviewed fifteen German cases reported in German medical journals. Duchenne identified the stages of the progress of the disease.

One of his major contributions to understanding the condition came from his interest in muscle pathology. Duchenne invented a tool for obtaining muscle tissue from patients. This allowed him to make observation on the muscle tissue without requiring a body to be available for autopsy. The most fundamental abnormality in the muscle tissue that he noted under microscopic investigation was the hyperplasia (enlargement due to an increase in cells) of the fibrous connective tissue, which later turned into fatty tissue. Duchenne was unable to

explain how this increase in connective tissue occurred. His detailed clinical descriptions and thorough pathological analysis were an important steppingstone in the understanding of the complexities of the disease. Duchenne, however, did not note the familial traits of the disease.

Meryon and Duchenne were doctors practising in similarly large industrial cities during the same medically stimulating times. They identified the clinical parameters of DMD and made some initial steps in discovering its pathology however there was still an enormous amount of research to be undertaken to clarify the nature of the disease.

Duchenne's publications during the 1860's and 1870's were instrumental in increasing awareness of, and interest in, non-infectious muscle disease. The widespread dissemination of his publications undoubtedly contributed to an increase in research in this area. Emery and Emery note that the period immediately following Duchenne's publications saw a proliferation of accounts of similar muscle wasting conditions. These reports coincided with a growing scientific interest in nosology – the categorising of disease. Muscle disease was of significance as investigators tried to identify the cause of the condition. By the mid 1800's the cause of infectious diseases was becoming understood within the medical fraternity, but the cause of the muscle wasting disease described by Meryon and Duchenne remained a mystery.

After Meryon and Duchenne, the research of Gowers gives the most thorough analysis of pseudo hypertrophic muscular paralysis, the name given to DMD at the time. The introduction to his 1879 lecture and associated monograph remains a classic description of the condition (see the opening quotation to this thesis) and his conclusions regarding clinical presentation have hardly been improved upon (Emery & Emery 2011:99). His contribution to the study of DMD continues to be acknowledged through the eponymous Gowers' manoeuvre which describes the distinctive way in which boys with DMD get up after lying down. Gowers' 1879 monograph was based on 220 case studies. Twenty four of these cases were his own patients. Twenty were patients of his colleagues and 176 cases were from reported literature. His analysis was, therefore, based on a much larger number of case studies than the work of those who preceded him.

Gowers overlooked one significant feature; that mild intellectual impairment can be a symptom of the condition. He did make a radical discovery with regard to the histology of the muscle tissue. His contribution was to discover that the anterior horn cells were normal in this condition while in progressive muscular atrophy these cells present abnormally. This finding

is an initial step is the classification of muscle wasting diseases. However, his most interesting observations (according to Emery and Emery 2011:101) were a "focus on a predilection for the male sex and its familial nature." Gowers realised that heredity passed through the maternal line and that a mother's brother may also be affected and he noted the similarity in inheritance to haemophilia.

Gowers also pointed out that the premature death associated with pseudo hypertrophic muscular paralysis was unavoidable regardless of treatment. However, he identifies the need for appropriate exercise to prevent joint contractures and the possible use of mechanical aides to maintain walking for a longer period of time. The use of leg braces remains part of the accepted treatment of DMD. Gower's promotion of spinal jackets to impede the development of spinal deformity also remained a recognised form of treatment for some time. Although Gowers' work was the most comprehensive study of the disease at the time it contained some ambiguities that were clarified by later medical experts. It included case studies which are now known to be different types of muscular dystrophy.

Classification within the Muscular Dystrophies

Initial attempts to classify the muscular dystrophies were based primarily on the clinical presentation of the disease and differences in the way it manifested in children of different sexes. Up until the end of the nineteenth century there was little understanding of genetic inheritance. It was only as Mendel's principles of heredity came to be recognised and accepted that a clearer understanding was possible. It was not until 1943 that a study by American doctor Ade T Milhorat (1899-1997) attempted to classify the role of genetic factors in the disease. From the 1940's onward a plethora of studies attempted to relate genetic differences to clinical differences. These studies clearly show a shift in analysis – while clinical and pathological data remained important genetic factors became central. In fact, by 1960, genetic factors were not only important in understanding the cause of the disease but also in diagnosis.

The findings of Walton and Nattrass published in 1954 eventually led to the identification of five types of dystrophy; Duchenne muscular dystrophy, autosomal dominant facioscapulohumeral muscular dystrophy, limb girdle muscular dystrophy, distal myopathy and ocular myopathy. Aware of the need for genetic information Walton attempted, unsuccessfully, to locate the genes responsible for these muscular dystrophic conditions. Using existing genetic knowledge and technology Walton, researching in 1956 and 1957, was

not able to identify the responsible gene/s. It was the work of mathematical geneticists, C.S Chung and Newton Morton in 1959 that further explained the genetic transmission of the muscular dystrophies. Using discriminant analysis, Morton and Chung concluded that there were four different groups of muscular dystrophy; dominant facioscapulohumeral, X-linked Duchenne, recessive limb girdle and isolated limb girdle.

Further Significant Discoveries

With each scientific breakthrough in understanding of how one aspect of the human body works came a greater appreciation of the complex, intricate systems involved. For example, the advances in microscope technology during the nineteenth century led to a detailed understanding of plant and animal cells (Brown 1831 and Schwann 1839, cited in Emery and Emery 2011). In 1869 German scientist Miescher-Ruesch researching cell formation described a new substance, which he called "nuclein". It is now known as DNA (Deoxyribonucleic acid) (Wolf 2003). These developments are part of the archaeology of knowledge about DMD, to apply a Foucauldian metaphor; they are the fossils and artefacts that lie beneath the current publications regarding current best treatment guidelines (Bushby et al 2010a and 2010b) which are highly valued by clinicians and patients alike.

In 1944 Avery, MacLeod and McCarty working at the Rockefeller Institute in New York discovered that this DNA contained the cell's genetic information (Emery and Emery 2011:177). This discovery was followed by the publication in 1953 of Watson and Crick's paper that explained the double helix structure of DNA. Since their pioneering publication a wealth of research on the role of DNA has been undertaken and contributed to further knowledge about DMD.

By the late 1950's there was widespread acceptance that DMD had an X-linked mode of inheritance which meant that healthy females could carry the mutant gene. This led scientists to search for a test which could detect carrier status. In 1959 two Japanese biochemists, Professor Setsuro Ebashi and Professor Hideo Sugita, published the results from their investigation into serum levels of creatine kinase activity in dystrophy. Their results indicated that testing for this enzyme was most useful in the diagnosis of DMD. Testing for creatine kinase showed preclinical cases of DMD had elevated levels of creatine kinase of up to 100x the usual levels. In Becker muscular dystrophy similarly high levels of creatine kinase were noted. Most significantly female carriers of DMD also had high levels of creatine kinase. These findings meant it was possible for female carriers with affected relatives to determine

their status before having children. However, further research indicated that the creatine kinase levels were raised in only seventy percent of female carriers. Thirty percent of healthy female carriers would not show up using a CK test. Despite much research being undertaken to discover a more effective test to better diagnose female carriers with affected relatives, no successful test was developed until the DMD gene was identified in the mid-1980s (Emery and Emery 2011:156-8).

Prior to the identification of the DMD gene in 1985 the most important step in understanding DMD was made by Alan D. Roses, an American biochemist. His contribution was the discovery of defective muscle membrane in DMD. The exact nature of the defect could not be determined by biochemists. The new approach of reverse genetics was required to understand the exact cause of the defective muscle membrane and before this could happen the defective gene had to be isolated (Emery and Emery 2011:171-174).

Isolation of the Duchenne Gene

It was in the early 1980's that research to isolate the Duchenne gene began in earnest. Dr Don Wood was the scientific director of the American Muscular Dystrophy Association at that time. He realised that the existing scientific knowledge meant it was timely for the investigation to begin. Dr Wood was instrumental in the decision that the Muscular Dystrophy Association (USA) made to allow money from an annual telethon to be set aside to fund research to isolate the Duchenne gene (Schmeck 1988). Three laboratories were contracted to analyse family blood samples. Louis Kunkel, a newly qualified PhD from Harvard with an interest in the X chromosome, became involved. At the time it was known that there were at least 30,000 human genes but none had been linked to a specific disease.

Because DMD manifested mainly in boys, with only a few exceptional female cases, scientists knew the faulty gene lay somewhere on the X chromosome. Kunkel decided to compare the genes from healthy family members to members who had Duchenne. In his comparison he was looking for a missing piece of genetic material in those with DMD. The genetic information provided by a boy called Bruce Bryer was important. Bruce Bryer had multiple genetic disorders including DMD because he was missing a large piece of his X chromosome. Study of the boy's genetic material meant that the Duchenne gene could be located within Bryer's particular missing piece of chromosome.

In 1985 Kunkel announced he had isolated the gene responsible for DMD. This was an exciting scientific breakthrough. It was the first time in history a gene entirely responsible for a disease had ever been discovered (Schmeck, 1988). Whilst Kunkel is credited as the scientist who isolated the Duchenne gene other researchers in Europe and Canada were also attempting to find the gene responsible for Duchenne muscular dystrophy. Shortly after Kunkel's announcement that the Duchenne gene was located at Xp21.2, Worton, a Canadian, published the same findings. Worton's work was, like that of Kunkel, based on a comparison of abnormal X chromosome disorders. Research into those exceptional female cases of severe DMD was used as a starting point in the search to isolate the Duchenne gene. While some girls with severe symptoms of Duchenne muscular dystrophy were found to have Turner's syndrome (in which all or part of one of the X chromosomes is absent) studies in the 1970's indicated that some girls with severe symptoms proved to have an X/autosome translocation (when part of the X or sex chromosome transfers to an autosome or homologous chromosome).

Emery and Emery note that, "the breakpoint on the X chromosome was always in the region of Xp21. The most likely explanation was, therefore, that the translocation had in some way disrupted the normal gene at Xp21 which thereby resulted in the disease" (2011:185). In collaboration with a visiting paediatrician from Belgium, Christine Verellen, Worton studied the significance of this X/autosome translocation. Verellen had just described a case of a girl with Duchenne muscular dystrophy and an X/ autosome translocation. The two began work to map the breakpoints on the translocation. Their work indicated that there was a gene deletion on the X chromosome at p21.2.

The initial discovery of the gene led to a great optimism that an effective genetic treatment would soon be available. Pat Furlong, founder of the support group Parent Project Muscular Dystrophy and a long-time advocate on behalf of affected families, poignantly recalled the effect the publicity about the discovery of the Duchenne gene in 1985 had on her sons. The media-hype around the discovery suggested that a genetic treatment would be available within the year. She recounted, at a Duchenne conference, one of her boys retrieving his calendar and circling the date when he believed he would be able to play Little League, like all his friends.

Although the isolation of the gene has not lead to an immediate cure, it has led to a greater understanding of the condition. Kunkel had made the probes of his research available for

international study. Investigators throughout the world used his probes and it soon became apparent that the Duchenne gene was far larger than scientists had initially anticipated. Most genes are 2,000 base pairs in size. The Duchenne gene had 2.5 million base pairs. The dystrophin gene is 90 times the size of most genes. It is its size which makes it susceptible to random mutation and explains an issue which had puzzled scientists for years – why so many cases of this genetic mutation occur spontaneously.

Furthermore, the use of Kunkel's probes also showed that, "up to 70% of cases of Duchenne and Becker dystrophies had gene deletions or, rarely, duplications. The remaining 30% have now been shown to be caused by a variety of point mutations" (Emery and Emery 2011:193).

Discovery of Dystrophin and its Function

Once the Duchenne gene had been isolated, scientists needed to discover exactly which protein the gene coded for. It was the work of Eric Hoffman that was important in ascertaining the protein that the Duchenne gene produced. Hoffman joined Kunkel's research team in 1986 and focussed on protein identification. Hoffman used the technique of reverse genetics to identify the protein that the Duchenne gene coded for. This was another first in the study of DMD.

> Duchenne muscular dystrophy was the first disorder in which the protein defect was identified by molecular techniques in a disease in which there was no prior knowledge as to its basic cause. It was therefore a triumph for reverse genetics". (Emery and Emery 2011:195)

Hoffman and his colleagues called the protein dystrophin. As those with DMD have a gene mutation their bodies do not make this protein. Hoffman et al (1988) discovered that,

> [...] dystrophin was virtually absent in Duchenne dystrophy, abnormal (in size or amount) in Becker dystrophy, but normal in all other muscular dystrophies, including limb girdle, Emery-Dreifuss, and oculopharyngeal muscular dystrophies, as well as in a variety of neurogenic disorders. (Emery and Emery 2011:196)

This discovery helped in the clarification of the different dystrophies. As a result of this work it became clear that some patients believed to have limb girdle muscular dystrophy or spinal muscular atrophy, did, in fact, have Becker muscular dystrophy. Further work was required to detect exactly how dystrophin worked.

It is now known that, dystrophin is found mainly in the skeletal and heart muscles and that small amounts are found in the nerve cells in the brain. In skeletal and cardiac muscles,

dystrophin is part of a group of proteins that work together (a protein complex) that strengthens muscle fibres and protects them from injury as muscles contract and relax. Parent Project Muscular Dystrophy uses the metaphor of a shock absorber to explain the job of this protein complex. Dystrophin links muscle fibres to membranes. Without dystrophin muscle fibres are less resistant to mechanical stress. The dystrophin complex acts as an anchor, connecting each muscle cell's structural framework (cytoskeleton) with the lattice of proteins and other molecules outside the cell (extracellular matrix). The dystrophin complex may also play a role in cell signalling by interacting with proteins that send and receive chemical signals (NIH National Library of Medicine 2016).

Kunkel was aware of the similarity of dystrophin to other cytoskeletal proteins but it was Kevin Campbell of the University of Iowa who identified that dystrophin was part of a protein complex (Emery and Emery 2011:196-8). Hundreds of mutations in the DMD gene have been identified in people with the Duchenne and Becker forms of muscular dystrophy. Most of these mutations delete part of the DMD gene. Other mutations abnormally duplicate part of the gene or change a small number of DNA building blocks (nucleotides) in the gene. The DMD gene mutations almost always result in a stop codon. The chain of amino acids that makes up the protein dystrophin cannot continue to be built if the codons do not make sense. In other words the three nucleotide "letters" do not code for an amino acid.

Mutations that cause Becker muscular dystrophy, which typically has milder features and a later age of onset than Duchenne muscular dystrophy, usually lead to an abnormal version of dystrophin that retains some function. Mutations that cause the more severe Duchenne muscular dystrophy typically prevent any functional dystrophin from being produced. Skeletal and cardiac muscle cells without enough functional dystrophin become damaged as the muscles repeatedly contract and relax with use. The damaged cells weaken and die over time, causing the characteristic muscle weakness and heart problems seen in Duchenne and Becker muscular dystrophy.

Mutations in the DMD gene also cause a form of heart disease called X-linked dilated cardiomyopathy. This condition enlarges and weakens the cardiac muscle, preventing it from pumping blood efficiently. Although dilated cardiomyopathy is a sign of Duchenne and Becker muscular dystrophy, the isolated X-linked form of this heart condition is not associated with weakness and wasting of skeletal muscles. Researchers are not certain why some mutations in the DMD gene cause X-linked cardiomyopathy instead of muscular

dystrophy. They believe that some DMD mutations affect a version of dystrophin that is specific to heart muscle (NIH National Library of Medicine 2016) .

Boys with DMD do not make dystrophin so as their muscles move the cell membranes tend to tear and are replaced with fat and scar tissue. The gene for DMD led scientists to identify a protein in muscles that no one had ever heard of or seen before. Although the discovery of the gene did not provide an instant cure, the discovery did allow females with affected male relatives to have their carrier status effectively tested.

Treatment of DMD

In 2010 a group of experts, led by Kate Bushby, Professor of Neuromuscular Genetics at Newcastle University produced two management documents for best practice in the care of Duchenne muscular dystrophy (Bushby et al 2010a and 2010b). Eighty-four international experts in the diagnosis and treatment of DMD were chosen to represent a broad range of specialties. They independently assessed various recommendations and methods used to manage DMD. In total they considered over 70,000 different, possible strategies. The final guidelines represented those treatments and strategies the group agreed were the best practice for managing DMD. The best practice guides identify management strategies in seven areas of medical specialisation; neuromuscular treatment, rehabilitation management, orthopaedic management, pulmonary management, cardiac management, gastrointestinal management and psychosocial management.

Neuromuscular management focuses on maintaining muscle strength. Currently the best way for those with DMD to maintain muscle strength is by taking exceptionally high doses of steroids. The recommendations (Bushby et al 2010b) are 0.75mg per kg per day for prednisone and 0.9mg per kg per day for deflazacort. Medsafe New Zealand notes that the recommended dose of prednisone for maintenance (i.e. regular, longterm daily dosage) in children is 0.125 to 0.25mg/kg daily (Medsafe 2016). This means children with DMD are taking about six times the usual dose. Steroids are the only drugs known to slow the decline in muscle strength. The benefits of steroid use include the ability to walk for longer, which means that children can participate with others more easily and the longer they are on their feet the greater the delay in scoliosis (curvature of the spine) and breathing problems. This in turn reduces the need for spinal fusion surgery, when a rod is inserted next to the spine to improve posture and breathing, which is a painful and distressing procedure. However, there

are also negative side effects of steroid use including; stunted growth, delayed puberty, a distinctive cushingoid (round) face, weight gain, increased appetite and osteoporosis. There are ways some of these side effects can be managed, for example testosterone can be given to trigger puberty and calcium supplements may assist with bone density. Another serious side effect for some children is that these high doses of steroids can have a major impact on their moods and emotions.

Rehabilitation management concerns the maintenance of muscle extensibility and delaying or avoiding joint contractures. Rehabilitation involves a range of experts; physiotherapists oversee a programme of stretching which should become part of the family's daily routine, occupational therapists, orthotists, wheelchair assessors, other seating specialists and on occasion orthopaedic surgeons will be involved in this management. The aim is to avoid or delay the characteristic joint contractures which reduce movement and independence.

Orthopaedic management addresses ways to avoid or, if necessary, treat scoliosis. The current high dose steroid regime means fewer people with DMD are developing scoliosis and hence fewer require spinal fusion surgery. However, many of the young men I spoke to had had this surgery as the current high dose regime was not recommended when they were younger. Every effort is made to avoid scoliosis because it reduces lung function. This means boys with DMD need regular checks of their spine, spinal X-rays when they become wheelchair-dependent, and the installation of correct seating systems in the wheelchair, and surgery if the degree of the spinal curve becomes greater than 20^0.

Pulmonary management focuses on looking after the breathing muscles. Usually boys do not have trouble breathing or coughing while they are still walking. However, as they age the breathing muscles are affected and the risk of chest infections increases, often due to an ineffective cough. This can be addressed with breathing techniques that can be taught by a physiotherapist and manual cough assistance or a mechanical cough assist machine. Commonly young men will require a ventilator at night, then, if they experience increasing daytime tiredness (due to a lack of oxygen in their blood), they may start using one during the day as well. Regular monitoring of lung function is an important part of this management.

Cardiac management aims to detect and treat the deterioration of the heart muscle. Cardiomyopathy commonly occurs in DMD and often without the symptoms associated with other muscular deterioration. Regular monitoring of the heart is important because there are heart medications available to treat abnormalities.

Gastrointestinal management emphasises appropriate nutrition. At different stages both being over and underweight are common concerns. Often children are overweight when they are taking high dose steroids that increase appetite and their ability to exercise is compromised by physical weakness. In older people difficulty in chewing and swallowing can lead to weight loss. Maintaining a healthy diet is important and at a later stage some people will opt for a gastro feeding tube. Constipation can also be a serious problem and fluids must be sufficient to prevent impacted bowels. Constipation can also be addressed with laxatives. Dieticians and speech and language therapists (who can assist with difficulties with swallowing and choking) are specialists who should be involved in this aspect of care.

Psychosocial management focuses on behaviour and learning. Along with all the physical problems associated with DMD, problems with behaviour and learning are also common. Children with DMD have increased risks of poor language development, poor comprehension and poor short-term memory; they may have difficulties with social interactions, anxiety, and controlling anger. There are also increased risks of Autism spectrum disorder, attention deficit hyperactivity disorder and obsessive compulsive disorder. In many cases boys eventually catch up to their peers with their learning delays. For example, boys who learn to read more slowly than their peers do become competent readers and may go on to complete tertiary studies. However, some boys with DMD also have intellectual impairments that they do not grow out of (Bushby et al 2010a).

Treatment developments – Future Hope

These management strategies are currently the best way of addressing the degenerative nature of DMD. At the moment there is no way to stop the muscle degeneration that characterises the condition. However pharmaceutical treatments are currently being developed and it is hoped that they will significantly delay the onset of symptoms or prevent muscle degeneration altogether. These treatments give some hope to the DMD community. Of particular interest to some New Zealand families is a treatment, ataluran also called translarna, being developed by a pharmaceutical company called PTC Therapeutics .This is a treatment that seems to override certain types of genetic mutation during the translation of DNA through the messenger RNA process. PTC Therapeutics say,

> We believe that ataluren interacts with the ribosome, which is the component of the cell that decodes the mRNA molecule and manufactures proteins, to enable the ribosome to read through premature nonsense stop signals on mRNA and allow the cell to produce a full-length, functional protein. As a result, we believe that ataluren

Three New Zealand families are involved in the phase III trial of this drug. Translarna™ is already available in the European Economic Area for boys with DMD who are aged over five year and are still ambulatory. However the cost is prohibitive. The actual cost remains confidential but I have been told a commercial price of around $300,000 ($NZ) per annum would be a realistic estimate.The treatment is effective for only 13% of Duchenne cases. The potential for effective treatment offers future hope, but the pricing regime means this particular treatment is unlikely to be immediately available for New Zealand children.

Chapter Conclusion

This clinical history of DMD is an example of Foucauldian archaeology, showing how knowledge about DMD developed in a particular social and historical context. The first recorded descriptions of muscular dystrophy by Bell and Conte in the 1830's align with Foucault's analysis of the formation of medical knowledge as an aspect of scientific endeavor (Foucault 1973). The categorisation of the different muscular dystrophies by von Leyen, Erb, Ladouzy and Dejerine (in Emery and Emery, 2011) was part of a wider scientific endeavor to order all types of illness into set categories; this nosology became a prevalent part of understanding illness. The importance of the autopsy in the work of Partridge, Little and Meryon reinforce Foucault's point that at a particular historic moment death became an instrument for understanding life. The role of the autopsy, as a research tool, gave way to the role of the gene. Since the 1940's genetic research has dominated findings into DMD. Genetic research is a highly specialized form of knowledge (Simpson, 2000). Thus it is clear that knowledge about DMD is part of an historic process. The biomedical epistemology is embraced by scientists, physicians and families. It is the harbinger of hope for DMD. It is also just one epistemology among several in this discussion of life lived with DMD in New Zealand. This particular epistemology is imbued with a social authority that gives it more influence than some of the other epistemologies, or ways of knowing, associated with this condition. This authority reinforces the biological value of life that Fassin argues currently predominates in the ethics of governance.

Chapter 3: Research Design and Methods

Research Design

This section describes the construction of my research field, my use of flexible methods and the co-production of information, the ethical dilemmas of this particular project and the development of my analysis. In designing this research I followed Gupta and Ferguson (1997) who highlighted the need for critical reflection on the selection of fieldwork sites by anthropologists (1997:2). They noted the inadequacies of the traditional (Malinowskian) single, bounded fieldwork site, designed to study small-scale societies, for the analysis of the contemporary, complex, postcolonial, world.

Construction of the Field

The field that I describe here is one part of a "multistranded methodology" (Gupta and Ferguson 1997:37). I construct the research field through various sites. The sites create, "a mode of study that cares about, and pays attention to, the interlocking of multiple socio-political sites and locations" (1997:37). The research is based on fieldwork with the muscular dystrophy community. My field is not a geographically discrete location but a thematic location that exists within and beyond New Zealand society. It is centred on the families that live with DMD and those who interact with them. In order to understand what it means to live with Duchenne muscular dystrophy I am creating a field that goes where the families go and beyond that to those entities and networks in which the families are enmeshed. These families are surrounded by a variety of advocacy and support agencies and service providers, some of which have funding or advisory relationships with national government. Through the Muscular Dystrophy Association my field also links to international advocacy and research groups, the World Muscle Society, Australasian Neuromuscular Network (ANN) and Treat Neuromuscular Disease (TREAT NMD), Project Parent Muscular Dystrophy (PPMD) and Muscular Dystrophy UK. These organisations are involved in the latest scientific research and treatment for DMD. So my field (following Madden 2010 and Gupta and Ferguson 1997) is a conceptual place.

My research considers the lived experience of DMD in New Zealand. DMD is a rare condition and NZ has a relatively small population base of 4.5 million (Statistics New Zealand 2014a). A recent national Muscular Dystrophy prevalence study was undertaken by

Auckland University of Technology. Data collection was completed in July 2016 so findings are still to be published but researchers inform me that there are 103 people currently diagnosed with DMD in New Zealand. This figure is in line with the findings of an international literature review that preceded the study and estimated that the prevalence of DMD in New Zealand was between 1.7–4.2 per 100,000 (Theadom et al 2014: 267). Given the relatively small numbers of people involved I have been able to conduct nationwide research. The national body that supports people living with neuromuscular diseases, the MDA, agreed to support my research. The organisation itself became an integral part of the research field. I volunteered in the National Office, I attended social events organised by the regional branches and the Membership Manager contacted MDA members to see if people were interested in participating in interviews. My field also involved attending conferences and workshops and other events attended by people with DMD such as powerchair soccer matches. My field was primarily New Zealand based but had tendrils that extended to international networks.

Co-production of Knowledge

As I spent increasing amounts of time with people involved in various ways with DMD I gradually developed a complex understanding of what it means to live with DMD. My research methods were flexible and I was able to clarify new information and ideas by revising interview questions and through discussions with various members of the community. The more I learnt, the more relevant and appropriate my interviews and discussions became. The research field for this project reflects my understanding of DMD. The field comprises different, distinct sites, some of which were pre-existing networks. This chapter explores three distinct aspects of my fieldwork; the MDA national office, social events attended by families directly affected by DMD and interviews. In the description of each fieldwork example I demonstrate the way my understanding of what it means to live with DMD developed through conversations and observations. The data collected through participant observation and the data collected through the interview process were mutually informative, as I demonstrate below.

Ethical Dilemmas: Emotional Intensity and Anonymity

Madden (2010:33) states that ethnographic research is infused with ethical considerations. He notes that there are ethical decisions to be made at every stage of the research process;

designing research, conducting research, analysing research and writing up research all involve ethical decision-making. This certainly reflects my experience of undertaking research. There have been many ethical dilemmas in this research and I consider these in context as I describe aspects of the research. However, an over-arching comment about the emotional intensity of this research is salient here. DMD families are involved in an inevitable spiral of grief. The gradual nature of the weakening of the body's muscles means that those who have DMD are constantly losing physical abilities. They do not just lose the ability to walk but gradually the ability to use their arms and then their hands and then to control their speech. One mother said, "The grief never ends." Constantly the boys, or men, and their families are aware of new physical tasks that they struggle with. And ultimately these young men face a premature death. Given this context the ethical considerations common to anthropology are magnified in this project. For example, in Chapter Ten I discuss the end of a life. I have needed to consider the impact of this part of my thesis on the young man's family and friends who remain active in the DMD community. How will the parents feel if I talk about the death of their son? Will I open up additional experiences of grief? I feel that I should ask their permission to use the information I collected. But, I wonder, is it appropriate to ask parents to read my analysis of the death of their son? However, is it ethical to write about that death without consulting them? Given the particular circumstances and the familiarity of many in the community with those circumstances, I cannot guarantee anonymity. How should I address this ethical issue of confidentiality?

The small size of the community and the hereditary nature of DMD is a particular concern as some families have known and supported each other across several generations. This makes anonymity particularly difficult. I either had to change the details of people's stories so radically that they became fictional accounts based loosely on fieldwork data (a strategy adopted by Bluebond-Langner 1978) or I consult with informants and ask for permission to use personal information in a way that may allow those who know a family's background to identify their input. I decided on the latter.

Researcher Empathy and Subjectivity

Here I acknowledge my own role in the research process. It is now well acknowledged in anthropological methodology that the researcher is integral to the research process (Madden 2010). I reflect on my role as a researcher in relation to my emotional reactions to the deaths I mention above as an example of strange role I found myself inhabiting as part friend, part

colleague and part researcher. Part insider and part outsider. While I focus here on death and grief to explain my insider/outsider position there were many fieldwork experiences that were not to do with grief.

My fieldwork experiences include that which Rosaldo (1989) refers to as an "emotional force", which Behar (1996) refers to as "anthropology that breaks your heart", and which Turner and Bruner (1986) refer to as "the anthropology of experience" .The grief I experienced and witnessed helped me to understand the emotional burden that accompanies individuals and families living with DMD. The analysis I adopt in this thesis is, however, not about me and I do not assume that the sadness I experienced is the same as the sadness research participants experienced (Beatty 2010). The insider/outsider position I inhabit in this research is central to my analysis of all the data I collected. Tett offers a succinct recap of this dichotomy, "Anyone who has been immersed in Anthropology is doomed to be an insider/outsider for the rest of their life; they can never take anything at face value but are compelled to constantly ask: why?" (Tett 2015:50).

Here I consider this insider/outsider polarity specifically in terms of three of the deaths that happened during my fieldwork. In first instance I was clearly an outsider, I did not know the young man well, my involvement resulted through my positon as a researcher and my emotional reaction to his death was muted. I was very sad to hear of his death and especially some of the circumstances leading up to it but I was not affected at an immediate and personal level. With the second death I had become an insider. The man who died was both a colleague and a friend. I went to visit the family home with a group of work mates and we all cried when we saw him in his coffin. I watched a livestream of the funeral as I had a terrible hacking cough and did not want to spread it among the many people with neuromuscular conditions who I knew would be at the funeral. It was a strangely lonely experience and I wished I could have attended the funeral and grieved properly with all the other mourners, many of whom I now knew well. As I write this thesis this man is never far from my thoughts so often I feel sad thinking of him. The third young man who died during my fieldwork had a sudden and unexpected death. I had met him many times during my research and his parents had become friends. He was a teenager and like most teenage boys he did not spend much time chatting with me, a middle-aged woman whom he knew as someone who worked at the MDA office. His parents however I knew very well. The tears I shed were as much for their loss, as for their son. I knew he was the centre of their world and how devastated they would be to lose him so suddenly. In each case I shared the grief of the wider community but the

exact nature of my emotional reactions varied according to the type of relationships I had with the person who had died and their family.

Whilst this study could be described as studying "at home" (Madden 2010: 45-54, Jackson 1987) as it is New Zealand based research and I am a New Zealand based researcher, I am certainly not an insider to this community. I have no experience of living with a disability or of having an immediate family member with a disability. However, I did not feel like a complete outsider. I have a shared experience of living in New Zealand. I have participated in the New Zealand education system, I have been employed here and I have raised a family here. I have a shared understanding of sociocultural values and traditions, the taken-for-granted cultural patterns (Tett 2015:28) with some of the families involved in this research. For example many of the families wanted their child(ren) to attend a school where the child(ren) would be happy and be able to reach their potential. Obviously the context in which that happens is very different for children with DMD, but the underlying value is shared.

In some ways I was an insider studying "at home". I have an understanding and familiarity with New Zealand society and cultural norms. But in other ways I was clearly an outsider. One of the repeated messages of my interviews was that only people who live through DMD can really understand it.

> No one really knows what you have been through. Like, I ended up working with a woman whose son also had DMD and we only needed to look at each other to know. But no one else really knows what you are going through. No one understands unless you have been through these things. No one else can know and you can't expect them to. There is no way they can.
>
> (From an interview with a woman who was both a sister and a mother of men with DMD)

While my understanding of the lived experience of DMD is now much better than it was four years ago, I remain an outsider. Participation was determined by the families who live with DMD. Generally those families who agreed to be involved with this research were happy to be interviewed and to chat to me at Christmas parties, soccer matches, camps and other events. They had major commitments caring for themselves or their children as well their usual daily routines. It became clear that I could not have an intrusive role. Families did not want that and I was also uncomfortable with the idea of intruding inappropriately into family life. The people and families I spoke to understood that I was a university researcher studying

what it means to live with DMD in New Zealand. They may not have had much knowledge of what anthropologists do but were happy to talk to me. They answered my questions and when I explained that I was interested in collecting their stories and their experiences they seemed happy to provide these

Methods

"The assumption that anthropology has a valuable contribution to make is first of all rooted in the idea that the discipline's emphasis on long-term and close-up research makes it ideally suited to provide a grass-root perspective and to communicate the complexity of social phenomena. Because of its comparative approach, it is also well positioned to unearth various kinds of ethnocentrisms. Moreover, these elements contribute to an anti-establishment position that tends to critique power. These are some of the key elements that 'we' tend to value in anthropology, and which convince us of the underused but vital potential of our discipline" (Pelkmans 2013:400).

The research methods I have used have, as Pelkmans (2013) suggests, enabled me to; collect detailed, nuanced, grass roots perspectives, analyse repeated themes, ideas and data that challenge the taken-for-granted assumptions (or ethnocentrisms) of the dominant, able-bodied majority, identify complex social phenomenon and contextualise understandings.

Participant observation involved attending Muscular Dystrophy Association (MDA) regional branch camps and activities, volunteering at the MDA National Office. I also undertook 41 semi-structured interviews. Thirty one of these were with people who had DMD or were family members of people who had DMD and ten were with other people connected to the DMD community. In two cases children were present when I arrived to interview parents and, in these cases, I adapted the interviews to focus primarily on service provision. I was careful not to ask questions about end of life issues as I had no idea how much information those children had about their prognosis. In one case the child present suffered from the intellectual impairments that can accompany DMD and was unlikely to understand much of the interview content.

Table 1. Different types of interviews undertaken for this research project

Category of interview	Number of interviews
Adult men with DMD interviewed alone	6
Adult men with DMD and family present and contributing to the interview (includes one 17 year old)	6
Parents only of deceased men over 18 years	3
Parents only of children under 17 years	13
Parents with child present	2
Parents only of adult with intellectual impairment	1

Allied workers (various)	10

I focussed my participant observation on public events. I did not conduct participant observation inside family homes. When I planned my research I thought conducting participant observation in the family home would be a good way of understanding what it means to live with DMD. As the organiser of a busy household with two teenagers, a young child and a partner working long hours, I can fully appreciate why the presence of the researcher at the breakfast table would be awkward. Even a well-intentioned one who was happy to hang out the laundry or do the dishes would just be in the way. I would feel as if my parenting and house-keeping was being assessed and I anticipate that others would have similar reactions. However, as I learned more about DMD the main reason I decided not to pursue the idea of researching the private world of life with DMD was because it seemed like an additional form of objectification. As I entered the community of DMD I came to realise that all the equipment that comes with the later stages of the condition is quite alienating. People not involved with DMD find it hard to see beyond the wheelchair, the straps that hold the young men upright and the masks or sipper tubes that assist with breathing. These young men are surrounded by technology. While this equipment offers increased freedom and improves opportunities for leaving a confined home environment, it also alienates those not familiar with it. The discussions of Haraway (1991) regarding possibilities created by body-focussed technology and her use of the cyborg metaphor offers one way of examining this reliance on technology (see also Gibson (2015) for a critique of this discourse in relation to disability studies). However, here I focus on way these machines emphasise the marked category of disability. These young men also speak softly. This is associated with their muscle degeneration. In addition they are frequently accompanied by family or support workers. This means many of their transactions are conducted through an intermediary. I noticed a common experience of objectification and isolation. Young men with DMD already tend to be objectified in ways others are not. Repeatedly they told me that people talk to their support workers rather than to them, and ask about them as if they are not there. They do not like being seen as the person in the powerchair (which often happens). In one interview a young man told me he wanted to get a sign that said, "I can hear you!"

Having a high level of disability that requires a lot of equipment means these young men are in a 'marked category' (Brekhus 2008). To add to this burden of difference by constructing the young men as research objects within their own family home just felt wrong.

I combined my clerical voluntary work at National Office (see details below) with my reading in the field of Disability Studies to understand some of the macro level issues that face this particular community. My observations and conversations at social events helped to identify issues central to individuals and families and I clarified and sought elaboration on these issues in interviews. The interviews themselves revealed much that was important for this research. The reason that anthropologists engage in participant observation is so that they can observe what people do, as well as learn what people say they do. This research is perhaps biased towards what people say they do, rather than observation of what they actually do. I was able to observe in the public arena but less so in private homes. Most of my interviews were in private homes but these were formal visits, rather than observations of daily life.

As relationships develop over time I have become "friends" with members of the community on facebook and been invited to join or follow various Facebook groups relating to disability generally or Duchenne muscular dystrophy in particular. I have become friends with those who made requests to me. I did not make requests because I did not want to put those generously giving their stories to me in an awkward situation. Social media is recognised to be an area fraught with potential misunderstandings and conflict (Wang et al. 2011), I do not know if my posts have unwittingly alienated any of the people from the DMD community with whom I am friends. Facebook, including messaging, has however been a useful way to maintain and develop relationships with a few key members of the muscular dystrophy community. It has also allowed me to maintain an informal connection with others.

Much of my research resulted from my interactions with the Muscular Dystrophy Association. I discuss the significance of this organisation to my research. I then go on to discuss some of the insights I gained through attending social events and finally discuss the way the information that was collected through the interview process helped to clarify the areas I consider in this thesis.

Fieldwork Site 1: The MDA

The Muscular Dystrophy Association is a registered charity. It is governed by a National Council made up of elected MDA members and Branch representatives who act in a voluntary capacity. Maori representation on the National Council is not mandated by the organisation's constitution but it is increasingly important to the organisation and during my

fieldwork there was a noticeable shift with the participation of Maori members on the National Council, the employment of Maori staff and the development of relationships with Maori kaumatua and kuia (elders) who have mana (an indigenous concept akin to prestige and authority) to offer advice and support to the organisation. The National Council appoints the CEO and determines the strategic direction of the organisation. The CEO ensures that the strategic decisions made by the National Council are implemented. The four regional branches are independently registered as separate charities. The regional branches operate in a similar way to the national organisation, in that each is governed by elected representatives and solicits its own funding for maintaining the branch office, paying the salary of the office managers, the regionally based fieldwork service and other activities and events. There is a representative from each regional branch on the National Council. Although the national organisation and the regional branches are separate entities they work together.

In my initial proposal I acknowledged that the MDA would be part of my research field and the facilitator of it and that I would need to be, "respectful of the support and facilitation that they provide." This seemed like an appropriate comment to make and I imagined that working with the MDA would be reasonably straight forward. I had no idea how complex my relationships with the organisation and various staff members would become.

I volunteered from Sept 2012 until June 2014. I spent at least one day a week at National Office during this time (and sometimes more days depending on the tasks that needed completing). I was also employed in January 2015 for six months as a part-time grants writer while a review of how the organisation could optimise performance was conducted.

Prior to my research I had little experience of the Not for Profit (NFP) sector, or the tensions inherent in this sector. As I conducted my research and volunteered at the MDA office it quickly became apparent that the MDA was just one small cog in a weighty machine of charitable endeavour. One of the MDA's marketing managers (there was only one Marketing Manager at a time but there were several during the period of my research and work with the organisation) returned from a workshop and announced that, "New Zealand has the highest per capita ratio of charities." Data on the Fundraising Institute of New Zealand (FINZ) website suggests that New Zealand does not actually have the highest per capita ratio but is "significantly over serviced by Registered Charities by comparison to other jurisdictions" (Austin 2015). Despite its relatively small population, New Zealand supports a similar number and range of charities as other OECD countries. This means that the general public

are constantly being solicited to support 'good causes' and charities are encouraged to review and innovate their fund-raising ideas. FINZ is an integral part of this NFP fundraising industry which constantly targets charities with new fundraising strategies and ideas. These strategies are well researched and evidence based, often building on international innovations, but are also part of the corporatisation of the charitable sector. This corporatisation is characterised by a reliance on a managerial model (Klikauer 2013) in which generous salaries are paid to highly qualified managerial, marketing and accounting staff. The rationale is that this cost will be offset by the expertise these employees bring to the charity, raising the organisation's profile, marketing the services the charity offers, improving income generation, ensuring honesty in accounting and managing income and expenditure appropriately. This is an international approach to the provision of NFP services. It is clearly prevalent in the New Zealand charitable sector and the MDA was just one small player in this pervasive cultural milieu.

At the time of my research there was an increasingly competitive funding environment (Yang et al 2014, Webster 2015, Hall 2015). Staff at the National Office had to both solicit funds and provide services to members. Initially I was interested in the latter. I wanted to know what services the MDA provided to members. I imagined that this would give me some insights into the issues and concerns of people living with neuromuscular conditions. I soon learned that service provision was reliant on income generation and that a lot of time and effort was devoted to the generation of funds. In my first few weeks of volunteering I found myself counting collateral for the annual audit. I was counting information booklets and thousands and thousands of bowtie brooches that were given away during the annual appeal week. This week was known as Bowtie Week and was the main fundraising and awareness raising week. I often left the office wondering why I was doing all this counting and how on earth it was progressing my research. One of the things I was doing, as well as learning about the bureaucracy, economics and politics of charitable organisations from the 'other' side – the side that families do not see - was building relationships with some key National Office staff. It was through these relationships that I came to gain some incisive and valuable insights about what it meant to live with DMD. It was through the friendships that I built that I came understand some of the underlying issues facing families living with DMD.

Staffing at the National Office went through many changes during the course of my research. There were maternity leaves and other resignations and reappointments. The National Office has a small, permanent staff of about eight, although roles changed and amalgamated during

my research. During my three and half years of interactions with the National Office I have worked with 19 employees. At times there was a sense of transience in the workplace. However, there were two staff who were long-term employees, they were very familiar with the members and their conditions. Both became friends and developed an interest in my research. Each regional branch (with the exception of the small Southern branch) employed fieldworkers and an office manager. These staff were also familiar with the members and knowledgeable about the problems that families faced. Some also became friends and were invaluable as a source of information and as a way of checking my ideas and thoughts.

The Marketing and Membership Assistant and the Programme and Service Advisor at National Office were long-time staff members with comprehensive knowledge of the members and their conditions. Their knowledge about disability was based in different epistemologies but both have been fundamental to the development of some of the ideas in this thesis. The Marketing and Membership Assistant, Kerry Hills, was guarded and initially suspicious about my agenda in undertaking this research. The Programme and Service Advisor, Miriam Rodrigues, was positive and enthusiastic about my research from the start. Through my relationship with Kerry I learned about the social model of disability, whilst Miriam's expertise was in the alternative medical model of disability. I discuss the importance of these two key informants further in Section Two of this thesis.

The other staff at National Office were involved with finances; the Business Accountant (sometimes titled the Business Manager), Accounts Assistant, Marketing Manager and Grants Fundraiser. They were employed because they held different sorts of expertise, their backgrounds were not in the field of disability (either social or medical models) but rather in the accuracy of accounts or the strategies of marketing. The Accountant and Accounts Assistant were responsible for maintaining accurate records of the revenue; what came in, where from, and how it was spent. Accurate reliable systems of income and expenditure are essential for auditing processes, maintaining charitable status and the overall viability of the organisation. In order to work effectively these staff did not need to be familiar with neuromuscular conditions or develop relationships with the members. They needed to be accurate and methodical. However, those who I got to know well explained that they wanted to work in the NFP sector for philosophical reasons. They wanted to work for an organisation that made a difference to people's lives, rather than just made a profit. Someone who had previously worked in the building industry said he had been glad to be offered the position

with the MDA because he thought it would be more valuable and satisfying to work for a charity than for a commercial business.

The Marketing Manager and Grants Fundraiser did need to understand neuromuscular conditions in some detail and the Marketing Manager particularly needed good relationships with members as their stories were central to marketing strategies. The Grants Fundraiser needed an understanding in order to write grant applications that were convincing. Grants were solicited to address the specific needs of MDA members and in order for the applications to have a chance of success the applications had to explain why these needs were important. However the primary skills these staff needed was the ability to generate income. Several of the staff who filled the Marketing Manager role had come from corporate backgrounds. They were very much focussed on branding, raising awareness about the MDA, and assessing new and creative strategies for fundraising, attempting to balance the potential benefits of such strategies against their costs. Staff also had to cultivate and maintain positive relationships with their existing donors. This expertise fell within a marketing and managerial epistemology. It too can be regarded as an area of expertise.

My voluntary work at the National Office demonstrated the importance of both the social model of disability (as articulated by Kerry) and the biomedical model of disability (as represented by Miriam) for people with DMD. I also became familiar with a managerial epistemology that was embedded in the charitable sector. I became aware of the difficulties a fairly small charity faced in trying to negotiate a path through the demands and expectations of an increasingly professional NFP environment. The trusts that fund many charitable endeavours in New Zealand increasingly require evidence based information for applications to be successful. The Ministry of Health funds some of the MDAs information service and has similarly stringent reporting criteria. Trusts require charities to provide statistical evidence to support grant applications. This means that charities need to have databases that allow them to easily pull out the relevant statistical information. They need to assess their services. How effective are the services they offer? How do members rate these services? They need to be able to find how many members live in rural areas, or are classed as children or young people, or meet other specific criteria. All services need to be counted. How many welcome packs were sent out in a particular financial year, how many member queries did the Membership Manager and the fieldworkers respond to, how much mileage did each fieldworker undertake? In many regards charities operate like businesses and they need

robust systems in place. To operate like businesses they need appropriately qualified staff and these staff are paid at rates comparative to the commercial sector.

The staff needed different types of skills and knowledge to fulfil their roles. These different epistemologies or ways of knowing were underpinned by different experiences, values and ideas. Predominantly the MDA, as the national body, was focussed on providing expertise; understanding the latest neuromuscular research and its implications for members, training and oversight of the fieldworkers, maintaining links with biomedical specialists and international neuromuscular organisations, and providing information to people working with those who have neuromuscular conditions. The regional branches placed a greater focus on the immediate needs of members. They organised events such as camps, Christmas lunches, coffee mornings, parent support groups and the administration of the fieldwork service. The incompatibility between the biomedical knowledge that the National Office wanted to develop as its area of expertise and the everyday needs of the (often) marginalised members has been noticed by others:

> Furthermore, patients' associations' investments in the science of the present to hopefully promote future health can sometimes take place at the expense of meeting the current economic, social or educational needs of those presently affected by illness (Stockdale 1999; Stockdale and Terry, 2002). (Novas 2006)

However, this incompatibility was complicated by the pervasive corporatisation of the charitable sector. This was evidenced at the MDA National Office by a plethora of jobs undertaken by managers, Business Manager, Membership Manager, Marketing Manager, National Service Leader, Programme and Service Advisor, in addition to a Chief Executive Officer (six managerial positions in an organisation with just eight staff). There were also complicated lines of reporting and supervision. Some aspects of this cultural shift in the NFP sector are clearly appropriate. Transparent accounting processes, for example, are important as is the need for charities to generate sufficient income to provide quality services. At the time I was volunteering at the National Office the MDA seemed to have over-subscribed to the managerial approach. Much of this has changed with the arrival of a new CEO who is more familiar with the day-to-day struggles of members, and who streamlined the organisation and reduced the number of managers.

Fieldwork Site 2: Social Events of the Regional Branches

Attending social functions, camps and fundraising activities formed the basis of my participant observation. As I attended various social events – ice skating, powerchair soccer, mid-winter functions, Christmas parties and birthday parties, DMD day events – it was clear that the same families tended to be involved in these events. In the course of my interviews I met other families that chose not to participate. My participant observation was, therefore, not representative of all families with DMD. Mainly I attended those events organised by the Northern branch as this is the area in which I reside. The families that regularly attended were not only families who had a member with DMD; the social events were held for people with any neuromuscular condition. However, these social events did give me insights into some of the ways that life with DMD is distinctive although I note that I was observing special occasions rather than ordinary aspects of daily life. However through this participant observation I came to a better understanding of many of the issues I explore in this thesis; a do it now attitude, concerns about employment, health system funding, the emotional impact of diagnosis, the importance of a supportive group and the emotional as well as the physical challenges those with DMD face.

Some of these issues became apparent early on in my fieldwork as the first event I attended, a Christmas lunch organised by the Southland branch, demonstrates. I attended this function with a National Office staff member. We travelled to the function together, having been met at the airport by a DMD family. The mother, Rita, was very welcoming to me. She talked at length to the National Service Leader. This was a fairly new position and as a long time member of MDA and a National Council representative there was much Rita was keen to discuss. I noticed that the she talked a lot. She was very enthusiastic for the recently appointed National Service Leader to learn about the issues that faced families with neuromuscular conditions. Rita seemed very knowledgeable, I noticed. I soon came to learn that the literature that referred to expert patients (Epstein 1996, Rabeharisoa and Callon 2004, and Terry and Terry 2001) was describing many of the families I would meet. This was also my first time to meet a young man, Declan, with DMD. It was much more difficult for him to talk to us. This was due partly to the noise inside the van. His chair was locked into the back of the van. And the van was quite noisy. Declan was also softly spoken and as he had spinal fusion he was not able to turn his head to talk to us. That was something else I was to get used to, the need to position myself in front of people who had undergone this surgery as they were unable to turn their heads to address the person speaking to them. It was also important

to watch the face because as DMD progresses the muscles that facilitate speech weaken so the men cannot project their voice. For older men it is often quite hard to hear what they are saying. As the muscles that control the tongue weaken speech is not only quiet but also impaired as the tongue cannot be controlled easily. I realised later that the pattern of communication on that first journey with a DMD family was typical. The father was concentrating on the driving, the son sat quietly in his chair and the mother talked effusively about Duchenne muscular dystrophy and the local health system.

The local hospital was facing some dilemmas as they dealt with end of life care. Rita felt that in some cases the young men had really had enough and were ready to "let go" but often the families were not ready. She thought that in a couple of recent cases the families had wanted interventions that the young man had not really wanted. Perhaps partly as a result of this conversation, within a year the MDA had organised a workshop with international speakers Cynda Rushton, Professor at John Hopkins School of Nursing, and Gail Geller Professor at John Hopkins School of Medicine, to address living wills and palliative care. The workshop, however was in Auckland and not the rural South Island community where this conversation took place. The problems of the provision of health services for people living in rural areas are widely acknowledged and this is particularly salient for those with rare conditions (see, for example, Park, Scott, Benseman and Berry 1995) and research participants repeatedly mentioned this issue.

I came to realise that I could learn a lot from parents, they were often experts in the social barriers their sons faced, in the biomedical diagnosis and latest treatment of the condition and in the emotional issues their sons faced such as coming to terms with their isolation, the desire to lead a fulfilling life and leave a legacy. However it was clear that while often more accessible, theirs was a second–hand experience. This research does rely significantly on this second-hand experience. Often parents, especially mothers, talked to me effusively about living with muscular dystrophy. The young men themselves were often more reticent. While some of this may have been to do with DMD, in that these young men are often fairly socially isolated, I believe that it was also a reflection of wider New Zealand social norms. The work of McLaughlin et al (2008) contextualises the role of parents with a disabled child(ren) and the recurring findings they identify in relation to a British study were also characteristic of the parents I met (2008:12). DMD is a condition which can affect any family so the families I met were a diverse group and some were clearly affluent but others faced similar economic challenges to those mentioned by McLaughlin. The requirements of care

often compromised opportunities for employment. The disruption to education as a result of medical appointments was universal, parental stress and social exclusion were also characteristic.

The Christmas lunch was attended by MDA members with a range of conditions. The members lived in small towns and rural areas and travelled to the lakeside restaurant which was half way between two main centres. It was an overcast day and the lake was grey and choppy. The lunch was attended by eighteen predominantly middle-aged folk. They were Southland people, quietly conservative, meeting old friends for lunch. The conversation was that of long-term acquaintances with discussions about children and grandchildren and the well-being of absent members.

One mother talked to me about how impressed she was with Auckland's public transport. Her son had undergone major surgery at Auckland's Starship hospital and they had relied on public transport while in the city, presumably before the operation. Given the very serious nature of the surgery I was surprised that they used public transport. Her comments revealed the "get on with it" attitude that was characteristic of many of the families I encountered. This mother had travelled by herself with her son to a big city that they had not been to before and were unlikely to visit again any time soon. Despite his very serious health issues they decided to see as many of the sites as he was able to manage. She also mentioned her concerns about what her son would do when he finished school. He was a teenager and not keen to stay on until the end of year 13. The family lived in a rural area and she was concerned about the employment options especially given his disability. This too I came to learn was a major concern with all families – although it was a special concern for those living in rural places with fewer sedentary options. He said very little. He was the only teenager at this lunch and looked politely bored. His mother clearly valued the support of the group. This was another trend I noticed, also addressed by McLauglin et al (2008:118) who discuss in detail the way groups can either exclude or include disability and the benefits that some families gain from a sense of belonging and inclusion.

Some of the issues that were exposed at this Christmas function were clearly obvious at the time, concerns about employment and the role of palliative care for example. Others became apparent with hindsight as I engaged more and more with the community; the special role of parents, the need to adapt communication strategies to suit this particular group, the "get on with it attitude" in which people want to make the most of every good day and finally the

importance of a supportive group. As I note above, not all individuals or families with DMD choose to be involved with the activities organised by their MDA regional branch however, I did notice that most families had some kind of supportive network around them. For some this consisted of family, existing friendships and community networks. In very few cases I discovered situations where people were exceptionally isolated, they did not seem to have support and were marginalised in many ways. Participating in a supportive network seems to offer real benefits to people with DMD.

Fieldwork Site 3: Families with DMD

The interviews for this research took place between November 2012 and July 2014. During this period, service provisions were undergoing change. The constancy of service change was part, and often a destabilising part, of the experience of people with DMD.

I travelled all over New Zealand to conduct interviews, from Whangarei in Northland to Invercargill in Southland, from Hawkes Bay and Christchurch in the East to New Plymouth in the West. I conducted mainly face-to-face interviews but also did several phone interviews. I tried to organise my interviews in batches. For example, I travelled to Hawkes Bay, Wairarapa and Manawatu on one trip. I did several day trips to the Waikato and the King Country, to Whangarei and to the Coromandel. I had two trips to Dunedin and Southland and two to Christchurch and the Canterbury area (see Figure 1).

Figure 1. Map of New Zealand

Working with participants in their own homes gave me insights into daily routines. Insights from early interviews helped inform my later interactions, again indicating the advantages of the flexibility and sensitivity of anthropological methods.

The majority of interviews were conducted inside the family the home, however I met one family at a cafe when they travelled to their nearest hospital for a medical appointment. This was so I did not have travel such a long way to see them. Two interviews I conducted with allied professionals also took place in cafes. Usually the interviews took over two hours and as a general rule I ended the interview with a view to the hours I was going to spend transcribing. Once people started talking it was like a tap without a washer. The information just poured out; bad experiences and good, people who had been supportive and people who had been horrid, family dynamics, grief and loss and pride and joy. Each interview was very

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different. The experiences that people described and the personalities of the interviewees were all quite disparate.

My interview techniques improved during the course of this research as I became more comfortable discussing sensitive issues like dying and leaving a legacy. At first I had not really felt comfortable directly addressing these issues. However as I reviewed interview transcripts it became apparent that these were issues that a few participants wanted to talk about. I developed skills in asking open-ended questions. When participants talked about issues relating to death, for example if a brother had already died, I would ask, "How do you feel about that?" This approach left it up to the participant to decide how deeply they wanted to address their feelings or concerns.

Despite the differences between each interview, there were some commonalities. These commonalities are discussed in detail in other parts of this thesis, but briefly the most significant are; support networks, pain and discomfort as part of life, a focus on living and the overwhelming expressions of frustration with bureaucracies.

In each interview families and individuals living with DMD discussed those people who formed a part of their support network. I asked about this and in every case having a team of people that supported the family was important. These were organic networks formed of family, friends and others who demonstrated a level of compassion far beyond anything that their wages would justify.

Through my visits with young men living with DMD I came to understand more about the embodied nature of the condition. Although the young men tended to be fairly stoic about their conditions, it became apparent that they lived with a level of ongoing pain. Of all the difficulties individuals with DMD face, their personal experience of frequent pain was something I noticed a certain hesitancy to discuss.

Not many young men indicated that they wanted to discuss end of life issues. Most were far more interested in talking about issues relating to life than to death. Even Kerry, who knew that I was interested in the issue of a living will (the instructions that people leave about the interventions they want in case they are unable to communicate these themselves), did not discuss his with me. It was only as a result of his death that I learned he had written one. The fact that few of my interviewees raised the topic does not mean that it was not something they were thinking about. Rather it shows some of the cultural code that exists around death and

dying in New Zealand. I was reluctant to raise it in interviews in case it caused upset or distress. Some of the young men had a tendency to answer directly and succinctly the questions I asked and not to elaborate on their answers. So in these interviews I was not going to learn about anything I did not directly ask about. In three interviews however, the young men did show a willingness to discuss end of life issues. When I followed up their comments with open-ended questions I learned a lot about the acceptance they had that their lives would be short but they were all far more concerned with the living that they had left to do, than the inevitable dying.

Another over-riding commonality was the frustrations that families and individuals experience. Interviewees expressed frustrations with repeated experiences of systems that did not work for people with DMD. In some cases schools were sites of frustration in others it was the health system or the bureaucratic process through which support workers were allocated. Whilst the sites of the frustration varied, the experience of the needs of people with DMD falling outside established systems of support was common.

Chapter Conclusion

I discuss specific findings from my interviews in Parts Two and Three of this thesis. The general points I make here are about improving my techniques to facilitate discussion, and encouraging initially shy young men to elaborate on what was important to them. What I found was that these young men have a great passion to live fulfilling lives, despite the barriers and frustrations they experienced. It was a pleasure to record these interviews, both with young men and the parents of children, and to hear the stories they generously shared.

Together these three fieldwork sites provided me with a wealth of data, only some of which is included in this thesis. The data that is not explicitly referred to in this thesis was nevertheless invaluable, providing a comprehensive view of life lived with DMD. In the following chapter I consider another aspect of this project which is essential to understand the way life is lived with DMD in New Zealand. A knowledge of New Zealand's distinctive health and disability system is a pre-requisite for understanding the data analysis in Parts Two and Three.

Chapter 4: Rationales for New Zealand's Health and Disability Provisions

Funding: Ministry of Health and Accident Compensation Corporation

National and local health and disability systems are central to the Duchenne muscular dystrophy assemblage. Given the complexities of these I devote this chapter to explaining the systems as they are important actants in the creation of the local biology of DMD in New Zealand.

People whose impairments are caused by a medical condition (such as DMD) receive health and disability services funded by the Ministry of Health. People whose impairments are caused by an accident receive their services through both the state and private health care systems but via a different funding system, the Accident Compensation Corporation (ACC).These two systems result in markedly different levels of service provision for people living with similar types of disability. People whose disabilities are caused by a medical condition are disadvantaged by this system.

Examples of this inequality that were important to people living with DMD include the provision of a mobility vehicle, the number of hours support workers were paid to assist people with the activities of daily life (activities such as morning routines of getting up, showered, dressed, having breakfast, assistance with eating and using the bathroom), the provision of disability equipment and the modification of housing to accommodate disabilities. Mobility vehicles are routinely provided to people whose disabilities are caused by an accident but there is no provision for people whose disabilities result from medical conditions and whose services are reliant on Ministry of Health funding. During the course of my research I interviewed one family whose teenaged son stayed home during the winter. In summer months he could travel to some places on the bus, but his parents did not want him to get caught out by bad weather and risk catching a cold in the winter. In earlier times they had a van but it had failed its warrant of fitness and the tie-downs (that secure the heavy powerchair into the van) were no longer safe. The family were eligible for assistance to purchase a hoist (a lift to raise the chair into the van) but could not afford to buy a suitable van in which to install the hoist. Mobility vans are particularly expensive as they have special features such as a high roof so that the van can accommodate a person sitting in their powerchair and tie-downs to hold the chair securely. Those using powerchairs cannot sit in

regular vehicle seats, they are hoisted into the back of the vehicle and the chair is locked in place for the duration of the journey. The family had applied to a trust which was established to exclusively address this significant shortcoming in service provision. However, the trust was oversubscribed and their application was unsuccessful. This meant their son suffered social isolation and was not able to participate in the types of activities that other boys with DMD enjoyed. The inequitable funding of mobility vans was the issue that triggered a legal challenge to the two tier system, as mentioned below.

Another example of the inequitable nature of the two tier system is the issue of household modifications. One person specifically mentioned the number of times each impaired individual who required special modifications to the bathroom was eligible to have a bathroom installed (i.e. how often people were able to move house). People who are in receipt of ACC funding can move house and a new bathroom will be installed under the provisions of their ACC funding. People who receive Ministry of Health funding will receive just one modified bathroom in their lifetime. Clearly this limits the ability of those with severe disabilities who generally do not work to move house.

This inequality was widely commented on by many people involved in this research; families, regional branch staff, National Office staff, support workers themselves, clinicians and allied professionals. It was an underlying issue of concern to many. The two systems are firmly embedded in New Zealand social structures through legislation and health and accounting systems. A major challenge to this inequitable system had been mounted by multiple sclerosis sufferer, Melanie Trevethick, who went through the Human Rights Commission, the Human Rights Review Tribunal, the High Court and the Court of Appeal to challenge the inequalities inherent in the two systems. Repeatedly the courts found that the definition of disability in the Human Rights Act (section 21 (1) h) does not include cause of disability. Therefore the inequality in service provision between ACC and Ministry of Health systems does not contravene the Human Rights Act. The cause of a disability is not a prohibited ground of discrimination.

When I began my interactions with the DMD community in 2012 it seemed that Trevethick's passion and commitment and the support she had garnered from many disability support groups had peaked. The decision of the High Court in 2008 to dismiss her claim that the two system approach was "discrimination contrary to s.21 (1) h of the Human Rights Act, 1993" had deflated the momentum her case had created. The many people who commented on the

unfair situation that exists did so with a sense of resignation, it was another unfair blow to endure, another example of the way the lives of people who experience DMD are dismissed as unimportant and less worthy than other lives. Possibly the key issue to arise from the Trevethick versus Ministry of Health 2008 Court of Appeal decision was the last point of the Court's analysis that, "Mr Miller (Trevethick's lawyer) candidly accepts that what the applicant considers are the resulting anomalies are matters she wants to bring to the legislature's attention" (Court of Appeal 2008). The issue is political, not legal. The remedy lies with politicians, not the courts. The judiciary can only consider what the statutes say to determine if policies are lawful and in this case the statutes do allow the two separate systems to co-exist with unfair outcomes for disabled people based on the cause of their disability. The inequality that exists between the two systems results from the values and principles that underpin them. ACC was based on principles of community responsibility and Ministry of Health funding is based on a principle of fiscal responsibility.

Key to this particular analysis is the original Royal Commission of Inquiry that led to the report, *Compensation for Personal Injury in New Zealand,* 1967 (commonly referred to as the Woodhouse Report). The Commission of Inquiry, led by the chairperson Justice Owen Woodhouse, was briefed with the task of reviewing "the law relating to compensation and claims for damages for incapacity or death arising out of accidents (including diseases) suffered by persons in employment" (Woodhouse 1967:30). The Warrant setting up the commission specifically asked the commission to address accidents, not the wider health system. It was set up to examine the inadequacy of compensation benefits paid to workers (which was having a deleterious impact on the justice system as injured workers sued those they held responsible for their injuries). It was not focused on disabilities or impairments more generally. The report recommended a completely new 'no-fault' approach to compensation for personal injury. It recommended a scheme to provide 24-hour, no-fault insurance for all personal injury. The commission recommended that the scheme should cover injuries that occurred both in the workplace and outside the workplace and resulting from motor vehicle accidents. In return New Zealanders should give up the right to sue for damages arising from personal injury. The report recommended that the scheme be funded by three levies; one on motor vehicles and drivers, another on employers, and a third on the self-employed. This is clearly a distinctive model of insurance-style funding. It contrasts significantly with the way other disability services are funded – primarily through general taxation.

The report also recommended that the scheme be based on five basic principles. These principles offer a snapshot of an inclusive social era. They were community responsibility, comprehensive entitlement, complete rehabilitation, real compensation and administrative efficiency (Woodhouse 1967:39). These principles advocate that the scheme itself, its provisions, the way it actually works should value all citizens and their contributions to society. Despite the many changes that have occurred to ACC over the years, it was originally designed to be an inherently fair system. These ideas were reiterated 45 years later in a National Business Review interview with Sir Owen Woodhouse in which he notes,

> The social responsibilities which underpin ACC ought never to be tested by clever equations, or brushed to one side by economic dogma. In the end, they depend on decent fellow feeling and the ideas and ideals that support it. (Vaughan 2012)

Unlike ACC, the Ministry of Health does not have a set of clear principles upon which its services are based. There are various visionary documents, which advocate for the rights of people with disabilities, acknowledged within various statutory frames but these are not binding upon the implementation of services. Examples include the United Nations Convention on the Rights of People with Disabilities, The New Zealand Disability Strategy 2001, and the Disability Action Plan 2015. Rather services are balanced between human rights ideals and pragmatic economic decisions. This discrepancy was highlighted by Melanie Trevethick during her legal challenge to the inequality between the two systems.

> "There are obvious inequities between ACC and Ministry of Health funding that blatantly disregards our human rights," says Trevethick. "The New Zealand Disability Strategy objective 7.5 encourages equity of funding and service provision for people of similar needs regardless of cause of their impairment. All I'm asking is that the Government allows those of us disabled by illness to lead a dignified life, just as if we'd been disabled by accident". (Trevethick quoted by Carlin 2007)

Furthermore the Woodhouse report also states that logically the same approach based on the valuing all citizens should apply to those incapacitated by sickness and disease as well as those incapacitated by accident,

> It may be asked how incapacity arising from sickness and disease can be left aside. In logic there is no answer. A man overcome by ill health is no more able to work and no less afflicted than his neighbour hit by a car. In the industrial field certain diseases are included already. But logic on this occasion must give way to other considerations. First, it might be thought unwise to attempt one massive leap when two considered steps can be taken. Second, the urgent need is to co-ordinate the unrelated systems at present working in the injury field. Third, there is a virtual absence of the statistical signposting which alone can demonstrate the feasibility of the further move. And

Despite the Commission's original support for an equitable, inclusive system, the 2003 to 2008 campaign by Melanie Trevethick which garnered support from many in the disability sector including, eventually, the Human Rights Commission, support from Ruth Dyson when she was Minister for ACC in 2003, more recently from Andrew Little as opposition ACC spokesman and constitutional lawyer and former Prime Minister Geoffrey Palmer who both spoke at a 2012 ACC conference, the two systems remain in place. Those whose disabilities are caused by an accident receive a vastly superior service to those whose disabilities are caused by a medical condition.

When Melanie Trevethick's case, that argued that the Ministry of Health had acted in a discriminatory manner towards her (this was discrimination based on the cause of her disability), reached the Court of Appeal not only did the Court find that the definition of what was included as discrimination under the Human Rights Act, 1993, did not include the cause of a disability but furthermore even if the cause of a disability was included in the definition of discrimination in that Act, the inequitable system that Trevethick was challenging, would still be legal. The Court of Appeal noted that section 5 of the Bill Rights Act allows for discrimination if it can be reasonably justified in a free and democratic society. In other words the judges were saying that the two tier system which in which one group of citizens receives a comprehensive package of services and another group of citizens receives a vastly reduced, and much harder to access, package, an arrangement which is seen as incredibly unfair by people living with DMD, is reasonable and can be justified. The Court of Appeal did not elaborate in their analysis as to why this is a reasonable and justifiable situation so I can only surmise that the Court was acknowledging the right of parliament in a democratic regime to determine tax rates and the allocation of budgets. It seems that the judges were acknowledging that such a major review of the provision of service needed to be mandated by policies signposted by political parties giving citizens the opportunity to support or oppose what would be a major and costly change to the current system.

It is little wonder, then, that there was a general sense of resentful resignation to the inequitable system at the time of my research. Melanie Trevethick had taken her case as far as she could, the Court of Appeal turned down her request to appeal the decision made by the High Court because the system which she (and others with disabilities caused by medical conditions) experienced as discriminatory was not discriminatory according to the statutes, it

was not legally discriminatory, and even it had been legally discriminatory this would still have been acceptable. While the judges who reviewed the case alluded to the complexities of a free and democratic society, those living with DMD are left with the sense that their lives are marginal and considered unimportant by the justice system.

History of Ministry of Health Funding for Disability Services

In 1972, when the ACC Act was passed there was bipartisan support for a fair and inclusive system – support for the scheme from both the Labour and National parties. The Act was based on the value that it was right to acknowledge "decent fellow feeling" over and above "economic dogma and clever equations" (Woodhouse quoted in Vaughan 2012). The provisions of ACC were available "to all citizens" (Woodhouse 1967:39). The Woodhouse report and the ACC Act, 1972, reflect a very different social milieu to the current era. The idea of community responsibility was widely accepted. The bipartisan support for this policy and for other inclusive policies introduced at that time reflect social values different to the values of individual and fiscal responsibility that currently dominate political discourse. The inclusive values of the 1960s and early 1970s are part of a larger historical trajectory. A key element of this trajectory includes the 1938 Social Security Act, passed by the Labour party of Sir Michael Joseph Savage. Savage is widely credited as the architect of the welfare state in New Zealand.

The Social Security Act, 1938, was based on the principle that every New Zealand citizen had a right to a reasonable standard of living. It introduced free hospitals and a comprehensive array of welfare benefits for families, the elderly, invalids and the unemployed. This targeted provision for social assistance has remained the dominant model of social security in New Zealand. The principle that every citizen had the right to a reasonable standard of living and that the state should provide assistance by targeting it towards those who did not have a reasonable standard of living remained current until the early 1970's (McCarthy 1972). The 1950's, 1960's and early 1970's were prosperous times in New Zealand with hardly any unemployment. However the economic challenges of the 1970's and 1980's saw unemployment rise and budgets for the provision of state welfare stretched. These challenges include the oil shocks of 1973 and 1978-9 (New Zealand relied on imported oil and price increases had a negative impact on the economy). The United Kingdom's decision to enter the European Economic Community in 1973 saw a reduction of

exports to New Zealand's previously major trading partner. By 1976 New Zealand was in recession. Unemployment and the cost of living increased.

Welfare ideology also underwent change both in New Zealand and overseas. The controversial policies of President Reagan of the United States of America and Prime Minister Margaret Thatcher of Britain were radical implementations of classical liberal economics, characterised by policies to; limit the role of the state, promote privatisation, create a low-tax government and advocate for family autonomy (Centre for Policy Studies (CPS) website 2014).

Margaret Thatcher became the leader of the UK Conservative Party in 1975 and the Prime Minister in 1979. Reagan who also supported political philosophies of a free market, unencumbered by state regulations, was elected into office a year later in 1980. Similar free market policies were introduced to New Zealand, in 1984, by the fourth Labour government of David Lange under the auspices of Roger Douglas, Minister of Finance. Known colloquially as Rogernomics, the policies of Douglas aimed to address the economic problems facing New Zealand. These included; a deteriorating economic performance, falling standard of living, comparatively high level of inflation. Douglas believed that the protectionist economic policies and subsidies supported by governments of both the left and right since the 1930 needed radical review. Like Thatcher and Reagan he agreed with the neo liberal economic philosophies of Friedrich Hayek and Milton Friedman and introduced policies to deregulate the marketplace, remove subsidies, decrease income tax and introduce a goods and services tax (GST). He operated a tight monetary policy to manage New Zealand's high rates of inflation, which included devaluing the dollar by 20% (Aimer 2012). Douglas railroaded his economic policies through in one political term (three years). His policies were challenging and controversial both for ordinary citizens who were subjected to sudden change and to some of his Labour party colleagues (especially those on the left). However the policies themselves were not reversed by subsequent National or Labour governments. The policies of the next Minister of Finance, National member Ruth Richardson, continued to embed neoliberal ideology in New Zealand's political landscape, particularly in the provision of state welfare. Since 1984 New Zealand has seen a continuation of neoliberal governance including the sale of state assets, and reforms to state-provided services. One key aspect of this new neoliberal political rationale was the importance of competition in the market place. Monopolies were seen as suffocating innovation (Centre of Policy Studies website 2016). These newly dominant economic policies relied on consumers having choice. These ideas

were applied to all types of services including health services. Seldon's 'Point of View' article in the 1967 Lancet was an early neoliberal discussion of the ways in which competition and diversification could be facilitated in Britain's National Health System. Arthur Seldon was an influential and early contributor to the neoliberal ideas that influenced the politics of many Western democracies from the late 1970's to the present. He co-founded the Institute of Economic Affairs (the IEA) which challenged the increasing power of the government long before it was popular to do so.

> The political fiction that medical care is 'free' has been a barrier to humanity, choice and expansion. In the 1970s and 1980s … consumers ... will be ready to pay more for diversified, responsive, personal health services than for a National Health Service. It is from the millions spent on everyday consumption that the additional money for medical care must come. Doctors will attract it when they prepare themselves to be paid not by the taxpayer via the politician but by the man or woman they serve, the patient (Seldon 1967:677).

Although there are distinctive differences between Britain's National Health System and the health system in New Zealand, notably in New Zealand patients pay a part charge to visit their doctor, the newly popular libertarian economics were applied in similar ways to both systems. Neoliberal rationales were, and continue to be, promoted by organisations such as the IEA and CPS in Britain and by organisations such as the Maxim Institute and the New Zealand Initiative (formerly the New Zealand Business Roundtable and the New Zealand Institute) (Smellie 2012) here in New Zealand. The bureaucratic difficulties encountered by New Zealand's DMD community today have their roots in these historic changes to the underlying drivers of political decision making. The welfare policies of the 1930s to 1970s were based on the socialist hypothesis that the government needed to directly ensure that citizens had a reasonable standard of living whilst the free market policies from the 1970s are based on the liberal economic hypothesis that a free market place would better ensure people's standard of living.

This hypothesis has been strongly challenged and subject to enormous debate (for example, Harvey 2005, Larner 2000 and Leitner et al 2014). Larner (2000) argues that a simplistic dualism that presents social welfare on the one hand and neoliberalism on the other fails to acknowledge and therefore address the complexities of neoliberalism and its hegemonic capture of social values. This is a valid critique and by noting this shift in the ideologies underpinning the work of New Zealand's governments during the twentieth century I am setting a scene rather than ignoring the complexities of such a shift. As Larner notes, "Only

by theorizing neo-liberalism as a multi-vocal and contradictory phenomenon can we make visible the contestations and struggles that we are currently engaged in" (2000:21). In Section Three I unpick and explore the contestations and struggles that result from the implementation of policies driven by neoliberal understandings. I examine the resistance to aspects of policies and examine how these resistances can be reformulated within such policies.

Path Dependency

Economic theories of how to improve society were implemented in a pragmatic fashion, adapting to systems that were already in place. Laugesen and Gauld (2012) discuss how this tends to happen in practice. They note the relevance of path dependence theory and 'incrementalisim' in the history of the New Zealand health system. Path dependence is "the theory that policy change is influenced by pre-existing policy arrangements that are deeply embedded (Pierson 2002, Wilsford 1994)." These "deeply embedded" arrangements in the health system involve long-standing institutions with specific histories, with their own power dynamics; existing conflicts and/or allegiances between professional groups, services, management and optimal service provision.

Path dependency means that policy changes are made where change is likely to be politically achievable (goals will be acceptable to the range of institutions and interests) and technically achievable (changes will not completely disrupt service continuity). These may include small changes at the margins of the system, or the creation of new national arrangements to promote improvements in quality and patient safety (Laugensen and Gauld 2012:11-12).

So added to the philosophical changes about how and the state can best improve the standard of living for citizens and provide services, the current health system, with its heavy reliance on meeting criteria, has been constructed in a pragmatic fashion where the changes most acceptable to existing 'power holders' are implemented ahead of more challenging changes. The tinkering-at-the-edges approach alluded to by Laugensen and Gauld and others (Immergut 1992, Lindblom 1959) is perhaps a partial explanation for the constant changes to the disability services experienced by Duchenne families (piloting programmes such as 'Enabling Good Lives and Choices in Community Living' part of 'The New Model' discussed further in Chapter Nine). Those with the least power in these embedded arrangements and institutions are likely to be patients with rare disorders. Policies which

address the multiple layers of bureaucracy that families have to meet are therefore unlikely to be prioritised in either the incremental 'tinkering' changes or the structural changes that Laugensen and Gauld refer to.

Health System Reforms

Criteria based assessments which are often used to deny services to people with DMD have resulted from four major structural transformations of the health system since 1983. As these reforms have involved lengthy processes of consultation and reporting, taskforce review, Royal Commissions and ministerial reports, the health system has been under almost constant review and reform for the past 30 years. Reforms that cover such a long period of time are complex and do not have a single motivating factor. While the acceptance of neoliberal governance has been significant there has been a smorgasbord of other overlapping issues that have shaped the reforms (see Quin 2009, Laugensen and Gauld 2012, Upton 1991). Throughout the reforms improving efficiency, equity and health outcomes have been the overriding concerns. Two structural changes have been particularly important. Firstly, the introduction of population based funding (PBF) in 1983. PBF meant that the health system has had,

> to live within annual central government allocations as determined by a Population-based funding formula (PBF). This provides a dollar amount per person per region, weighted for various characteristics, including ethnicity, socio-economic status … and the composition of urban and rural based population". (Laugesen and Gauld 2012:151)

Secondly, in 1993 the changes, recommended in the reviews of 1980s and early 1990s, to separate the purchase of health care from the provision of service were implemented. This followed a 1991 a Ministerial Taskforce, chaired by Roderick Carr, which resulted in the paper, *Your Health and Public Health* (Upton, 1991) that relied on the 1988 Gibbs report *Unshackling the Hospitals* (Gibbs, Fraser, and Scott 1988) and the *1986 Choices for Health Care: Report of the Health Benefits Review* (Scott, 1986).

Population based funding (also called capitation) replaced an earlier funding model that, "determined funding on the basis of prior allocations and expenditure and total populations served" (reimbursement) (Gauld 2009:34). This earlier model was widely acknowledged (Smith 1981) to be inequitable as small hospital boards in predominantly rural areas with static or declining populations received an unfairly large portion of the health pie. The justification for PBFs was to improve equity in health care. The shift from a reimbursement

model of funding to a PBF model contributed to the current system of health funding now characterised by fixed budgets.

The purchaser provider split was another important contributor to the fixed budget approach. Initially this split was driven with a market model in mind but following the election of the 1996 coalition government this was changed so that health provision was to be "business like" rather than "for profit" (Quin 2009:14). This purchaser provider split is significant. Gibbs et al. (1988:20) found basic accounting practices in hospital management were woeful.

> The New Zealand hospital system is characterised by very poor management information. Management and costing systems are almost non-existent. … Simple operating statistics are difficult to obtain and are often inconsistent and out of date.

These findings, of which the taskforce said, "We find this absolutely extraordinary. No enterprise should be conducted this way" (1988:20), along with other equally surprising findings such as the point that,

> … none of the people who make decisions are aware of the cost of the resources they use or misuse. Therefore they cannot take the costs into account when considering different courses of action. … Many doctors believe they alone should determine what procedures are undertaken, when and where, with no regard for budgetary approval or total cost (1988:21).

This led to the recommendations by Gibbs et al. that the structure of health care (and hospital funding in particular) should be dramatically changed. Separating the purchase of health care from its provision would result in clear and transparent payment for measurable outputs. For the new system to work, accounting and statistical information would need to be collected and used. The new system would be based on contracts agreed between the purchasing agent and the service provider. Service providers would need to be very clear about how much their services cost so that they could negotiate reasonable and realistic prices with the purchaser. This in turn would enhance transparency in the way the health dollar was spent and ensure efficiency.

This move institutionalised the notion that greater accountability and transparency would facilitate efficiency (Laugesen and Gauld 2012:104). It involved the use of contracts to specify the relationship between purchasers (at the time these were Regional Health Authorities, RHAs) and service providers (Gibbs et al. 1988:30). While many aspects of the health reforms advocated by the 1991 taskforce were not implemented as envisioned, or were implemented but later adapted, this fundamental change to the funding of health services has

remained. This underlying idea that it was better to separate the purchaser of services from the provider was an idea that was modelled on liberal economic theory. Other anticipated reforms that would have seen a more consolidated move towards the free market were not eventually implemented (for example risk-adjusted health insurance, a shift to private hospitals and voucher systems to increase consumer choice, Laugensen and Gauld 2012: 103). However the idea that contracts facilitate clarity, accountability and efficiency have been accepted as "common-sense" or what Bourdieu would refer to as "doxa" (Bourdieu, 1977) and are now embedded in the structure of the health system.

The relationship between the purchaser and service providers are legally clarified via contractual arrangements. Providers have to demonstrate that they are fulfilling the expectations and requirements of the purchaser. In order to do this, service providers must ensure that their services are fulfilling their contractual obligations. The services they provide are based on auditable assessment criteria. The assessment criteria that service providers rely on are based service specifications produced by the Ministry of Health. These documents are used to assess eligibility for services, but eligibility is not enough to guarantee access (New Zealand Treasury 2012). Access is also limited by fixed budgets. Fixed budgets were initiated with the shift to PBFs in 1983 and have been extended throughout the health system, supported by the move to new public management systems (rather than funding being based on clinical decision-making and reimbursement) and policies which focus on rationalising costs. This means the provider who must operate within a fixed budget becomes a gatekeeper to service provision. Service providers must use their discretion to decide how much of their fixed budget it is reasonable to spend on any one eligible person. Just because a person is eligible to be assessed does not mean that their needs will be fully met. The health reforms of the past thirty years, in which neoliberal economic philosophy combines with ideas about equity and quality of service, conforms to Foucault's description of the strategies that governments of complex capitalist states rely on to govern populations. The assessment and eligibility criteria that characterises the Ministry of Health funded system (and some of the recent changes in ACC) is an example of the increasing surveillance and monitoring, or the "meticulous attention" (Foucault 1991:92) that the government relies on to provides services. Chapter Nine discusses the problems inherent in this model for those with high levels of need such as people with DMD.

Chapter Conclusion

The current system is inequitable. The two tier provision of services to people with disabilities offers a significantly better quality of life to people whose disabilities are caused by an accident. Those whose disabilities are caused by a medical condition feel discriminated against. I have examined the history of these two systems and the underlying values that inform the current situation. The history I outline in no way justifies or diminishes the inequality that exists in New Zealand. I have included this history because it contributes to a distinctive local biology (Lock and Nguyen, 2010) of DMD in New Zealand which is referred to in Parts two and three of this thesis. Some disabled people receive services that are based on the principle that the community has a responsibility to sustain citizens experiencing physical incapacity. This responsibility exists regardless of the cause of the injury and includes an acknowledgement that bodily impairment is a loss in itself. Other disabled people receive services based on the principles of liberal economic theory that contracts facilitate clarity, efficiency and accountability. The first set of principles are based on ideas of social responsibility the second on ideas of economic responsibility. These different underlying principles result in inequitable outcomes. They are important indicators of the way lives lived with DMD in New Zealand are valued.

Having now addressed in this first section of my thesis introductory information, the research proposition, essential material about DMD, my research methods and some background information about the structure of New Zealand's health and disability system, I now synthesise this information with the results of my fieldwork to address the messages that this anomalous biosocial group receive about the way their lives are valued within the New Zealand society.

SECTION 2

VALUING LIFE: MEANING, FULFILMENT, PURPOSE AND THE POLITICS OF LIFE.

Section 2 Introduction

On a crisp, sunny, autumn day, I drove in a rental car around the rural roads of the South Island. Living in Auckland it is easy to forget just how stunning the scenery in other parts of New Zealand is. On this beautiful morning the roadside trees still had their autumn colours, there was little traffic and the distant mountain peaks created an idyllic background. I stopped for morning tea in a quirky café that sold organic muffins, fair trade coffee and the works of local artists. I was lulled into a sense of relaxed enjoyment, a liminal traveller. I was on my way to interview the parents of a ten year old boy with DMD. I drove along a road that ran parallel to a river, poplars and willows grew along its banks and dairy cattle grazed in the paddocks. It felt like a quintessential New Zealand journey: a pleasant drive on a magnificent morning. Perhaps it is etched in my memory because it contrasted so dramatically with the emotional turmoil, the grief and anger, which I encountered at the family home. Although the family had had their diagnosis for four years, our discussion about that process triggered some raw memories. The father, Richard, presented as a typical "Kiwi bloke". He worked outdoors, he wore shorts and gumboots, he talked about rugby, and his language was forthright and colourful. The couple were successful business owners and much of their success was based on determination, hard work and physical labour. Although forthright and outspoken, Richard cried when recalling his son's diagnosis. Not only was the diagnosis itself a shock (the couple were not aware of any family history and had little knowledge of muscular dystrophy) but they felt "abandoned" by service providers.

Laura   I remember when it got confirmed that he had muscular dystrophy everyone left, deserted us. We had no support whatsoever, everyone left

Kate   Who left?

Laura   Everyone. It was like, he's going to die. We can't help them. They were gone. … I would hate people to go through the process the way we went through the process. Because we felt abandoned. We did. We felt that because there was no cure for our son we were abandoned.

Apart from a neurologist whom they saw once a year, Richard and Laura felt there was no one that knew about DMD to help them. This included their GP, their first physiotherapist, the social worker who contacted them and even their fieldworker.

Richard   I know it is a small population here and not many people have muscular
             dystrophy and even fewer have DMD but you'd think there would be a few more
             people that have a few more clues.

Their son, Zane, had not met his milestones and had raised levels in a CK test. This is a test
which measures the amount of creatine phosphokinase (CPK or CK) present in the blood.
When tissue is damaged, creatine phosphokinase enzyme leaks from tissue into the blood. It
can indicate several conditions including muscular dystrophy. The test is not diagnostic; a
follow up muscle biopsy confirms the diagnosis. The family had to travel to a large hospital
in a different DHB for the muscle biopsy as their local hospital was not able to manage the
additional anaesthetic risks that accompany DMD.

Richard   We had to take him to (large hospital) because they wouldn't put him under here
             for the muscle biopsy. Which doesn't give you a lot of confidence for the health
             system here, when they say that the anaesthetists aren't good enough to do it.

At their first social work appointment, when they were expecting to hear some advice about
what they could do, they were clearly shocked that all the social worker had to offer was a
form to fill out to collect $50.00 a week disability allowance. "As if $50.00 a week is going
to make it all better." Repeatedly service providers would ask them how they were feeling but
no one was able to suggest concrete constructive steps they could take to assist their son. It
was not until a physiotherapist with knowledge and expertise in DMD was employed by their
DHB some years after their diagnosis that they felt supported.

Laura   For the first years after Zane got diagnosed we'd go in every 6 months. She
             (physiotherapist) would do tests on him and it would be, "See you in another 6
             months." So it was not about doing any physiotherapy. It was about recording,
             ticking boxes. "I've seen him, good as gold, OK Bye." But with Wendy we do six
             weeks blocks of physio, two weeks of hydro. It is all on.

Richard   So when we met Wendy (the knowledgeable physiotherapist) she gave us *hope*
             and that is what we were looking for.

Recalling the terrible situation when they had the diagnosis but did not know how to help
their son caused Richard's grief to resurface. It was the absence of hope, the powerlessness
and despair that was so distressing.

In this second section of my thesis I address some of the ways in which a life lived with DMD is distinctive. I highlight the importance of activities that imbue life with meaning, fulfilment and purpose – that give hope as illustrated in the vignette above. I also consider the grief and suffering that accompanies DMD. I examine different ways in which life is valued and understood, and following Fassin (2012), I highlight social and political ways that hope is enacted in daily lives.

The concept of social suffering was defined in 1997 by Arthur Kleinman, Veena Das and Margaret Lock. Importantly they challenged the idea that suffering was primarily an individual and medical issue noting that "Social suffering results from what political, economic, and institutional power does to people and, reciprocally, from how these forms of power influence responses to social problems" (Kleinman, Das and Lock 1997:ix).

DMD is a health condition which like the other examples of social suffering referred to by Kleinman, Das and Lock (1997) is also a political and cultural matter. Using the concept of social suffering to analyse DMD highlights that DMD is not simply a medical condition but also a political and cultural matter. While DMD may seem more routine, affecting only a very few people, compared to the atrocities of the case studies included in *Social Suffering*, it is nevertheless an example of social suffering. While I certainly situate my work within an anthropology of suffering I also analyse my findings in relation to the developing areas of anthropological interest; value, morality, imagination, well-being, care, hope and change (Robbins 2013:457). Suffering remains central, but, following an anthropological trend discussed by Robbins (2013), I also seek to examine the ways people work on themselves and their social relations to improve their situations. My task here then is to explore the ways people imagine good lives and enact their hopes.

Mattingly (2014:9) describes the same phenomena in the longitudinal research she has conducted with African American families who have chronically ill children. She describes one of the mothers involved in her research as having the aim of creating a good life for her son and considers this desire in broad moral terms "The good life for humans is not merely about surviving but concerns flourishing … leading a 'life worth living' or a 'good life'".

She goes on to explain the value of anthropology in analysing such moral issues (which are traditionally associated with philosophical rather than anthropological discourses),

An ethnographic focus on how people attempt to realize lives they consider morally worthy, even in the most blighted and unpromising circumstances, has something to contribute to anthropology. It calls attention to the way people consider and evaluate their lives in light of notions of what is ethically good or right… the cultural point is that moral striving seems to matter a great deal to people in all sorts of societies. What constitutes the good life may vary widely from society to society, but it is difficult to imagine any community where this does not matter. (Mattingly 2014: 10-11)

My discussions (in Chapter Five) about hope and the hopeful activities that those connected to the DMD community engage in mirror some of Mattingly's findings and analysis. Participating in hopeful activities that create "good" or purposeful lives are central considerations for individuals and families with DMD. Determining what constitutes a good life varies for different people and different families but the process of "becoming", of "cultivating virtue" (Mattingly 2014: 10), of developing a moral way of being, is a central consideration for this community.

This broader focus on living a moral life, a good or purposeful life, signals another important shift in medical anthropology, rather than a narrower focus on health (Fassin 2012). One way this is done is by considering the political "worth of lives", exploring the often unconscious, cultural values that inform decisions of government (e.g., Scheper-Hughes 1993). The second way life can be considered is through the individual's right to live. Fassin refers, for example, to the rights of the foetus (2012:111), the dying patient in need of expensive treatment (2012:110) and the increase in the number of migrants accepted by France on the basis of requiring life-saving medical treatments (in contrast to a reduction in the number of refugees granted asylum) (2012:112). He calls this a consideration of the "value of life", a biological understanding of life. In unpicking the distinction between these two ways of understanding life Fassin suggests there has been a "profound change in the recognition of the value of life, which has shifted from the political to the biological" (2012:112). This distinction between political understandings of the worth of lives and biological understandings of the value of life has been very helpful in allowing me to understand some of the cultural issues that frame the stories people shared. Both are significant considerations in this thesis. My research demonstrates that people living with DMD often face socially constructed obstacles that indicate that the political worth of their lives is devalued. However, simultaneously, there are also developments within a wider engaged or concerned community that support Fassin's analysis of a biological valuing of life. His point that this biological way of valuing life is taking increasing precedence in social understandings of disability is reflected in the stories people shared with me.

Chapter Five focuses on the ways individuals and families living with DMD imagine life and create and use hope to infuse life with meaning. The community's hopes for the future show how they value life. I review the concept of hope in the literature and in the context of both the social and medical models of disability. Here the social model is understood as a model of disability which prioritises participation in society. It is participation in ordinary activities which offers boys and young men an immediate experience of hope. The medical model is a useful contrast for considering the hope that exists in the research area. As mentioned in Chapter One people within the DMD community value medical research. It is through this research that people hold out hope for a future free of the muscle degeneration that characterises the condition.

Chapter Six acknowledges that suffering is an inevitable part of the condition. This suffering is both physical and emotional. Using information from interviews and from observations, I examine the impact of; diagnosis, boys losing the ability to walk, how DMD can affect families, the experience of physical pain and finally I discuss the repeated experience of death that this community endures. The way in which the community responds to the pain and suffering is socially constructed. I analyse what this case study reveals about the way 'New Zealand society' values life and quality of life.

In the final chapter in this section I examine participants' approaches to reproductive technologies. In discussing whether or not to have children and how to manage reproduction, men and women with DMD in their family reveal different and deeply felt attitudes and values towards life and the quality of life. Following Fassin (2012:115) these chapters allow me to contest the untested, and sometimes unconscious, assumptions that surround those who live with DMD and how these assumptions influence the ethics of government.

I examine suffering as an embodied and social reality but go beyond this and describe the ways members of the DMD community imagine ways their lives could be better and the steps people take to achieve the improvements they imagine. In examining the different ways hope is enacted by this particular group of people I intend to do justice to the ways people with DMD live for the good (Robbins 2013:459) and anticipate that these efforts contribute to a growing body of work about hope within anthropology (see Kleist and Jansen, 2016, for a special issue of the journal History and Anthropology which addressed hope).

Chapter 5: Hope

As I began researching DMD the comprehensive Bushby et al (2010a and 2010b) guides had recently been published and they were welcomed by many in the DMD community for the surety they provided. They clarified the current best practice for the treatment of DMD. These key documents also formed part of a wider, dominant discourse focussing on the biomedical aspects of the condition. Anthropological literature also highlighted the importance of the biomedical model for people with the condition. Rabeharisoa and Callon (2008) show that for those with DMD the widespread 'social versus medical' model of disability was not a useful dichotomy. For this particular patient group the two models intertwine. Yet the focus of most peer reviewed literature tended to be the Duchenne body and the way in which the Duchenne body is different to other bodies. I soon realised that there was another equally important focus for the DMD community that was largely overlooked in the literature. Repeatedly what I was hearing and seeing was not a distinct focus on the body, the focus was on getting on with activities and daily life. This often included a consideration of the body but the focus was on an activity or task. Issues that were raised during fieldwork by people with DMD or family members were about the activities in which people engage, or want to engage, and the adaptations they had to organise in order to participate in those activities. They had ideas about the things they wanted to do and positive attitudes towards making those things happen. They tended to get on and do things now, as tomorrow and "later" are tentative, no one can predict how someone with DMD will be in the future. The individuals and families involved in this research wanted to take advantage of the good days to participate in activities. The flip side of this was that they found bureaucratic barriers that prevented them fulfilling their aspirations to be phenomenally frustrating and this is discussed further in Section Three of this thesis. Here I deal with various manifestations of hope. I address the immediate hopes of those with DMD and their families, the hopes of clinicians and researchers and the way hope is incorporated at a structural level through interactions between DHBs, crown entities and pharmaceutical companies. This chapter is about different expressions of hope that all relate to New Zealand's DMD community.

Anthropology of Hope

As anthropology is a discipline which studies the cultures, the values, the beliefs and behaviours, of people all over the world there could hardly be a better discipline to explore

hope. However, notable in the recent anthropological writing on hope is the view that anthropologists have historically tended to overlook hope in their analyses of culture (Appadurai 2004, Crapanzano 2003 and 2004, Hage 2003, Jansen 2015, Mattingly 2010).

Crapanzano (2004) and Appadurai (2004) offer different explanations for the apparent lack of hope in anthropological discourses but both go on to advocate for the benefits of the inclusion of hope in anthropological analysis. Appadurai advocates for the study of hope as way of examining how "people actually navigate their social spaces" (2004:84). By focussing on hope Crapanzano says anthropologists will open their horizons and maintain anthropology as a moral science that "explores the ways in which people regulate and evaluate their association with one another at both communal and intimate levels of life" (2004:4).

I concur with Jansen (2015) that my analysis is not to do with hope as method or a new field of anthropological knowledge but rather I am concerned with "the hopes embodied with the evocations of 'normal lives'"(2015:46). Amongst the participants in this research there was a widespread desire to take part in the normal activities of life. This was also a finding of the research of Good et al. (1990), Hage (2003) and Mattingly (2010) who write about the role of hope within ordinary, daily activities. Good et al. (1990:72) for example, demonstrated that hope was integral to the lived experience of American oncologists. They asked fifty one oncologists in Harvard teaching hospitals how they valued hope, and if and how hope was incorporated into their clinical practice. The oncologists all valued hope, not so much in relation to extending life, but more so in improving patients' wellbeing. The interviews reflect the complexity of the meaning of hope to these oncologists: hope that the patient enjoy the richness and colour of daily experience, hope to control pain, hope to be secure in the physician-patient relationship, are the minimum the physician can offer. Hope to affect the quality of life, perhaps to influence the magnitude of side effects, to give treatment options their best chance is the most that might be offered.

The idea that hope has a wider role to play in patient well-being than simply in relation to longevity is central to my analysis. Participants engaged in positive activities for a fulfilling, meaningful life. The discussions in this chapter demonstrate the importance of optimism and positivity for families and those who work with them. Hope for families with DMD has an intrinsic value for daily life – not just in terms of biomedical outcomes. Good et al's (1990) research is useful in emphasising the "complexity of the meaning of hope." They identify one particular cultural belief that relates to hope – the importance of individual will, a positive

attitude, or as Good et al. describe it, "popular notions of psyche-soma" (1990; 76). Mattingly (2010) explores other expressions of hope. Her argument is that for marginalised communities hope is as much about building community and mutual support as it is about individual success. Hope is associated with overcoming racial and economic divides, recognising common humanity and struggling for common good (2010:17).

> An African American vision of hope is not an easy commitment to "blind optimism" but an arduous struggle against fearsome obstacles has become part of a national story, though one often hidden from public sight. (2010:15)

These anthropological accounts offered a key to analysing my own findings. The point that hope can be enacted both individually and collectively, and that hope is not some distant aspiration but a constant and persistent driver or shaper of how people live their daily lives reiterates my own findings.

Hage (2003) alerts us to different ways that the concept of hope is understood, which reflects my own experience of working with this concept,

> Not only is the language of hope associated with aspirations as different as 'hoping for an ice-cream' and 'hoping for world peace'; there is also considerable difference between hoping as an affective practice, something that one does, and hope as an affect, something one has. There is also a considerable difference between hope as a momentary feeling and hopefulness as an enduring state of being. (2003:9-10)

Despite the difficulties of this amorphous concept Hage (2003:3) argues that it is central to the operation of society, "… the most important thesis developed in this work is that societies are mechanisms for the distribution of hope".

While the primary role of government is often understood as planning and organizing the economy, collecting tax and providing services to citizens, or maintaining national security and law and order, Hage argues that underpinning all these functions, of government, is the distribution of hope. Hage defines hope as, "a set of subliminal beliefs that individuals hold that that makes them feel that their life has a purpose, that they have a meaningful future" (2003:12). Whilst his focus is not on people living with a disability his analysis is applicable to my analysis of New Zealand's DMD community. Following Bourdieu, Hage argues that society provides opportunities for purposeful life and meaningful futures. It is society that distributes the social opportunities for self-realisation, or hope (2003:16).

According to Hage, people are not primarily motivated by accumulating capital but rather by creating life that is meaningful to them or by perpetuating their own social being – having a

life that is meaningful, satisfactory, fulfilling. It is society that creates opportunities for living a meaningful life. "It does so by offering people the opportunities to make a life for themselves, to invest and occupy and thus create and give social significance to their selves" (2003:16).

Hage links his analysis of hope closely to the role of the state in distributing hope and this is particularly apt for his topic – the experience of migrants and refugees entering (or being denied entry to) Australia – where the role of the state is paramount. Hage's analysis of the role of the post or neo colonial state (countries like Australia and New Zealand) in distributing hope is accurate as a broad-brush overview. Increasingly marginal members of society are left with a sense of entrapment, often criminalised and without a sense of opportunity or meaningfulness in their lives. However unlike the indigenous and the noticeably 'other' migrants of Hage's research, people with Duchenne muscular dystrophy are positioned within an ambiguous subjectivity – both marginal and discriminated against because of their disability but also (sometimes) part of mainstream families, well connected, articulate and valued by their communities. Furthermore people with DMD are part of the disability sector which is a focus of government concern. The notion of individual responsibility that underpins so much social and economic policy in New Zealand does not apply to people with serious disabilities in the same way that it applies to other citizens. There is some recognition that people with disabilities face additional challenges in participating and contributing to society. This recognition results in some additional social support that offers hope, as demonstrated by the Office of Disability Issues within the Ministry of Social Development, the Disability Rights Commissioner position within the Human Rights Commission and the oversight of New Zealand's commitment to ensuring the rights of people with disabilities by the United Nations. The existence of these statutory bodies suggests that the disability sector is one where the government wants to be seen to be doing well. Within a changing society where the opportunities for many citizens to access social hope are shrinking, participants in this research were able to access some hope demonstrating that the way hope is distributed in society can benefit some marginalised groups. People with disabilities experience marginalisation yet in New Zealand there are opportunities for this marginal group to access hope. Following Mattingly (2010) I argue that hope is found not only through an individual psyche but also through community and mutual support, and following Novas (2006) I further argue that this particular community enacts a political economy of hope.

This chapter explores two examples of enacting hope – powerchair soccer and the New Zealand Neuromuscular Disease Registry. The work of Novas (2006) is useful for the discussion of the biopolitical nature of hope. My discussion about the enacting of hope through medical research confirms Novas's (2006) position that biosocieties (in this case the MDA) are involved in biopolitics (in this case the establishment of the New Zealand Neuromuscular Disease Registry to facilitate drug trials) and that this results in biovalue contestation. Biovalue is the generation of health and wealth derived from biological samples. This contestation occurs because currently the interests of research owners (pharmaceutical companies) tend to be favoured over the interests of individual participants and patient support groups (Novas 2006:301). Novas' (2006) analysis is highly pertinent to the establishment of the New Zealand Neuromuscular Disease Registry which I consider here. The hope for an effective treatment is enmeshed in a series of complex relationships which speak to the provision of genetic technologies (testing services and drug treatments), the roles of companies and markets, the competing agendas of wealth and health. Community activities, like powerchair soccer that offer hope to people with DMD, involve different types of governance and different financial provision. The costs are usually met through fund-raising activities. Fund-raising activities include for example, soliciting charitable trusts (often redistributing the profits from gambling machines), sponsorship from businesses and other organisations, receipt of bequests, as well as activities like sausage sizzles and bake sales. Participants accepted that they should be responsible for securing funding for optional, lifestyle activities. This contrasts with their attitudes to the provision of what they considered essential equipment and services (see Section Three of this thesis).

The current face of capitalism in New Zealand often requires a family to receive two incomes to maintain a standard of living that facilitates the provision of a fulfilling or purposeful life, a life lived with hope. For families with a member with DMD the opportunities for two parents to participate in the paid workforce tend to be limited. For families struggling financially it can be challenging to find affordable opportunities for the family member with DMD to participate in. However despite the difficulties that families faced, both economic and through the condition itself, seeking out a meaningful and fulfilling life, seeking out hope, was a common quest.

As I noted in Section 1, when I discussed the fieldwork undertaken at the MDA National Office, I developed relationships with office staff and two of these relationships were of particular significance. Kerry Hills and Miriam Rodrigues were key informants. They were

friends with each other and also came to be good friends with me during the course of my fieldwork. They are also representatives of two of the epistemologies central to this analysis. Each of these epistemologies reveals something of the hope that is key to the lives of people living with DMD.106

Kerry and Powerchair Soccer

Kerry was both an MDA staff member and a member of the organisation living with a neuromuscular condition. When I started my voluntary work he was the only staff member at National Office with a neuromuscular condition (not DMD but another serious neuromuscular condition). His input has been invaluable to this analysis. Kerry had a constant battle to lead an ordinary life. Spending time with him, both in the office and later visiting his home and at muscular dystrophy social events, revealed the constraints that people with severe neuromuscular conditions deal with all the time. Many of his personal issues were the same as those of people with DMD. I feel it is important to acknowledge Kerry's input into this thesis because he opened my eyes to some of the issues I discuss and particularly the significance of hope. Kerry became a good friend and his death in July 2014 was a huge blow. I was still new to this world of neuromuscular disease and I did not fully realise the precariousness of life.

Many of the things I learnt through my relationship with Kerry were to do with the significance of living a purposeful life. There were many ways that Kerry engaged in living a purposeful life. He was employed, and thus did not seem as isolated as some with DMD were. He maintained close relationships with his family and a busy social life but the issue I focus on here is his involvement with powerchair soccer. An Auckland team had recently been established and he had become involved in both playing and organising the team. He enjoyed playing but his commitment to the administrative side came from a desire to provide a forum for younger players. Kerry undertook his administrative roles (he was at various times treasurer and team manager) primarily from a sense of duty. He did not really enjoy the administrative roles to the same degree as he enjoyed playing. At one stage he wanted to resign from his voluntary position as he was finding all the administrative meetings and discussions too time consuming.

On one particular occasion I walked with Kerry while he took his assistance dog Bella for her lunchtime exercise. It was a warm day and we strolled round a small park near the office

while he discussed his dilemma with me. He was finding all the discussions and social media messaging that his role on the team entailed too time consuming, but did not want to let down a very good friend whose dream the team was. I remember thinking just how admirable Kerry was when he told me he was staying because the team gave opportunities to children with disabilities to be like other children. He said he wanted to make sure that children on the team had a chance to go to school on Monday morning and join in with conversations about weekend team sports. "Perhaps," he said "they too will be able to tell the class they have been awarded player of the day." Kerry, having experienced being a child with a disability, knew just how important participation of this type of was. Establishing the team had involved a lot of work, much of it had been undertaken by a young man with DMD, a good friend of Kerry's. But as the team grew so did the work load. Kerry had initially been happy to offer his support. However, Kerry was also developing a busy and exciting personal life in addition to his formal employment and his commitment to the soccer team. He wanted to devote his spare time to other activities but was driven by a desire to provide purpose, fulfilment and hope to children using powerchairs to continue his administrative roles with the team. This is an example of the importance hope and mutual support has for the DMD community. Appadurai's call to examine aspirations in order to understand how people actually navigate their social spaces (2004:84) is relevant here. The aspirations of men with neuromuscular conditions to establish and run a sporting activity so that others with similar disabilities could participate in a pleasurable and fulfilling activity demonstrates one way of navigating the social space this community inhabits. This social space is often characterised by physical limitation and social isolation. Children with DMD are excluded from most sporting activities unless opportunities for activities like powerchair soccer are available. It also seems pertinent to note that as hope is a concept associated with future benefits it often discussed in relation to children.

It was not only Kerry who appreciated the importance of team sports, the parents of several other Duchenne boys also reiterated how significant the sport was. One family explained how concerned they had been about their son as he became increasingly isolated, wanting only to play on his computer and losing touch with friends who developed new interests that he was unable to participate in. They described the enormous change getting involved with powerchair soccer had made to his life. It gave him something to look forward to each week, increased his social networks, and provided a forum that gave him pleasure and enjoyment. As with the vast majority of activities undertaken by boys with DMD, participation in the

sport involved the family. Similarly, other parents noted how finding a sport which their boys can do helped enormously with their confidence and the pleasure they gained from being able to do something well, improving both their confidence and their quality of life.

One young player used his eligibility for the Make-a-Wish opportunity to get a top of the range sporting powerchair. Make- a-Wish is a charity whose mission is, "To grant the wishes of children with life-threatening medical conditions to enrich the human experience with hope, strength and joy" (Make a Wish 2016). Each eligible child is able to have one dream fulfilled and in this case the boy's biggest wish was to play in international powerchair soccer competitions. For this he needed a particularly powerful and manoeuvrable chair so that he could compete at the top level. This is a clear example of hope in action. This boy is a skilled player and the opportunity provided by the sport, gives him a sense of pride in his ability. The type of hope that participation in powerchair soccer offers is the type of hope that Good et al (1990) discovered in their work with oncologists – a broad idea about making life better. The hope that this powerchair soccer example reveals is the hope for positive and rich experiences. Those involved in pursuing this dream took account of the physical limitations of DMD and envisaged ways of working with this to provide opportunities for people to participate in a sporting activity, as a form of inclusion. This example demonstrates the importance of optimism and positivity. The committee continued to pursue the dream because the fulfilment of the dream offered an improved quality of life for those who wanted to play soccer.

Powerchair soccer provides DMD boys and young men with a normalising activity. Team members participate in a sport and develop skills through coaching sessions, they attend practices and matches, they participate in tournaments both nationally and internationally (during my fieldwork a New Zealand powerchair soccer team attended two Australian based tournaments). The players experience the highs and lows of their team winning and losing, they have the opportunity to be awarded 'player of the day'. The spread of the sport and the establishment of new teams (there are now four other North Island teams as well as the original Auckland team that Kerry was involved with) is testament to the value that the sport provides to people using powerchairs. It is exciting and fun. Some players, like the boy whose dream it was to have a new Strikeforce powerchair, are highly committed players, others are more social players. It is an activity that enhances the quality of life of people living with conditions like DMD by providing pleasure and removing the focus away from activities that they cannot do and focussing instead on what they can do. Kerry was well

aware of just how important this was for children who are so often left out and unable to participate in many social encounters. As such, the sport conforms to Mattingly's (2010) understanding of hope as being about mutual support and overcoming barriers. This was an initiative that was imagined and driven by a man with DMD who sought the co-operation of others in the disability community to assist him in transforming his dream into a reality. The barriers were familiar to those involved in the organisation of the team. The committee consisted of those who had a direct experience of this type of isolation. The committee worked together to provide an inclusive sport that addressed some aspects of isolation and as such were a mutually supportive group challenging boundaries. Powerchair soccer was, following Mattingly (2014), a moral laboratory. Through the activities that were involved in establishing and maintaining a powerchair soccer team people were in the process of becoming moral, they were acting in ways that they considered ethical. Their actions were not just for their own benefit but was for the benefit of a wider community.

Of course not all boys and young men with DMD were interested in playing powerchair soccer; other participants discussed other activities that similarly enhanced the quality of their everyday lives. One participant composed electronic music using digital technology. This was something he was able to do to on his computer and the creative aspect offered this participant a sense of fulfilment. He also worked with a singer and travelled to an Auckland music studio to record his music for a CD. I interviewed brothers who designed websites. One brother did the coding and the other the graphic design. One man was learning to use a 3D printer. At the time of our interview he was designing devices that would attach to the arms of his powerchair and could hold remote controls and his mobile phone in a position that allowed independent use.

It is through engaging in such activities that participants demonstrate a subliminal belief that their life has a purpose, that they have a meaningful future - that they have hope. In presenting such an analysis I acknowledge the point made by Hage (2003) about the challenges of using hope. While these examples demonstrate that people living with DMD imbue their lives with hope and meaning this does not mean that they experience hopefulness as an enduring state of being. In the following chapter I examine some of the aspects of the condition that counterbalance this focus on hope. However, I now consider a different type of hope. In the following section I examine aspects of hope that are characterised by a longer trajectory.

Miriam and the Medical Model

The insights about living with DMD that I gained through my friendship with Kerry contrast with the type of knowledge that Miriam shared with me. Whilst the knowledge that Kerry provided could be categorised as fitting into a social model of disability, Miriam's expertise was more in-line with the medical model. Miriam prized research and explained that the reason for this was because for so many years there had been little medical help for Duchenne patients. Now, however, the latest research looks promising and the international community is constantly improving treatment options. She felt that by maintaining her relationships with the international community and keeping up to date with the latest research she was able to help people in New Zealand. On reading an early draft of this chapter Miriam commented that she prized research because it offered hope. She said that for so long there has been so little hope for Duchenne patients but now she feels that there is hope. The production of pharmaceutical treatments is a slow process but finally is seems that there may be hope on the horizon.

Miriam was constantly involved in research projects, conferences and publishing in peer-reviewed journals (see Rodrigues et al. 2012, Theadom et al. 2014, Rodrigues et al. 2015, Bladen et al. 2013). She was the catalyst for a number of research projects undertaken in conjunction with the MDA during my fieldwork. This research was facilitated by a bequest received by the MDA in 2008 specifically for "research purposes to establish causes and provide effective treatments." The organisation established a separate trust for the management of this bequest, the Neuromuscular Research Foundation Trust (NRFT). The majority of the trustees of this trust are clinicians and academics (MDA Annual Report 2013). In 2010 the NRFT began considering research proposals (MDA Annual report 2013). During the course of my fieldwork the NRFT supported research applications that were undertaken in conjunction with the MDA and research that was independent of the MDA. Miriam was an advocate for, and heavily involved in, the projects that were conducted in partnership with the MDA. One of these projects was the establishment of the New Zealand Neuromuscular Disease Registry. I consider this as an example of hope enacted by a complex assemblage of actors concerned with neuromuscular diseases.

Miriam developed relationships that have proved important to the New Zealand neuromuscular community. She had a key role in the establishment of the New Zealand Neuromuscular Disease Registry. She has a background as a genetic counsellor and has continued to develop an expert knowledge of the biomedical science relevant to neuromuscular conditions. This is all highly specialist and detailed scientific knowledge which can be described as a particular area of expertise. Miriam is able to translate the dense language of this particular area of expertise into accessible information for people in the neuromuscular community.

This biomedical, genetic expertise is valued by many different actors within the MDA community including many MDA members, management, employees and National Council representatives. The MDA supports people living with rare medical conditions and the members want staff who are experts in neuromuscular disease. Scientific expertise in neuromuscular disorders differentiates the MDA from other disability NFP organisations. Not only does the organisation want to provide this scientific expertise but to keep abreast of the wealth of research being undertaken in the field of neuromuscular disease. Genetics and genomic research is at the forefront of pioneering biological discoveries and hence biomedical knowledge about conditions like DMD is increasing. Miriam has been a vital source of information and assistance in this thesis and her ability to translate dense genetic research findings into detailed but accessible information has been invaluable.

Current pharmaceutical developments offer hope to people living with DMD and other rare neuromuscular conditions. This is a different sort of hope to the hope that Kerry enacted as he facilitated the establishment of powerchair soccer. That was to do with hope that had an immediacy: once established, powerchair soccer was immediately available to people. This is a hope for the future, for an effective treatment that will benefit people in the future. Novas (2006) notes that often patient support groups that decide to pursue long term goals, such as investing in future treatments, cannot prioritise more immediate issues such as current economic, social and educational needs (Novas 2006:302). DMD developments in genomic research and pharmaceutical trials suggest that perhaps one day there may be effective treatments for these conditions. Maintaining expertise in this area is valued by many members and by the National Council. It is also a positive area for the organisation to be involved in and invest in. Maintaining and sharing this expertise is clearly achievable. Staff with expertise in neuromuscular conditions can attend conferences and network with others in international forums. They can report new research findings in the MDA's quarterly

magazine or on the website or respond directly to individual queries. Support for research that may eventually lead to an effective treatment is widespread among neuromuscular support groups internationally (Callon and Rabeharisoa 2008, PPMD, TreatNMD, CureDuchenne, The Duchenne Family Support Group – DFSG, Muscular Dystrophy Foundation Australia). This support fits with the cultural dominance of a biomedical understanding of the value of life (Fassin 2012).

The Registry

The establishment of the New Zealand Neuromuscular Disease Registry was an important milestone for the MDA. The registry was launched in September 2011. The purpose of the registry is to facilitate research – particularly pharmaceutical research with the long-term goal of effective treatment for neuromuscular conditions. The MDA website describes the purpose of the registry,

> The NZ NMD Registry provides an important enabling tool for clinicians and clinical trial sponsors to quickly identify participants suitable for each study, particularly those therapeutic strategies that target specific genetic defects. Potentially eligible participants will be informed about new trials and studies though the Registry's network. (MDA 2016b)

The hope for an effective treatment, that will be available to those with DMD, is a dream for all members of the DMD community. The establishment of the registry was a practical step towards the fulfilment of this dream. Miriam described the process through which the registry was set up, identifying other people and organisations that were influential.

> In 2007 I [Miriam] came across a European based network called TREAT-MND and one of the things that TREAT-MND was doing … They were setting up registries for people with these rare muscular diseases to facilitate clinical research. And there are a whole lot of issues around conducting research with rare diseases, one of them being that they are rare, it is difficult to find people, and so you can alleviate that by having these registries where you can quickly contact a number of people with these conditions. TREAT-NMD recognised that in any one country there's perhaps never going to be enough people with a particular condition so they knew they needed to have a network and they needed to have registries in many countries. I think that initially they were European focussed but that very quickly opened up to more of a global sort of a thing.

The team that was involved in organising the New Zealand Neuromuscular Disease Registry decided to create a registry that had a broader scope than other registries associated with TREAT-NMD. They decided to structure it so that it could store the necessary information for all neuromuscular conditions – not just DMD and SMA which was the aim of TREAT-

NMD at the time. Miriam discussed how her collaboration with the Office of Population Health and Genomics in Western Australia, through a contact she made at a conference in France, was a catalyst for the establishment of the New Zealand registry. The contact who was similarly involved in establishing a registry in Australia and was a little further down the track shared useful documents,

> Something which is absolutely beautiful about these meetings is how collaborative and sharing of information everyone is. And I handed Hugh a little data stick and he downloaded a whole lot of information on it, all their letters to government about getting this funded and that kind of thing and we decided that we could definitely work together to achieve this for both New Zealand and Australia.

Through this collaboration Miriam also became aware of the advantages of creating a multi-condition registry in New Zealand. The Australians had set up separate registries for each condition and for each additional registry time-consuming bureaucratic procedures had to be followed.

Following the work of Callon and Rabeharisoa (2008) that identifies the advocacy of French DMD families in encouraging and facilitating medical treatment and the work of the PPMD in similarly supporting medical research in America, I asked Miriam about who was driving these disease registries. She noted a kind of synergy that occurred following the establishment of TREAT – NMD. MDA members who followed international developments, particular clinicians – neurologist Richard Roxburgh and paediatric neurologist Rakesh Patel – and Miriam herself simultaneously realised the possibility for a New Zealand based registry to link into this international network. There was a groundswell of support for initiatives to push research from the lab bench to the patient. Miriam noted that this involved more than just the establishment of registries.

> It wasn't just registries that were setup. They (TREAT-NMD) looked at everything that was required to translate research into therapies. And they addressed every single one of those aspects so they also setup a database and on it they record where in the world there are clinical trials sites that are interested in doing clinical trials with people with neuromuscular conditions and here in NZ we have one of those a trial sites registered as well. They … networked in with a bio bank.

Miriam also noted that strategies to avoid duplicating research and to ensure that negative results were published were also undertaken. So the establishment of the New Zealand Neuromuscular Disease Registry was just one part of a much larger and comprehensive plan to address neuromuscular disease.

All of the work that surrounded the setting up of this registry was undertaken because diverse groups of people shared a hope that eventually there may be a treatment for these rare, genetic neuromuscular conditions. The establishment of the registry is a clear example of Mattingly's analysis of hope (2010) in which people work together to overcome barriers. The people involved in the establishment of the registry came to the endeavour from different backgrounds and with different expectations. Two MDA members in particular had mooted the idea about New Zealand setting up a registry, one was a man with DMD and the other was a woman with SMA and throughout the establishment process they rang to ask how the registry was progressing and to maintain their support and engagement with the initiative. Given their familiarity with neuromuscular issues and the advanced stage of their own conditions it is unlikely that they were expecting to personally benefit from the work of the registry, in which case their hope was for long term benefit for those coming after them. Others who were involved in the registry and shared the hope that ultimately it would contribute to effective treatments included clinical and database experts, supportive decision makers at charitable trusts and at Auckland DHB and people who accepted roles on an Advisory Committee.

The registry, as a form of hope, has a long trajectory. The pathway for the development of medicines to treat genetic conditions is long and slow. For this reason the registry is an example of a different type of hope to powerchair soccer, which has an immediate benefit for those with DMD. This long trajectory is well understood by those familiar with pharmaceutical research and development (Park, Scott, York and Carnahan nd.). Even among these cautious experts there has been recent excitement about treatment developments. Currently a pharmaceutical development that treats DMD is available overseas. This treatment is called Translarna™ (Translarna) and a second treatment, Eteplirsen has proved successful in phase 3 trials and is undergoing the approval process in the United States. I discuss Translarna in detail because it is being developed with the participation of DMD boys, including some from New Zealand who have been enrolled into a drug trial via the New Zealand Neuromuscular Disease Registry. This case study of Translarna demonstrates the relationship between hope and political economy. It is also a classic Latourian assemblage. The role of the drug is a central actor in a network that also involves individual research participants, the MDA, the registry, various clinicians, a pharmaceutical company, Medsafe and the crown entity PHARMAC. All these actors intersect in the anticipation of an effective treatment for DMD.

The registry has been used to facilitate two pharmaceutical trials. One of these trials is to study the suitability of a drug that is currently approved to treat other aspects of bone health but which researchers believe may also improve the bone health of people with DMD. In the other trial three children with DMD have participated in trials for a drug called Translarna (also called ataluren) developed by PTC Therapeutics. The trial is complete but the three participants are on an open-label extension trial of the drug. They continue to receive Translarna while the company takes it to market. PTC continues to provide the drug to patients who had already assisted them by participating in the trials and they will continue to monitor and gather data on the drug.

Translarna

The history of the development of Translarna is an interesting case study of the numerous actors involved in pharmaceutical development. Central to the assemblage is the product itself, the drug Translarna. By the time the drug reaches the stage of human trialling it has already been subjected to years of scientific development. So part of the assemblage includes all the historic efforts of research scientists studying in the field of genetic mutation and for this particular drug in the field of nonsense mutations. These scientists primarily include university based researchers so there are numerous institutional based relationships within and between university departments studying nonsense mutations that link to the assemblage. Nonsense mutations are responsible for about 13% of DMD cases. Hope manifests throughout this assemblage. It is central to the work of neuromuscular support groups such as PPMD (see for example the EndDuchenne.org newsletter of PPMD January 2015 and March 2016 (Parent Project Muscular Dystrophy 2016b and 2016c)), Duchenne Family Support Group (UK), and the Duchenne Foundation in Australia and many other similar support groups around the world who share advice and strategies for getting effective pharmaceuticals fast-tracked through the various national and international approval processes. Individual families and children advocate for the drugs including Translarna to be approved with petitions, parliamentary protests, letters and appeals. Again hope is an action shown through concerted efforts, persistence and refusal to give-up. The company that produces the treatment also has a dual vested interest in getting it to market. In the case of Translarna the drug is manufactured by PTC Therapeutics whose slogan is Translating Science: Transforming Lives with the tagline PTC is committed to bringing hope to patients

with rare and genetic disease. The message from the CEO of PTC on the company website is embedded in the discourse of hope.

> Working in orphan diseases like Duchenne muscular dystrophy, cystic fibrosis, and spinal muscular atrophy positions you right in front of the patients and their families. You have the opportunity to understand the challenges that these families face every day. The possibility of leading the team that tries to discover drugs that alter the course of these diseases and hopefully change the lives of these patients gives our work incredible purpose; it inspires and motivates all of us at PTC. It helps everyone understand why we are always so persistent; every day matters to the patients and their families. It makes PTC a cause as much as a company. (PTC Therapeutics 2016b)

The CEO is a former university professor with expertise in the area of post transcriptional control of gene expression of RNA. He set up PTC to translate scientific efforts into medicine. To do this has been enormously costly. PTC's Consolidated Statements of Operation for the year ended December 31st 2015 show a net loss of $170,447,000 (i.e. 170 million US dollars) (PTC Therapeutics 2016c). Thus the company has an interest both in producing effective treatments that benefit patients (in this case boys with DMD who are still ambulatory) and in returning money to the company.

As well, the assemblage includes those involved in the phase 2b and 3 trials of the drug. These trials operate out of accredited hospitals. The three New Zealand families involved in the trial travel to Westmead Children's Hospital in Sydney, Australia, but other hospitals around the world are also involved. The efficacy of the treatment is measured by walking tests, and monitored for any side-effects. There are many people and organisations involved in running the trials and in getting the boys to and from trials. This was especially the case for the New Zealand boys involved as they had to travel overseas for several days every two months. Clearly many people were pinning hope on these trials.

In addition to those who were directly motivated by hope are others whose bureaucratic roles may act as barriers to hope, for example the various national organisations responsible for deciding whether or not to approve Translarna for prescription. In the US this role is fulfilled by the Food and Drug Administration (FDA), in the European Union it is fulfilled by the European Medicines Agency (EMA), in Australia by Therapeutic Goods Administration (TGA) and in New Zealand the role is undertaken by Medsafe. These are the organisations responsible for determining the safety of drugs and deciding which ones meet their own stringent standards of benefit and safety. Translarna received EMA accelerated approval in August 2014. Despite the success of the drug in trials and its legal supply in Europe the FDA

did not accept the PTC's application for Translarna in February 2016. In Australia and New Zealand Translarna has not yet received TGA or Medsafe approval.

In New Zealand a new product, like Translarna, needs to be approved by Medsafe for it to be available for sale in New Zealand. However, in order to receive a government subsidy which will make it affordable to ordinary people, the product needs to go through another assessment procedure. This assessment is undertaken by PHARMAC. PHARMAC was established in 1993 as a crown entity (controlled by the government but legally separate from it) to decide which medicines and related products are publicly funded in New Zealand and to what level. PHARMAC operates as a crown monopoly negotiating the best price for medicines. As it purchases for all subsidised medicines in the country it is able to negotiate competitive prices, especially for drugs whose patents have expired and which are available from a range of generic manufacturers. However, for drugs like Translarna which are new to market and still on patent (no generic version is available) and which are targeted at a small number of patients it is harder for PHARMAC to negotiate on price. If the price of the drug is high PHARMAC is less likely to fund it. PHARMAC's 2013/14 Annual Report shows an overall pharmaceutical spend of $806.9 million and the May 2016 budget announced an extra $124 million over the next four years. This money was to be spent only on new medicines and was not entailed in PHARMAC's new responsibilities to fund medical devices. PHARMAC is an effective drug purchasing system which receives international recognition (Gleeson 2013). However the criteria by which PHARMAC decides which drugs to fund have inequitable outcomes for people who need expensive drugs to treat rare conditions. In response to strong lobbying by the New Zealand Organisation for Rare Disorders (NZORD) a new trial fund of NZ$5 million was established to address this inequality and in May 2016 four drugs that treat rare disorders were funded.

The aim of the trial fund was to see if PHARMAC could negotiate competitive prices for drugs to treat rare disorders. Those who have been aware that the PHARMAC assessment system was not able to meet the costs of the treatments that are currently being developed for rare diseases were pleased that this trial was introduced but they noted from the outset that the funds being allocated were insufficient. NZORD has maintained the position that a figure of $(NZ)20 to $(NZ)25 million is required (Piper 2015).

What this assemblage demonstrates is the conflicting agendas that exist along the path which a drug travels as it moves from the lab bench to the patient. For many involved in the process

hope is an underlying value. Hope informs the advocacy of people living with rare neuromuscular conditions. It informs the practice of laboratory scientists researching the precise nature and effect of genetic mutations and looking for ways to overcome these effects. Hope underpins the work of the clinicians who treat those with the conditions and who, like the neurologists involved with the set-up of the New Zealand Neuromuscular Disease Registry, are also involved in research projects. Hope was fundamental to the work of Miriam in establishing the registry and it was influential for the decisions made by the MDA board to support this initiative. The three families that travelled every two months to Australia to participate in the Translarna trial hoped for benefits, not only for their own child, but for all DMD children with a nonsense mutation. These parents did not know if their child was receiving a placebo or Translarna. PTC Therapeutics, the company that is developing Translarna also seems to be motivated by hope. Drug manufacturers are often viewed sceptically by those who benefit from their products. They are seen as being motivated by a profit agenda, rather than a health agenda (Novas 2006 and Timmerman 2015).

Novas's (2006) discussion about the generation of health and wealth, through a concept he calls biovalue, is important here. He argues that patient organisations, like the MDA, who aim to fast track the development of effective treatments challenge some traditional ideas about science (Rabinow, 1996) but also that their focus on the production of health is "intimately bound up with the generation of wealth" (Novas 2006:303). In this case-study hope brings people together. The actants in this assemblage are working together to overcome apparently insurmountable obstacles. The establishment of the New Zealand Neuromuscular Disease Registry was really the easy part of the equation. How DNA produces proteins, what goes wrong when the genetic instructions do not work and finding solutions to these problems has been the work of dedicated professionals over many years. Similar stories both from New Zealand participants and from the international community describe the huge hopes people had for finding a cure for DMD when the Duchenne gene was first identified by Kunkel in 1987. One family who talked about the death of their son in 2009 described how initially, during the 1980's, the DMD families had high hopes that a cure was imminent but gradually as the boys died their hopes died too,

Judith   Ron (husband) was actually vice president many years ago, in Dunedin. We had a lot of hope back then. We were waiting for the break-through any minute.

Ron   And in the end they (other children with DMD) all died away and Morris was the only one left.

Judith   But we did have a lot of hope back then.

Now the intricacies of protein manufacture within the body are understood in much greater detail and the science about just how various genetic mutations result in the lack of dystrophin are understood in detailed and complex ways. This slowly developing picture of how molecular biology works provides researchers with the information they need to hypothesise possible ways to address the effects of various genetic mutations. The hopes of the community are still focussed on finding effective treatments but the increasing knowledge about molecular biology means this hope is now understood, at least by clinicians and expert patients, in realistic ways. While families with new diagnoses tend to believe that there will be a cure in time for their child, those who have had the time to familiarise themselves with the drug trial programmes know how painfully slowly advances are made.

The lack of effective medicines was the first hurdle that the DMD community addressed and after a long, slow path it now seems that the indomitable hope of the wider community (including families, clinicians, researchers, patient support groups and pharmaceutical companies) maybe realisable. It seems that there are going to be effective treatments that delay and perhaps avoid the serious symptoms of DMD. The cost and subsidies for these new medicines is the next hurdle that the community needs to address. Already alliances to overcome this barrier are developing. Patient support groups are working together. NZORD has played a major role in lobbying PHARMAC and raising awareness of the inequalities that people with rare disorders face under the current system. The MDA has also made submissions to PHARMAC and supported the position of NZORD. Other support groups whose members will benefit from the new biologics (for example some cancer treatments) are likely to support efforts to extend PHARMAC's ability to fund these drugs. PHARMAC is a crown entity and as such is part of a democratic system of government. This means that it reviews its assessment criteria and solicits public input to the review process. It also has a consumer advisory committee "to provide the Board of PHARMAC with input from a consumer or patient point of view on matters related to PHARMAC's activities" (PHARMAC 2016). These activities are further examples of opportunities for hope to be enacted and reiterate Novas's (2006) point that hope is an integral part of the biopolitics of health.

Hage's arguments about the shrinking state and its impact on hope need to be considered carefully in relation to PHARMAC. The ability of PHARMAC to subsidise pharmaceuticals is increasing as its budget increases. However the volume of new medicines currently in production means that demand will continue to exceed supply. The availability of treatments which people will be unable to access may give the impression of a shrinking state and a feeling of loss of hope may well eventuate. However the situation as it currently exists suggests that PHARMAC is listening to community concerns and is exploring ways to address these concerns. The government has increased the amount of money available to PHARMAC to subsidise pharmaceutical treatments, although Bill Rosenberg and Lyndon Keene argue that this increase comes at a cost to other parts of the health system (Rosenberg and Keene 2016).

Finally, the role of PHARMAC in transferring wealth to the shareholders of pharmaceutical companies needs to be noted. One quiet voice, whose position within the New Zealand Translarna network demands a diplomatic approach, mentioned that while the new biologic treatments are understandably expensive Translarna is not like these drugs. It is derived from an existing antibiotic called gentamycin. It is not like some biologics that have to be administered by injection or through an IV infusion. It is a chemical drug which is administered orally. Even given the costs associated with research (such as flying New Zealand children and families to Australia and funding their accommodation expenses), they cannot understand why the anticipated price bracket is so high. Which returns us to Novas (2006:303) who suggests that the interactions between patient organisations whose goals tend to be about making health accessible and affordable and firms and markets whose goals tend to be around wealth creation is the site where the political economy of hope is situated.

Chapter Conclusion

Following the observations of Robbins (2013) about reorientations within anthropology, my intention in this chapter has been to examine some of the specific ways in which this particular community – those living with DMD in New Zealand – enact hope, how they imagine a good life and strive to achieve it. In pursuing this intention I have contextualised hope within a fairly recent body of literature. Anthropologists who have noticed that this important aspect of socio-cultural reality had been somewhat overlooked by the discipline agree that in rectifying the absence of hope in our field we are addressing a topic that allows us to examine how communities and societies change (Appadurai 2004 and Crapanzano

2004). Enacting hope is a key part of the DMD experience. Here I have addressed it with reference to two distinct case studies. The establishment of the Auckland Powerchair Soccer team is an example of the enacting of hope within the social model of disability. There were many other examples of this social model of hope. It was a fundamental part of many of the interviews I conducted. I heard about people's pleasure and pride in their academic achievements, their love of gaming and the significance of the Armageddon event for some. Some found a sense of belonging and hope through being part of a church community. Planning travel and going on trips was one young man's passion. He would spend long hours researching accessible activities and each year his family would load up their van and head off on an adventure. Many were avid sports fans. One family had organised a rugby world cup tour with their son with DMD. Other young men were fans of wresting and boxing. The pleasures that these young men enjoyed were often unexceptional but reflect the type of hope that Good et al. (1990) identified in their work with oncologists, hope about living a full and meaningful life, not just hope for longevity. This example also illustrates Mattingly's (2010) findings that marginalised and discriminated groups enact hope by working together and finding mutual support to overcome barriers to social participation and her later work (2014) on the ethical considerations of these activities. Mattingly argues, and I concur, that these ordinary, everyday activities are examples of moral strivings people as people attempt to live good, purposeful lives. People and families worked together to set up the first North Island team, to organise a venue, sponsorship and equipment. It was not an entirely harmonious process but an attitude of hope underpinned the work involved.

Similarly the desire to find an effective pharmaceutical treatment for DMD – an example of enacting hope which is embedded in the medical model of disability and is about increasing the longevity of those with the condition – caused a range of people and organisations to come together, to offer mutual support, and work on the multiple small steps necessary to fulfil a long-term vision of hope. This process is ongoing as new alliances and new strategies are devised to address issues of affordability. The role of the state in distributing this hope (Hage 2003) is important. It is the state that regulates the structure of PHARMAC including the special contestable fund for rare diseases. It also allocates the budget PHARMAC is able to spend on pharmaceuticals. The role of patient groups is also important in lobbying both PHARMAC and pharmaceutical companies, especially in those cases where patients have provided resources for the development of wealth generating products. This reaffirms Novas'

(2006) position that the political involvement and participation in science by patient groups (like the MDA) is based on the hope of advancing effective treatments.

The circumstances of lives lived with DMD are often described in grim terms. The impact of gradual but constant muscle decline and the inevitable premature death is the central focus of much biomedical literature. The focus on this degeneration is also common in material produced for general audiences (such as condition descriptions found on support group websites). This discourse involves an inherent lack of hope. The condition itself is described and widely understood as one of hopelessness. However for those living with DMD the reality of daily life involved enacting hope. Finding ways to create meaningful and purposeful lives, given the increasing limitations of the body, was central. Powerchair soccer is a form of hope that is owned and operated by people with disabilities. The New Zealand Neuromuscular Disease Registry is an enactment of hope that is part of a complex international assemblage. The ownership or governance of this type of hope is spread more widely. It is not directly owned and operated by people living with disability. It aligns with Fassin's argument about the prioritising of a biological understanding of the value of a life. It is a different type of hope. It has a longer trajectory and a less certain and less immediate outcome. However the hope for an effective treatment was a dream shared by all members of the DMD community. In line with an emerging anthropological discourse about hope, this study shows the ways those who face situations of despair and suffering imagine and enact hope. This body of literature offers examples and ideas of different ways of being that challenges and enriches the anthropology of social suffering. DMD is not just about increasing weakness and the spectre of death, and structural exclusion from a meaningful life. It is also about living life, embracing opportunities, imagining and creating positive futures. It is through enacting hope that people who live with bleak prospects find meaning and purpose in life. I argue that hope is a fundamental aspect of humanity.

The importance of hope for DMD families cannot be overstated. However, along with the need for hope (as was so eloquently stated by the family who felt "abandoned" when they received their initial diagnosis but found great relief when they met a physiotherapist who could offer them hope) there is inevitably sadness and often depression. Hage (2003) in his discussion of hope mentions the difference between hope as a practice – what one does – and hope as an affect – what one has. The examples I provide here are examples of hope in practice. They are examples of actions that show people clearly have some hope. But as Hage notes this does not indicate that they hold hopefulness as an enduring state of being. In the

following section I consider the inevitability of suffering that is associated with DMD. Those living with DMD enact both hope and sadness.

Chapter 6: Pain

The focus of this chapter is the sadness, pain and grief that is inevitably associated with DMD. The chronic pain and grief is distinctive because it starts with the initial diagnosis of a child and remains, in some form, until the end of life, both for the child and in some ways also for parents and siblings. Grief and pain are enduring phenomena of DMD. While medical experts tend to distinguish between the physical sensation of pain in the body and the emotional pain associated with anxiety, anguish and depression, for the DMD families these types of pain are inextricably entwined. Furthermore, the suffering of the child often has profound effects on the emotional health of parents. Whilst clearly there are some fundamental, universal aspects of the biology of DMD, the experience of the condition is not universal: it is informed by distinctive socio-cultural phenomena.

Social suffering is a prevalent topic of anthropological research. Wilkinson (2013) notes that the anthropology of social suffering is committed to, "understanding how people's pains and miseries are caused and conditioned by society (Bourdieu et al 1999, Dejours 1998, Kleinman, Das and Lock 1997, Scheper-Hughes 1992)" (2013:124). My study of DMD sits rather awkwardly within this paradigm. Unlike many other chronic conditions (TB, diabetes, HIV/AIDS, Rheumatic Fever, for example) it is not a disease that co-relates with poverty, social disadvantage or social dislocation. However, social phenomena affect the way families experience DMD and its accompanying pain. Here I utilise the ideas of Lock (1995 and 2001) and Lock and Nguyen (2010) about local biologies to explain how DMD within New Zealand is a distinctive experience. The experience of bodily pain is universally attributed to a biomedical, genetic explanation by individuals, families and health professionals alike. But the emotional pain that this chapter also discusses has its roots in both medical and cultural explanations. DMD in New Zealand is a distinctive experience because of the distinctive way services such as genetic diagnosis, specialist medical services, subsidised medicines, supportive technologies and emotional and social assistance are provided. DMD is also defined by community acceptance of disability, the way peers react, the Human Rights Act 1993 that prohibits discrimination on many grounds, including disability and, as I discuss here, cultural attitudes to motherhood and the way services are provided. Together the biological, cultural and institutional realities provide a distinctive local biology of the pain of DMD in New Zealand.

In developing a theoretical frame I bear in mind some of the critiques of similar research, particularly those from disability studies, and points raised by Wilkinson (2013) and Mattingly (2014). The focus on pain in this chapter is not intended to reinforce the pity that impaired people often find challenging and demeaning, (Beatson 2004:393-396 and Morris, J. 1991). In a discussion of identity politics Shakespeare (2013:106) highlights that,

> The bulk of disability studies literature casts disabled people in a negative light, and discusses the woes of bodies that don't work, in a society that does not care. This is not only depressing for disabled readers, but it also does not accurately capture the lives of disabled people and their families, which are almost always more positive than these accounts suggest.

My intention in addressing suffering as an aspect of life with DMD is not to depress readers or to ignore the positive aspects of life. Rather it is to reflect the reality of the pain and grief associated with DMD. The continual, gradual decline in strength causes ongoing grief that is distinctive to the condition and informs and influences the way individuals and their families understand or experience aspects of daily life. This thesis would not be a true reflection of the experiences of the families that participated in this research if this aspect of life with DMD was not addressed.

One tranche of literature that examines pain and suffering seeks to understand pain at a philosophical level. Wilkinson (2013) notes that anthropological accounts that explore human suffering have tended to be,

> Devoted to documenting the subjective viewpoints of those in distress, with a focus brought to the immediate dilemmas of their 'lived experience', to exposing adverse social conditions and events that visit harm upon people's lives. The overriding aim is to bear witness to human affliction so as to raise moral objections to social practices, cultural conventions, legal decision or political processes that do harm to people. (2013:124)

This thesis certainly matches Wilkinson's description, it is based on the lived experiences of people with DMD. And in Section Three of this thesis I also address social systems that "do harm" to people with DMD. Thus I have considered carefully Wilkinson's criticism that,

> most of the work in this area is more documental than analytical in its construction and design… very few have ventured to comment on how the experience of and response to suffering in local worlds may both exemplify and contribute to wider processes of social and cultural change (2013:124).

Wilkinson suggests that suffering plays an important role in the search for an essential meaning of life. He discusses the need humans have to make meaning of their suffering. His

point links to the analysis of Mattingly (2014) who argues that those involved in the lives of critically ill children develop ethical ways of living because they are repeatedly faced with difficult and complex decisions about making good lives for their children. These decisions are different to those made by parents of healthy children and mean that parents of the chronically ill must learn to live different types of lives, lives that Mattingly describes as morally worthy. Wilkinson (2013) examines shifting ideas in sociological discourse as religion and religious belief have been subsumed into a rationalist discourse during the twentieth century. In the past Christianity offered a way to understanding suffering, which was to do with having trust and faith in God's purpose, in divine providence. Although people (in industrialised, capitalist countries) have tended to move away from finding succour through religion, Wilkinson argues that suffering continues to play an important role in the way that people make meaning in their lives. Mattingly concurs with this view and uses ideas from philosophy and anthropology to construct her position that lives which are intimately connected with suffering become more intensely engaged with ethical action. These lives become "moral laboratories": parents have to repeatedly undertake decisions and actions regarding the best care for their child. These decisions are complex and require parents to consider many angles and possibilities as they consider how best to make a good and happy life, for a child or for themselves (2014:9).

In my research everyone I spoke to accepted the rational, genetic explanation for their condition. They understood that DMD was caused by was a random genetic mutation, that it was a rare occurrence and that they were terribly unlucky to experience it. It affected the way they lived their lives completely. People accepted this and adapted to it, which does not mean they liked it. It is not what anyone would have chosen. I heard many parents say how unfair they felt it was for their child to have to go through the difficulties that the condition bought. But they also accepted that this was their experience and they just had to get on with it. The rational, medical explanations for the cause and treatments of DMD was universally accepted but as Wilkinson (2013) and Mattingly (2014) note the acceptance of a rational explanation does not exclude the possibility of DMD changing people's world view. Having DMD, or having a child with DMD changes the way people understand their lives. I show how this idea of life becoming a moral laboratory applies to some of the decisions and actions that parents describe in this account of the suffering associated with DMD.

Some participants were religious, they regularly attended church and they talked about their religious faith giving their lives meaning and purpose,

I tend to view the value of life from a social and spiritual perspective now. The greatest hope for me as a Christian is to meet God and be called good and faithful. (From an email from a young man with DMD)

For some families their spiritual beliefs helped them not to feel resentful, but rather to "count their blessings" (although this idea of gratitude was not limited to those who actively attended church). The funerals I attended included references to an afterlife, in terms of running free, of mobility, of leaving behind their pain and meeting with friends and relatives who had already died. These spiritual discourses did not impact on the fundamental and widespread acceptance of the rational, biomedical cause of people's suffering. Good et al (1992) suggest that religious transcendence is no longer a useful frame for understanding suffering. While such a religious approach may no longer be as widespread as it once was, it was certainly powerful for some involved in my research.

The biomedical model was universally relied upon to explain the cause and symptoms of DMD. However as Charmaz (1991) notes, transcending chronic pain remains important and the young men had to cope with their pain. Chapter Five, Hope, shows that experiences of pain do not define life. Those with DMD want to have fulfilling and meaningful lives despite the pain they suffer. By finding ways, in difficult and complex circumstances, to lead good lives, to morally strive to lead purposeful lives (Mattingly, 2014) individuals and family members are attempting to transcend pain.

Good et al (1992:13) note that the language of suffering has changed from moral and religious vocabularies and is now situated in "the proliferation of rational-technical professional argots that express and constitute suffering in physiological, public health, clinical, psychological and political terms". They note that these secular discourses have replaced talk of yearning, misery, aspiration and transcendence with systematic, routinised, quantified talk about biomedical and psychiatric and legal and policy issues. Pain and suffering are usually not discussed, or understood, in terms of the human spirit, or a moral burden or defining existential experience. Pain is now shaped by the pain clinic and pain medication. Mattingly (2014) demonstrates that while the language of pain may have changed, pain itself still has an important role to play in defining morality – an ordinary, everyday type of morality – where the decisions people make about how to live with suffering are moral, ethical deliberations. This analysis applies to my own findings. This chapter on the pain associated with DMD addresses how the experience of and response to suffering in a particular local world both exemplifies and contributes to wider processes of

social and cultural change (Wilkinson 2013:124). It does this by providing an example of the acceptance of the genetic difference as a rational explanation for suffering and contributing to the increasing acceptance that physical and emotional pain co-exist in complex ways (Morgan 2002, Morris, D. 1991 and 1998) and are dependent on social phenomenon (Lock 2001 and Lock and Nguyen 2010). Morgan says that,

> The cultural articulation of pain as a bio-medical disorder has far-reaching implications for the way we represent the experience of suffering. As David Morris observes, it has led to an unshakeable belief that physical and mental pain are different experiences, and that 'real' pain always has an organic cause (1991: 28ff.). However, once we challenge the idea that pain is simply a matter of peripheral nerve endings and electrochemical codes, a different bio-cultural perspective opens up where contexts of meaning become fundamental to the experience and interpretation of pain. (Morgan 2002:314)

Following Morgan, I argue that pain is not just a function of having a body and while somatic pain becomes an increasing feature of life for people with DMD, those with and intimately connected to DMD experience forms of emotional and mental suffering that are embedded in powerful social, political and economic processes. While the symptoms of sleeplessness, stress, anxiety or depression may be treated with medications, they result from both biological and cultural phenomena. The experience of pain, in the context of DMD can be understood in terms of Lock's concept of local biologies.

> …the biological and the social are co-produced and reproduced, and the primary site where this engagement takes place is the subjectively experienced, socialized body. The material body cannot stand as a black-boxed entity, assumed to be universal, with so much social, cultural, and political flotsam layered over it. The material and the social are both contingent—both local. (Lock 2001:69)

Much of the emotional pain of DMD is cumulative so the discrete topics within this chapter; diagnosis, losing the ability to walk, enduring chronic pain, the impact on family members and finally the repeated experience of death for the neuromuscular community, are academic categories. One characteristic of the condition is that new experiences of pain occur in the context of an on-going history. New pain is like an additional block on the tower, a new paving stone on a path. The actual pain may be new, but it occurs within a very familiar context.

Diagnosis

The second part of this thesis commenced with Richard and Laura's account of the diagnosis of their son Zane. In that account Richard and Laura focussed on the sense of abandonment

they experienced when Zane was diagnosed. They lived in a remote, rural part of New Zealand's South Island. Although they could drive to the nearest urban centre which had a small hospital (140 beds) in about an hour, complicated treatments required them to undertake a five hour drive to a larger hospital. At the time of their diagnosis none of the professionals in any of the services they dealt with seemed to know much about DMD. Whilst some of these professionals appeared well-meaning (as evidenced by repeated enquiries about how the couple were feeling), Richard and Laura wanted the professionals to be invested with greater expertise and able to clarify what they should expect and more importantly what steps they could take to assist their son. Given the small population of their DHB – it serves a population of 146,000 – and the rarity of DMD it is perhaps not surprising that the professionals they initially interacted with were not familiar with the condition. Although the grief they felt on first receiving Zane's diagnosis was shared by all families who described this part of their journey, their feelings of abandonment were distinctive. Other parents described being lost in a bureaucracy, which is slightly different to the complete abandonment that Richard and Laura described. Commonly parents wanted to know that they were providing the best possible care for their child and this required expertise, support and a communication pathway between clinicians and families. Currently in New Zealand there is no specific service to ensure that families receiving a diagnosis of DMD (or other similarly life-changing conditions) are supported and reassured about the services they receive. Rather families have to fit into existing systems. Sometimes MDA fieldworkers may be able to offer support but fieldworkers cannot necessarily access paediatricians and neurologists whose busy schedules are remotely organised by bureaucratic hospital booking systems. While these specialists may be happy to respond to the concerns of parents, the parents often do not know how to contact them and may have apprehensions around repeatedly questioning and not necessarily understanding specialist advice. The pain that families experience with diagnosis relates to their initial emotional reaction and can be compounded by feelings of powerlessness, the need to act in the best interest of the child but not being sure what that involves. Experiences with service provision can compound parents' grief and confusion. Being unclear about the best way forwards, not knowing which questions to ask or whom to ask, needing to develop positive and trusting relationships with professionals who are difficult to contact, were all common themes that families mentioned.

The following account comes from fieldwork undertaken in a large North Island town where the local hospital has a regular paediatric neuromuscular clinic on a bi-monthly basis. As part

of my fieldwork I attended an MDA Christmas function in this town. On arrival, there were six or seven mobility vans in the carpark. One van was unloading. This meant that a boy in a power wheelchair was being hoisted down on the rear lift, ready to zoom off inside. In another car there was a crying woman, a distressed looking man and two young children strapped into their car-seats in the back of the car. The mother of boy being hoisted out of the van went to talk to the family with the two young children. Later, as we sat down to our meal, I found myself seated near the family with the two young children. The mother, Anita, explained that they had only recently received their son's diagnosis of DMD which had come as a shock as they had no family history that she was aware of. The Christmas function was the first time they had met other families with neuromuscular conditions and Anita said seeing the teenage boy in the wheelchair and all the vans in the carpark had really made their situation concrete. It hammered home the type of future they were looking at. By the time she came inside Anita had calmed down, she was busy seeing to her two children, making sure the toddler sat reasonably still and ate his dinner and caring for the baby. She was keen to talk to people who were connected to this new community that the family had become part of. Later I interviewed her at her home. She acknowledged how confronting that first social event had been for her, "that was really hard for me… I got in and out of the car about three times, just bursting into tears, but once we got in there it was good."

The emotional impact of a diagnosis played out in the car park as Anita reacted to seeing the teenager with DMD. In every interview parents talked about the grief and despair that accompanied their initial diagnosis. Interestingly, the mother who had gone to support Anita when she noticed her crying in the carpark later told me that she had guessed that the family had had a recent diagnosis. She said that she could easily understand Anita's reaction, "That's normal. We all do that." During my interview with Anita it became clear that her family's experience immediately following the diagnosis of their son was different to that of Richard and Laura. Anita and her husband seemed well supported. Not only were they regularly seeing Rakesh Patel, a paediatric neurologist with a particular interest in DMD, the family had changed GPs and moved to a practice with a supportive GP who was interested in learning about DMD and they had also linked into a local counselling service which they found to be invaluable.

> They are amazing. They are absolutely beautiful people. It is just like a lovely house. There is no receptionist. They just welcome you in and make you a cup of coffee and it is just like going to someone's lounge and they just chat away. They sit on the floor with your kids and chat away. But they are qualified to talk to you. They have a music

room downstairs so if you are having a tough time of it they will take the kids down there to belt it out while you have a wail and a cry upstairs. If we go to a meeting and we want someone to come with us they will come with us.

The family was emotionally supported and also in the care of trusted medical practitioners. Nevertheless the grief the family experienced was raw and profound, as demonstrated by Anita's reaction in the car park.

At the time of our interview Anita shared a concern that she and her husband had. Their son Lukas had been prescribed with high doses of steroids from his first appointment at the paediatric neuromuscular clinic when he was only 18 months old. She had since received the DMD Best Practice Guide for Families which said steroids were only recommended once a child turned 2 years. Anita was clearly worried about this, especially given the serious side effects that are associated with the extremely high doses of steroids that Duchenne boys are prescribed. Appointments at the paediatric neuromuscular clinic were organised by a referral or a recall booking system. Theirs was a reasonable question but they did not know how to find the answer to it. The GP did not know what the specialist's reasoning for prescribing outside the guidelines was. The family wanted to understand why there was a difference between their son's treatment and the guidelines but were not sure they needed, or would be eligible for, another clinic appointment to have this clarified. The situation was different to the "abandonment" experienced by Richard and Laura, but Anita and Douglas felt lost in a bureaucratic system as they attempted to ensure that they were doing the best for their son. The political reality of a remote booking system compounded the anxiety that the couple felt. While this example is, perhaps, a minor concern, Duchenne families are repeatedly exposed to similar situations. The frequency with which they are confronted by bureaucracies that make no allowances for their particular circumstances, adds to their emotional burdens.

This situation is similar to another couple's whose diagnosis was particularly unsatisfactory. Not only did it take a long time and many tests before the cause of their son's health problems were identified but once his condition had been identified as muscular dystrophy the two specialists involved in his care disagreed as to whether the child had Becker's or Duchenne muscular dystrophy. Eventually the family requested a referral to genetic services and at this point the particular complications of their son's diagnosis was explained them.

Fleur: So in Dr Maui's mind he does present clinically with Duchenne's and in Dr Wu's
          mind he presents more with Becker's. So seeing the geneticist was so helpful and
          it is a shame that it took years for that to happen, because she just basically said

they are extremes of the same thing on a sliding rule you can have mild Duchenne's and extreme Becker's and it is going to end up meeting in the middle. … And Dr Wu, obviously he is very optimistic and every time we see him, he says, "He's fine, he is a great young lad, you've got nothing to worry about." And he is the paediatrician, and when we see Dr Maui, he is a lot more pragmatic and looks at treatments. And I think in Dr Wu's mind there really is nothing we can do to help Vincent, he's close to retirement, and when he started there was nothing you could do. He thinks the steroids are dreadful and Vince should not be on them. And every time we see him he tells us that, and he writes letters to Dr Maui saying perhaps we should reduce the steroids. And meanwhile Dr Maui is telling us they are the reason Vince is doing so well. But Dr Wu says the reason he is doing so well is because he's got Becker's, not Duchenne's. So we are just going round in circles.

Kate   That must make your job as parents making decisions about what medications Vince should get quite difficult.

Glenn   It was difficult

Fleur   Well really hard, because you have to go by the expert. We knew nothing about muscular dystrophy. So we've had to do our own research and come to our own decisions and feel that we have made informed choices. You can't be guided by your medical team when they are telling you two different things.

This family critiqued the health system and wrote to the hospital to suggest the introduction of a more co-ordinated approach.

Kate   So a more cohesive system or an in-charge practitioner might have alleviated some of that (problems with piecemeal nature of the process)?

Harry   We wrote to them suggesting that they have someone co-ordinating the care for Duchenne

Kate   And do you feel that now that the care is more co-ordinated?

Fleur   No, I co-ordinate the care. It is co-ordinated because we've learnt how the system works, or that the system doesn't work. And you have Special Education (services provided by the Ministry of Education) doing the physio and school

occupational therapy and then you have Wheelchair Solutions doing wheelchair assessments and you have (the local) Hospital with your paediatrician and you have Starship Hospital with your neurologist and your local GP and all of those people have a part to play and none of them work for the same organisation and they don't communicate with each other at all, apart from sending each other letters with little digs about what they have got wrong. Truly.

The grief associated with a DMD diagnosis was invariably profound. One mother mentioned how she cried each time the quarterly MDA In Touch magazine arrived and she saw the pictures of children in wheelchairs for the first few years after her son's diagnosis. Other parents described the day they received the diagnosis as "the worst twenty four hours", "it was just awful", "I was completely devastated". It was always described as devastating, heart-breaking and painful. It was also vividly recalled. In all cases families remembered the day they received the news, where they were, who was with them, the shock and the overwhelming emotions they went through. While this grief is universal it can be compounded or alleviated by particular local circumstances such as the availability of medical expertise and supportive counselling services. These services affect people's experiences of diagnosis and their grief.

The stories of diagnosis that I have considered, Richard and Laura who felt abandoned and without hope, Anita and Douglas who worried about the prescription of steroids and Fleur and Harry who had to negotiate conflicting medical advice and disjointed service provision, were all attempting to decide on a course of action which was best for themselves and their children. They, along with the other parents I spoke to, were engaged in, "a complex reasoning task that engenders ongoing moral deliberations, evaluations, and experiments in how to live" (Mattingly 2014:4). Their lives had become "moral laboratories".

Losing the Ability to Walk

I learnt first-hand about the physical and emotional pain that occurs when boys lose the ability to walk when I attended the 2012 annual children's camp run by one of the regional branches. My participation was facilitated by the fieldworker who collected me from the airport and took me, along with several of the children attending the camp, to the most beautiful alpine location. The scenery was stunning. The large campground was surrounded

by native bush and had imposing South Island mountains for its backdrop. The accessible accommodation had a commercial style kitchen with a large dining area and a large common room. There were plenty of dormitory style bunkrooms and two families stayed in neighbouring cabins. The camp was for children with neuromuscular disease and most parents did not attend. One family did attend. It was their first year at the camp and their boys had significant sensory impairment as well as muscle degeneration. To ensure there was good communication between the boys and others at the camp the parents decided that they would stay in a nearby cabin but let the boys stay with the other children in the large bunkhouse. A second family also stayed in a cabin. Wendy was a physiotherapist with a special interest in DMD. She had trained overseas where she had had her own clinic attended by DMD clients. She was very knowledgeable about the condition and knew those children attending the camp who lived in the area served by her DHB. She attended in her own time, in a voluntary capacity and brought with her two sons aged ten and fourteen.

Altogether there were eight children - seven boys and one girl – at the camp, ranging in age from seven to fourteen and with various forms of muscular dystrophy. There were also four able-bodied children who came with their parents, all boys. Along with these twelve children there were six adults who all had significant life experience; we were all parents and in some cases grandparents. In addition there were six physiotherapy students. A group of physiotherapy students in their final year of study at the University of Otago attend the camp each year. This long standing arrangement was a win win situation, the students gain valuable hands on experience working with children with rare muscle conditions and they help with supervision. The children bonded warmly with the students.

For me the experience was something of baptism by fire. Seven of the eight children with conditions, were still able to walk. Two of the boys had manual wheelchairs. One had received his just a few days earlier and his mother was not confident about his ability to use it appropriately and decided not to bring it to camp. The other child brought his to camp. All of the boys in the dormitory I was supervising were losing their strength. The boy who had not brought his manual chair to camp, Rory, was, in fact, significantly weaker than the boy who had brought his chair. Rory frequently fell over; he could not go long distances without collapsing to the ground. It was hard to watch this. By the time I attended the camp I was familiar with the literature about DMD, I had met parents at a DMD conference in Australia and they had spoken about how difficult the constant loss of strength and ability was. This was the first time I had actually witnessed it. Rory was really hurting himself physically when

he fell. There was a large grassed area outside the dining room ideal for team games like cricket or soccer and on our first evening at the camp the boys went to play on this grassed area while dinner was prepared. Several times Rory crumpled to the ground and then struggled to get up. He just did not have the strength in his muscles to keep up with the other boys. The falls hurt and he was clearly upset and frustrated; he wanted to participate in the group, to keep up with the others, but he could not. He cried often. His behaviour was angry, difficult and oppositional. He had an existing relationship with the fieldworker and Wendy, the physiotherapist, but even they struggled to calm him down. Rory's physical and emotional pain were entwined in complex ways; his muscle degeneration was the cause of much of his emotional suffering.

The campground included a play area with a climbing frame, swings and a slide. In the morning, before the group's jet-boating activity the boys went to this playground. They really wanted to play there and set off with some of the physiotherapy students. Most of the boys did not have the muscle strength to climb the ladder for the slide. Some of the strong, young students piggy-backed the boys to the top of the slide and slid down with them while others waited to catch the children at the bottom. However the short walk to the playground, the supported use of the equipment and the return to the bunkhouse was a tiring outing for the boys. Rory fell several times. His last fall really hurt, he lay crying on the ground and could not get up. Some other campers near to the playground offered to drive him back to the bunkhouse but he refused their suggestion. Finally I fetched the manual wheelchair that belonged to another child for him to return to the bunkhouse. He did not want to use the wheelchair but he was persuaded to do so by several of the students who offered to push him. Despite the pain, both physical and emotional that Rory experienced, the provision of equipment (the manual wheelchair) is an example of a particular service that mediates the experience of DMD. The provision of disability equipment results from a complex array of social, political and economic processes which are considered in detail in Section Three of this thesis. What this example illustrates is the impact of local biologies on the lived experience of DMD. The provision of the wheelchair is one particular local factor that defines the experience of DMD in New Zealand and other similarly resourced countries. Although initially reluctant to use the wheelchair, by the end of the camp Rory was reluctant to share it with its rightful owner. Using the wheelchair reduced his falls and the associated pain and allowed Rory to keep up with the other boys. This provision of technology changes the experience of DMD by alleviating pain and psychosocial suffering.

Bedtime involved unexpected struggles. Once the boys all had their pyjamas on, teeth brushed and were ready for bed Wendy and some of the physiotherapy students came in to stretch the boys' legs and to make sure they had their splints on. The boys with DMD all had a pair of splints which they are ideally meant to wear during the night. Some of the splints were made of a moulded, hard plastic; others had a soft cushioning fabric around the plastic shell. They are highly recommended for the middle stage of DMD and are designed to postpone or eliminate the onset of ankle contractures which, along with other aspects of DMD, reduce a child's ability to stand and walk (Bushby et al 2010a). None of the boys wanted to wear their splints and all tried to say that they did not have to wear them at home. We ended with the youngest of the boys, Junior, who had only just turned seven years old, sobbing because at home he only had to wear them after dinner, while he sat still watching TV or playing on his computer until bedtime. He did not have to wear them for sleeping because it was too hard to fall asleep with them on.

My participant observation allowed me to begin to understand some of the issues that families face. I was able to explore some of these issues further in interviews. Several parents mentioned that the splints were a site of conflict. Boys with DMD find it difficult to drop off to sleep, regardless of how tired they are (Bloetzer et al. 2012). The parents may have had a challenging time dealing with the behavioural issues that ensue from the loss of strength. Often a child's emotions of anger, anxiety and sadness are compounded by the incredibly high doses of steroids they are prescribed. Parents and fieldworkers mentioned not only how the boys have to learn to manage all the difficulties of their condition but also what they colloquially referred to as "'roid rage". The splints are uncomfortable, especially in the summer when they make the boys feet and legs hot and sweaty in bed. For some families after a long day of encouraging positive behaviour, the bedtime battle of the splints was one they did not need. Like Junior's family they engaged in moral deliberations (Mattingly 2014) to find compromises that worked for them. The emotional difficulties that these children deal with are well recognised in the literature (Hendriksen et al 2011). The combination of muscle weakness (which makes physical activity more tiring) and the difficulty that Duchenne children have falling asleep causes physical exhaustion. This exhaustion together with other emotions common to boys with Duchenne muscular dystrophy; fear and anxiety about what is happening to their bodies, sadness or anger as they start to realise that they will not be like other children. These emotions can be compounded by the mood swings associated with the extremely high doses of steroids that the children are prescribed. The steroids are effective in

keeping the boys ambulatory for longer and assist in reducing scoliosis and the need for traumatic, painful spinal fusion surgery. For some boys their angry outbursts can further isolate them from other children. During one interview a couple described the relationship between exhaustion and anger,

Ian   … but he was shattered by the end of the week. He was tired and we'd get yelled at.

Ella   His behaviour would really deteriorate and it would get to the point where everyone would get hurt basically.

Ian   And we noticed it would start on a Wednesday so we would keep him home on a Wednesday … And that really helped for a while. But then the school came out bitching and moaning that kids weren't going to school. Not us, but kids in general. After that they'd put it to the Ministry of Education. They did not care that he was grumpy at home, he was fine at school. He'd get to the point where he'd sock you.

Ella   He threw a metal crane at his brother's head and cut his head open. That sort of thing. We found it so upsetting to see him like that. Just so tired he lost the plot.

Ian   I've had to restrain him on the bed with my arms and legs wrapped around him just to calm him

Ella   so he does not hurt himself or anyone else, it was horrible.

Ian   He has been a bit better for the past six months but it is still there, the anger and the tiredness underneath it. But he's been better since we got his sleeping sorted out.

It was this type of behaviour that Rory displayed at camp. This is an example of the way biological and social factors co-produce the pain of DMD (Lock 2001:69).

Two and half years later, in April 2015, I met Wendy at a neuromuscular conference in Auckland. During our catch up she mentioned how vastly improved Rory now was, as he had come to accept his condition. One of the other boys, Toby, was, she said, now much like Rory had been at camp. Toby had reached the same stage of needing to rely on his wheelchair more and more, and like Rory was reacting angrily to this change. Although not all boys

exhibit this behaviour many do and it is recognised as characteristic of DMD (Hendriksen et al 2011).

Watching Rory struggle with the challenges of his weakening body was sad. I could see both the physical pain he experienced when he fell over and the emotional pain and frustration he experienced as he was unable to keep up with the other boys. However this pain is just one of the inevitable stages of DMD. The physical pain of repeated falls is transient but the grief that individuals go through as they lose ability remains constant, it is an ongoing part of life. During one conversation a mother mentioned that her son was no longer able to lift a full cup to his lips and the sadness she felt as he gradually lost his strength. Her point was that each time they got used to one new difficulty there would be a new challenge as he lost strength. She said poignantly, "With muscular dystrophy the grief just never ends." Parents weigh up different complex factors as they work out how best to care for a child with DMD. As this analysis of the pain that occurs as children lose the ability to walk illustrates, the decisions that parents make are fraught with ethical dilemmas. They need to decide how to manage the exhaustion, the difficulties with sleeping, the expectations of schools (around attendance in this case), the advice of physiotherapists (regarding the use of splints), whether to permit the use of a new, expensive, wheelchair without the parental supervision at weekends away from home. These are all examples of the ordinary and everyday ethical decisions that parents repeatedly face, in which they need to consider how they are creating a good life, a flourishing and purposeful life for their child.

Enduring Pain

Through my visits with young men living with DMD I came to understand more about the embodied nature of the condition. Although the young men tended to be fairly stoic about their conditions, it became apparent that they lived with a level of ongoing pain. Of all the difficulties individuals with DMD face, discussing their personal experience of frequent pain was something I noticed a certain hesitancy about. However I learned during my interviews that the young men often experience pain in their neck, back and hips from sitting in their chairs. These young men cannot stretch or change position to relieve their discomfort so pain can be a problem. A young man I interviewed explained that this pain was the reason he tilted his chair back several times during our interview. Tilting his chair helped to relieve the pressure and discomfort. During another interview I talked to a seventeen year old after a day at school. His Dad told me he usually went straight to bed after school but had stayed up

because I was visiting. The reason he often went to bed so early was to alleviate his pain. In another interview a sister described her brother's sense of humour and how he joked about his "druggie teeth" because the morphine he relied on to manage his pain had damaged his teeth. Two of the young men I interviewed mentioned the pain that travelling caused. Both needed to travel, one because his home was a long way from the hospital appointments he had to attend, the other because he needed to travel to his parents' home on alternate weekends as his support worker package did not allow him to remain living independently full time. Both mentioned that the jolting of the van caused pain. Coming to terms with chronic pain both physical and emotional is part of life with DMD. I also noticed a kind of black humour amongst those with DMD when they joked with each other about the type of pain relief they had been prescribed. The young men had a nuanced knowledge about what the prescription of various painkillers indicated about the level pain involved. On occasions I noticed they would gently banter with each other about the drugs they had been prescribed, "Oh, just tramadol, can't be too bad then." As tramadol is a medication for moderate to serious pain, the comment was clearly made in jest.

Whilst this pain is part of life as the condition progresses, generally most young men expect to be out and about in their communities engaging in those activities that are possible in spite of the pain they may be experiencing, especially in warm weather. Avoiding anything likely to cause ill health, such as cold and wet weather, is common because a common cold can quickly progress to a serious pneumonia infection for this particular group (especially in the absence of a cough assist machine). This is likely to involve an emergency admittance to hospital and potentially a stay in critical care. Young men describe pneumonia as an awful experience when each breath is a struggle. It is exhausting, it is painful and being in hospital has particular anxieties for those with DMD (as discussed in Section Three). The degenerative nature of DMD means that these men face many painful health concerns; as children they may face operations to treat muscle contractures particularly of the ankles, the high doses of steroids that children are currently prescribed involve a range of risks including osteoporosis which increase the possibility of bone fracture, spinal fusion surgery is an extremely painful operation that involves the insertion of a rod (or rods) and screws to straighten spinal curvature (scoliosis), muscle wastage can lead to bowel problems including chronic constipation and impacted faeces which is associated with acute stomach and back pain, untreated scoliosis also causes discomfort and pain. So physical pain is a part of life with DMD and with it comes anxiety and emotional suffering. The publication of the Bushby

et al diagnosis and management guides in 2010 provide some surety with regard to the treatment of these common symptoms but the disjointed nature of medical services within the New Zealand health system causes additional emotional stress for individuals and their families. This is recognised by some involved in the treatment of DMD and currently attempts are being made to establish a multi-disciplinary clinic specifically for the paediatric care of children with neuromuscular disease.

The Bushby et al publications are an indication of an increasing international awareness of neuromuscular disease and new ideas and developments in treatment. This awareness results from the work of neuromuscular support groups and the work of researchers and clinicians. The guides themselves are a reflection of Fassin's (2012) argument that a biological valuing of life currently holds a dominant position in our cultural understanding of health and disability. The inconsistent provision of health services is evidence that the political worth of lives is not given such precedence. This is reiterated by the requirement of the Ministry of Health that any plans for a new multi-disciplinary clinic would need to be budget neutral, in other words a new system that alleviates some of the emotional burden imposed on families and fulfils the advice of the diagnosis and management guides, cannot be more costly than the current system.

The guides recommend that treatment should be provided by a multi-disciplinary team for all DMD patients, not just children. The problems of inconsistent regional care, the problems that occur when patients transfer from paediatrics to adult services (unlike paediatrics there is little or no follow up of missed appointments in adult services) would be addressed by the provision of a nationwide expert service that managed the monitoring and care of all people with DMD. In this regard the New Zealand health system is part of the local biology of DMD. The fragmented nature of the service provision when appointments at different hospital departments are not co-ordinated, there is often little follow up of missed appointments, and there are insufficient checks that people are being appropriately monitored, severely compromises the care that people currently receive. Heart or lung function may be in decline but if routine monitoring is missed treatment regimens cannot be implemented. The fractured nature of the provision of neuromuscular services creates a distinctive local biology of DMD in New Zealand which often negatively impacts on health outcomes and the physical and emotional pain for people living with DMD.

Impact of DMD on Family Carers

DMD also impacted on the health of some family members. Extreme stress is not an uncommon experience for Duchenne parents and incidents of self-harm were acknowledged within the community, although no one disclosed this to me in their own interviews. During my research three mothers mentioned stages when they had been reliant on sleeping tablets or anti-depressant medication and in three other cases mothers had just not been able to continue being the primary carer. The mothers who mentioned their use of medication had not suffered complete breakdowns and recognised that while the medications they used were helpful they also needed to examine other strategies for managing their anxiety and depression. Two mothers in particular discussed with me the decisions they had made to allow someone else to take over the day-to-day care of their son. In one case the mother, Christine, described experiences from a much earlier era. Her son with DMD, Derek, was born in the 1960s. At this time there was no assistance available to families and the expectation was that children would go into residential facilities when their needs became too complex for families to manage. Christine described how traumatic her son's diagnosis had been as she was told at that time that the only facility that would be able to care for him was in Hawkes Bay. She had lived her entire life in Whangarei so all her support networks were in Whangarei. However, by the time Derek required care there were facilities available in Whangarei. This mother explained that she had married at a young age and soon had her first child, Derek. The couple had adopted a second son a few years later. The marriage did not last and although she was still clearly upset about leaving she explained that she felt she had little choice at the time. She was unable to take her boys with her as she had no accommodation, no job and no way of providing for them. The boys stayed with their father and for a few years he managed the care of them both. However by time he was 11 Derek was living in a local residential centre for children. When he was too old to stay there he moved into an adult residential facility. Even though thirty five years had passed since Derek went into residential care Christine still found it difficult to talk about. She did not want to discuss exactly what had caused her to leave her family, but tellingly she worried that I would think badly about her for doing so. This indicates Christine's awareness of normative, social judgements about "abandoning" a vulnerable, disabled child. She noted,

> we were offered counselling through victim support (after the accidental death of another family member) and I said no. But I think probably twelve months after Derek went it would have been helpful because it actually took me about 15 years to come right, to stop feeling like a victim.

Another mother, Maxine, also referred to her decision to leave her son with DMD in the care of his father. In this case the mother explained how very stressful life had been as she had two boys with DMD and other children. Her eldest son died at the age of fifteen and she had managed to care for her family through his gradual decline and his death. After he died she had a routine appointment with a specialist in relation to the younger brother with DMD. This specialist complimented the way she had coped and said, "You'll be fine next time too." This remark, that was well-meant, triggered an emotional collapse as the realisation that she was going to have to endure all the grief and all the difficulties again overwhelmed her. Her emotional and mental health suffered. Ultimately she decided she needed to live by herself, separate from the family.

The impact of DMD on other family members is an integral part of the story of life with DMD. The condition is difficult for those who have it, it also dictates many aspects of the lives of other family members. Mothers often become primary carers and frequently family life revolves around accommodating the needs of individuals with DMD. Families who have a child with DMD face a difficult juggling act to meet the needs of all family members because meeting the needs of the person with DMD is demanding. Some of this difficulty is to do with the condition itself, the gradual deterioration and the increasing level of care needed. But some of this difficulty is to do with the bureaucratic nature of service provision in New Zealand. Families feel that they are constantly battling systems and this compounds the stress they are under. Even when I was talking to the families that appeared to be organised and in control of their situation, it did not take long before the stories about the grief at what was happening to their child and the frustration they were experiencing with complex bureaucracies came out. In this particular milieu it is easy to understand how some mothers become overwhelmed. In this research it was mothers who discussed with me their emotional and mental health in detail (rather than fathers).

These disclosures by mothers who felt unable to continue with the daily care of their boys with DMD were generously shared in the hope that their stories might benefit others. In the account of the provision of support workers (in Chapter Nine) a similar story was gently alluded to by other members of the family concerned (not the mother herself). The two cases discussed here include marital breakdowns and the women involved did not elaborate on the details of such personal issues. While marital separation is now socially acceptable (in New Zealand) in these particular cases the separation was inextricably linked with a much less socially acceptable decision to leave a child with disabilities. Both mothers said they did not

feel that they had any other choice at the time. Christine, whose son lived in a residential facility from a young age was still distressed about the impact her decision had on this child. This distress was despite the fact that at the time institutional care was routine. Her decision to participate in this research had not been easy, revisiting these memories was painful and challenging. Maxine's son was well cared for by his father in a family situation. The family received sufficient support to allow her son to remain at home, to go to school, and to plan for tertiary study. Maxine focussed on the steps she had taken to regain her emotional stability and how she was using her experiences to help others. She accepted that her son was now well cared for, even though she was not able to fulfil the role of primary care giver.

These two cases demonstrate that improvements have been made in the provision of services to people with disabilities. Children are no longer institutionalised although residential care remains a possibility for adults (see Chapter Nine).The personal accounts these women so generously shared illustrates the stress that DMD can add to a family. While the grief associated with constant, gradual muscle loss is currently inevitable, the provision of support to families facing particularly stressful or critical circumstances is a socio-cultural phenomenon subject to the vagaries of political decision-making. Clearly support needs to be made available before the primary carer reaches crisis point. Mothers themselves seem aware of their own vulnerabilities, as these insightful comments from another interview demonstrates,

Sinead   … we'll never stop fighting for Conrad, I'll never stop fighting for my brother, and quite frankly I would be just inclined to fight for somebody else if they needed me, because someone's got to, that's how I feel. It's very hard when you've got to be so strong, and, people think I'm just like this concrete block, I'm not.

Jay   Yes. No one tells you what you need to know, no one tells you where to go to get what you need.

Sinead   And no one comes and holds your hand either.

Jay   You only find out by getting into the right places and finding out who you've got to talk to and the language you've got to use, and sometimes that language might be me on the end of the phone saying look if you don't sort this shit out there's going to be trouble.

Sinead   Or me threatening or being really harsh. Like at school I used to feel like everyone hated me, that everyone would see me coming and go "Oh get out of her way,". I felt like they all hated me. I was very self-conscious when I was younger about being a nice person and now I'm nothing like I was. This disease has turned me into a very abrupt hard person at times and that's not me at all, is it?

Jay   No.

Sinead   I'm soft, really I'm soft. But, you know, it does really affect you, and I break down all the time. Like I'll be all tough and stoic and then I'll have a weep. It's like say instead of six appointments I might have ten and it's just too much.

Jay   And you call them melt downs.

Sinead   Call them melt downs. I'll go for three months solid and then I'll fall apart, massively fall apart and then I'll be back up again. I remember reading once that 80 per cent of Duchenne parents break up within the first little while of diagnosis and then you've got the percentage of Duchenne mothers who are on medication for depression. I thought, "Oh my God, I've been on and off stuff for years, it's so true". And that's the thing that I worry about the mums, because people don't know, they just don't know, they don't understand what it's like. I hope the doctors get in quick, see that mum is falling apart, and help her.

This account from Sinead not only reinforces the need for better family support but also highlights the benefits that might accrue from the provision of a nationwide service that could support families both medically and socially. The account that she and Jay give of their experiences also demonstrates that they are acting morally, battling not only for their own son, Conrad, but prepared to do the same for other DMD families because "someone's got to". Verkerk et al. (2015) argue for a focus on "family ethics", that is an ethics where families, however they imagine themselves, are at the forefront of ethical considerations; an ethics where the limits of imagining people as autonomous isolates is recognised. They note, just as Jay and Sinead did, that the responsibilities of familial caring are "given" rather than chosen, and that family members may try to provide all possible care for the one with high needs, even when that is harmful to other family members, including themselves, as Sinead revealed had been the case for her.

The accounts of Christine and Maxine similarly reinforce the social suffering that is part of DMD. Christine and Maxine found it difficult to talk about leaving their families. All three women - Sinead, Christine and Maxine - experienced psychological distress partly because of the autonomous, individualistic nature of many families in New Zealand. They were overwhelmed by the expectation that they could manage the care of their sons, the challenging interactions with bureaucracies that are part of DMD, and their other responsibilities. The entwining of the biological reality of DMD, , the fractured nature of the health system requiring multiple interactions and neoliberal ideas about personal and family responsibility resulting in limited assistance to struggling families, all combine to create a particular local biology in which overwhelmed mothers are treated with medication for anxiety and depression. A solution for the individual body, rather than the social environment.

Repeated Experience of Death

Finally I consider the repeated experience of death within the DMD community. During the course of this research I attended three funerals. The first was the funeral of the young man Anthony, whose death is discussed in Chapter Ten, the second was Kerry's and the third was for a teenager who died six months before his eighteenth birthday. When Anthony died Kerry was devastated. Death is always a sad event but for Kerry, who was only 30 at the time, it was an experience similar to that of some very elderly people who outlive all their peers. When Anthony died Kerry told me that they had been part of a close knit trio of friends and now he was the only one alive. Whilst Kerry's immediate family appreciated the emotional impact of Anthony's funeral, others perhaps did not. Those more recent members of the neuromuscular community and even his colleagues at the MDA did not understood that he was grieving not only for the loss of his friend but also for part of his youth.

These funerals were not only opportunities to farewell individuals; they also bought the neuromuscular community together. One of the saddest things I saw during my research was a group of Duchenne mothers gathered around Anthony's coffin after the funeral ceremony was over. Everyone else had gone from the chapel, where the service was held, over to a reception area where refreshments were being served. Anthony's family left first and most other mourners followed. The boys and young men with DMD left with their fathers and other carers, but the mothers lingered by the coffin, crying with each other. It seemed to me that they were crying for their own grief and their own sons as well as for Anthony. This was

reinforced when one of them, wiping her eyes with a tissue said, "this is such a cruel disease" as she made her way over to her son and husband in the reception area. Unlike other activities that bring the community together (sport, camps and Christmas functions for example) these funerals not only celebrate life but also reiterate the inevitability of an early death. At one funeral I stood for a while with the father of a woman who had a neuromuscular condition. "We have been to too many of these," he said. "Over the years so many of Sue's friends have died too soon. So many. She finds this very difficult. "

No matter how moving the funerals were, no matter how poignant the memories that friends, family members and carers shared were, no matter how much love and affection was evident, there was an additional dimension to these funerals with the presence of others who will inevitably also die young and those who love them. While the grief was primarily for the life lost it was also the grief of a wider community mourning their own sad circumstances.

Chapter Conclusion

This consideration of pain and suffering as an aspect of DMD situates the degenerative nature of the condition firmly within a biomedical model. It acknowledges that the physical and emotional suffering is inseparable as much of the emotional suffering is inevitably tied to the physical pain. The pain of struggling for each breath during an acute respiratory infection cannot easily be separated from the anxiety caused by the respiratory infection. Young men with DMD know that respiratory infections are a common cause of death. However the emotional suffering can be compounded by the way services are provided. The provision of a cough assist machine, for example, reduces the incidence of colds developing into pneumonia (as discussed in Chapter Eight). Pain and suffering are co-produced through the biology of the condition and a combination of social, political and economic factors. These social, political and economic factors determine which services and treatments will or will not be provided.

Furthermore, the surety and security that the Diagnosis and Management guides offer contrasts with the insecurity of service availability and this seems to validate Fassin's suggestion that the biological paradigm has greater cultural validity than the political worth of life (2012:111-112). These social signifiers of the different ways DMD lives are valued have important implications for this community, both at the beginning and the end of life. In the following chapter I consider the beginning of life as I discuss the accounts about

reproductive decision-making that the families shared with me; here I close with a final reflection on the significance of suffering as a qualifier for social understandings about the value of life as life becomes increasingly fragile.

Following an account of a life-threatening respiratory issue, one young man disclosed that one of the reasons he hated being admitted to hospital was because he firmly believed that some staff he encountered did not think his life was worth living. He believed they could not see past his disability, his wheelchair, his ventilator, his carers, and the fact that he needed help with eating and toileting. "They just look at us (people with high needs) and think we are not really living." So as well as suffering because of the condition and because of the vagaries of service provision, cultural attitudes such as discrimination are an additional part of the "flotsam" of the local biology of pain for people living with DMD.

Chapter 7: Selective Reproductive Technologies

The selective reproductive technologies considered here are both intricate biotechnologies and complex moral dilemmas. The use of these technologies decides, in the words of New Zealand's Toi Te Taiao; The Bioethics Council (2008) "who gets born" (and who does not). Discussions about the use of these technologies with carrier women and their partners' offers important insights into the way those intimately involved with DMD understand its impact on the lives of their brother and sons. This chapter offers a window onto the different ways some members of the DMD community in New Zealand understand and value lives lived with DMD. The biology and genetics of DMD interacts with treatment and reproductive technologies and the personal, social and cultural context to produce the local biologies of DMD in New Zealand.

The decisions that couples make about having children provide important indicators about values within the DMD community towards hope, towards suffering, towards lives lived with DMD and towards selective reproductive technologies (SRTs). The ethnographic data I collected about reproductive decisions reveals a kaleidoscopic response to similar circumstances. Unlike the previous chapter where families had similar, shared stories about the pain relating to predictable, biomedical symptoms as well as similar accounts of emotional suffering and grief associated with these symptoms and with socially constructed phenomena, in this case family stories varied remarkably, despite some of the similarities of the wider context within which decisions were made. The most dominant factor that informed the decisions that couples made was to do with personal integrity and the strength of people's own values and experiences. The decisions people made were the ones that they were least conflicted about, given their experiences and the options they had at the time.

Fitzgerald et al (2015) provide a useful analysis of the historical, philosophical and legal context for the provision of selective reproductive technologies in New Zealand. They apply a Foucauldian biopower analysis to a particular flash-point of bioethical debate in New Zealand. In describing their use of a biopower analysis they say

> We use bio-politics in the Foucauldian sense as the "strategies and contestations over problematizations of . .. human vitality . . . forms of knowledge, regimes of authority . . . and [desirable, legitimate and efficacious] practices of interventions (Rabinow and Rose 2006:197) ". (Fitzgerald et al. 2015:401)

They focus on the moral reasoning that two different support groups publically adopted in their statements about selective reproductive technologies and on the personal (or private) views that members of these support groups discussed during interviews. They consider these reasonings in the context of the approach to selective reproductive technologies adopted by the state. Selective reproductive technologies are to do with the selection of particular traits (often to do with disability or sex) rather than to address infertility. The justification for the state provision of these technologies is based on ethical notions of principalism which, following Beauchamp and Childress (1979), they define as the "consideration of autonomy, beneficence, non-maleficence, and justice in assessing the ethical harms and benefits" (Fitzgerald et al. 2015:401). Adherence to these values of principalism are evident in the guiding principles of the Human Assisted Reproductive Technology Act 2004, which is the legislation that established ACART (Advisory Committee on Assisted Reproductive Technology) and ECART (Ethics Committee on Assisted Reproductive Technology). These committees are charged with guiding the state on the provision of ethically complex reproductive technologies. These principles are summarised on the ECART website as; the importance of the health and well-being of children, promoting and preserving human health, the safety and dignity of present and future generations, the importance of informed consent, an awareness of, and accessibility to, information about a child's genetic origins, the respect and consideration for Maori needs, values and beliefs and finally respect and consideration for different ethical, spiritual and cultural perspectives (ECART 2016).

As well as this acceptance of an ethical approach that embraces autonomy and informed consent (rather than coercion or denial) the provision of state reproductive services involves other material and historic factors. The 1987 exposé of the uninformed, non-consented, experimental treatment of women with cervical cancer at New Zealand's National Women's Hospital was a cataclysmic event for medical bioethics in New Zealand (Bryder 2009 and Nie and Anderson 2003). The experiment included non-treatment leading to the potentially avoidable deaths of some women (Coney and Bunkle 1987). As a consequence of this event legislative requirements were introduced through the Health and Disability Commissioner Act, 1994, to ensure patients were provided with information and were able to make autonomous decisions about medical treatment. There was also a prioritising of ethical considerations within the curriculum for medical students. Fitzgerald et al. (2015) argue that these changes entrenched the principalist approach towards bioethics and made it difficult for

other philosophical positions to gain traction (in particular a human rights discourse that acknowledges the rights to citizenry for people with disabilities).

However material factors such as developments in reproductive technologies march ever onwards and as new technologies become increasingly utilised their ethical implications are increasingly debated. The particular ethical or moral significance of reproductive technologies is widely acknowledged in anthropological literature (for example, Franklin and Roberts 2006, Kaufman and Morgan 2005:329, Rapp 2011 Scully 2008). In the specific New Zealand context within which I conducted research, Fitzgerald et al. (2015) note that the suggestion of a routine introduction of an first trimester screening for Downs Syndrome triggered a challenge to the dominance of the principalist values in the provision of selective reproductive technologies. They explore the role of two support groups, one of which advocates for the principalist underpinnings of selective reproductive technologies and one which challenges the values of autonomy and informed consent by focussing instead on the rights to citizenship of people with disabilities. The New Zealand Organisation of Rare Disorders (NZORD) publically advocates for selective reproductive technologies to be provided. NZORD represents members with rare conditions that may involve on-going suffering, conditions similar in some aspects to DMD. They hold a principalist position that couples who know they might pass a genetic condition of suffering to a child should have the opportunity to avoid doing so, if they want to. Saving Downs, the other group, argues that the routine introduction of a blood test in the first trimester of pregnancy that included a screen for an increased risk of Down Syndrome was an act of coercion by the state, the final outcome of which was likely to be an increase in the termination of pregnancies affected by Down syndrome. Saving Downs argued against the inclusion of a Down screening test as a routine part of antenatal care on the grounds that such a screen discriminates against a particular disability. Fitzgerald et al. (2015) analyse the public positions of these two groups and the private positions that members discussed in interviews. They discovered that the public dichotomised positions of each group contrasted with a much more nuanced attitude to SRT expressed during interviews. This finding of the complexities of ethical decision making reflects my own research findings and is also noted by Lock and Nguyen,

> "People interpret what fate has dealt them in the form of genes in numerous ways …. An outsider who tries to generalize about such responses is, it seems, doomed to failure."(2010:319)

My research took place within this environment of a raised awareness of bioethical concerns about selective reproductive technologies. However, whilst interviewees expressed strong personal views, they were not involved in the types of philosophical and political challenges of NZORD and Saving Downs. The MDA, also, was not lobbying on this issue. It did publish information about a fairly new reproductive technology, pre-implantation genetic diagnosis (PGD) in their In Touch magazine and one regional branch offered a workshop for members interested in this topic but there was no member interest.

The fact that the couples I spoke to and the MDA did not adopt a strong stance with regard to the concerns addressed above suggests (and is verified by research findings) that the status quo worked well enough for them. This is alluded to by Fitzgerald et al. (2015) who point out that despite the apparent differences in the positions of NZORD and Saving Downs in fact both are based on "an aspirational social ideal of the provision of choice to citizens as an appropriate position from which to regulate selective reproductive technologies" and that "a cultural ideal of choice (is) a shared symbol of a decent society" (2015:414). Fitzgerald at al. note that a range of diverse organisations are involved with bioethical decisions relating to selective reproductive technologies and all accept ideas about choice but construct the meaning of this term differently. Saving Downs promote the rights of pregnant women to choose not to have a test that screens for Down syndrome, and if they do choose to have this screening test, argue that they should also be provided with comprehensive information including information about the choice to have a child with Down syndrome. In contrast, the state argued that the latest in best practice community testing (including the inclusion of routine Down syndrome screening) ensured access to meaningful choice. "The point is that while all of our own study participants might be observed externally and from a distance as engaging in a choice, the term "choice" masks some very complex situated moral reasoning" (2015:414). The public positions and advocacy of NZORD and Saving Downs affected the way selective reproductive technologies are provided by the state. The political positions these organisations adopt and their interactions with the state challenge regimes of biopower. I suggest that accepting the status quo is also a form of biopoolitics. The absence of contestation, the absence of challenges to regimes of authority in the New Zealand situation, where such interventions are commonly adopted, indicates that the existing provisions are at least adequate. The choices that the current system provides to DMD families include the choice to use selective reproductive technologies to avoid having a child with DMD as well as the choice to conceive naturally and risk having a child with DMD. Couples who receive a

DMD diagnosis when pregnant with a subsequent child are also offered choices about whether or not to test a possibly affected pregnancy and choices about continuing with the pregnancy if it is affected.

No one I spoke to objected to these choices being available. But all were keen to explain the moral reasoning that they applied to the choices they made. Fundamentally the people who discussed their reproductive decisions accepted the existing system, whether they opted to use selective reproductive technologies or not. Furthermore, both the discourse around autonomy and informed consent and the discourse around the rights to citizenry for people with disabilities were discussed and validated in the interviews I conducted.

While selective reproductive technologies loom large in anthropological literature and appeal to anthropologists exploring a range of range of theoretical approaches; Foucauldian biopower analysis (Fitzgerald et al. 2015) biological determinist versus social constructionist understandings of embodiment (Lock 2001) challenges to normative kinship and gender relations (Levine 2003 and Strathern 1992) for the families who participated in this research selective reproductive technologies were not a significant site of political engagement. There were other more pressing areas where biopolitics was enacted (the establishment of the Neuromuscular Disease Registry, for example) and the immediacy of issues such as the provision of cough assist machines, support workers and mobility vans were of greater concern to the community. However the information that families shared about these reproductive technologies adds to existing knowledge about the particular local situation in which DMD families make ethical decisions and this is important for future policies about the provision of these technologies. Scully (2008) distinguishes between a conventional ethics of disability and disability ethics. The more conventional, or traditional, ethics of disability is the philosophical consideration of the way disabled people should be treated. This consideration is often undertaken by people without direct experience of disability and concerns among other issues the selective reproductive technologies considered in this chapter (prenatal testing, abortion and pre-implantation genetic diagnosis). Scully advocates for an alternative disability ethics which she defines as,

> Look(ing) at the embodied effects of impairment … working from people's experience of disability to see if and how it colours their perceptions, interpretations and judgements of what is going on in moral issues, especially in moral issues that have direct relevance to disability. (2008:11)

In some ways the information that this chapter is based on does this. All the families who talked about their decisions around reproductive technologies were talking about their own experiences with disability as a parent or sister. For one family the availability of PGD and their successful use of it reflected Franklin and Roberts' (2006) findings that PGD can be a technology of hope. Other families talked about the benefits that diversity has for society. But in all cases the decisions couples talked about were couched directly in terms of their own experiences of living with a family member with DMD and their own values. The wider political debates, triggered by Saving Downs, which were current in the media at the time I undertook this research and which Fitzgerald et al. (2015) refer to as "public and highly politicized processes of moral reasoning" were not mentioned as being of concern to this particular cohort. These were families facing an array of difficulties, challenges and complex decisions but the moral reasoning or the ethical dilemmas they addressed with regard to reproduction were undertaken within the private realm.

The lack of engagement with the politics of reproductive technologies indicates a theoretical acceptance of the availability of selective reproductive technologies. This particular community acknowledges the possibilities and potentialities of biotechnology but when faced directly with decision-making in relation to selective reproductive technologies also takes into account other compelling factors. The stories that people shared highlight the interactions of nature, culture and technology (Lock 2001) but they do not challenge conventional ideas about family and kinship (Strathern 1992:17). Levine, for example, argues that reproductive technologies can shatter classical ways of thinking about kinship (Levine 2003:184) but the selective reproductive technologies that were discussed by families with DMD reinforce, rather than shatter, the connections between biology, genetics, family and kinship. These families pondered preimplantation genetic diagnosis (PGD) a technique that reinforces conventional ideas that kinship is about "natural", biological or genetic human connections, connections between a married couple (or a couple in a relationship similar to marriage) and their children and hence biological connections to other family members – grandparents, aunts, uncles and cousins. Families were not discussing options like surrogacy or donated genetic material which are examples of reproductive technologies that challenge biological ideas about kinship. These were couples that wanted children who were biologically related to them. The development of PGD and the state provision of this technology for families with DMD (and other similarly serious conditions) reinforces social and cultural ideas that kinship rests on biological closeness. Other reproductive technologies

challenge this understanding but one factor common to the stories that couples shared with me about their reproductive decisions (regardless of the decisions that couples made) is that biological or genetic connection between parents and children continues to be a fundamental consideration. The findings of this research show that for New Zealand families making decisions about reproduction in the context of a significant risk of DMD, biological connectedness and genetic relationships remain extremely important.

This claim does not refute the research of others (Ragoné 1996, Strathern 1992) that demonstrate that biology can be over-ridden by social and cultural factors in determining kinship. Clearly this is the case and three of the families involved in this research mentioned the inclusion of children who were adopted. In each case the adoptive status of the child was mentioned as an after-thought, the child was intrinsically part of the family, the care of the child, the worry, the grief and the pride expressed demonstrate that family and kinship consist of relationships other than biological or genetic closeness. Nevertheless the fact that parents only discussed prenatal testing, terminations and PGD during interviews and not surrogacy or IVF using donor eggs when asked about their decisions about family planning does suggest that normative ideas about biologically connected children continue to play a very important role in the decisions people make about creating families – even when those decisions are impacted by serious hereditable, genetic disease.

The ethnographic data about how couples with a known risk for DMD went about planning their families explores how the decisions they made speak to the themes of this thesis; hope, grief, personal experience , the local biologies of DMD and the way lives with DMD are valued. The interviews also respond to the discourses identified by Fitzgerald et al. (2015) as important to debates about the provision of reproductive technologies – covering attitudes to autonomy and informed consent and the rights of the disabled. This chapter is divided into the following topics; adoption, natural conception with prenatal testing and termination, natural conception without testing, and PGD.

Adoption

In the three cases of adoption that were mentioned during this research, adoption was not a strategy for avoiding DMD. In two cases the families were not aware, at the time of the adoption, that they had a child with DMD. In one case, after the couple had their first child and before he was diagnosed with DMD, the husband had become infertile as a result of

contracting an illness, so the couple decided to adopt. In the other case the couple married and the father adopted his wife's son in order to formalise kinship relations within the family.

The third case was slightly different, a boy with DMD, Scott, was adopted by a couple who already had two adult children. The mother, Maureen, worked at a day-care centre for at risk children where she formed an emotional bond with the boy and his birth family. A process of initially fostering this child led, eventually, to a family decision to permanently adopt him. In this case the couple had a pre-existing, biological family and it was because of social circumstances that all members of that family supported the adoption of Scott. Clearly Scott's diagnosis with Duchenne was a factor in the family's decision to adopt. It was evident that his birth mother was not able to care for him and because of his DMD Maureen and the family worried that he would be a hard child for adoptive services to find a permanent home for. They knew that they were able to offer Scott a safe and loving home, and wanted to do so.

In contrast, another family knew that adoption was not for them. In this interview I talked to a mother and daughter (Lynne and Tracy) who were describing the decisions they had made about having children. Lynne had lost both her brother and her son, who was born in the 1980s, to DMD. Tracy had a young daughter and had undergone prenatal testing and terminated affected pregnancies. She was hoping that PGD would be a workable option allowing her to have a second child.

Lynne   Well Neil (husband) and I had talked about adopting when we were younger but I had felt that I could never treat another child the same as my own. He probably wouldn't have minded but I just couldn't consider it.

Tracy   Yes, that just was not an option; we would just have had Bridget (daughter) if it (PGD) did not work out anymore.

The cases where families mention adoption reiterates that, just as Malinowski noted in 1930, social and cultural influences can "over-ride the biological tie" (Malinowski 1930). Clearly both biological connectedness and social connectedness are valued by different members of the DMD community. This is not surprising because the DMD community, a community primarily connected by a random genetic mutation that can affect any family, reflects the diversity of wider New Zealand society, including diversity in values about ways of forming families.

Natural Conception with Prenatal Screening and Termination

The decisions that people made about pre-natal screening show the complex ethical concerns that families dealing with 'the sharp end' of reproductive technologies face. During interviews people discussed a variety of considerations, implications and ethical issues that prenatal screening raises. A view expressed by two women, both of whom had adult sons with DMD, was that it was wrong for any woman who knew she was a carrier of DMD to knowingly risk having a child with DMD. I interviewed a mother and her adult daughter who agreed with this view for themselves but acknowledged that such decisions were completely embedded in personal experiences.

> People do not understand. Unless you have been through it or know something about it they have got no clue. Absolutely no clues. I mean all these people that talk about ethics and what have you, they have absolutely no clue and never will have until they have been in exactly the same situation.

Other women and couples were not comfortable with invasive tests and terminations. Those women who decided against prenatal tests discussed a range of complex ethical considerations. This group included women who were not aware that they might be carriers until they were already pregnant with a longed-for and planned second child. Reservations about testing and termination were also expressed by families where the mother knew her carrier status prior to having any children but the couple wanted to have their own biological children. Also, people's attitudes were not static and in two interviews women who knew they were carriers and had decided to conceive naturally and not undergo prenatal testing indicated that, as a result of their subsequent experiences, they might not now make the same decision. This, too, reinforces the importance of lived experiences in ethical decision making. In those cases where couples discussed their decisions not to undergo testing, there was no rejection of the availability of testing. People acknowledged that the risk of DMD created difficult circumstances for couples planning to have children and that only those who were in the situation could really decide on what to do. This goes back to the point raised by Fitzgerald et al. that "a cultural ideal of choice (is) a shared symbol of a decent society" (2015:414). The two mothers who both felt strongly that it was wrong to knowingly risk having a child with Duchenne and that carrier women should test and should terminate an affected pregnancy, were not advocating forced abortions, rather they were expressing their own incomprehension that others would make different decisions. Their strong views (and similarly those of the mother daughter interview) were based on their own experiences of

loving and caring for their son (and brother) through an entire life lived with DMD. However, others with similar experiences came to completely different points of view. For all families their ethical analysis depended on the way they interpreted their experiences, the conclusions they have come to about life and death. Deciding what to do was not an abstract 'choice' but a decision based on their experiences of living with DMD.

The story Tracy told about her experiences with selective reproductive technologies, having made the decision to avoid having a child with Duchenne Muscular dystrophy, demonstrates the grief and sorrow that committing to this decision can entail. While the current legislation did give her the option to make a decision not to pass on the type of suffering she witnessed her brother go through, the actual way in which she experienced this choice involved a different type of suffering. This suffering was partly to do with the grief associated with terminating much wanted pregnancies but also results from how this is intertwined with local social and cultural contexts in which she made her decisions. This particular social suffering results from the co-production of her own heart-felt decision not to have a child with DMD, the availability of genetic testing services through international arrangements, the vagaries of New Zealand's abortion legislation and an unfortunate experience with a counsellor.

Tracy was pregnant when I interviewed her. She had had five pregnancies in eight years and had a daughter who was three years old at the time of our interview. Tracy had known her carrier status since she was a teenager. When she was 15 the doctor found her carrier status was recorded in her medical notes. Tracy was determined not to have a child with DMD and had known before she got married that having a family would not be straightforward for her. This was known and accepted by her husband, his parents and her own family, including her brother, John, who had DMD. She knew exactly what the condition involved. She was three years older than John and she was clearly very close to him for his entire life. He lived until his late twenties. She said that she had never doubted her decision not to have child with DMD.

Tracy   we are both really close with our families and they have been such a big support for us. No one has ever doubted our decisions about everything. I mean it was our decision but it was good that everyone supported us.

Lynne (Tracy's mum)

> But when you are a sibling of someone with DMD it is like it is not a choice. You just know what you have to do.

When she first started trying for a family Tracy was not aware of PGD, so as far as she knew her only option was prenatal testing. She had discussed this with her GP before becoming pregnant and was reassured that it was all reasonably straight forward. She was told that she would be best to have Chorionic Villus Sampling (CVS) rather than an amniocentesis test because it can be done at an earlier stage of pregnancy. If the CVS test returned a positive result then a termination would be available. CVS and amniocentesis are similar tests. Both these tests are undertaken after conception and before birth. Both use a needle with a syringe to remove a sample of the tissue or the fluid that is found in close proximity to the foetus. In each case the sample contains cells that are identical to those of the foetus. These cells are then cultured before being tested to see if they are affected by DMD. With CVS a sample of the placenta is collected, with amniocentesis amniotic fluid is collected. Tracy was told that having a CVS test meant that if it was a positive test (i.e. if the foetus was affected by DMD) the timing should enable her to have a "straight forward" medical abortion. Various procedures are used to perform abortions. A very early stage abortion can be done by an early medical abortion (EMA) when a woman takes two medicines that cause the uterus to expel the pregnancy in a manner similar to a miscarriage. An EMA is only available in the very early stages of pregnancy so it was not going to be an option for Tracy. A surgical abortion is usually used between seven and fifteen weeks of pregnancy. The woman is given either a local or general anaesthetic. A small tube is inserted into the uterus via the vagina to remove the pregnancy by suction. It usually takes about five to ten minutes. Another option is a medical abortion which uses the same medicines as an EMA but the drugs used are stronger and take longer to work. A surgical dilation and evacuation (D&E) is used after fifteen weeks of pregnancy. A late stage abortion, sometimes termed a partial birth, may be used in exceptional circumstances and involves the process of induction, labour and delivery.

Tracy's GP had not realised that the genetic testing to determine if the cultured cells were affected by DMD had to be done in Australia. Sending the test to Australia added to the timeframe. The test was positive; the foetus was an affected male. This test should have been returned in two weeks but there were some complications which meant the pregnancy was about fifteen or sixteen weeks before a termination could take place. (This was Tracy's recollection without recourse to her medical notes. It seems likely the pregnancy was possibly even more advanced as at sixteen weeks she would have still been within the legal time frame

for a surgical dilation and evacuation.) Tracy's GP who needed to refer her for the termination had been on holiday and there were other bureaucratic delays. The obstetrician who was involved in Tracy's care with this first pregnancy was shocked and upset to discover that Tracy did not realise that she could not have a "routine" medical or surgical termination. Due to the time frames she was actually going to be induced, go through labour and deliver. She suggested Tracy see a counsellor. Tracy's experience with the abortion counsellor was distressing.

> … it was a horrendous experience the first time. When the doctor, who was really lovely, and was like, 'Oh my God, I can't believe you did not know you were going to have to go through this'. I think she (the obstetrician) thought she was doing the right thing and she sent me down to see the counsellor. It was hideous. She (the counsellor) had no fucking clue. I rang Mum bawling my eyes out because she was telling me what it was going to look like what size it was... and I did not want to know. …. I was not some retard (sic) who got pregnant and did not want to be pregnant. Do you know what I mean? It was hideous and I'll never forget it. I was like, "Stop talking. I do not want to hear you". I ended it. I was like; I'm going to ring my Mum. Mum and Dad were in (a town some distance away) so my Mother-in-law came down to be with me because Daniel (Tracy's husband) was at work. Everyone thought it was just a formality appointment.

Tracy never doubted her decision not to have a child with DMD. She continues to be completely sure that this is the right decision and has constantly been supported in this by her husband and all of their parents. Her brother, who had DMD, also supported this decision. Tracy and her husband went through the delivery with their mothers waiting outside the room.

Tracy   We did not want them to be inside when the babies were born. That was just something that had to happen. They did not need to see that. They did not need to be there for that.

Lynne   It was just terrible

Various aspects of the health system contributed to the grief Tracy experienced in her decision not to have a child affected with DMD. The need for genetic material to be tested in Australia and the referral that was required by the GP, added to the time frame and hence to the more difficult and traumatic type of abortion that she eventually had. These are examples of local biologies. At the time that Tracy needed this genetic test some DHBs relied on assistance from Australia. The specific expertise in testing for a rare genetic mutation was not, then, available for Tracy within her DHB. New Zealand also has distinctive legislation

regarding the termination of pregnancy (McCulloch 2013). For an abortion to be legally performed two certifying consultants (doctors) must agree that continuing the pregnancy would result in serious danger to a woman's mental or physical health. Saving a woman's life, foetal impairment and incest are also given as reasons for providing an abortion before twenty weeks of pregnancy. After twenty weeks of pregnancy foetal abnormality in itself is not a ground for abortion. The particularities of this legislation (the need for a doctor's referral for the abortion to be legal) also added to the length of time Tracy had to wait. Given Tracy's thoughtful and considered position, the approach taken by the counsellor added unnecessarily to her grief. The account Tracy provides of this counselling session demonstrates the significance of direct experience and that experiential knowledge is more powerfully relevant to the decisions of women who know they carry the DMD gene than the abstracted generalisations about foetal development offered by the counsellor. The experience that Tracy described sounded more like an anti-abortion lecture than professional counselling.

Tracy and her husband then went on to have a second pregnancy and this too was an affected male so again Tracy underwent a late stage termination through partial birthing. The family then moved to a new town and received their treatment through a different district health board (DHB). About a year after moving Tracy was pregnant again and at 11 weeks had a CVS test. Tracy received a phone call the following morning to tell her that this baby was a girl, which was wonderful news for the family. Sex testing of CVS material is a simpler process than the need to culture cells and genetically test those cells for the presence of DMD. Two years after the birth of their daughter Tracy found herself pregnant for a fourth time. This was an unplanned pregnancy but again an affected male. However, Tracy found that her care with new staff at the new DHB was better. The termination in this DHB was by surgical dilation and evacuation so Tracy did not have to endure a third partial birth. She wondered if the surgeon should really have performed the D&E as she thought the pregnancy was pretty marginal for this type of abortion.

At this point Tracy and her husband had decided not to try to become pregnant again on their own, if PGD was not available they would happily accept the one child they had. They could not face anymore abortions.

Tracy	It used to really annoy me. People were like, "would you not just try again yourself?" I was like, "NO! I will not try again myself. We have been through enough."

Lynne and Tracy also talked about the decision not to have any ritual or funeral, although they knew of other similar length pregnancies when couples had decided to name the baby and have a funeral,

Lynne	… we did not want to know about anything like that. It has just been pushed aside and Tracy said she couldn't have carried on if she had thought too hard or too much about those things."

Tracy	I don't celebrate the anniversaries or think about it at the time or anything. The first time they gave me photos but I will never look at them. I think I threw them away because I know I will never look at them."

Lynne's comment, " I think you draw a line where you can cope", is a powerful indicator of just how difficult the enactment of the decision not to have a child with DMD was for Tracy and for the wider family. Clearly the decision not to pass on the suffering associated with DMD was a heavy burden for Tracy and her family. Each pregnancy involved the potential creation of an array of family connections potential grandmother, aunty, sister, brother and so the loss of that potential kin is grief for an extended family network and this is not a type of loss that receives much social validation.

Natural Conception without Testing

While Tracy was determined to stick to her decision not to have a child with DMD and to endure whatever grief this entailed, the account shared by Ngaire shows a completely different reaction to the idea of prenatal testing. Ngaire and her husband, Geoff, had two children when they received the DMD diagnosis for their son, Thomas. They had no family history prior to his diagnosis and by the time his diagnosis was made a second child, Jasmine had been born. As a result of Thomas's diagnosis Ngaire realised she was at risk of being a carrier but the couple proceeded with a third child before the mother had her carrier status tested. Building on things she had already mentioned, I asked if they had considered prenatal testing.

Kate   And you were offered amnio but you did not want a bar of it?

Ngaire   Yes, that was not for me. Just the thought of it made me feel sick.

Kate   So the thing that made you feel sick was the procedure itself?

Ngaire   Yes, the thought of a great big needle. I did not like the idea of sticking something near her.

Kate   I guess the reason they offered you amnio, the implication is that if it comes back as a positive result, if the pregnancy is going to be affected by Duchenne's then you can terminate, basically.

Ngaire   Yes, and I did not think I could do it (terminate) so there was probably no point in doing it (amniocentesis) anyway.

Her concern seemed to be with the procedure itself rather than with concerns about selecting against DMD. It was an embodied reaction, which prioritised protection of the foetus. I asked if she had been worried that the baby might be affected with DMD. "I can remember (distant relative), saying, "Are you worried about it?" And I said to her, "Whatever will be, will be." For Ngaire it seems that protecting a life that already exists took precedence over the risk of a disability and this attitude towards valuing life was shared by families who described how difficult it had been to receive a diagnosis of DMD when pregnant for a second time.

Three families with no previous history were expecting a second child at the time of diagnosis of the first child. In every case couples said they were strongly advised to consider having an amniocentesis test. Families reported that their paediatrician, obstetrician, midwife and even their neurologist mentioned the availability of this procedure when they were pregnant with their second child at the time their first child was diagnosed. Parents understood that the reason for these recommendations were so they could terminate if the tests came back positive. The repeated offer of SRT did not necessarily come from the same medical professional. However, the repetition of the message suggested a medical endorsement of SRTs. The parents were clear that given these second pregnancies were desired, were planned and that they had been so excited to find out that they were pregnant, there was no way they were going to terminate them. Variously parents said, "that just did not sit well with us," "we had no intention of terminating so there was no point in the amnio, especially with the risks." An amniocentesis test has a risk of causing a spontaneous abortion

of about 0.5% or 1:200 tests (Auckland District Health Board Te Toka Tumai, 2016). In one case the mother talked about feeling the baby moving and kicking and "being alive" and how this, combined with other factors, made even the suggestion of termination upsetting.

Others noted how special their first child was.

> "He is a dude. I know how beautiful my two children are now. I wouldn't not want to meet one of them."

> "If we hadn't experienced Eammon, if this makes any sense, and we were about to have a child for the first time and someone said they might have muscular dystrophy, I might have said yes (to screening). But the reality is that we have lived with a terrific kid. And no matter what the future holds, you would not undo him, so I would say no."

> "He is a gift, no different to any other child. And none of us are here for long, whether it is 20 years or 80 years."

These comments emphasise how much the child with DMD is loved and valued by its parents and how a decision to test and terminate a subsequent pregnancy seems (to these parents, at least) to violate that love and affection. For parents who do not have a family history and have not experienced the later stages of DMD, the repeated offers of amniocentesis were challenging. Not only did the idea of prenatal testing seem to devalue their existing child but they had also bonded with the second pregnancy. The connection between these parents and their potential baby, was part of the reason couples did not want to consider termination.

One couple also mentioned wider concerns about the social significance of selecting against diversity,

> "Children with muscular dystrophy or Downs (syndrome), they all bring something to society. And I think diversity is a fantastic thing to celebrate and I think it is really sad that we have reached a point where we want people to be the same."

Scully (2008) elaborates on the complexities of this position. She acknowledges that there are some who do not support the termination of any foetus but differentiates those operating from a disability critique (Chipman 2006, Mahowald 2007, Shuster 2007, and Vehmas 2003) who are opposed to prenatal screening, not because they object to termination per se, but because they see it as discriminatory. They claim that selection against genetic impairments is discriminatory. Scully cites Adrienne Asch, the bioethicist who, "has long argued that an impairment is often no more predictive of the happiness or worth of a future life than a characteristic such as gender or social class; if we do not consider it ethically acceptable to exercise prenatal selection in terms of those characteristics, we should not automatically

consider it right to do so for genetic anomalies that happen to be detectable before birth." (Scully 2008:799). This position is similar to that of the New Zealand advocacy group Saving Downs (mentioned above), and the couple I interviewed. They alluded to prenatal testing being discriminatory but did not use this language. However, their comment shows the diverse range of ethical concerns that families facing similar circumstances take into account in their decision-making processes.

Some of the women I spoke to who knew they were carriers of DMD, because they had been tested following their brother's diagnosis, had gone on to have children without using selective reproductive technologies. One woman was tested and erroneously told that she was not a carrier but her first born son was affected. She and her husband had gone on to have a second child several years after his diagnosis. The couple described the decisions they made regarding the second child.

Megan   By the time I had Leo I was in my 30s and my obstetrician said that basically, he had nothing against that sort of thing (testing), but I was in my 30s and the test for me wasn't the amnio, it was the other one which was very invasive and I had a huge risk of miscarriage because I was already older. And it was, you know (hesitates). I had been trying to get pregnant for a long time…. I didn't want to do it (testing) because I wasn't risking that.

Dave   We did look at the pre-implantation genetic diagnosis, but that was only available overseas at the time.

Megan   Yeah, it was really expensive.

Dave   And it was going to be...

Kate   What year was Leo born?

Megan   2008.

Dave   When we were looking at that, Frankie was only two or three.

Megan   There's a seven year gap between them, so... yeah Frankie was only little and we just couldn't afford it.

Dave   And it was 20 or 30 grand, before we flew over and stayed there.

Megan   So we were completely out of the picture straight away. So yeah the other option was the testing and I just, oh no, I just ... apparently it's quite painful, the one that they said I would have to have, very painful and incredibly invasive.

Kate   Was that called chorionic villus sampling, CVS, and they can do it earlier than amnio?

Megan   Yes, that's what it was, but yeah oh no, I just… You know, it just doesn't agree with me, no it's not my thing at all. So to me it was like well I've already done this, dealt with this with my brother, doing it with Frankie, well what's another one, seriously. It's not like I hadn't dealt with it before. To me it wasn't a big deal. You know, easier said than done though.

This account reveals several issues with regard to the use of selective reproductive technologies. Clearly this couple did not object to all reproductive technologies and would have considered PGD, had it been available. They were not saying it was wrong to select against genetic difference. Megan's remarks about the risks associated with CVS were not statistically accurate. Her incorrect recollection of information relating to risk is similar to the findings of other research and in this case Megan was remembering information she had received many years prior to our interview. Again the interview demonstrates a case where that attachment to the existing pregnancy takes precedence over the risk of DMD and that the decisions are being made within a wider life context (Megan's age and her experience with DMD). During our interview Megan talked often about how her experiences with her eldest son compared to the experiences of her brother. She was well informed about the implications of her reproductive decisions.

Megan's account of the risks, and the invasive nature of prenatal testing using CVS takes place five years after Leo was born and so she was recalling the information which, given the numerous interactions the family have since had with health services, surely seemed historic. Statistically her narrative is not accurate but it does reveal much about the factors that influenced the decisions she and her husband Dave made. Mujezinovic and Alfirevic (2007) publishing at the same time that Megan was considering using PGD, compared the statistical risk of miscarriage following CVS with the risk following amniocentesis. The rates were in fact very similar. "Total pregnancy loss was marginally higher 2% (CVS) compared with 1.9% (amniocentesis), which is to be expected given the difference in gestational age when procedures are performed" (Mujezinovic and Alfirevic 2007:692).

Currently (2016) the advice offered to pregnant women by the Ministry of Health is that CVS has a risk of miscarriage of 0.5 – 1% (1:100-200) (Auckland District Health Board 2016). Rapp (2004) discusses misunderstandings that arise as genetic counsellors translate the language of both genetics and statistics into lay terminology and how common it is for pregnant women not to accurately remember the information that they are given. Rapp also notes that the explanations of risk that are provided are about very specific risks relating just to particular biotechnological procedures. They do not, and cannot, take into account other social and life circumstances that may affect the decisions women make (Rapp 2004:66-73).

Megan describes the CVS test that was recommended to her as being "very invasive" and having "huge risks of miscarriage". As a carrier of DMD, Megan had a 1:4 chance of having an affected child. This risk increased to a 1:2 chance once the sex of the foetus had been confirmed as male. It seems likely, that Megan's decision not to use CVS was not primarily to do with the much smaller risk associated with miscarriage following this procedure. Rather it was to do with her general concerns around testing and termination, with which she was clearly uncomfortable. Not only did she want to protect the pregnancy from harm ("I wasn't risking that"), she also felt by terminating her second pregnancy she would be sending a message to her son with DMD that she would have terminated him if she had known he had DMD. As both Megan and Dave evidently adored this child, this was an important ethical consideration. In refusing a prenatal test Megan was, amongst other things, affirming the value of Frankie.

Megan   I had already had Frankie (with DMD), so to me it was like, well, how awful to have a test and then go. 'Oh well no you're not happening (about the pregnancy), but sorry Frankie, you just slipped through the gap, mate, you got through it, you got lucky.

Another point arising from this interview is that although PGD was not available in New Zealand when Megan and Dave first investigated this as an option, by the time Leo was conceived PGD was available. but the interview suggests the family were not aware of the significance of the Human Assisted Reproductive Technology Act 2004, and the subsequent work on eligibility and funding. The context within which the family was making difficult decisions had changed in important ways and despite being linked into DMD health services (as a result of the needs of their eldest son) they were not aware of these changes. The establishment of a multi-disciplinary paediatric, neuromuscular clinic (a current goal of

specialists, the MDA and parents) would be an appropriate service to provide this kind of communication. Such a clinic would be able to identify and inform known carrier women of reproductive age about new biotechnologies available to them.

Although Megan said having a second child with DMD would not be a "big deal", her later comments about the relief they felt when Leo received a negative diagnosis suggest that it was a difficult decision.

Megan   So we had already decided that the game plan was we would find out the sex so we could prepare ourselves, and of course I'll never forget the day they said it was a boy … so then we organised for cord bloods and thank God our game of Russian roulette worked, because he is fine. I didn't stress too much, but the cord blood, you know, that was a nervous wait.

Kate   How long did the results take?

Megan   Oh it was pretty quick

Dave   A day and half

Megan   A day and a half, but they had a bit of a mess up, the way they actually told us the results. First they mixed us up with another family, we knew that family. They got such a fright. "What the hell are we being tested for that, for?" And then they sorted that out and then told us as if we had just had a test for the 'flu. We were just horrified about the way they just came and rattled it off and we said, "Hang on a minute, you better go and double check that, that doesn't sound very important." And I sent her back…. No idea, poor woman.

Dave   She just came in and said, "He's fine."

Megan   She came in and said everything is fine and then went to walk out of the door and I said to her hold on, called her back and had a bit of a go at her, so she went and saw her superiors. I demanded that they ring the paediatrician. I wanted to hear it from someone important… who could relate it to me. I mean we are very facts and figures, we are quite clued up.

Dave   It wasn't hard to see because Frankie was sitting there in his wheelchair.

Megan   So the whole thing was just awful and by the end of it I think I had upset the whole maternity ward.

Dave   They upset you too. There's been a bit of that.

Megan   They had rung up, they had rung the paediatrician twice I think for me. I think I spoke to her, I can't remember, and about three people later and I had a huge apology and rah de rah and then after that I think we all just collapsed in my room, Frankie included, and all just burst into tears didn't we, while Leo slept through the whole thing. He was fine, but the rest of us, it was very, you know, it's such a dramatic thing to be just relayed like that. I was pretty angry about the whole ... and then it's funny, I redid the test, I redid it when he was three months because I was just absolutely, you know, the good stuff didn't happen to me. I don't get good luck, so I'll want it rechecked, double check, triple checked. So by the time I think he got to about four months old, we were pretty convinced, yeah. And he's certainly very healthy.

Megan's use of symbolic language including gambling metaphors "a game plan", "a game of Russian roulette", and her comments about luck "good stuff doesn't happen to me" and "I don't get good luck" is significant. It suggests a world view that doesn't just rely on rational risk-assessment. Close analysis of this interview reveals the many layers of values and beliefs that impinge on reproductive decision making.

Decisions about the use of reproductive technologies were not absolute, immutable ones as shown by a family who initially decided against prenatal testing but for a subsequent pregnancy decided they would test. Brenda knew she was a carrier, she and her husband had an affected son and decided to test the next pregnancy.

Kate   Because I was just wondering about your initial decisions when you first had Charlie you knew there was a risk? Was amniocentesis or CVS offered to you?

Brenda Yes.

Kate   And you chose not to.

Brenda We just chose not to, we just wanted him regardless.

Mark   With Charlie, we knew the risks and we were probably a little bit, I mean how old was I when Charlie was born? Early twenties?

Brenda   Yeah, young

Mark   So young and you know, "Nah it's not going to happen to us."

Kate   Despite having already lost your brother?

Brenda   That's right, yeah.

Kate   And you knew you were a carrier?

Brenda   Yes, I found out when I was about 18 I think, when I first met Mark, he was actually there for the results for my sister and I.

Kate   Were you both carriers?

Brenda   No, just me. That was hard.

Kate   It must have made the pregnancy feel quite...

Brenda   Well it wasn't exciting for our parents. They were all kind of, well especially for my parents, that worry was there. I found that quite hard because usually the first grandchild is … you're just having the baby, you know, so it was all very, no hype. I found that quite sad. …   So yeah that for me was really quite hard, because it's supposed to be exciting.

Mark   And for my parents, for my side, same for me, didn't really understand ... knew it, but didn't understand it, that sort of thing. And when Brenda got pregnant, this genetics guy from Wellington came up and said "here's the test" and we said "no" and they asked us why and we said the reason is because you do the test, if you find out it's a boy with Duchenne's there's really one option and we're not taking that option so there's no point doing the test. So that was fine. And my mother really cracked a fit on that one. She said why, should be doing the test, blah, blah, blah, yeah.

Brenda   They found it quite difficult. Having a child with special needs was quite huge for them because they thought it was not just physical, they thought it was intellectual as well. And I just don't think they wanted to have that in their

grandchild. It was real hard, yeah. But even with Sally we had to keep that fairly quiet.

Mark   We kept that quiet until we had done the tests and found out, so we didn't tell everybody until ... and it's a girl.

Brenda   And then with Sally it was, "Can you afford her?"

Mark   Yeah, that was the first thing it was, "Can you afford her?"

Mark   And then it was just deciding when the timing was right for Sally and we decided we would do the test, what they do at 14 weeks or whatever it is?

Kate   Is that amniocentesis or …?

Mark   It's another one. It's something like that. So we did that with the intention at the time of if it was a boy and it had Duchenne's we would terminate. I'm glad we didn't have to go down that path because I don't think we would have, who knows.

Brenda   Religious wise too. I just thank God that we didn't have to make that decision.

Mark   We didn't have to make that decision.

Brenda   He gave us the girl. A bit of a miracle.

Mark and Brenda, like Ngaire, Megan and her husband Dave and the families who were pregnant at the time of their first diagnosis, were all clearly uncomfortable terminating a pregnancy. This interview shows how attitudes towards SRTs change over time. Mark suggests that their initial decisions were based on their youthfulness, and their optimism that "it's not going to happen to us", that sense of being bulletproof that accompanies youth. This was combined with their religious faith. I asked Brenda about her comment "religious wise" and she said that the family was Catholic, and that she, especially, was. This was reflected by some of the metaphors she uses in her speech. That Sally was a girl, she describes as a "miracle" and when talking about some of their difficulties she used the phrase "a heavy cross to bear". As they had initially been so reluctant to accept SRT, even when the genetic counsellor came to discuss it with them, the couple's change in attitude supports my analysis that attitudes of people living with DMD towards reproductive technologies are based on their lived experiences. As the younger sister of a boy with DMD Brenda had some

experience with the condition but caring for her own child offered a different insight. Lived experiences also impacted the decisions of the family who thought they may have been more accepting of the offer of amniocentesis if they had no experience of having a child with DMD.

Similarly when I interviewed Ngaire, she asked if I was going to visit another family with two affected sons. She said she could not imagine how hard things would be for that family and mentioned both managing on a daily basis and the grief when one dies before the other. I was initially surprised by this comment as her family were at risk of this same outcome, given their refusal of prenatal testing. On reflection I realised that when they had proceeded with a third child they would not have known how hard life was going to get. The challenges they came to experience, with barriers in the education system and a systems-centred, rather than person-centred, health service which was unable to provide optimal care for Thomas, were far in the future. At the time they decided to proceed with a third pregnancy their knowledge of DMD would have been quite different and possibly more hopeful. Ngaire's empathy for the additional grief the family with two affected sons will have to endure suggest that since her own decision not to test 14 years earlier, she had come to understand in a more experiential way the sadness associated with DMD.

For families with a history of DMD there is quite a different reaction towards the announcement of high risk pregnancies. This experience of sadness is what informs the reactions that Brenda and Mark describe within their wider family as the news of Brenda's pregnancy is received with such a muted response. The usual joy and excitement is replaced by anxiety, concern and even judgement from the wider kin group (especially grandparents). Megan and Dave demonstrate their own anxiety by narrating in detail the upset and misunderstanding that occurred when they received the news that Leo did not have DMD. For Megan and Dave the test result was loaded with so much significance they could not accept the casual way they were initially informed. For over nine months they had been waiting to learn the answer to this life-altering test and they needed to be informed formally and by a person who fully understood the significance of the information being imparted.

The birth of a baby is significant for many reasons and one of those reasons is a about hope for the future. The nature of that hope is quite different if the child has DMD. Megan and Dave were well aware of this as evidenced by Megan's comment acknowledging that dealing with a second child with DMD would be, "easier said than done."

Pre-implantation Genetic Diagnosis

PGD is a technology embedded in hope. The experiences of Tracy and Daniel with this technology reiterate the findings of Franklin and Roberts (2006) that PGD is a technology of hope. I'm not sure the family would agree with Franklin and Roberts' statement that "all reproductive technologies are 'hope technologies' (Franklin 1997) in their orientation toward future progeny, scientific progress and triumph over adversity" (2006:213). Tracy's experiences with prenatal testing and terminations were sites of grief rather than hope. However, their success with PGD, demonstrates that this particular reproductive technology did offer hope for "future progeny, scientific progress and triumph over adversity" (2006:213), as the following account shows. After their first termination Tracy and Daniel heard about PGD and requested an appointment to discuss their suitability.

Tracy   I waited about six months for an appointment to see the fertility clinic in (city) to find out about it. But I really felt we were quite brushed off and told to go away because I was young and I could get pregnant. So they pretty much said go away and try again. Take your chances. Because they only got funded for three people in the whole region to have PGD done.

PGD, or pre-implantation genetic diagnosis, is similar to IVF (in vitro fertilisation or "in glass" fertilisation) when an egg and sperm are fertilised outside the body (in a glass petri dish) and fertilised embryo are implanted back into the mother's uterus. With PGD an additional stage is added to the process. Once each fertilised zygote consists of eight cells, one cell is removed and tested for a specific genetic mutation (in this case DMD) and only those that are unaffected are implanted.

Tracy and Dave followed the advice of the fertility clinic, and after conceiving one girl and terminating two other pregnancies decided that they were not going to try again on their own. Having decided against any further terminations or adoption, Tracy and Daniel made a request to their new DHB for PGD. PGD is technologically complex, time-consuming, emotionally challenging and also exhausting (Franklin and Roberts 2006).

Tracy   … fairly soon after (the third termination) we went to the GP and asked to be referred to X (city with a large hospital that had a fertility clinic) for PGD. And we got an appointment at the fertility clinic within 3 months, which was amazing. And it was really a year to a year and a half later that we actually started the

whole process. And then with that we did two full cycles with the drugs and everything. The first full cycle we only got, ended up with, 6 eggs and 4 fertilized and only one that was healthy and that was implanted and did not take. And then the next month we did the full cycle of drugs so it was all over with pretty quickly and out of that I think there were eight eggs which resulted in 4 fertilized eggs and all 4 were healthy, which they said was amazing. To have that odds. And then one was transferred and did not take and out of the three remaining there was only one suitable for freezing. So that was this one. Last Chance Charlie.

The family was obviously thrilled at the time I interviewed them, after all the grief and loss, that they now had a second reproductive success story. At various times during the interview Lynne said,

> "And I still can't believe this one (current pregnancy). When Tracy said, "Well it has worked!" It is still hard to believe it. When Tracy had a scan and showed me that picture up there (a picture of the scan was on the fridge)it sort of became more real."

> "Yes it is amazing. I still can't quite get my head round what they can do. It is quite incredible."

This family's excitement and anticipation was tied to a number of experiences. Although Tracy and her family were in full agreement that DMD was to be avoided at all costs, Tracy had paid a high price for achieving a family without the condition. The terminations were difficult and Tracy said several times that she could not think about them too much,

Tracy   I guess you do feel guilty a little bit. Especially the third one, I did feel guilty. It just seemed worse than the others somehow, I don't know why.

Lynne   It was hard.

As well as the grief associated with the three pregnancies that did not go full term, the family also had to deal with their much-loved son and brother succumbing to the final stages of his DMD during this time. It is clear from the interview that DMD has been a blight for this family. Lynne's brother was the first in the family to have DMD, although her mother must have been a carrier. No one realised this until Tracy's brother, John, was diagnosed. Tracy was born before John and was also a carrier. Lynne has experienced the loss of her brother and her son, her daughter was a carrier, her daughter had terminated three desired pregnancies which were all affected boys, her granddaughter may be a carrier. For this family PGD was, as Franklin and Roberts note (2006:108), a treatment for grief, Lynne in particular

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was annoyed that her granddaughter, Bridget, could not be tested until she was old enough to understand and consent to the test.

Lynne   And you are worrying about it for however many years and they may not be. And then you've done all that worrying unnecessarily.

To finally have a pregnancy that was free of all the worry of DMD was a relief and an experience of hope. Hope is generally difficult to define (see Chapter Five) but is particularly so in relation to PGD, as noted by Franklin and Roberts,

> One of the major problems resulting from the importance of hope to technological improvement, and particularly to new reproductive and genetic technologies, is its unaccountability. Hope is not easily explained nor readily interrogated. It is neither accessible nor assessable, and it is frequently unmanageable. Indeed, as both a noun and a verb, hope eludes easy classification. It does its work primarily as a condition. (2006:215)

All births are about future hope. When a child is diagnosed with DMD the grief parents universally experience is partly about coming to terms with the loss of the hopes they held prior to diagnosis and coming to terms with new ways and new types of hope. For Tracy, Lynne and the rest of the family the knowledge that the baby whom Tracy was carrying was not at risk of the suffering they had experienced with her brother John offered, not only a relief from worry, but also hope for a healthy child.

Chapter Conclusion

For the couples in this research deciding whether or not to use prenatal testing was ethically complex and emotionally gruelling. Decisions were embedded in complex networks of kinship, grief and hope and were never straight forward. In every case a child or brother with DMD was central to the discussion. The commitment by families to those affected by DMD cannot be overstated. The one universal aspect of the kaleidoscopic decisions couples made about SRTs was the high value and high regard with which families held those who had DMD. Even though some families reject the idea that the care they provide is exceptional and point out that the commitment with which they act is not optional, I found the relationships within many families to be extraordinarily supportive. The devotion of parents and siblings to enhance, as far as possible, the lives of their affected sons and brothers was a significant characteristic of many in this community. It was this commitment which led to completely different reproductive decisions. For some families the closeness and bonds between parents

and children prevented the parents considering termination. The value of the existing pregnancy and the hope that this pregnancy entailed (whether or not it would be affected by DMD) took precedence. These families did not want to risk any harm to the pregnancy from invasive tests and were not going to terminate, partly because of the implications such a termination would have for the existing and much loved sibling. In contrast for Tracy and David, despite the distress of repeated terminations, avoiding passing on the suffering they knew Tracy's brother had endured in his life, took precedence.

In discussions about SRTs there was a general acceptance of the current system that allows people some autonomy in their reproductive decision making. The idea that SRTs discriminate against disability were alluded to by one couple, but they did not suggest that others should not be able to access testing, terminations or PGD. The fact that neither individuals from the DMD community nor the MDA engaged in a public or political way with the debates about the provision of prenatal testing described by Fitzgerald et al. (2015) as "public and highly politicized processes of moral reasoning" suggests two things: firstly an acceptance of the status quo which provides known carrier women with two funded cycles of PGD and secondly a possible reluctance to engage in an emotionally loaded and polarising debate that has direct links to close family members.

The increasing availability and success of PGD enables couples, where the female is a known carrier, to avoid the grief of terminations, the anxiety of not testing and the decisions by some sisters to avoid having children altogether. However aspects of service provision, information and communication need to be addressed so that women who may benefit from such procedures are aware of their availability. The very personal experiences which underpin the decisions that couples made (relationships with much loved brothers, for example) explains in part the private nature of reproductive decision making amongst DMD families.

Often debates about SRTs are framed around ideas of choice. Fitzgerald et al. (2015) point out that the idea of choice is central to the provision of reproductive technologies in New Zealand and this research reinforces that this value is accepted by many families where there is a risk of passing on DMD. However, in a final nod to semantics, families who are at the 'sharp end' of SRTs are not making abstract 'choices'. These are difficult and ethically complex decisions and in making these tough decisions people rely on their own lived experiences. Scully (2008) challenges the conventional approach of bioethics because it does not consider that those living with an impaired embodiment may contribute to the moral

debates of issues relating to disabilities. This chapter is based not on the views of people with DMD but the views of their mothers, sisters and the partners of these women. Their decisions are based their intimate knowledge of the impairment of a son or brother. The decisions cannot be separated from the embodied experiences of DMD that couples have been a part of. The impaired embodiment of DMD does not contribute to these decisions; it is the foundation of these decisions. There were two factors of overwhelming significance – the desire to have a child biologically connected to both parents and the parents' experiences of the effects of DMD on their close relative.

Finally this chapter can be understood as examining a range of experiences, values and services that impact on SRTs which in turn influences the number of children born with DMD in New Zealand. The factors considered here, therefore, influence a local biology of DMD. Different couples had different experiences of DMD. Some couples were deciding on the use of SRTs while a diagnosed son was young and they did not have the lived experience of the end stages of the condition. Other couples were very familiar with how adult life would be for a child with DMD. In addition to this experiential understanding, couples also had their own personal values about testing and termination and were influenced by wider social values. Fitzgerald et al's (2015) discussion about choice is an example of the social values that impinge on the environment in which people make decisions regarding SRTs. The availability and delivery of services is another significant factor. While PGD is available to families where the mother knows she is a carrier of DMD, this is not necessarily something carrier women were aware of. Megan and Dave were unaware of the legislative changes that occurred as they investigated their options and I heard of other sisters who did not know that PGD was even possible, let alone available to them. Awareness of PGD is likely to increase as more women like Tracy share their stories. For example, in a recent MDA *In Touch* magazine there was another PGD success story told by a family with a history of a different neuromuscular condition – not DMD. The services of genetic counsellors and abortion counsellors also have roles to play in shaping people's understandings of their options. The way services are provided in important. Genetic material is no longer routinely sent overseas for testing, so timeframes while still difficult have improved since Tracy had her first CVS test.

These factors all combine to create a particular local situation for the provision and uptake or refusal of SRTs in relation to DMD in New Zealand. In some ways these are all quite measurable and assessable aspects of decision making. What is not so easily measurable is

the grief, suffering and hope that are entwined in the decisions that couples make. For Tracy the grief associated with her brother's suffering was so severe that Tracy was determined not to have a child with DMD, despite the significant cost to herself. For her family PGD offered hope, an antidote to their grief. Other families decided against SRTs – accepting children conceived naturally and rejecting CVS or amniocentesis. For these families testing or terminating a pregnancy because the foetus was like the child they already had was wrong. Both in cases where SRT was used and where it was not used, this research indicates that those with DMD were central to the decision making process and were highly valued by their families.

Section 2 Conclusion: Valuing Lives Lived with DMD

In this second section of my thesis I have considered some of the ways in which lives lived with DMD are valued. Central to this reflection has been the complex way in which suffering and hope are intertwined, somewhat like an image of a double helix of DNA. "A DNA molecule consists of two strands that wind around each other like a twisted ladder" (Gene Ed 2012). In this metaphor, see Figure 2, the backbone of each strand is made either of hope or pain. The bonds that hold the strands together are: the biomedical treatments and technologies that Fassin (2012) refers to as the biological "value of life"; the complexities of systems-based services provided by the state, Fassin's (2012) political "worth of lives"; and Lock and Nguyen's (2010) local biologies; and the bonds of family and community support.
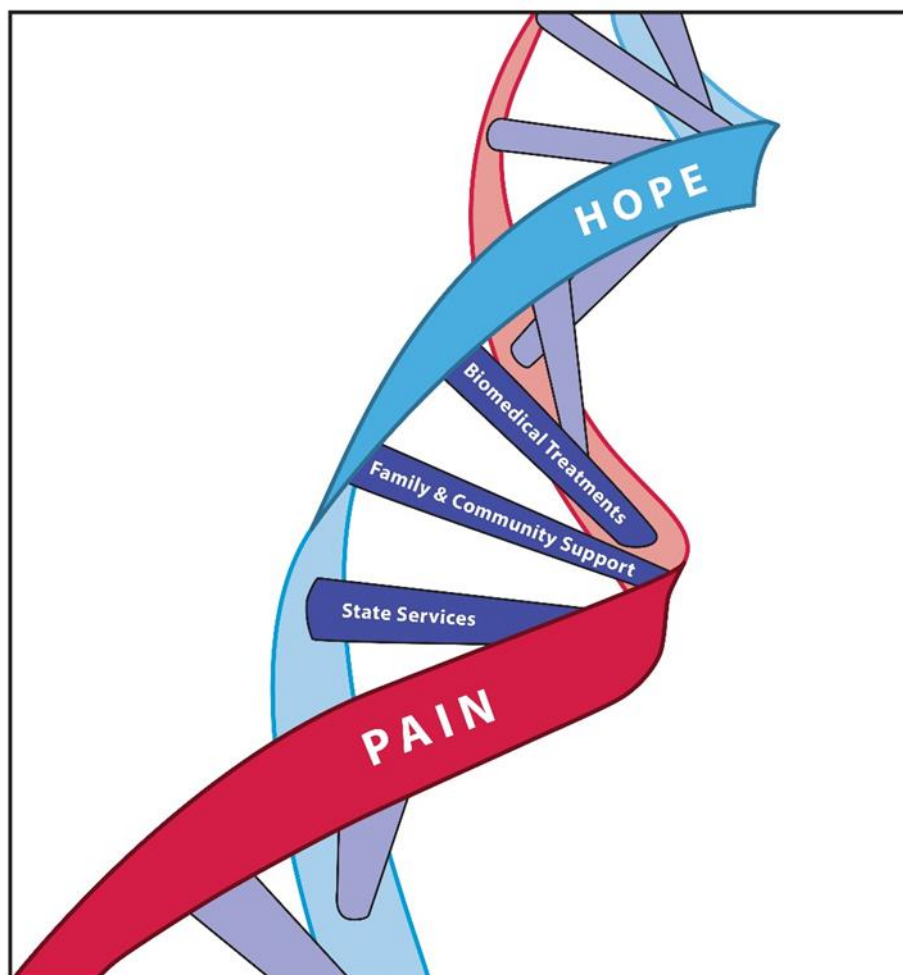

Figure 2. The double helix of hope and pain.

Hope is essential, without hope those with DMD face inevitable depression and despair. And people do live with hope, hope that each day will be a good day that the activities which people want to engage in will be accessible, both physically and in other ways; ways that are

to do with social inclusion, people's attitudes and prejudices. In framing hope I have addressed theoretical developments in anthropological research. The ideas developed by Appadurai (2004) and Crapanzano (2004) that hope is an aspect of culture that has long been over-looked by anthropology is pertinent to my research. I could have written a thesis that focussed on social suffering, the cultural dominance of biomedicine and biotechnologies, and the difficulties of bureaucracies and audit culture with no mention of hope. Such a thesis would have been an accurate analysis of much of my data but it would not have been a full account of my research. Addressing hope has been a challenge because, as Hage (2003) and Franklin and Roberts (2006) note it is an amorphous concept, difficult to define. However, in considering hope in relation to this particular community I ponder whether it is not only central to this ethnography but perhaps even a fundamental aspect of humanity.

Pain and suffering are also unavoidable parts of the DMD experience. I argue that this is due not only to the genetic reality of the condition but also to the particular local environments that shape and inform the individual experience of condition. The inevitable muscle degeneration caused by the lack of dystrophin is a shared arena of hope and action. Genetic research and the development of pharmaceutical treatments is the site where a range of experts share a common goal. The cultural ascendency of the biomedical paradigm is widely accepted by this particular community. It is through the successful development of biomedical treatments for DMD that pain and suffering may be alleviated.

The support of the biomedical paradigm is part of a wider cultural phenomenon as noted by Fassin (2012) in his analysis of the biological "value of life". The wider cultural ascendancy of the biomedical paradigm offers hope to the individuals and families directly affected by DMD and to those that support them – the MDA and medical specialists (for example). The international research and the strategies that the MDA and associated New Zealand specialists have implemented to support this research, demonstrates that a network of international biomedical experts, patient support groups and other allies value lives lived with DMD. The pain that is caused by complex service provision and the reluctance to fund person-centred care (which is considered in detail in Section Three of this thesis) shows that this value is not universal. Service provision that affects the suffering of individuals and family members is a component of local biologies of DMD. In some cases services relieve suffering (for example the counselling service that was mentioned by Anita, and Wendy's knowledge about how physiotherapy could benefit Zane) but, as I discuss in the following section of this thesis, 'Bureaucratic Entanglements', the ways in which education, health and

community services are provided can increase distress because they diminish the "worth of life" with DMD. This increase in distress is an indication that lives lived with serious genetic conditions are caught in the crossfire of ideological frames. Service delivery is a site of contradiction where the values and discourses of fiscal responsibility and fixed budgets tend to take precedence over values and discourses around human rights and social inclusion.

The lack of value that seems to be associated with service delivery contrasts completely with the way those families that participated in this research valued their sons and brothers. Within a family context the person living with DMD was often surrounded by a close and loving kin group. The way that families describe caring for a person with DMD and the way that young men with DMD describe their bonds with their parents, grandparents, siblings and cousins demonstrated how the exceptional circumstances of DMD sometimes created exceptionally devoted families. In some cases the requirements of care overwhelmed mothers (primarily it was mothers who acknowledged these difficulties) but the sense of devotion remained. This finding challenges the common depiction of family members with disabilities as burdensome. McLaughlin (2012:402) notes, "This narrow approach has been criticized by the disability movement as yet another version of disability as tragedy". Families that participated in this research did not view the person with DMD as a burden but as an individual who was sometimes happy, upbeat, funny, sometimes cross, angry, fed up, sometimes focussed and busy. The disability may have changed roles and dynamics within the family but the person with the disability was just an ordinary person with all the everyday ups and downs of life. Similarly the parents had the same ups and downs, times of stress and grumpiness, times of laughter and happiness, times of getting on with work and times of being tired. Research participants were neither heroes nor victims but they were closely connected, interdependent, they understood each other's needs with a great degree of familiarity. Families made arrangements for ensuring that the needs of the person with DMD were met in various ways but underscoring the arrangements was an appreciation for individual personhood. Quirky behaviour was an in-family joke, achievements were celebrated, grumpiness was met with eye rolls, these were very ordinary families who loved each other. Family members with disabilities were "full and valued participants in family life" (McLaughlin 2012:409). This devotion was the primary motivating factor in the decisions that couples made about selective reproductive technologies. The ways that couples made decisions always centred around the experiences of a much loved family member (be that a brother, or a son, or both). This way of valuing life is not directly addressed by Fassin (2012) in his discussion of a dichotomised

biological / political analysis but is fundamental to a comprehensive understanding of how lives lived with DMD are valued.

The complex relationships between hope and pain, between biological, political and familial values regarding lives of people with DMD are beautifully captured by the words of Raymond Mok, a young New Zealand man with DMD, who recently wrote the following piece for Duchenne awareness day, 7th September 2016. This blog post is reproduced with his permission.

> *World Duchenne Awareness Day 2016: victim impact statement edition*
>
> *Today is World Duchenne Awareness Day. I want to share with you what it's like to live with Duchenne muscular dystrophy (DMD). I say "what it's like", not "what it is", because people with this condition are diverse. Living with DMD can be very different depending on the society the person lives in, the resources accessible, the severity and stage of their condition, and certainly their aspirations.*
>
> *I live with my enemy. It's inside of me. It has helped me learn to see a different set of scenery, to appreciate life, and to feel others' joys and pain. DMD is my enemy because it's causing harm to my body and it brings me pain and discomfort. It has contributed to my back pain and it makes breathing difficult. I'm literally suffering from it although it doesn't always feel that way. It's frustrating not being able to do things but I don't see that as suffering.*
>
> *The purpose of this post is not to moan, but to raise awareness. So here is what I would say to DMD if it is a person:*
>
> *I hate you because you took my brothers-in-arms, but I tolerate you because I got stronger despite your constant debilitation. I have even accepted you, but I have to tell you that it's not resignation. I was once depressed because of you, but fortunately I eventually managed to find a way to stop you hurting my spirit. You can reduce my physicality, but you can't kill me spiritually. I am fighting you. I may seem to be fighting a losing battle. However, with the love from my God, my family, my friends and my comrades of common cause, I can and I will fight you. There are days when the thought of giving up on life comes to my mind. But I simply cannot give up because it's just not my nature.*

I wrote a poem for you:

Hey there bloody monster,

Your initials are a palindrome.

Will you ever slow down

When you introduce the symptoms?

You make me,

You break me,

You drive me crazy.

I'm stronger,

I'm better,

I'm growing weaker.

You took my ability to run, then to walk, and then to raise my arm. It's getting harder to breathe, to eat and even to do shit. Nowadays I get a headache if I'm off my ventilator for a few hours. However, I must tell you that I will never give up.

I love my family. I want to be there for them. I don't want my parents to farewell me. I'd rather farewell them. I don't want to break their hearts, even though it's not under my control. I do the best I can to live well.

There was a time when I was very worried about my future and I was too focused on myself. But then I found some new purposes in my life. I wonder if you know that you reduced my selfishness. I live with the hope that God be pleased with me. I live to share my story with the hope that somebody will be encouraged just as others have encouraged me.

Raymond Mok (Mok 2016)

SECTION 3

BUREAUCRATIC ENTANGLEMENTS

Section 3 Introduction

This section of my thesis is a complex micro-level study of the diverse bureaucracies that individuals and families with DMD engage in. I adopt a Latourian analysis to reveal the inequalities and marginalisation that people experiencing DMD live with on a daily basis. The complex networks of care I discuss here are entangled in the structure of the health system and the quasi business model of service provision. The repeated exclusions that families experience reveal a mismatch between the vision of service provision and the financial constraints imposed upon these services. This ethnography demonstrates that only people with the condition and their immediate family can be fully aware of the social exclusion people with DMD experience. My hope is that this ethnography provides glimpses of this experience; but it will inevitably fail to convey the second by second, minute by minute, daily, weekly and yearly grind.

People with very high levels of disability repeatedly fail to receive adequate levels of service but due to the fragmented nature of service provision providers are not aware of this. Each service provider is only aware of the issues in their own field of expertise, whereas families are holding multiple tigers by their tails. Each actor brings a certain epistemology, a way of knowing, a form of expertise to the assemblage. Here I consider the experiential knowledge of families, the professional and skill-based knowledge of service providers and the governing and management knowledge of those administering key resources. These are contested epistemologies but those in governing and management roles who control resources have a particular authority. The way in which this authority is wielded has serious implications for those living with DMD and offers a clear message about the political worth of lives lived with DMD (Fassin 2012:112).

In order to illustrate the compounding problems that interacting with bureaucratic systems creates in the lives of DMD families I discuss three examples that arose in one interview. The family in question, Harry who had DMD, his primary carer, Margaret, and his grandfather, Bruce, were dealing with various issues and their associated bureaucracies at the same time. This family was not unique in this respect. All families had similar stories of dealing with several bureaucracies at one time, as the following comments from another couple, Olivia and Dean, parents of four young children, one of whom had DMD, make clear.

Olivia   I find it frustrating even with our family, getting them to understand what day to day living is like. You are giving them daily medicine and ringing people and

filling in forms and trying to stay on top of things as well as managing family life. I find it very, very hard and getting people to understand that a little bit more. The attitude I get is "well, what is so hard about your life?" People just assume everything is fine.

Dean   So Dad might pop round after work and the house will be a mess and he'll say, "So what have you done today?" And she will have sat at the computer all day to do this, this and this. We had to get a computer because Olivia needs it to run Stan's life.

Olivia   I'd be lost without it

I have used the issues that Harry's family faced because they discussed underpinning rationales as well as service provision per se. The under-pinning rationales they were familiar with included an acknowledgment and even an acceptance of the fiscal focus of the Ministry of Health and a familiarity with disability rights and the implications of the United Nations Convention on the Rights of People with Disabilites. They attempted to follow pathways of appeals and lobbying that some other informants just found impenetrable. There were many, many other bureaucratic frustrations that were mentioned during my fieldwork which are not analysed or addressed in detail here. The following examples merely give a glimpse of the numerous sites where families encountered frustrations.

The family whose experiences of diagnosis begins Section Two, Richard, Laura and their son Zane, had discovered in their expert physiotherapist, Wendy, a committed ally, someone who understood their situation and whose knowledge offered them hope. She was employed by their local DHB. Like all children with Duchenne muscular dystrophy, when Zane started school he was eligible for Ongoing Resourcing Scheme (ORS) funding. The Ongoing Resourcing Scheme provides funding so that students with the highest levels of special needs can learn alongside their peers. It is through this source of money, supplied by the Ministry of Education, that children with DMD receive funding for things like teacher aide assistance and any modifications to the school environment that may be required such as ramps, lifts and accessible toilets. Once a child is under the ORS funding scheme their physiotherapy and occupational therapy will also be provided through the Ministry of Education by Special Education staff. While Zane would eventually require the assistance of a teacher aide and modifications to the school environment, his parents were upset to learn that by applying for ORS funding they would no longer be eligible to receive physiotherapy treatment from

Wendy. Richard and Laura were dismayed to discover that the one DMD expert they had found in their local area would no longer be able to treat Zane.

Families that required their homes to be modified for wheelchair access and the installation of a special bathroom also encountered frustrations with the bureaucracies they had to engage with. The solutions that were "permitted" were not necessarily the ones that worked best for the entire family. Even in cases where the families' solutions cost the same amount, or where families were able to pay the additional costs, they found that they had to accept the "authorised" solution or receive no assistance at all.

One family was informed by the agency that provided their support workers that the physical transfer of a young man with DMD was a health and safety issue when only one support worker was in attendance. The agency made a decision to provide two support workers half as often without proper consultation or review of the allocated support worker hours. The decision had a major impact on the family, not just on the way they managed and organised daily activities but also their sense of embattlement and vulnerability.

Bureaucracies within institutions were also sites of frustrations. Healthcare in particular was repeatedly noted as very difficult. Due to the complex nature of the condition people have many appointments with many different departments. For children the lack of co-ordination means frequent absences from school. For older patients the lack of co-ordination means that recommended routine monitoring often does not occur in a timely fashion, interventions may be missed, which in turn may affect life expectancy. My research provides some accounts of this and the results of the MD Prev study demonstrate similar findings. This study was undertaken by Auckland University of Technology National Institute for Stroke and Applied Neurosciences in conjunction with the MDA. Data collection was completed in July 2016 and publications are forthcoming.

Some families had problems within the education system, although generally there did seem to be a more co-ordinated approach in the schooling sector. The commonly mentioned Individual Education Plan (IEP) meetings, for example, demonstrate a systemic awareness that communication and co-ordination are important. These are regular meetings where those involved in the education of the child meet together to ensure that there is a suitable plan to address the child's particular requirements. However, some parents noted that sometimes the needs of the school seemed to be prioritised over the needs of the child. For example, one family was annoyed that the part-time teacher aide was not available when their son had

science, as this was when he needed help with practical experiments. However, the aide was available when their son needed less help, for example during English or Maths, when he could manage well once his laptop was in place.

These are just a few of the bureaucratic frustrations that families mentioned. As well as problems like these, caused by impersonal, institutional procedures there were also examples of frustrations caused not by policies but by people. Often this took the form of bullying or exploitation of a participant's vulnerabilities; support workers that threatened violence or stole property or money, teaching staff who resorted to turning off powerchairs for disciplinary reasons and in one case then forgot and left a child sitting in the playground at the end of playtime, a child receiving a detention for not buying a pencil from the school stationary shop even though the shop was not accessible, schools where parents felt unwelcome and as if the principal wanted them to enrol elsewhere. Although these experiences added to an environment of general frustration they were not mandated behaviours, and while they indicate an unpleasant and widespread culture of bullying in New Zealand (see also Balanovic 2016), such behaviours are not socially endorsed or accepted. Bureaucracies are socially endorsed and culturally accepted as appropriate ways to manage society (Wedel et al. 2005:34).

This consideration of the impact of repeated experiences of dealing with bureaucracies is divided into three chapters. The first chapter, Chapter Eight, examines the provision of cough assist machines. Based on an actor-network analysis of the various actors and their motivations I offer a biopower analysis of the contested environment within which the provision of these life-saving devices are negotiated. The second chapter, Chapter Nine, looks at the provision of wheelchairs and support workers. In contrast to cough assist machines, the provision of these services is covered by existing Ministry of Health contracts and policies therefore I use a governmentality approach and an anthropology of policy approach, following the work of Shore, Wright and Pelò (2011) and Wedel et al. (2005), to examine the way exclusions are mandated and challenged. Chapter Ten examines the rigid funding regimes that determine how people with DMD are cared for when they are admitted to hospital. The ethnographic information that Chapter Ten is based on does not come from the interview that I refer to for the other three issues (cough assist machines, wheelchair allocation and the provision of support workers). This chapter reinforces the point that policies and bureaucratic decisions are more than simply frustrating and devaluing for people living DMD. The rigid funding regimes enforced by Ministry of Health discussed in this

chapter are life-threatening. The socially endorsed and culturally accepted policies and bureaucracies considered in this section of my thesis can be read as a gauge for the way lives lived with DMD are valued in New Zealand society. These aspects of health – these policies and bureaucracies which are as much technologies of health as blood tests, pharmaceutical medications and vaccination programmes (Lock and Nguyen 2010:362) – expose the evaluation of the worth of lives, "the practical ways the in which people's lives are considered, protected and cared for, or conversely abused and sometimes eliminated" (Fassin 2012:111). My analysis is grounded in the experiences of Harry and his family.

This is an interview that I drove to a provincial town to conduct. I left Auckland in the rain and travelled through some torrential downpours on my journey, the kind when it rains so hard the windscreen wipers cannot keep up. In typical New Zealand fashion by the time I reached my destination a hot sun was starting to break through the clouds and the pavements were steam drying.

Harry did not live in this provincial town but had travelled from his home for an appointment at the hospital. I met Harry, his support worker, Margaret, and his grandfather, Bruce, who had all made the two-hour trip for the appointment. Their van was old and rickety and made a lot of noise. They had had an early start from home, rattling and bouncing down the highway in the van. Harry was fixed into the back of the van in his unsatisfactory wheelchair with its poor suspension. His back pain was noticeable by the time they arrived at the café. Bruce was in his seventies. It seemed like a long day for an older man.

I met the family at a café after their appointment. It was one they knew well as they often visited it when they came down for their various appointments. I refer to the group of people I met at the cafe as a family. They presented themselves as a kind of family unit even though Margaret was a paid carer; they were united in their commitment to battle for what Harry needed. Before meeting me at the café they had been to the hospital for an appointment at the physiotherapy department to learn about the cough assist machine that had been gifted to the hospital the previous year.

Harry was quite reserved to start with but Margaret made up for that with her enthusiastic story-telling and Bruce was considered and thoughtful with his contributions. It was a relaxed, free-flowing conversation, which covered many topics; here I highlight those parts of the discussion that related to their experiences with bureaucracies: the acquisition of a cough

assist machine, the wheelchair bureaucracy and the bureaucracy related to the provision of support workers.

Chapter 8: Acquisition of a Cough Assist Machine

Following Foucault (1978:138-145) and Rabinow and Rose (2006:197) I adopt a biopower approach to examine the difficulties Harry faced in accessing a cough assist machine. I examine the "strategies and contestations" (Rabinow and Rose 2006:197) that different organisations and people employed in debates about the provision of these machines. Despite the three years that have passed since I first interviewed Harry, the provision of these machines is still not guaranteed and other individuals and families continue to engage in similar struggles to the one outlined here. As noted in the introduction the security measures of biopower (mechanisms that offer protection from accidents and anomalies) are designed to optimise an average state of life, not an individual life. The mechanisms of biopolitics that are established to address a general risk of physical anomaly, variations within the population, a chance or random event, are not designed to meet the individual needs of people with DMD. This means that the DMD community has to negotiate, strategise and contest regimes of authority and forms of knowledge in order to acquire these life-saving machines.

Cough assist machines stimulate a natural cough and are valuable in treating colds and reducing both the incidence and the severity of chest infections. (Phillips et al. 2014). As the muscles of the thorax weaken, coughing to clear mucus becomes more and more difficult. If mucus sits in the lungs, the risk of pneumonia increases. Cough assist machines are portable electrical devices that use a mask or mouthpiece to "puff" air into the lungs. The pressure of the puff of air can be controlled by the settings on the machine. In order to use the correct setting, so the puff of air is not too forceful (causing damage to the lungs) or too weak (is not effective in creating a mucus-clearing cough), the patient needs to have their lung function assessed by a trained professional. After emitting the puff of air that is breathed into the lungs the cough assist machine quickly reverts to a negative pressure causing the air to leave the lungs which creates a cough. Again, the setting is adjusted to control the force of the negative pressure and hence the speed with which the air leaves the lungs. Cough assist machines are highly sought after by people with DMD but there is no nationwide guarantee of access. Some families have their own machines, some families borrow machines from the MDA, some families borrow machines from their DHB. Others have to use the DHB machine in the hospital. These machines are life-saving and are expensive, Phillips et al. (2014) indicate a cost of $12,000 per machine. The reason that provision of these machines is inconsistent is

partly to do with the regional structure of the New Zealand health system. Each DHB is responsible for the allocation of its own budget. The Ministry of Health negotiates services that the budget must cover and these are agreed to in an annual plan and are regulated through legislation (the New Zealand Public Health and Disability Act 2000, the Health Act 1956, and the Crown Entities Act 2004 and various health and disability strategies). However, these requirements are not directive about the provision of particular technologies. The inequitable provision of cough assist machines is also demonstrative of technological "creep". This commonly happens when sophisticated and expensive items become increasingly routine, and exceptional or trial purchases gradually become part of standard care practices.

Harry and his Cough Assist Machine

At the time of our interview, Harry did not have his own machine and his DHB did not have machines available for loan. The DHB did have a machine that had been given to the hospital as the result of a fundraising initiative undertaken by a local community who had a child recently diagnosed with DMD. This DHB would not lend out its machines. If he needed to use it, Harry would have to be admitted to the hospital which was about two hours away from his home. Harry and his support team were developing strategies to find ways that he could use a machine at home in the hope that any cold he caught would not progress to a severe chest infection requiring hospitalisation and all the complications that came with such an admission (see Chapter Ten). They were in communication with a cough assist machine agent (whose son also had DMD) and who was keen to offer the machines at the cheapest possible price.

The following interview data is included because it demonstrates how familiar the family is with the financial rationales around the provision of equipment.

Margaret   The fiasco with the cough assist machine is another thing we have been involved in which is what we have finally, after a year, got to see at the hospital here… But we can't have any kind of use at home. You can only be hospitalised with it. You have to be in hospital to use it.

Harry   And the whole point of having it is to avoid going into hospital. Because a couple of nights in hospital is the same price as one of these machines.

Bruce   Unfortunately they are very expensive. It was quite interesting today to see one for the first time.

Harry   They are about $10,000. But two nights in intensive care is about the same, anyway. So it is a cost savings thing.

Margaret Anyway we have got this guy coming up to see us on the 24th May. He sells them privately. His son has got DMD. He went through all this with the cough assist, so in the end decided to buy his own and that is how come he recommends it, not from a salesperson point of view but because it does actually work. And they have got the two system. Harry has already got the machine at night for the breathing, the C- pap, for breathing, but he (the agent) was saying that he has got one that does both. The funding to get something like that is quite difficult because it is a medical piece of equipment so they'll say Ministry should fund it.

Harry   They should fund it.

Margaret But they don't want to. But it does not make sense because the machine that Harry has got is about the same. They fund that (C-pap) but they won't fund this (cough assist and C-pap combined).

Bruce   Well it is a wee bit more.

Margaret Well we were told that when we got that one from Auckland hospital. A nurse basically said, "Look after it, it is worth about $9,000". And when I spoke to the agent I said, "Can you give me a ball park figure so we have some idea of what we are looking at?" And he said, "Not more than $10,000." So they are about the same price and they are doing the job of two machines.

Kate   So have you found a way of getting funding for that?

Harry   Not yet. Because obviously we need to make sure it works first

Kate   And then are you going to look for ways to get funding?

Margaret Yes, but it is going to be a mission because, as I say, it is a medical piece of equipment that they don't like funding. Apparently in the United States they recognise how important they are and they are just part of the standard care.

Kate   And does it require a certain amount of training to use it, is that right?

Margaret   I think once you have got the settings right.

Bruce   One of the things I took from today's meeting was that when you use the machine for the first time the inward pressure for the lungs needs to be monitored very carefully. It is not just a case of put it on and away you go.

Margaret   Because Harry's lungs are obviously not used to being fully expanded. So when they are being assisted by a machine for the first time we have to be really careful. But it settles though.

This dialogue shows that family have undertaken their own research regarding cough assist machines. They know that they are provided as part of routine care in other countries, they mention the United Sates in particular. The health system in the United States is funded differently, often through insurance. The issue for New Zealand families is about gaining public funding for these machines. The efficacy of the machines is not in doubt, they are well recognised as life preserving for people with DMD (Muscular Dystrophy UK 2015). The family are also familiar with the arguments of health economists and offer their own analysis to demonstrate that providing these machines would save money. Harry assumes that if he is unable to access one of these machines at home he would probably end up in hospital and likely be admitted to an intensive care ward. This assumption is based on a clear biomedical knowledge of his condition. It is a highly probable outcome (Muscular Dystrophy UK 2015). Harry's statement that providing one of these machines would cost about the same as two nights in intensive care is also accurate (Phillips et al. 2014). He has clearly undertaken some research to know this. Similarly Margaret is also correct in her comparison of the cost of the machine that has already been provided – the C-Pap machine – and the machine that better meets the needs of Harry – the machine that provides C-Pap and cough assist functions. A stand-alone cough assist machine is an expensive piece of equipment but the dual function machine is not hugely more expensive than the ventilator which is currently part of standard care in New Zealand. Furthermore, expensive equipment like these machines are re-used. If the machine that includes both cough assist and C-pap were provided to Harry, he would return the C-Pap machine which could easily be serviced for another patient whose needs were better suited to it.

To acquire a machine exclusively for Harry through the agent would mean applying for grants and/or fundraising. Having now worked as the MDA Grants Fundraiser I can concur with the concerns of the family that they would find it difficult to access funding, primarily because philanthropic trusts will not accept individual or personal applications; applications need to be made by a registered charity.

Harry felt it was ridiculous for the DHB not to allow him to use the cough assist at home, as one of the advantages of the machine was to avoid the unnecessary complications of a hospital admission (see Chapter Ten). I later discovered that the decision to keep the machine at the hospital and not permit patients to use it at home had been made by the MDA regional branch. The branch owned the machine; it had been gifted to them following a fund-raising initiative by a small community who had recently received the news that a child in their community had been diagnosed with DMD. The regional branch entered into an agreement with the DHB to manage the machine and the requirement that the machine stay in the hospital was included in this agreement. I was unable to discover why this was one of the criteria of the agreement.

Harry's interview demonstrates the particular knowledge that resides with people who have DMD and their families. Harry and his supporters are clearly expert patients. They know that Harry, who was 21 at the time of this interview, will be experiencing increasing incidents of respiratory problems. DMD is a predictable and progressive condition and respiratory problems are a serious part of it. They also know that this piece of equipment reduces the number of respiratory issues that develop into pneumonia and require treatment in the intensive care ward of the hospital. Harry, Margaret and Bruce do not mention the personal pain and suffering that is associated with pneumonia. The trigger for their quest to obtain a cough assist machine had been Harry's hospitalisation with a chest infection, so Harry clearly knows how distressing this is. But he does not focus on this at all in the discussion. He and Margaret and Bruce use the arguments that they believe will be most effective – those of cost benefit and health economics. As well as being expert patients in the medical sense, they are also well versed in arguments that are used to provide or withhold services. They are experts in leveraging proper patient care. This reflects their numerous experiences with various service providers.

The Cough Assist Machine Assemblage

A number of distinct groups were involved in Harry's cough assist machine network and also in the wider network concerning equitable, nationwide provision. The groups or assemblage that were relevant to Harry's situation were; Harry and those most immediately concerned with his well-being (his family and Margaret), the MDA regional branch, MDA National Office, Harry's DHB (and particularly that part of the DHB responsible for respiratory care), the agent who sold the machines and more tangentially the community group involved in the initial fund raising. Similar groupings or assemblages are also involved in the wider network involved in ensuring nationwide access. Again those with lived experience (the individual with the condition and his immediate support network whose concerns are primarily the well-being of that particular individual), the patient support groups at both local and national level, the DHBs, the equipment suppliers, and importantly also, those ultimately responsible for providing the funds for the health system and negotiating the annual and longer term plans with the DHBs – the Ministry of Health policy advisors and contract and relationship managers. Central to these assemblages is the interactions between these human actors and the technology, the cough assist machines.

In these interactions, aspects of Latour's actor-network theory (ANT) are relevant. Cough assist machines are actors in the assemblage, they are the catalyst for the relationships and the negotiations that ensued, "any thing that does modify a state of affairs by making a difference is an actor" (Latour 2005:71). The machines are responsible for making a difference in other actors' courses of action; policies, ministers, DHBs, budgets and bureaucrats. As such they become participants in this assemblage, in this case they are "at the origin of social activity" (Latour 2005:72). By following the interactions of this particular assemblage I use Latour's ideas to uncover what actors do and how and why they do it (1999:19). This assemblage demonstrates the way that relationships within networks effect change. For each action within in the cough assist machine assemblage there was a change in the relationship between the participants. Latour argues (1999:17) that the strength of ANT is that it changes the focus of social analysis, explanation and understanding from the unsatisfactory agency/structure, micro/macro dichotomy prevalent in earlier methodological approaches towards a consideration of circulation and movement. People understand why they act as they do, by focusing on these actions, rather than moulding them into pre-existing theoretical frames, ANT tries to avoid imposing pre-existing sociological ideas onto the data. The network aspect of ANT refers to "the summing up of interactions … into a very local, very practical,

very tiny locus" (1999:17). "When one explores the structures of the social one is not led away from the local sites … but closer to them" (1999:18). The point, Latour says, is to "learn from actors without imposing on them an a priori definition of their world-building capacities" (1999:20).

The push from the various individuals who would benefit from the provision of this technology causes both the regional and national support group to examine and understand the importance of the machines. Individuals often know about this technology as a result of their connections to international muscular dystrophy support groups and through online friendships with others with the same condition living overseas – giving the assemblage an international connection. Prior to the development of cough assist machines people were assisted to cough manually by a carer compressing their chest to push the secretions out. The technique can be taught by a respiratory physiotherapist. This procedure is uncomfortable and as lung function decreases it is not as effective as the use of a cough assist machine (Philips et al. 2014:130, and Gomez-Merino and Bach 2002). As they continued to have recourse to manual (rather than mechanical) cough assistance, the DHBs appeared reluctant to fund cough assist machines, so initially the support groups covered this gap in service provision and in doing so met the needs of the members. The MDA and local branches, as registered charities, are able to source funding from avenues that are not available to individuals. However once some cough assist machines were sourced and provided to members for their exclusive use the MDA identified risks connected with their actions (risk analysis is part of the managerial approach – another regime of authority). Firstly there was the potential criticism, based on the value of fairness, that some members were advantaged over others, as not all those who would benefit received a machine. This led to the first change in MDA policy regarding the provision of the machines. Rather than providing a machine for the exclusive use of a member the machines were kept by the MDA and lent out to members who needed them – that is, lent out on a short term basis, when a member who had difficulty coughing caught a cold. They were no longer permanently given to members to keep at home. The rationale was that this policy better addressed the equity concerns. All members would be able to borrow a machine, should the need arise. Secondly, the machines required maintenance and servicing and some parts needed to be replaced. The MDA was better able to monitor this when they kept the machines in a pool and had direct contact with them, rather than when they were kept in the homes of members. Thirdly the setting of the machine needed to be supervised by a physiotherapist. An accurate measure of the lung function of

each individual user was important to correctly adjust the setting at which the machine would be used. This meant that the MDA and the user needed to develop relationships with respiratory physiotherapists at the local DHB. The MDA changed its policy in a strategic way to manage the risks associated with providing essential medical equipment to members. However this change led to a new range of problems.

Partly because of the success of cough assist machines and as a result of other improvements in the care of young men with DMD, average life expectancy is increasing and this in turn increased demand for the machines. On occasion there were not enough cough assist machines to meet demand. This caused re-positioning within the network. The staff member at the MDA who was responsible for the distribution of the machines (and this role was filled by various people during my research) was really being asked to make a clinical decision for which he was not trained – he did not know who had the greater need of the available machines and his distress at being asked to make such vital decisions challenged the new policy. A messy situation thus existed with some MDA members having machines that they had been given to keep at home for their exclusive use, and some members relying on pool machines, and some being able to use a machine provided by the regional branch either in the hospital or to be loaned for home use from the hospital. At the time of my interview with Harry the National Service Leader at MDA was arguing that the providing cough assist machines was, "not our (MDA) core business." Increasingly the National Office staff and the National Council saw the provision of cough assist machines as the responsibility of the DHB.

To shift the provision of the machines from a small and deeply engaged support organisation to a large and somewhat disengaged DHB was not straight-forward. Clearly the MDA could not guarantee that the twenty DHBs in NZ would all purchase sufficient machines for all the people living in their region to have guaranteed access. The MDA needed to lobby on behalf of their members to get the DHBs to provide the machines. This lobbying would inevitably take place in a competitive environment. The finite allocation of money available to each DHB has many demands placed on it.

To further complicate matters, at the time that the MDA decided to focus their efforts on lobbying the DHBs to provide the machines instead of fundraising for the machines themselves, some of the machines that had been given to members for their exclusive use were becoming obsolete. The MDA received information from the supplier that they were no

longer going to manufacture some of the essential tubing and that eventually the machines would need to be replaced.

So the MDA delivered two messages about the cough assist machines at the same time. One message explained the notification from the manufacturer that some existing machines would need to be replaced as parts wore out. The second message was that a strategic decision had been made to lobby the DHBs to provide the machines. The fallout from this double message was significant. Some families who had a machine on long-term loan, who thought that they had a guarantee of access, were extremely upset as they now thought they were about to lose their machine. Others pointed out that not only did they not have a machine on long-term loan but that their DHB did not have any machines either. Existing concerns about cough assist machines were escalated.

The anger that some families expressed when they thought they were going to lose their cough assist machines was a distinctive and embodied reaction. It was a raw anger that has a degree of power, resulting from the personal, lived, direct experience of receiving the message that your life, or the life of your child, does not matter.

These are families that have a long experience of not meeting assessment criteria or having to endure long and complicated bureaucratic battles to access services. They know that cough assist machines can make the difference between surviving a chest infection and not surviving it. They also know that the need can arise at any time. Many of these families knew that they may well need one long before the transition in provision that was being suggested would be implemented. Timeframes for changes to health care systems and timeframes for people living with Duchenne Muscular dystrophy are not compatible. The information that members received regarding the change in provision caused an immediate angry reaction.

A heated Facebook exchange illustrated the way relationships within the network facilitate change (Latour 1999). Members were angry that the organisation that was supposed to represent them seemed to be repeating the message that they often heard elsewhere, that their needs were too hard, too complicated and too expensive. The exchange demonstrated the way some people living with DMD resort to anger as a form of power to achieve a result. Several mothers I interviewed talked about how they resort to anger to access essential services. One described how she could feel anger taking over at times when she was faced with an immovable, overwhelming bureaucratic injustice. Some families have learnt, through repeated interactions with bureaucracies that getting angry can work. Usually this was

described as an immediate reaction, rather than a calculated choice of strategy. It reiterates that these parents are not 'ideal' or compliant patients, they have learnt to 'talk back' to power. The personal toll of this repeated angry reaction is significant and some families talked about meeting other parents whose children had severe disabilities and not wanting to become angry and embittered like these parents.

However, the increasing demands from members (of which the angry reaction on social media was a part) caused the MDA to develop a clear lobbying strategy aimed at the Ministry of Health and the DHBs to increase the provision of the machines. The MDA employed someone on a short contract to produce an information package that could be used by respiratory departments in DHBs to assist them when requesting funding for a cough assist machine. This package summarised the research that demonstrated the effectiveness of the machines and included a cost benefit analysis related to the demands for intensive care beds. It included a letter of support from the MDA and used the membership database to calculate which members were most in need. It was a thorough and detailed analysis. The MDA wanted the fieldworkers to liaise with the respiratory physiotherapists in their areas to facilitate applications to DHBs for cough assist machines for members. The MDA was enacting biopolitics, it was challenging the regulatory mechanisms that were established to address the needs of the general population but which did not meet the need of people with serious neuromuscular disease. Its role as support group for people living with rare diseases meant it was in a difficult position facing conflicting expectations from members. It was expected to advocate for changes to the public health system so that this system better met the needs of its members (bearing in mind Foucault's point that such a system is designed for an average or overall benefit and not an individual benefit), as well, members expected the organisation to offer support to them as individuals, given that their needs repeatedly fell outside the parameters of the public, general system.

Actions of one part of the assemblage cause changes in other parts. The push from members to access machines led to an increasingly strategic approach being developed by the MDA. The development of a lobbying information package by the MDA in combination with the increasing familiarity with the machines in some New Zealand hospitals and increasing use of the machines internationally, may potentially increase DHB provision. The MDA has also been involved in discussions about cough assist machines with the Ministry of Health.

In May 2016 a meeting was held to discuss improvements to respiratory services. The catalyst for this meeting was the recommendations made by the Coroners Court in June 2012 in response to the death of a man with DMD. The man was "fully dependent on his Bipap machine and was unable to tolerate more than one or two minutes off the machine". A Bipap machine is a non-invasive ventilator, it is not the same as a cough assist machine. He died during an electrical power cut in the middle of the night. Although his Bipap machine had an alarm that was meant to alert the family that the machine needed to be plugged into a battery, they did not wake up until it was too late. The coroner recommended (among other points) that the DHB consider supplying NIV (non-invasive ventilation) machines with an inbuilt rechargeable battery for portable and home supply. The coroner forwarded the recommendations he made in light of this particular incident to the Ministry of Health, "to ensure that the recommendation is considered by all Health Boards". Four years later, in 2016, the Ministry organised a meeting to discuss this recommendation and other issues relating to patients with complex respiratory needs. The meeting was attended by an MDA staff member (Miriam), the Chairperson of the MDA National Council, neurological and respiratory specialists, specialist respiratory physiotherapists, staff from the motor neurone disease support group and a chief advisor and support person from the Ministry of Health. While the focus of the coroner's recommendations was the provision of a safer Bipap machines for home use, other issues were discussed.

Interestingly, despite the numerous conversations about cough assist machines to which I had been privy through my work at the MDA, in the course of many interviews, via debates on Facebook pages, through discussions at regional branch AGMs, at various workshops and conferences I had attended, at no point had any people with DMD raised concerns about other shortcomings regarding respiratory care. Members seemed to view cough assist machines as a "magic bullet" for respiratory care. So it was with great interest that I learned that the meeting organised by the Ministry of Health, following the Coroner's report, addressed a much wider concern with standards of respiratory care generally. As I completed a final draft of this chapter I rang Miriam to discover if there had been any concrete outcomes from the May meeting, wondering if there had been any developments regarding the provision of cough assist machines. So to hear that there had been progress in addressing other respiratory concerns but that cough assist machines, which were so highly valued by MDA members were not considered to be such a priority by those attending the meeting, was somewhat surprising. Miriam explained that a cough assist machine is just one tool in the toolbox of

respiratory care and that there are other tools that need to be developed in a more consistent manner. Miriam mentioned the inconsistent availability of skilled respiratory physiotherapists who can assess breathing problems and teach people particular techniques and breathing exercises to maintain lung function and delay the need for a cough assist machine. I was familiar with the literature (Finder et al. 2004) advocating these techniques but had not realised that people were not necessarily receiving these services and so not learning these techniques. The techniques are important because they help maintain sufficient lung function to keep a (normal and healthy) layer of mucus in the lungs moving so that this healthy mucus can protect the lungs from any inhaled particles. If lung function reduces, this mucus stagnates and the risk of chest infections increases. As lung function reduces there are other problems too such as headaches, tiredness, difficulties sleeping through the night and lack of concentration. So maintaining lung function is important and if it is maintained with these techniques there is less likelihood of chest infection. Official advice indicates that cough assist machines are helpful when a person is acutely unwell but they are a part of a much wider system of respiratory care. Because there was such heated debate about the cough assist machines within the DMD community, I had not appreciated that the wider system of respiratory care was just as ad hoc as the specific issue of cough assist machines.

On reflection the focus that families and individuals placed on accessing a cough assist machine, rather than on challenging the complexities of the general health system, makes sense. As the following chapters demonstrate negotiating complex bureaucracies is a familiar and wearying experience for families with DMD. Repeatedly people encounter rigid and seemingly impenetrable systems. Acquiring a cough assist machine, in contrast, gives the individual a guaranteed way to manage their respiratory health. Although the machines are designed for use during acute episodes several people and some advice (Philips et al. 2014, Chatwin 2008) described using them prophylactically to clear the stagnant mucus that accumulates as lung function deteriorates. They did this in order to reduce the chances of having a chest infection; they were not waiting until they had an acute chest infection to use the machine.

As a result of the Ministry of Health organised meeting a group of respiratory specialists and physiotherapists who understood the shortcomings of existing services identified the problems they were aware of. These were to do with some patients not receiving the monitoring they needed and not learning the techniques that would help them to maintain their lung function. While cough assist machines are recognised as being a valuable part of

respiratory care these specialists identified other services that people with neuromuscular weaknesses could benefit from. As a result of this meeting a research project to determine how well New Zealand services meet international respiratory standards of care (Finder et al. 2004) was initiated and respiratory physiotherapists from all three of Auckland's DHBs committed to improving the pathway for respiratory care in the greater Auckland region.

However the provision of cough assist machines is still not guaranteed and the competitive funding environment in which all health needs must be met within a finite and capped budget maintains a contested site for the provision of these machines. The cough assist machine assemblage was hotly contested because of the high value placed on the machines by people with DMD. While respiratory specialists may not see the machines as vital equipment people with DMD do, because cough assist machines give people control over their own health.

Analysis of the Cough Assist Machine Bureaucracy

Evident in this data about cough assist machines is the different perspectives and expertise that the different parts of the assemblage have. Those living with the condition demonstrate a sense of urgency and determination as they work through strategies to acquire a cough assist machine. They had a lived experience of being marginalised within the health system and were often sceptical about any genuine concern for neuromuscular conditions. This was a structural scepticism not related to their interactions with physicians and other health professionals who had expertise in their conditions and who frequently treated them with care and compassion.

The MDA was attempting to work strategically. It seemed to be moving from a concern with the needs of individuals to a focus on addressing structural issues. However, the language of the National Service Leader who argued that the provision of cough assist machines was not the organisation's core business was telling. This language indicated an acceptance of managerialism. Klikauer's (2013) analysis of managerialism argues that it has become a dominant ideology, a commonsense way of understanding, a Bourdieuian doxa, that has colonised many areas of modern capitalist societies, (Klikauer 2013:3). The discourse of a "core business" highlights the acceptance of a business model with inherent ideas about efficiency and a central focus on how best to allocate resources for maximum profit or benefit. As businesses grew from small operations in the eighteenth and nineteenth centuries into much larger and more complex phenomena, business management developed as a

'science' for controlling or managing these new large corporations. Gradually ideas about the effective management of businesses spread to other domains – in this case the charitable sector. Managerialism is "not about Rousseau's volonte generale (general will) of the people but about engineering-like approaches to societal problems that have been converted into technicalities" (Klikauer 2013:6). By co-opting a managerial paradigm the MDA was aligning itself with a 'commonsense way of understanding' that people living with DMD were familiar with. This was the rationale that was used by DHBs and other service providers not to meet (or fully meet) their needs. While the intention of the MDA was to address a shortcoming in the public health system, to change the system, to challenge the range of accidental incapacitations covered by the strategies of biopwer, so that publically funded DHBs would provide cough assist machines, the reaction of some members suggests that they felt further abandoned. Not only were they not properly cared for by their local hospital but now their own support group was relinquishing them to a system, which experience told them, did not necessarily prioritise their interests. The message that was received by the members who needed a cough assist machine was that the provision of this life-saving equipment was not considered to be an essential service (now referred to as a "core business") of the organisation whose raison d'être was to support them.

The respiratory specialists and physiotherapists who attended the Ministry of Health meeting following the Coroner's report were able to address the frustrations they had with a system which they knew failed to adequately meet the needs of some of their most vulnerable patients. They wanted to improve the provision of respiratory services nationally so that all patients whose breathing was compromised by neuromuscular weakness would receive care that met internationally recognised standards of care (Finder et al. 2004). The first step on this path was to research the current situation - how many people were receiving a comprehensive service and how many were not? The actual implementation of an improved system was clearly some way off, which is problematic for a person with DMD who is already losing lung function. Thus the determination demonstrated by Harry and other young men with DMD to find a way to get their own machine becomes understandable. Young men with DMD do not really have time for the typically ponderous paths of research, planning, consultation processes and budgeting that accompany improvements to systems of medical care. They are operating in a different, more urgent time frame.

The concerns and frustrations of the respiratory experts attending the Ministry's workshop on developing principles for providing respiratory care to patients with advanced progressive

neuromuscular disease resulted from their awareness that they were part of an inequitable system that had very real consequences for vulnerable patients. Respiratory care has a direct effect on longevity. The shortcomings in the provision of respiratory care for some people with DMD have biomedical impacts on their DMD. The effects of these inequalities are to produce different biological outcomes or different local biologies (Lock and Nguyen 2010:90).

Chapter Conclusion

The Ministry of Health and the DHBs have an interest in improving their services and health outcomes for the populations they serve, but this interest is always balanced against constrained budgets and fiscal responsibility. The timeframe of the meeting organised by the Ministry of Health, five and a half years after the death of a man whose ventilator could have better met his needs and four years after the coroner's recommendations, suggests a certain lack of urgency regarding respiratory care for this vulnerable group. While initial steps are being taken to establish a comprehensive respiratory care system, the complexities of the regional inequalities of New Zealand's health system, especially for those with rare conditions, are often difficult to overcome. The surety that a guaranteed cough assist machine provides contrasts to the uncertainty of such a system. Foucault's point that technologies of government, such as health services, are established to address a general risk and not a specific individual need explains the way the actors in this network talk past each other. The Ministry of Health and the DHBs are concerned with systems to provide general health benefits while Harry and others with DMD are concerned to find ways to get the equipment they need in time to use it. They know from experience that their needs repeatedly fall outside the boundaries of these general services. Harry's mission to acquire a cough assist machine occurred in an environment where there was a long history of bureaucratic systems not accommodating the needs of people with DMD, leaving families with a sense of exclusion and in this case the possibility of avoidable death. Harry's argument that providing the dual function machines for people to use at home was in fact a financial benefit to the DHB was valid but it still required the DHB to invest in new equipment, rather than to rely on existing treatments – the manually assisted cough facilitated by respiratory physiotherapists.

However, currently, for young men with DMD the repeated interactions with organisations whose over-riding concern seems to be caring for budgets rather than people (Fitzgerald

2004) create an ingrained sense that people with DMD are socially dismissed, their lives are not important, their lives are too expensive. The four-year gap between the Coroners Court report and the Ministry of Health meeting, the dearth of respiratory physiotherapists, the ad hoc 'system' of care, and the absence of 'fit for purpose' machines all convey this same devaluing story.

Chapter 9: Powerchair and Support Worker Bureaucracies

This chapter analyses the interactions between state mandated policies, which outline the government's strategies for the provision of healthcare for New Zealanders, and the decentralised, outsourced and fragmented bureaucracies involved in the implementation of these policies and their combined impacts on lives lived with DMD. As Foucault notes, part of the objective of a modern democracy is to provide health services for the benefit of the population. The purpose of these policies and the service providers governed by them is to address the issue of a general health risk and not a specific health need. The policies under consideration here are actors in networks of wheelchair provision and the provision of support workers.

The model of health services in New Zealand is based on the belief that the purchaser provider split, the decentralising of health care, the outsourcing and privatising of some services is the best way to get an efficient healthcare system that avoids unnecessary, wasteful financial expenditure. The government establishes which services are to be provided (through service specifications) and how much money is available to pay for them (via the budget). The Ministry of Health negotiates contracts with various organisations to meet these requirements (see chapter Four for a detailed analysis of this system). This system is well established and addresses issues of financial accountability. The public nature of the policies that frame this model should ensure that power is not hidden from view. However, I argue that because of the severity of their condition, people with DMD are overwhelmed by the complexity and fragmentation of the multiple bureaucracies that they engage with. These policies may be transparent for those implementing the health system but they are opaque for those attempting to negotiate their way through this system. The policies and the structure of outsourced bureaucracies act as Foucauldian technologies of governmentality (Foucault 1991) and as Shore et al. note they can be understood as "effective forms of domination" where "power is hidden from view" (2011:9). The structures of service provision and the polices to which service providers must adhere occur as part of New Zealand's democratic process.

This chapter demonstrates that Foucault's analysis of the importance of the provision of general healthcare for the population as a whole, rather than for the individual, (Foucault 1997) enables a system where the exceptional requirements of people with DMD repeatedly fall outside the limits of service specifications, eligibility criteria and budgetary constraints.

While these limits are public, the power inherent in them is hidden because it is so difficult for individuals and families to pinpoint the exact location of control. It seems almost impossible for families to know where the power actually lies. Is it with street-level bureaucrats (Lipsky 1980) such as agency employees who assess the eligibility of disabled people for funded services, or is it with others within an incoherent, hierarchical system of contracting and reporting? Despite the ideological transparency of the system, individuals and families were not clear about where to address their appeals, how to argue the special nature of their requirements and explain why their case should be given additional resources. The mantra of public accountability and transparency was undermined by the experiences of people with DMD.

Individuals and families with DMD invest unbelievable amounts of time negotiating the multi-level governance systems which "ha(ve) come to be an idiom for describing the modern regulatory state" (Shore et al. 2011:10). The time devoted to finding a path through this complicated, confusing maze is time that cannot be invested in the meaningful activities of life (see Chapter Five). Furthermore the services that individuals and families request are often services to maintain basic health. These are not frivolous requests for luxury items. Harry wanted a powerchair with better suspension because the one he had contributed to his significant back pain. When an improved powerchair was denied he tended to stay in bed rather than sit in his uncomfortable chair. This increased his social isolation and depression. The system of powerchair provision impacted on his biological experience of DMD (depression). Government policies around wheelchair eligibility, in conjunction with the outsourcing of assessment and supply, form part of the local biology of DMD in New Zealand (Lock and Nguyen 2010). Similarly the provision of adequate hours of support worker assistance was essential for Harry's physical, social and emotional well-being, his sense of security that there would be someone available to monitor his ventilator, to help him with eating, drinking and the other activities of daily life. Support worker assistance was also needed for his mother's emotional well-being.

The belief underlying the funding of public services in New Zealand is that transparency and accountability is ensured by policies such as service specifications, eligibility criteria and fixed budgets. My analysis of bureaucratic entanglements suggests that these policies are primarily transparent for funding agencies and service providers – rather than the recipients of service.

This chapter examines the way wheelchairs are provided for people with disabilities and then it examines the way support worker hours are allocated. My analysis of these services not only exposes the obscurity of policies but also shows how biological and social phenomena interact to create a particular local reality that has biological significance.

Harry and his Wheelchair

It was clear from his arrival at the café that Harry was in some discomfort in his wheelchair. I noticed that Harry was repeatedly tilting his chair. I asked him if he was OK, wondering if I should cut the interview short. But he said that this is just how life is for him. His wheelchair is not comfortable and he thinks he needs a different one with better suspension or to have the suspension on his current chair altered. He explained how he felt that his concerns were dismissed by the wheelchair assessor he dealt with. She had just returned from a six months leave during which time no-one was employed to cover her absence. As Harry was in such pain the family had been into Allied Medical, a specialist retailer of disability equipment, who had made some suggestions of how they could perhaps sort out the issues with his pain and chair. So Harry and Margaret entered the meeting with the wheelchair assessor with some ideas of solutions for Harry's pain. They felt quite positive. The assessor was not happy that they had sought independent advice during her absence and they felt she did not listen to that advice. In fact they felt that rather than being concerned about Harry's pain she acted more like a gatekeeper of the Government's coffers. Harry said, "I felt Iike I was asking her to spend her own money."

At the time of our interview Harry and his team were under the impression that the only chair she would recommend was a chair for indoor use only, otherwise he could keep the one he had. I was flabbergasted to hear this. By this time I had been involved in the DMD community for six months and I had never heard of a chair that could only be used inside. I sought the permission of Harry to mention this to the MDA fieldworker and office manager. Both indicated that this was not a unique situation. One replied, "This has happened with other members and frankly it's ridiculous and tantamount to putting somebody on house arrest when we should be encouraging them to get out and be part of their community." The other said,

> I have heard of this happening to another of our members because he is no longer in school or paid employment. He had to prove he was doing at least 20 hours of voluntary work in order to be funded the wheelchair he needed otherwise they were

I checked with Mobility Solutions in Auckland, the organisation that is responsible for assessing need and recommending powerchairs in Auckland. The staff member I spoke to was clear that the mobility base of the preferred chairs does allow the powerchair to be used outside. She said, "no-one should be confined inside because of their chair." Preferred chairs are selected by the funding bodies Accessable and Enable who research which chairs are good quality, long lasting and that can be repaired and upgraded if required. They research which products offer best value for money for most requirements. According to the wheelchair assessor I spoke to, preferred chairs will probably suit about 80% of users. The other 20% might need something more specialist and the process allows for the negotiation of this.

It seems that there exist diametrically opposing ideas about the same issue. Harry's team, and staff from a regional branch office were under the impression some people were being recommended indoor-only chairs while the staff member at mobility solutions was quite sure that this could not happen.

The Wheelchair Assemblage

In order to explore this misunderstanding and other issues of wheelchair provision I discuss a number of organisations. Much like the cough assist machine assemblage, a wheelchair involves a complex system of interactions. Key parts of this assemblage include; companies involved in the sale of products, wheelchair assessors, government contracted service providers, the Ministry of Health and the national Disability Support Services (DSS) and individuals and families requiring powerchairs.

Private companies that provide the products are an important part of the assemblage. These organisations promote and supply products that have the potential to enormously improve the quality of life of people living with impairments. The two main companies that sell wheelchairs in New Zealand are Allied Medical and Invacare. Disability expos such as Show your Ability that are held around New Zealand are forums where the latest technology can be seen and tested by people with disabilities. A very sophisticated range of technologically advanced equipment that facilitates independence is on show at these expos. They include; adapted vehicles, powerchairs that can climb stairs, beach wheelchairs, computer or mobile phone applications (apps) that assist with tasks such as checking who is at the door, opening

and closing curtains and adjusting heating within the home, and many other assistive devices. These types of technology tend to be expensive. For example a TrackerPro which is a mouse replacement device that enables people with little or no hand movement to perform all mouse functions by moving their head costs NZD$1,650.00 and a dynamic arm support costs NZ$4,600.00.

Wheelchair assessors, such as the staff at Auckland based Mobility Solutions or Seating To Go in the central North Island, are skilled occupational therapists or physiotherapists who have undertaken further training to specialise in identifying the wheelchair and seating needs of people with complex needs. In addition to their expertise in wheelchair and seating assessment they must also be familiar with the Ministry of Health eligibility criteria and the fiscal constraints of the equipment providers. Some wheelchair assessors are employed by DHBs, for example Mobility Solutions is part of Auckland District Health Board (ADHB), but some are employed by private companies, such as Seating To Go.

The two companies, Accessable and Enable, are responsible for managing government contracts to provide disability equipment and modification services (EMS). Accessable Ltd, provides equipment and modification services in the top half of the North Island, the region north of Meremere. It is a privately owned company with two private shareholders. Enable New Zealand is the other provider, it serves the rest of New Zealand, south of Meremere. Enable New Zealand is also registered as a limited company but its shareholder is MidCentral District Health Board. Despite the differing types of ownership, both companies commit to the same service specifications with a detailed reporting schedule that includes monthly, six monthly and annual reports (section 11.4 of the service specification) that must be submitted to the Ministry of Health. Accessable and Enable research, source and provide best value for money equipment, and employ EMS advisors to work with EMS assessors to ensure that the solutions that assessors design can be met through the funding and equipment available.

The Ministry of Health is responsible for the management and delivery of health and disability services. It administers the regulations upon which the health system is based. Disability Support Services, funded by the Ministry of Health, plans and funds services specifically to facilitate autonomy for people with disabilities. It provides equipment and modification services (EMS) for people with disabilities. It does this by contracting Accessable and Enable to offer a cost-effective service. The Ministry of Health establishes exactly what services the contracts with these providers will deliver. These service

specifications are public documents and are posted on the Ministry of Health website. The Ministry of Health also clarifies the essential eligibility criteria which people with impairments must meet in order to receive equipment (Ministry of Health 2014, Equipment and Modification Services, Equipment Manual).

Problems with Eligibility Criteria

The misunderstanding about indoor-only wheelchairs seems to be related to the essential eligibility criteria established by the Ministry of Health. Accessable and Enable base their decisions on whether or not they will support the applications from assessors, such as the team at Mobility Solutions, based on these eligibility criteria. These criteria (listed as 2.3 of the Ministry of Health, Equipment & Modification Services, Equipment Manual 1, 2014 version) state,

> Funding for equipment can be considered where it has been identified as being the most cost-effective intervention and is essential for the person (independently or with assistance from support people) to do one or more of the following:
>
> • get around, remain or return to their home
>
> • study full-time or do vocational training
>
> • work in full time employment
>
> • work as a volunteer
>
> • be the main carer of a dependent person
>
> • communicate effectively.

One of the key areas for assessors is the difference between essential and desirable. The assessors must provide information about essential disability-related need which fit the criteria. For DMD clients the focus tends to be on facilitating the first criteria. The powerchair mainly allows them to get around more safely in their home. The person I spoke to at Mobility Solutions clarifies that in New Zealand there are very few chairs that are defined as being suitable for indoors only. "These chairs would only be considered if there were significant access issues indoors and a more 'standard' power chair did not fit in the environment. All the preferred option power chairs can be used outside."

There was miscommunication about what the first criteria, "get around, remain or return to their home" means. Harry and his support team, and the regional branch staff, were clearly under the impression that this meant the chair would only be good enough to be used inside

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the home and this was not the case. The most basic preferred option chair can be used on footpaths and it can manage small kerbs. This level of powerchair would struggle with a gravel path, a playing field, or an unsealed road, for example. It could be used to go to a shopping mall, an accessible café, or along a well-maintained footpath. Harry understood that the suspension of the type of chair he was using was not as good as the suspension of some other chairs and he believed that a chair with better suspension would alleviate his pain. The Ministry of Health's eligibility criteria focus on people's employment and education status in measuring their eligibility for the type of suspension a chair offers, but not their level of pain. So while there was miscommunication regarding what the capabilities of an "at home" powerchair were, the criteria in and of themselves are problematic. These criteria demonstrate the creep of workfare criteria which are traditionally associated with financial assistance such as benefits or tax credits into the provision of services funded by the Ministry of Health.

Most equipment provided through the Ministry of Health is not dependent on a person's educational or employment situation. For example stents, hip prostheses, surgical mesh, pins and rods are not provided on a workfare basis. Only the equipment provided through Disability Support Services (DSS) is assessed in this way. DSS provides a pool of ring-fenced money to be used exclusively to assist people with disabilities to participate in their communities.

The implicit message is that disabled people who are in employment or education have a greater social value than those who are not. A person engaged in education or employment will be eligible for a powerchair that can go up and down uneven kerbs, across grass surfaces and generally permits greater social interaction. Not only is this inequitable with regards to other health services it is also a double form of disadvantage given the high levels of unemployment that exist within the Duchenne community. The criteria can be viewed as a form of governmentality – one small strategy among many that instils among people with disabilities the desire and disposition to govern themselves, to contribute to their own care through employment. As such the criteria are a blunt tool that ignore the difficulties those with DMD face in attempting to participate in employment.

The Disability and the Labour Market Findings from the 2013 Disability Survey clearly show that disabled people are more likely than non-disabled people to be unemployed (even though 74% of unemployed people with a disability said they would like to be in employment) (Statistics New Zealand 2014b:5). Furthermore, the findings show that it is those whose

disabilities are concerned with agility and mobility who are less likely to be employed as compared to those with intellectual, sensory or psychological impairments.

People living with DMD have very limited mobility so their employment opportunities are limited. In the course of my fieldwork I met only one young man who was working for pay and one who had been employed in a call centre soliciting donations, on a temporary contract where the employer was a disability organisation. Generally young men with DMD do not participate in the paid workforce so once they complete their education they are not eligible for equipment that enhances their independence and opportunities to participate in their community. They are limited to equipment that is essential to get around in their home. The eligibility criteria applied to Harry did not address the role of equipment in alleviating pain.

The policy documents about how the money allocated to Disability Support Services can and cannot be spent is clear – The Ministry of Health Equipment and Modification Services Equipment Manual 2014 contains 74 pages of such detail and it is only one of thirty contract documents listed under Disability Support Services on the Ministry of Health website. But the process is not necessarily clear to those seeking equipment and modifications. This is partly because of the policies keep changing and also because the policies have more than one purpose. As well as clarifying the way need is assessed, these documents also cement into policy the financial limitations of DSS. For example, the "Meeting Access for Criteria to Services" (point 2.3 of the 2014 Equipment Manual) needs to be read in conjunction with the EMS Prioritisation Tool (point 3 of the Equipment Manual) which states,

> A prioritisation system is … in place to ensure that those eligible disabled people who meet specific access criteria who have the greatest need for services and the greatest ability to benefit from equipment and modifications are able to access the available funding.

This means that even when a person meets the eligibility criteria (listed in 2.3 of the Equipment Manual) their requests will be considered in light of the requirements of other people with disabilities. The prioritisation tool was being trialled during my research to see if a more equitable way of allocating equipment could be implemented. An evaluation of the trial has now been completed and the Ministry of Health (in a 2015 update) has accepted the recommendations of the evaluation company to embed the prioritisation assessment into its formal assessment process. The prioritisation tool was "developed to prioritise access to EMS services based on client need, risk and ability to benefit" (Ministry of Health 2015). This prioritisation of equipment for people with disabilities is similar to the system used to

prioritise elective surgery (Chan et al 2016). Both measures are based on the idea that a methodical prioritisation is appropriate when demand for service exceeds supply. The limits to supply are not questioned. The approaches offer a more equitable way of assessing priority. Both adopt a consultative approach that allows for those seeking the assistance of the health service (people needing non-emergency surgery and people with disabilities) and experts providing the assistance (clinicians and therapy assessors) to contribute information for the prioritisation process. The EMS priority tool renders decisions about who receives services technical and 'scientific' and conceals the values the system is based on. This decision is made regardless of the cost of the equipment; the process is about prioritising recipients of service, it is about making sure the person most likely to benefit from the provision of equipment will be prioritised. This model fits with Pèro's examination of bridling, "the neoliberal state's efforts to bring under control or neutralise civic organisations and individuals, especially when these challenge its policies" (Pèro 2011:224).

The introduction of the priority tool is an extra layer of bureaucracy that creates a particular kind of governmentality (Foucault 1991). The priority tool is an additional step in the process of powerchair provision that was added after my initial interview with Harry. The rest of the process remains intact – fulfilling the eligibility criteria of 2.3 of the EMS manual, having a consultation with a wheelchair assessor to find a suitable solution and applying to Accessable or Enable for funding for the recommended solution and undergoing an appeal process where necessary. While the tool may address problems in one part of the system it also adds another layer to the bureaucratic maze that people experience.

This new tool and the other ways of assessing eligibility are strategies of governmentality. They are very specific, complex forms of power that incorporate meticulous measurements of the population. The development of the prioritisation tool also demonstrates how citizens can try to affect the policies that impact on them. Some of the problems with the system that Harry described were acknowledged by others in the powerchair network and a wider network of equipment users. The trialling and evaluation of the prioritisation tool is an attempt to make the system of equipment provision more equitable and transparent. Consultation with the Consumer Consortium (people representing national disability organisations) is one way that those governed by the DSS policies attempt to negotiate and contest powerful polices, as Pèro (2011) has discussed in another context. The prioritisation tool may facilitate a more equitable distribution of the limited resource – the 'first come, first served' approach that existed before this tool was developed will be replaced by a 'who will

benefit most?' approach – but the limited resources remain limited. The stress and suffering caused by multiple bureaucratic interactions is not addressed by this tool and it remains to be seen if the new prioritisation tool will address the exclusion that those with DMD often experience in their interactions with health and other systems of governance.

I argue that this new online prioritisation tool and other methods of assessing eligibility to receive resources primarily meets the needs of the managerial approach and the system of fixed budgets through which the health system is currently funded. In contrast to the situation studied by Fitzgerald (2004), this system of capped budgets and the efficient management of resources now seems to be ideologically embedded. Fitzgerald's ethnographic study of a New Zealand hospital identified two distinct types of carers within the hospital system, clinicians who combined technical expertise with care for the embodied patient and managers and administrators that envisaged themselves caring for the hospital. At the time of her fieldwork (1997-1998, a time of health system reform, see chapter Four) Fitzgerald notes conflict between these two approaches to care. This no longer seems to be so noticeable, it seems that the rationale of managerialism and fixed budgets is now firmly entrenched and socially accepted among many involved in the health systems.

> The evaluation by Synergia found that the aim of the pilot (the trial prioritisation tool) to establish a more fair and equitable prioritisation process is well accepted by clients, EMS Assessors, EMS providers and Ministry staff. (Ministry of Health 2012:5)

The prioritisation tool and other assessment strategies form part of the paradigmatic shift from a reimbursement system of health care to one of fixed budgets and in the course of my research no one challenged this new model of providing health services. People challenged their exclusion but not the system itself. This demonstrates the hegemony of the system and thus reinforces it as a form of governmentality. The new processes for providing health and disability services can be understood through a Foucauldian analysis; they are tools of governmentality which pay meticulous attention to the surveillance and economic control of the population (Foucault 1991:92). They include some input from those affected by the policies (the Consumer Consortium) so can be seen to fit within a consultative, democratic paradigm however the level of resourcing is not debatable – only the strategies used to implement the distribution.

The Disparity between Capped Budgets and Disability Rights

As well as Foucauldian issues of governmentality another consideration is the use of human rights discourses in the provision of disability services. The system of delivering health care and the particular criteria used to determine eligibility (especially those of education and workfare) do not align with many of the philosophical vision and mission statements that frame the work of the state-funded disability sector. For example the guiding principles of the United nations Convention on the Rights of People with Disabilites (UNCRPD) includes; "full and effective participation and inclusion in society, equality of opportunity and accessibility" (United Nations, 2006).

The New Zealand Disability Strategy 2001 (under review 2016) includes the following statement (2001:11) (abridged),

> The vision of this Strategy is a fully inclusive society. New Zealand will be inclusive when people with impairments can say they live in: A society that highly values our lives and continually enhances our full participation. This will happen in a country where:… the idea that society imposes many of the disabling barriers faced by people with impairments is widely understood and, therefore, legislation, policy and other activities enhance rather than disable the lives of people with impairments.(Minister of Disability Issues, 2001)

The ideas inherent in the guiding principles of the UNCRPD and the vision of the New Zealand Disability Strategy (2001) and other similar documents reflect an ideal where people with disabilities are included and valued and able to participate equally in society. This case study demonstrates that there is a gap between rhetoric and reality. This gap is where notions of equality and human rights meet notions of budgetary constraints and cost management.

For Harry and his particular situation (prior to the roll out of the prioritisation tool) where he wanted a more comfortable (and hence more expensive) powerchair so that he did not experience the pain caused by the jolting of the lower standard of suspension in his current chair, the issue was complicated by the poor relationship he has with the assessor. His DHB had only one assessor. Unlike people living in larger centres, he could not see a different assessor. His assessor declined his request because she did not believe it met the criterion (he was able to get around inside his home with his existing powerchair), so there was no possibility for an appeal or a review. He had very little room to change the situation, to engage in acts of resistance to this regime of biopower through contesting the decision. It might be expected that other disabled people would experience similar frustrations or

disempowerment with the workfare criteria. The opinion of the staff member at Mobility Solutions offers a possible explanation. She felt that the existing criteria are a satisfactory start and although she would prefer to see a greater focus on independence (as per the visionary statements), rather than risk management, she noted, "There is room for negotiation through an appeal process". The wheelchair assessor service is able to appeal decisions made by Accessable or Enable and they have on occasion appealed to the Ministry of Health. My discussions with other users of powerchairs reiterate the importance of the appeal process. One woman I spoke to used a powerchair and was also employed and a parent, so met a range of the eligibility criteria. She told me she had had to appeal for almost every piece of equipment including a higher specification wheelchair that allowed her to take her children to soccer and make the journey from her office to the nearby shops, bank and post office (a requirement of her job) on a regular basis (i.e. it was essential that she be able to do more than get around at home). The initial application from the wheelchair assessor was rejected by the funding body (in this case Accessable) but the assessor then appealed that decision and Accessable accepted the appeal and agreed to fund the equipment. This was also the experience of the second young man with DMD whose chair was falling apart but as he was not in education or working he was told he was not eligible for a better chair. In this case the assessor appealed the decision not to fund a replacement and the appeal was successful. In these cases there are people working in alliances, and there is more room for a power dynamic to operate and create a change.

Because Harry was part of a wider DMD community – he attended social events, conferences and workshops - he knew that other people who were in a similar situation to himself had better chairs. He could see them when they got together. As his request for a better chair was based on the pain he experienced he clearly felt that the situation was unfair.

The assessor's meticulous attention to detail is an example of the technology that Foucault calls governmentality (Foucault 1991). Her strict and uncompromising enforcement of the criteria met the budgetary demands of the funding organisation. Her position can be understood in light of this September 2015 Accessable reminder to staff to manage budgets, reinforcing the message that only essential needs could be met,

> "In the first two months, the demand for Equipment and Modification Services (EMS) has been exceeded by 19% ($640k). The spend for July and August is almost as much as the total overspend for the whole of the last 2014/15 Financial Year!

> This is going to be a tough year and we will have to do something differently and work smarter to make sure that peoples' needs are met within the available budget.

> All parties in the EMS funding process have an impact on demand and the budget; EMS Assessors prescribing practice and choice of solution, Accessable's advisory and procurement processes, and supplier's provision of equipment. With Accessable, EMS Assessors and Suppliers all working together we can better manage and maximise available funding. The Ministry of Health has been very clear that there is no additional funding, and we must find strategies to ensure peoples' needs are met within the available budget…

> Demand management is achievable (volume and cost of services). Remember EMS is not the default funder, so please; … ensure the person's need meets the access criteria and the need is essential and there are no viable alternatives – ensure that only the essential need is met by an EMS funded solution (Accessable 2015)

The message here is clear. The wheelchair assessors are an important part of a contracting system. The Ministry of Health, through Disability Support Systems has a contract with Accessable to provide services, including disability equipment, to eligible people living in a particular region. Accessable must meet their obligations within an agreed budget. The update reiterates the importance of assessors considering the financial implications of their assessments,

> Given where we are at in relation to the pressure on the budget already, we need your help! Please take extra care in establishing eligibility, access criteria and consideration of the most cost effective solution. Even small savings of $50-100 per request can make a huge difference (Accessable 2015)

Wheelchair assessors are in a challenging position, a place of conflict between the needs and expectations of their disabled clients and the needs and expectations of a complex intertwining of a registered (for profit) company and government (Disability Support Services) guidelines. Staff working for Mobility Solutions are DHB employees. They are not employed by Accessable Ltd or Enable New Zealand, yet part of their role is to facilitate positive outcomes for those companies. These companies operate in an environment where the government adheres to the underlying neoliberal principle that businesses provide better services than state agencies. This belief has been enacted in New Zealand society for over thirty years and the contracting out of government services to businesses is commonplace (see Chapter Four). According to this model of social organisation it is completely legitimate for those who own effective businesses, that meet their contractual obligations to the government (in this case these are clearly delineated in service specifications as outlined by Ministry of Health 2016a), to benefit from their business acumen.

Hidden Power of Eligibility Criteria

Despite the widespread acceptance of fixed budgets as a way to manage government service provision, debates about the distribution of budgets remains. Political debates about the level of funding are an inevitable part of the democratic system. In the 2016 budget (May 26th), the government announced $(NZ)42,296,000 each year for the next four years ($NZ 169 million over four years) for new health initiatives to be funded by Disability Support Services. In June the New Zealand Council of Trade Unions put out a Working Paper on Health. This paper argued that the increases within the Vote Health budget were not sufficient to maintain current levels of service given increasing costs, population growth and aging and specifically in relation to the increases to DSS the report notes,

> Centrally managed national services such as National Disability Support Services, National Elective Services, and Public Health services received $79 million below what they needed to cover cost increases and demographic changes, and are $90 million short when $56 million in additional services are included after taking account of $45 million in shifting expenses to DHBs and "reprioritisation". (Rosenberg and Keene 2016)

The paper argues that the money Vote Health receives has declined as a proportion of Gross Domestic Product since 2009/2010 and this in turn reflects reduced overall government expenditure. While the government states that the proportion of government spending on health has increased the CTU notes this proportional increase is because the proportion of government funding going to Vote Health has not been reduced as much as other services – not because it is receiving significantly more money. The report goes on to point out that one of the impacts of what it terms "chronic underfunding" is limited access to services (such as disability equipment).

> A Commonwealth Fund study (Davis, Stremikis et al. 2014) on the performance of the health systems of 11 comparable countries found New Zealand's health system performs well on what it actually does, ranking 3rd and fourth respectively on measures of quality and efficiency. And this was achieved despite New Zealand being ranked bottom on health expenditure per capita. But we perform poorly on measures of access to services, indicating significant issues with what our health system does not do due to inadequate service capacity. (Rosenberg and Keene 2016)

In light of this analysis, the prioritisation tool which has been designed and implemented to increase equality (so that those people who will most benefit from EMS are the people who will receive the funding) may also conceal increasingly limited access. Thus claims of

transparency are perhaps simplistic given the complex budgeting and funding environment within which these transparent policies occur.

The Provision of Support Workers

Harry, Margaret and Bruce also talked about many of the difficulties they had encountered as a result of Harry living independently in the community. Typically these were to do with bureaucratic gatekeeping, especially accessing the 24/7 care that Harry needs to live independently. As our conversation progressed Margaret explained why Harry lived independently. His mother had reached a stage of exhaustion that was of deep concern to them all. Bruce, especially, could see that the earlier situation when Harry's Mum (his daughter, Elizabeth) had been Harry's main carer as well as looking after other younger children was impacting her health, "she looked grey." Harry more directly said, "her brain crashed". It was clear from the way they spoke that the family could see she was suffering and that her suffering would only increase unless something else was put in place to support Harry. It was too much for one woman with young children to manage. As is typical for a young man with DMD, Harry needed to be turned two or three times during the night. The exhaustion caused by constant broken sleep is commented on frequently by parents. This mother needed the energy that is required to care for other young children.

The Impact of Duchenne Muscular Dystrophy on Family Carers

The impact of DMD on other family members is an integral part of the story of life with DMD. Families feel that they are constantly battling systems and this compounds the stress they are under. It is easy to understand how some mothers become overwhelmed. This is especially the case for women parenting alone and this is the situation that Harry's team described. (See further discussion of the impact of DMD on primary carers in Chapter Six)

Whilst Harry was living with his mother and siblings repeated requests had been made for more assistance but the Needs Assessment Service Co-ordinator (NASC) would not increase the hours Harry received. The family see this as a fault in the system,

Margaret   … and that is the system again, in not providing 24 hour care, let her (Elizabeth, Harry's mother) down as well.

Bruce   Well, the family is expected to step up, which is fair enough

Margaret   Yes, but as you say it is not fair compared to someone who is fully funded. Harry did not ask to be in this position, he did not ask for this condition. Just like someone in an accident did not ask to be in an accident, yet they get fully funded. It is like discrimination against people with a medical condition.

This quote highlights two important aspects of the assistance available to people with DMD. The first is the reference to "the system", namely, the way people who need help to live independently receive this help; the second is the reference to the different provision of support received by people whose disabilities are caused by accidents and who are covered by Accident Compensation Corporation (ACC). (See chapter Four)

The Support Worker Assemblage

The way the Ministry of Health pays for support workers to assist disabled people to live in the community (rather than in an institution) is highly bureaucratic. Just as the Ministry of Health contracts with EMS service providers (Accessable and Enable New Zealand) who work with wheelchair assessors (who may or may not be employed by independent businesses) who interact directly with disabled people, a similarly fractured and bureaucratic system exists when people living with disabilities want to access support workers. In this case, the Ministry of Health contracts with organisations called Needs Assessment Service Co-coordinators (or NASCs). NASCs assess how much assistance each eligible person is entitled to receive and then draws up a plan with the disabled person, or the family, to show how the allocation will work. There are various ways the assistance that the NASC decides to allocate can be provided. Often people will use an agency to provide the assistance for which they are eligible. When I began my fieldwork some people were using a system called individualised funding (IF). At that time it was a fairly new system and it seems to have become increasingly popular as more people develop the confidence to use it.

Under IF, rather than allocated funds transferring from the NASC directly to the agency, the funds are distributed by an IF host provider who assists the individual concerned (or their family) to allocate the funds in a more autonomous way. Some people employ their own staff and take on all the responsibility of employment, such as, paying salaries, tax, holiday pay, ACC contributions and so forth. Others prefer to choose their own staff but pay a fee to their IF host provider to undertake other parts of the employment process for them. The IF host provider ensures that the funds are used appropriately and reports to the Ministry. There are

some advantages to using an agency to provide support workers; staff are available to provide cover at short notice if someone is sick, the challenges associated with advertising and interviewing are met by the agency, the agency should deal with employment problems and conflicts. There are also disadvantages including a lack of control over who works with the individual. IF was established to address the lack of autonomy that some individuals experienced when the only option was agency-provided staff. There are many organisations and businesses involved in this system, or assemblage.

Those organisations and individuals involved in the support worker assemblage include; the Ministry of Health who provide the ring-fenced disability support funds to DSS, DSS who negotiate contracts with NASCs, NASCs who assess eligibility , draw up care plans and allocate funding packages, support worker agencies who provide workers to those people or families who prefer to utilise an agency, IF host providers for people and families who prefer a more autonomous system, people with disabilities, their families and the support workers themselves. In addition policy documents are an important part of this assemblage; service specifications, reporting documents, NASC assessment documents and subsequent plans and employment contracts. There are fifteen NASCs servicing twenty DHBs, so not each DHB has its own NASC. The NASCs can be private, for-profit companies, charitable trusts, or part of their DHB. Support worker agencies offer a staffing service to people who have been allocated assistance by a NASC. There are many of these agencies. Some are large, well-known organisations like the Salvation Army and Presbyterian Support and others are smaller, locally based operations. This service is provided by registered charities and private, for profit companies. No DHB provides this service. There are various structures through which IF host providers operate but all have a business-type model. Manawanui In Charge is one such organisation and although it is a registered company its shareholders are four disability sector organisations. Healthcare of New Zealand Holdings Ltd, in contrast, is a privately registered company that includes IF host provision among the services it offers.

Furthermore because this is a complex system that does not always work well for people with disabilities; changes and adaptations are made to it regularly. For example, "Enabling Good Lives" and "Choices in Community Living" (CiCL), which are both part of the "New Model", were new initiatives that affected the provision of support workers and were being trialled during my research. The disability system is constantly under review. "The New Model" is a new way of supporting disabled people to achieve their goals. It gives people

more choice and control over support and funding in their everyday lives" (Ministry of Health 2016b).

However, the support worker system usually works in the following way, and this was the system Harry relied on. The Ministry of Health contracts NASCs to assess the care requirements for individuals with disabilities. The contract the Ministry has with NASCs requires three things of the NASC; facilitating the needs-assessment process, service coordination, (which includes; giving information about options, planning and coordinating the supports in a support plan and allocating some DSS-funded support services) and finally budget management – NASCs must manage the Ministry-funded Disability Support Services in a fair and cost-effect way.

The government contracts an organisation (a NASC) to assess what an individual's needs are. Adults with Duchenne Muscular dystrophy have a high level of need. As their muscles weaken they need assistance with most tasks, in the past the condition was sometimes referred to as "creeping paralysis'" and this description conveys an idea of the very limited movement that eventuates as people age. These young men generally need constant attention. The state provides some assistance to families but the amount varies enormously. In some cases packages seem quite generous and in others they are clearly inadequate. Cases such as Harry's highlight the problems in the system. Family members are viewed as "natural resources" and the availability of such a natural resource will reduce the publically-funded assistance available through the state. In theory help is available but in practice it is not always forthcoming. Applying for and receiving a needs assessment, appealing decisions and the design and implementation of a support plan are all highly bureaucratic processes. But key to the decision about how much assistance a person will receive is that the needs of the individual and/or the family have to be balanced with the funding available – the finite sum of money received by the NASC to implement their Ministry of Health contract and meet service specification agreements.

Given how precise much of the bureaucracy around the provision of support worker hours is; it is anomalous that the way the hours themselves are calculated is unclear. The service specification between the Ministry of Health and the NASC is very detailed, planning and coordination of the allocated support is similarly meticulous. There are detailed requirements for reporting and compliance regimes both by the service providers and the NASC (Ministry of Health 2016a and c). Unlike all other aspects of the system, the way the hours themselves

are calculated is not explicitly mandated. This is partly because the amount of assistance each person will receive depends on the other needs the NASC has to meet. Allocation is determined by funding availability and not by needs per se (Holmes et al 2014). This leaves disabled people with the impression that the decisions are made arbitrarily and often unfairly.

Harry's Experience

As the assistance offered to Harry, while he lived at home, was not enough to significantly reduce his mother's stress Bruce organised a house in town for Harry to live in. His mother lives just a few kilometres away and maintains active involvement in Harry's life but is no longer primarily responsible for his day to day care. This is now provided by paid carers. The results of this change in living arrangements for Harry's mother have, according to Bruce, been beneficial, "she has come back to life again … the relief for her has been amazing."

Harry moved into an independent living situation in order to alleviate his mother's stress. As a result of no longer being primarily responsible for his daily care she regained mental well-being and continues to be involved in his life. However the problem of sufficient care for Harry remained. He needed to be reassessed by the NASC. Earlier his mother was considered a "natural resource" and because of this resource his hours were limited. His team assumed that his eligibility would increase significantly as she was no longer available. However they were disappointed in the hours that the NASC allowed. The family appealed the original decision which was to give them,

Margaret   40 hours, wasn't it? 40 something hours, 45 hours and we were told that if Harry needed any more hours than that he would have to go into residential care.

Harry referred to the United Nations Convention on the Rights of Persons with Disabilities during our interview and went on to say,"I don' think that they are allowed to do that, but that is what they threatened anyway." This is a clear example of how even within services designed to meet the needs of people with disabilities Harry's requirements were "too high". His needs could not be met by a system designed for an average need. It was an awful thought for him that at the age of 21 he might end up living in an aged-care facility which was the only residential care in his small town. He perceived the reference to residential care as a "threat", a disciplinary action to curb the expression of his needs.

A NASC, serving a similar region with a significant population of rural and small town residents, Northable, noted on its website that in 2006 they introduced a new service to address "a trend whereby young disabled people were moving to residential services out of the Northland region. A lack of resilience and resources within the family and fragmented disability support was thought to be significant contributing factors" (Northable Disability Services 2016). This is salient as it demonstrates that the circumstances Harry described were not an unfortunate one-off incident. At least one NASC recognised a "trend" of young disabled individuals moving out of their region to enter residential facilities. Perhaps these young people were moving out of region because they too were faced with aged-care facilities as the only local option for residential care.

The issue of providing personal care for Harry became serious. A NASC employee travelled to Harry's new home to discuss the issue. Bruce describes the meeting,

> It was really offensive because she had driven up from down here (provincial town) in her nice comfortable car and she was all dollied up. She came in a said, 'You are already getting the maximum number of hours (45) and if you want any more you'd be required to go into care in an institution and you wouldn't like that, would you?' … I asked what on earth was the purpose of her trip, if you came up here just to tell us, you're on the maximum and you can't have anymore, what was the purpose of this meeting if that is your bottom line? She was not very diplomatic about it. But then she got caught out. She had denied receiving our emails all about it but then had to produce the emails for something else and suddenly they doubled the hours.

The reason why the NASC changed the allocated hours was not clearly explained during the interview. This reflects a fairly common experience of having a decision to refuse a particular service overturned without explanation: a sign, perhaps, of gaps in the bureaucratic façade of impartiality. People often did not know why the change had occurred or which part of a bureaucracy had made the decision to accept the application or appeal. Usually people did not care too much at the point that support worker hours were increased, or an improved powerchair was agreed to, or a bathroom renovation included allowing for a separate toilet (for example). They did however, care and invest energy and time at the point of refusal.

At the time of the interview Harry received 116 hours of care a week. That is 4 nights (32 hours) and 84 hours during the day. Given that he requires care 24/7 that left 52 hours of care each week that the family had to provide for. Harry's mother and grandfather each do one night a week and the rest is covered by complicated and fragile arrangements.

The bureaucratic hoops that Harry and his supporters have had to jump through to get this level of care have obviously been emotionally scarring for them. They continue to be very suspicious and distrustful of the NASC. Although they can now manage to care for both Harry and his mother they do not do so in a confident fashion. They are aware of the risks in their current system and are wary of the reviews that are a regular part of the NASC procedure. This is a complex pieced together system that even with family input still needs private arrangements for Harry to live independently. It generates a sense of vulnerability as at any time it could break down.

The work of Kaufman and Becker (1986 and 1991) examining the treatment of stroke patients in the American health system makes similar findings. Their work considers the cultural value of different health services and they note the value attributed to the biomedical treatment of acute illness which contrasts to the devaluing of rehabilitation and ongoing support. Similarly the comments of Heurtin-Roberts and Becker also provide useful context. They note that,

> in the United States, at least, the dominance of biomedicine apparently precludes the attention of policy-makers to non-medical avenues that have the potential to effectively ease the burden of living with a chronic illness (1993:283).

The provision of support workers is enmeshed in a range of different understandings about care. Not only does the allocation of this service depend on financial resourcing, underlying this resourcing are values that reflect the points that these three authors make. The work undertaken by support workers is not biomedical treatment. It is about the worth of lives, not the value of life (Fassin 2012) and this type of work is not so highly esteemed. In New Zealand there is a system that allows for the public provision of support workers but the conditions of employment demonstrate that the work is not socially valued. It is low paid, often part time work with limited career pathways. One participant noted some of the problems in finding good support workers,

> Good support workers who are compatible with the supported person are not very easy to find. It's due to the funding restraints but also support workers' attitude (partly due to the society's attitude) and skills mismatch for "very high needs".

Harry has had problems with some of his support workers. This is a widespread problem for people with disabilities. Some support workers are marvellous and become like family members. Margaret would seem to be one of these. Harry, however, has had more than his

fair share of disastrous placements. Here we see Harry having to battle to be eligible for a service only to find the service unsatisfactory. He does not trust his service provider.

Dealing with the layers of bureaucracy is draining. Harry is assessed by one agency and has had to appeal and enter into a tense, threatening situation to get caregiver hours that are only manageable because Harry also makes private arrangements and relies on his elderly grandfather. Harry is now left in a vulnerable position, as was highlighted by a recent choking incident. Organising to get the personal care seems to have been an enormous job for Harry, Margaret and Bruce. Their dogged determination to get adequate hours reflects their lack of options. The alternative, an aged-care residential facility, was an unbearable, inhumane idea to them. They challenged the policies and decisions that exclude and marginalise people with disabilities, resisting these particular strategies of governmentality.

Pilot Schemes to Improve the Existing System

As noted, initiatives like Enabling Good Lives and CiCL are recent developments to improve the experiences of people with disabilities with service providers. Both these initiatives are part of a wider, more comprehensive pilot scheme called The New Model. This model has been introduced by the Ministry of Health to address the criticisms of the existing system (such as those experienced by Harry) that,

> … disabled people, their families/whānau and service providers … were generally unhappy with how services were being delivered. Services weren't coordinated and were often difficult to access; existing services and support were inflexible; and there was little choice about how support was delivered. (Ministry of Health, 2016b)

Rather than the disabled person having to fit the structure of various service agencies, under The New Model, the disabled person becomes central and agencies are required to meet the needs and goals of the person. The new model initiatives certainly appear to be improvements and offer a change in approach, if not more resources. A recent Ministry evaluation of CiCL notes that funding is an area of concern. The new initiative includes planning for a person's aspirations and goals and the evaluation states, "It is unclear what proportion of a person's funding package is allocated to necessities, such as support and equipment, and what is available to make other goals a reality." The evaluation also questions the flexibility of the funding under CiCL: Improving flexibility was one of the objectives of The New Model. A 2015 evaluation report of the New Model (Evalue Research 2015) highlights the expectation that adopting CiCL would be fiscally neutral. So while the brief is wider – disabled people

can include aspirational goals in their plans, the Ministry of Health is not intending to increase the allocation of money.

Others with DMD Who Lived Independently

The majority of young men with DMD remain living with their parents, partly because when they stay at home their care is guaranteed. In the course of my research I interviewed three young men with DMD who had lived separately from their parents at some stage (including Harry). I discussed the situation of a fourth young man, who died just as I began my research, with someone who had been involved in the appeal he made to his NASC to increase his support worker hours so he could live independently. In all of these cases the hours allocated for support staff were insufficient to enable the young men to manage to live independently and they and their families had make additional arrangements so they could have the fulltime assistance they needed. Rex's parents came to stay at his house on those nights when he was not able to have a support worker present. Simon, who wanted to leave his parents' rural home in order to enjoy the more accessible activities available in a larger city, returned to this rural home every other weekend. This was challenging as he suffered pain travelling in the mobility van on the long journey. I also briefly chatted to a young man at a conference dinner who was utilising the CiCL initiative to live away from his parents in a flatting situation with three other young people, one of whom had DMD. They were both at the dinner and I noticed that they shared a support worker. This is a strategy that two other young men who are similarly impaired discuss in the New Zealand documentary "Ryan and Blake Go Flatting" (Attitude Pictures, 2013). They pool their allocation for support workers and need only one person to be with them overnight. Such an arrangement works in larger population centres where there are likely to be several young men, some of whom may be interested in a joint living situation.

Chapter Conclusion

In some ways the support worker assemblage mirrors the wheelchair assemblage. There were various business and quasi-business agencies involved in contractual arrangements. Money flowed from Vote Health via DSS to assessors and providers. In both cases there is an assessment process to determine eligibility and to balance Harry's needs within the available funding. In other words, his needs compete with the needs of other disabled people for a portion of a finite sum of money. However, unlike the wheelchair assessment, there are no

publically available documents describing how the number of support worker hours should be determined. For powerchairs and other equipment there are the EMS guidelines that focus on risk-management (rather than independence) and contain a workfare influence. Once initial eligibility has been determined the prioritisation tool is applied. If the tool indicates that funding is available an assessment and solution process ensues. For support worker provision the decisions are made by the assessors based on home visits and discussions with disabled people and their families about their situation. The decisions take into account the "natural resources", "informal supports", "FFC" (family, friends and community) and then balance the individual's needs with all the other demands on a finite, capped budget. "Natural resources", "informal support" and "FFC" are all synonyms used by various NASCs to refer to the assistance provided by the family – usually parents. The implications of defining social relations, especially kin relations, as "natural" deserve their own analysis and discussion, but this will have to be in a different forum.

My data indicates that the allocation of support worker hours varies enormously. Some families who were quite well resourced (parents still married, both directly involved in the provision of care and at least one in full-time employment) had reasonably generous packages while other families living in much more marginal, straightened circumstances (sole parent, fully responsible for organising care, not able to participate in the paid workforce, living in a rural area) received packages that were clearly inadequate. There are many possible explanations for the inequality I found during my research; inequitable funding of NASCs, the cultural capital that some families bring to the assessment interview, the time in the annual funding cycle -- some people are assessed towards the end of the financial year when the bulk of the NASC's budget has been allocated. This inequality may also be to do with the lack of publically available guidelines for assessors, allowing for more interpretive, subjective decisions to be made. Further research would help to clarify this anomaly.

Young men with DMD want to live normal, ordinary lives but the way services are designed to meet a general, rather than a specific need, often curtails this possibility. Not only does inadequate service provision limit the ability of young men to participate in their communities, it also impacts on their health. Harry spent more time in bed when his wheelchair was uncomfortable which increased his isolation and had impacts on his mental health. Young men with DMD and their families manoeuvre within systems of care as best they can. Harry's case was particularly poignant as his decision to live independently was based on need rather than choice. In most cases (with the exception of those who pooled their

funding allocation with others who also needed fulltime support workers) families had to appeal their initial allocation of hours. This, combined with the lack of clarity regarding the provision of support workers, had been very frustrating – particularly in light of the visionary statements and inclusive, participatory discourse in which service provision documents are couched. Despite these visionary statements of social participation, inclusion and personal autonomy for people living with DMD, this detailed analysis of everyday life highlights the way technologies of governmentality control and limit of these visions. These technologies of governmentality include the prevalence of the purchaser provider split which results in multiple bureaucracies that families are obliged to engage with, policies that provide transparency for bureaucrats but not necessarily for people with disabilities, and the prevalence of strategies designed to facilitate self-management and self-governance which people and families with DMD find difficult to adhere to because of their anomalous situation. Invariably empowering discourses are constrained by practices of fixed budgets and further limited by policies that are ostensibly about transparency and equality but also entrench technologies of surveillance and economic control of the population. Here I have examined how strategies of governmentality and ideologies of efficiency and financial responsibility over-ride discourses of human rights, citizenship, participation and inclusion and ultimately serve to further marginalise a group of disabled people because of the exceptional nature of their requirements for services. This marginalisation is also shaped by the shifting ideas about the way life is valued that Fassin (2012) explores. The bureaucracies examined here address the political worth of lives. Although I have focussed this discussion on the technologies of care, the policies and appeals, the strategies and tactics adopted within assemblages, these technologies of governmentality and the resistances to them reveal the way lives with DMD are often socially devalued because of the needs of people with DMD are anomalous, even within the parameters of disability services. In the following chapter I continue my analysis of bureaucratic entanglements and examine the serious implications of rigid funding streams within the health system.

Chapter 10: Care in Hospital

This chapter focuses on a particular policy which illustrates that the exclusions that people with DMD experience can be life threatening. The rigid funding constraints that apply to the provision of care for men with DMD when they are admitted to hospital causes grave risks to their lives. The risks that the health system itself poses to the lives of these patients is overlooked because any poor outcome is inevitably attributed to their Duchenne muscular dystrophy, rather than to the structure of the health system. This chapter further demonstrates that Foucault's analysis of biopower as a form of power that "consists in making live and letting die" (1997:247) is highly relevant for a group as anomalous and marginalised as those with DMD.

Research participants repeatedly referred to the anxiety caused by the way their personal care was managed by the Ministry of Health when they went to hospital. People with severe impairments who rely on carers are not able to use their usual community support workers when they are in hospital. Support workers are funded through the ring-fenced Disability Support Services fund, which can be used to pay only for services that support community participation. Care that is required while a person is in hospital must be provided by the DHB. Each DHB addresses this issue independently with their own policies and procedures. Emergency departments are sites of particular anxiety but experiences of care on wards are also problematic.

This chapter begins by describing the personal experiences of people and families with DMD when they are admitted to hospital. This description notes that usual carers, particularly family members, provide the best care while hospitals struggle to provide adequate personal care for people with neuromuscular conditions. Some young men with DMD are at particular risk when admitted to hospital, notably those whose family members cannot stay with them. Men with DMD need carers who can provide their regular support and act as advocates when hospital staff lack knowledge about DMD. Not being able to rely on regular support workers increases the anxiety experienced when a person is admitted to hospital. Following this description of how people experience the hospital setting I go on to analyse the implications of the way the system is currently structured. My analysis develops ideas from the anthropology of policy (Shore et al. 2011) that policies which are intended to create transparent service provision are sombrely opaque. I note that recent vision statements in New Zealand's disability sector indicate a shift from a systems-centred approach to a person-

centered approach in service delivery. I consider this shift in terms of Foucault's discussions of biopower and governmentality (Foucault 1978), and in terms of Frank's (1995) analysis that such political changes without moral reorientations will only create more bureaucracy. However, the overriding significance of this chapter is that invisible barriers are constructed by rigid adherence to policy and that for this particular group such an adherence is potentially life-shortening. The anthropology of policy allows the perverse effects of "well-meant" and "well-intentioned" policies to be exposed. Research participants could see the serious, unintended consequences of this policy. The policy was an apparatus of power that support workers found hard to transcend. Shore et al. say, "declaring a policy to be 'official' endows it with a certain authority, …. It also endows a course of action with a dignity and morality that it might not otherwise possess" (2011:172). Prohibiting those best qualified from providing assistance to people with DMD in hospital is an example of a course of action that lacks dignity and morality. However, the implementation of a policy that legitimates this, sanctions this particular course of action and gives it authority.

The message that the current system gives to people with DMD is that in some situations their requirements are too complicated and ultimately they are expendable. Biehl (2012) notes, "care is made through the contribution of many hands … and disregard is a shared … process" (2012:261). Biehl describes the far more dire situation of Catarina whose muscular dystrophy plays out in the context of an economically marginalised Brazilian community with vastly reduced access to health services. However, his point that social disregard is a shared responsibility is relevant to the message of expendability that I discuss here.

Usual Carers Provide the Best Support

Parents or other immediate family members are finely attuned to the personal care needs of young men with DMD. As the majority of DMD young men live at home this care is part of daily life for these families. For those young men who live independently in the community most parents remain significantly involved in the lives and care of their sons. Parents and immediate family members are uniquely motivated in their caring roles.

Most DMD families do not expect DHBs to provide personal care. In the majority of cases when young men with advanced Duchenne's are admitted to hospital their families do not leave them alone. A parent will stay all the time to undertake personal care and to advocate on their behalf. Parents are of the opinion that relying on hospital provision of care would

increase the probability that their adult son would not survive the admission, as the following interview comments from a mother whose son had died following admission to hospital demonstrates.

Rhonda   I did not think the medical ward was the right place for Greg. And other parents have said this too, that they (staff) are so busy they (patients with DMD) are not monitored enough. If you get onto the heart ward it is a lot better because they monitor them all the time. But apparently they were full up so the medical ward was the only place……The doctor did say he'd seen how tired parents get looking after them, just like us, and they need to go home and have a sleep. But the thing is you can't leave them because they are at risk of choking. Because the nurses don't have time.

Similar comments were also made by another mother whose son had also died.

Shirley   Well people would say why don't you put him into care for a week? Blake would have died if you had put him into care for a week. People don't realise how fragile he was. But it was a small group of care-givers who knew him and knew us that you could count on. Life became so fragile in the end that I don't even think you realise. You are so busy doing it. Blake could not eat. He got really, really thin a few years before he died. I started making him this soup. Chicken soup with all the vegetables and everything you could think of and then I'd whizz it all up. And he'd drink it through a straw. He perked back up after that. And then he had to have his mask on all the time, he could not go without it for a second. Even his care-givers… it was really difficult at the end because he couldn't be without it even for a couple of seconds. So once they had transferred him out of his lift onto the toilet. It was a real race everyone had to be there to do their parts. It was like a well-oiled machine. Everyone knew exactly what their part was and if anything had gone wrong, it would have been all over rover. Anything could have happened to Blake at any time but you don't really think of that. You just carry on doing it because there is no choice.

These quotes highlight the very high level of care that these young men require. They need more than assistance with feeding and toileting. Parents are highly attuned to the assistance required; clearing secretions from the mouth, adjusting limbs, monitoring breathing masks and so forth. Although the quote from Shirley is about care in the home the same care would

be required in the hospital setting. Support work does not require a high of qualification but it is nevertheless essential work. As this example illustrates,

Cristopher   I can't call for attendance. I can't press the buzzer.

Kate   So you really need someone in hospital with you?

Cristopher   Yes Mum or Dad has to be there.

Grandparent   When he was in when he was 17 his Mum stayed in there all night with him. She stayed there in the morning and Dad would go in the afternoon. Mum would come home and have a quick sleep. And then she would go back up in the evening.

Kate   And you also had your brother here and he couldn't be left alone either?

Grandparent   No, so I used to come and stay here. And I would roll him during the night.

The general view that most parents hold is that leaving these young men to be cared for by the hospital staff would significantly increase their chance of dying. As one participant said, "When they have got Duchenne's they can go downhill really fast."

Hospitals Struggle to Support People with DMD

The following experience of this young man and his support worker is still common and shows how a lack of immediate care can quickly snowball into a serious event requiring an overnight stay.

Support worker   I did have to put in an official complaint at one point at the hospital to actually get people to sit up and take notice. Because we had waited six hours to see a doctor and they had already diagnosed him with having pneumonia and they did not even bring him any medication. And that was (a major) city.

Jonathon   Yes, they are notoriously unreliable.

Support Worker And then they said they were going to keep him in overnight on a drip and I said that is only because I have complained because you said he needed

urgent medication and we've been sitting here for over 6 hours and you have done nothing. You have not given him anything!

Jonathon   They were going to put me on an antibiotic IV drip and keep me in overnight but I said I wanted to go home so that is what we did.

Kate   You had to wait six hours for antibiotics?

Support worker And we weren't even in a proper ward, just a procedures room with no toilet, nothing, just a bed, not even a window. We were just stuck for six hours. I buzzed. And the nurse came in and said how can I help you? And I said you can help me by telling me what the hospital complaints procedure is. And then everyone fell over backwards.

Hospitals operate on the principle of Arthur Frank's (1995) "restitution narratives" which contrast with the "chaos narratives" of patients with DMD. Frank argues that restitution narratives are the prevalent discourses through which most patients and most hospital staff understand the purpose of hospitals. Frank argues that people's stories about health and illness are more than recounts of events. He considers them modern day myths that construct a shared reality and like fairy tales they are full of metaphoric significance. The underlying message of restitution narratives is that people recover. They become ill, they receive treatment, they recover. Illness is an interruption and patients are restored health. Frank argues that this narrative is powerfully constructed not only by sick people but also by hospitals. It informs their general practice. Their purpose is to restore people to health. However problems arise when restitution narratives are confronted by people who will not recover.

For people with DMD restitution stories are only ever partially relevant. They do experience serious bouts of ill health which require treatment and they do recover, but they do not recover like other people. They are still dependent on the assistance of others. Their illness narratives are those that Frank identifies as "chaos narratives" that describe their interminable and unrelenting physical fragilities and decline. DMD contradicts the restitution narrative of the hospital.

Some Young Men are Particularly at Risk

My research indicates that young men with DMD are particularly at risk when they are admitted to hospital and their family is not able to care for them. There can be a number of reasons for this situation. Where there are other family commitments it may not be possible for a parent to stay all the time, there may be other children to be looked after, possibly others with DMD. In some cases the health of the parent means the parent is unable to stay in hospital (see Chapter Six for discussion about the emotional/ mental health of primary carers). Sole parents face compounding difficulties particularly where their own mental (or other) health is fragile and/ or they have other dependents (with or without Duchenne's) to care for. There are a range of reasons why some parents do not stay in hospital all the time with their son (particularly an adult son).

Given the special care that those with DMD need as their deterioration progresses, these young men are particularly at risk. It is widely acknowledged by families that most general health professionals do not know how to look after those with the condition. Young men with DMD entering hospital without the support of parents or other immediate family face the added burden of complicated paths of communication. Where those providing care are not the next of kin communication becomes particularly difficult. In one case a young man with DMD was admitted to hospital with a collapsed lung and without the support of his parents. The pressure of his bi-pap machine was not altered to accommodate the fact that it now needed to support only one lung. This was of grave concern to his friends who repeatedly asked the nursing staff to check if this was right. They felt their concerns were dismissed (later an emergency department nurse explained that this would have been addressed although clearly the details were not explained to Gavin's concerned friends). Their status as friends rather than next of kin may have had a role in the communication process. Kerry told me about this situation. He described the worry and concern that both he and a support worker experienced, when the friend, Gavin, was admitted. The issue with the bi-pap was just one aspect of their concern. Primarily they were worried that the friend who needed a lot of assistance was alone, unless they were there and they could only be there during visiting hours. Gavin died in hospital and Kerry was realistic that given his fragility and the collapsed lung this may have been an inevitable outcome, but he continued to wonder if he had been more assertive, if the other friends had been older (Gavin's advocates were in their early twenties), more articulate, better able to challenge the hospital system, perhaps the outcome for Gavin may have been better. Kerry also voiced his opinion that when people with

advanced neuromuscular conditions enter hospital some staff impose their own ideas and values about quality of life onto people with disabilities. A similar point was made by another participant (see page 122) Kerry was not suggesting these were explicitly stated but he was quite sure that such an attitude did exist. His view refers to a Bourdieuian distinction between rules (for example the Nurse's Code of Conduct) and practices which are informed by their habitus – learned, reinforced attitudes and behaviours that become automatic over time, or 'body automatisms'(Bourdieu 1990: 68). Much has been written on the way people with impairments experience discrimination (see for example Coleridge 1993, Leach 2008, Morrris 1991, Novas 2006 and Oliver 1996), so Kerry's assessment of the attitudes of some hospital staff fits within this wider discourse.

People with neuromuscular conditions need extra care when they are in hospital but as this care is not well provided by the DHB, some people feel they have to break the rules by using their community support workers. During my fieldwork a Facebook posting was used to voice frustration that the workers whom individuals rely upon when at living at home are not allowed to work in the hospital. The advice from others, who had similar experiences, was overwhelmingly to not tell the agency about the hospital stay. This is not ideal. That people with DMD have to do this indicates that services are not being provided in a manner that respects the dignity and independence of the individual. This "don't tell" approach has potentially negative repercussions for the disabled person and their support workers. Neither agency workers nor those provided for under individualised funding are meant to work in the hospital. The inability of people to access their usual support workers, who are experts in their specific, individual needs, whilst in the hospital system has serious consequences for people with DMD. This can be seen in Ben's following interview comment:

> … a couple of months ago I started having heart issues. I knew I needed my medication put up. And I couldn't just go and see the doctor, I had to go through ED (the emergency department) and spend hours in the hospital. I spent hours and hours in ED and the doctors finally agreed to put my medication up slightly. And all it needed was for the specialist to say, "I know him, I'll put his medication up". It took ages, I only needed to see him for five minutes and I can't afford to miss meals and stuff. That is the problem with ED. I have set food at set times. And that is the trouble with the hospital, we are actually not allowed to access caregiving services (i.e. be fed by a support worker) because the nurses are supposed to do it. That is one of my biggest issues with going to hospital. The nurses don't want to help. Well I understand why they don't want to help – they are too busy. Technically we can't access our funding. I don't want to risk it.

Limited Health System Knowledge a Cause of Concern

Universally families and individuals are frustrated at the lack of knowledge about the condition within the general health profession, as the following part of my interview with Harry shows. At the time of this interview Harry and his family had just been to look at the cough assist machine at their local hospital.

Margaret   The other thing that is really surprising is the lack of knowledge in the hospital system about muscular dystrophy. I first phoned up when we went down [to the city where the hospital is] (rural family, required to travel to hospital).

Harry   Oh, the "does he have trouble walking?" comment!

Margaret   That was really funny, we had a phone call on the way, "Would he need assistance walking?" If we could get him walking while we are here that would be great! And in the morning we got woken up by the porter, "Does he need a wheelchair?" "No he does not need a wheelchair; he has bought a chair of his own."

Kate   That is something I have heard from families, the constant need to educate. So if you get really sick and you need to go to hospital you actually need someone there to explain your condition all the time. Every time there is a change in shift?

Margaret   Yes, otherwise they just abandon him and go off and see another patient and he needs someone there all the time. He can't be left in a hospital at all, by no stretch of the imagination.

Kate   So you need someone there to advocate on your behalf all the time.

Harry   I need someone who knows what they are talking about.

Margaret   And they get quite panicky don't they … "Oh, is someone staying with you ... oh… you might take up some extra staff time in here."

Harry   Well the doctors are alright, it is just the general staff they don't have any idea.

Kate   So when you say general staff, does that include nurses?

Harry   Yes

Margaret Most people don't know at all, do they?

Harry   No, most people do not know what to expect.

Margaret Like he had a blood test and the lady came in and said could he just lift his hand and hold that down like that. Well of course he can't do that, they are just on auto pilot, aren't they?

Kate   So this shows a lack of knowledge about the condition.

Margaret Today they asked him to cough, he can't cough.

Harry   That's right, "take a big deep breath" (Laughter).

Whilst the participants in this particular interview relied on a good sense of humour when explaining their surprise at how little nurses and others understand DMD, this same lack of knowledge had serious life-threatening implications for other participants.

Supplemental oxygen (a common therapy for many patients presenting with breathing difficulties) is known to be dangerous for patients with DMD. Best practice guides recommend non-invasive ventilation (the use of a c-pap or bi-pap machine) for initial treatment and where oxygen is deemed necessary the amount of CO_2 in the expiration breath or blood should be closely monitored (Bushby et al 2010b). This following interview segment reveals a serious lack of knowledge within a hospital of how to treat a DMD patient presenting with respiratory issues. In this case the young man was obviously very sick and the lack of knowledge and the fact that the health professionals did not listen to the patient or his family could have caused his death. This incident occurred on a weekend so the respiratory specialist was not at work. It appears that the staff did not consider the possibility that the patient himself was likely to have expertise in his own condition.

Christopher   So I just ended up in respiratory failure because of bad hospital management. They gave me oxygen.

Kate   And you should not have oxygen because you have Duchenne's?

Christopher   And they did not know that.

Grandparent   His mother and one of the teachers more or less held him up all night so he could breathe. Otherwise he would have died.

Kate	So one of your teachers was there?
Christopher	A family friend. So what happened was I was dehydrated so they gave me a litre of fluid an hour, so of course my bladder was not ready for that. And then I couldn't breathe. I couldn't go toilet. I got squashed, and couldn't breathe. Then they gave me oxygen which really was a no, no. And I knew but they didn't listen and that just pushed me into respiratory failure. My oxygen saturation went down and I was blue.
Grandparent	It was touch and go.
Christopher	They were basically breathing for me. Pulling me up and down.
Kate	So they didn't have a ventilator to help you, it was Mum and a family friend?
Grandparent	They (hospital staff) did not know what to do.
Christopher	They were confused. We were asking for them (ventilator) and they gave me oxygen. They gave in and decided to put me on a bi-level after 5 hours.

Despite this poor treatment which has had serious implications for Christopher's ongoing health, in that he has been unable to manage without a ventilator since this incident, he decided not make a complaint as he still needs to go to the hospital and wants to maintain relationships with the staff there.

This final comment demonstrates how another frequently mentioned issue – the exhaustion parents experience after several days of caring, advocating and worrying for their sick son – combines with the lack of awareness about just how vulnerable those with DMD become and how careful staff need to be, both with their medical treatment and the normal daily care of the body. Rhonda comments:

> I think it was lack of experience in that condition too. A lot of things like lifting, I remember when he was on the heart ward they were going to lift him without a hoist. And I had to say, "No you have to use a hoist or you could dislocate his arms." Just ignorance really, the nurses will just grab a patient and lift them without thinking. But that is just one of those things; people are going to be ignorant so you do have to be there all the time. I think I might have had two nights at home, but then you are just worrying about what is going on while you are away. But once you've done a couple of shifts you start to feel faint. And having to deal with a team of doctors, when you

are like a zombie it becomes really dangerous, you can't take things in and it is hard to advocate and the whole thing is horrendous.

Fieldwork Findings

Those who are familiar with DMD are highly aware of the risks associated with hospital admissions. This risk that members of the DMD community have when they go into hospital was bought home to me during my fieldwork when a man with DMD, Anthony, was admitted to hospital with a chest infection while his parents were away. I went into the MDA National Office to do some voluntary work and found Kerry in a distressed state. He had heard that his friend was in hospital and immediately knew this was bad news. He told me that without family members to advocate and care things could not go well. He wanted to go and visit his friend to see what was going on but his fear of contracting a respiratory infection prohibited this. There was no way Kerry was going to risk going to a ward of people with respiratory issues. Instead he was relying on others in the DMD community to keep him informed about how his friend was managing. Although they were not officially supposed to be there some of the usual support workers did attend to Kerry's sick friend during his illness. Some of his support workers did this regardless of whether or not they would get paid. They knew this was a crisis situation. Anthony needed constant monitoring and care and even though support work is low paid and the support workers had to pay to be at the hospital (parking and organising their own childcare) they knew he would not make it without them.

Other friends from the DMD community – parents responsible for their own sons and other family members – also spent time at the hospital. These parents had some relevant experience. They knew how to care for a person with DMD- suctioning saliva, monitoring ventilators, turning, toileting and so forth. Those support workers and DMD community members who were involved in caring for Anthony were not participating in the curative approach, in "modernist medicine" (Frank 1995:145) and the hegemonic restitution that accompanies it. These carers accepted Anthony's suffering. Frank suggests that "Modernist medicine has regarded suffering as a puzzle to be 'controlled' if not eradicated. Postmodern illness culture, lay and medical, recognizes a need to accept suffering as an intractable part of the human condition" (1995:146).

In following this line of argument the role of the support worker is incongruous within the hospital system. Two different models of care are involved. The purpose of one is to overcome suffering, to cure it, the purpose of the other is to accept suffering, to acknowledge

it and to relieve it as far as possible. However in understanding, accommodating and addressing Anthony's suffering and as time progressed these women became increasingly exhausted. Kerry asked me to find out why Anthony's usual support workers were not meant to be at the hospital and what was supposed to happen.

At this point I came to fully understand how difficult it is to negotiate with bureaucratic processes. As I was not directly connected to the individual concerned I could only ask about general principles and not this case per se. Part of the difficulty was that there were no next of kin able to be at the hospital so this young man had to advocate on his own behalf and he was too sick. I rang many DHB departments and help numbers most of which were automated voice machines asking me to leave a message. As this was a fairly urgent situation this was a frustrating process and few calls were returned. I called the Health and Disability Commissioner and the hospital chaplain to try and find who might be responsible for making sure appropriate support was provided. While the charge nurse on the ward and the hospital social worker were involved in talking to the patient about his support those supporting him said that the focus of these discussions was about his return home and not about who was going to look after him while he was in hospital. By the time I received replies to my calls and emails this young man had died. Given the advanced stage of his DMD his death was not surprising, but the awful anxiety of those trying to care for him without official sanction or a formal role as next of kin highlights a shortcoming of the rigid funding system. As with similar cases, this shortcoming of the health system remained unaddressed as the family and those who were involved in the last few weeks of his life, dealt with their inevitable grief.

The overwhelming opinion of the participants is that DHB provisions for ward staffing are simply not adequate for care of people with DMD and other similar conditions. These high needs patients experience the limits of state care which result from complex interactions between economic systems, staffing arrangements, bureaucratic edicts and moral practices of valuing life.

Duchenne muscular dystrophy is a life-shortening condition. This is well recognised by medical professionals (the neurologists and respiratory specialists involved in managing the health of young men with DMD, and other hospital staff), the young men with DMD themselves, their families and permanent support workers. The availability of support workers in the hospital setting will not alter the inevitable outcome of the condition. However their availability improves the likelihood of a person with DMD surviving each hospital

admission and as such the policies that constrain this are an aspect of local biology. The key biological difference that the availability of support workers familiar with the needs of the patient affects is life expectancy. The particular difficulties faced by men with DMD who are unable to be cared for by family members when they go into hospital increases the chance that they may not survive. Policy has a role to play in the creation of a particular local biology of DMD in New Zealand. This policy impacts on life-expectancy. It is part of "the shifting material reality, historically patterned by society, culture, economics and politics" (Lock and Nguyen 2010:108). While DMD is most often described in "standard biological terms" (Lock and Nguyen 2010:108) the way it is currently experienced in New Zealand is the result of "historical interactions between culture and biology". It is irrefutable that the social context of treatment does cause biological change and that change relates to longevity (Emery, Muntoni and Quinlvan 2015:222, Rahbeck et al. 2005, Rodger et al. 2012). The policy that prohibits the use of support workers, paid from the DSS appropriation, from providing assistance in hospital is a life-threatening example of a local biology of DMD.

Funding System

The current funding system with two clearly delineated funding streams exists partly as a result of welcome changes in attitudes towards disability. The New Zealand Public Health and Disability Act, 2000, shows disability is now understood as a social issue, rather than a purely medical or individual issue. The purpose of the act clearly states that it should, "achieve for New Zealanders … the promotion of the inclusion and participation in society and independence of people with disabilities." This legislative requirement results from years of advocacy by the disability community (Beatson 2004, Office for Disability Issues 2002). The disability community has engaged in challenges to strategies of biopower "a very real process of struggle; life as a political object (in this case a life with disability) was in a sense taken at face value and turned back against the system that was bent on controlling it." (Foucault, 1978:145).

The New Zealand Public Health and Disability Act, 2000, is the Act upon which the current health system is based. It provides for the ring fenced-money within the Ministry of Health to facilitate the participation of disabled people in society. This money is distributed by Disability Support Services (DSS). This ring-fenced money means that some services are dedicated solely to benefit those with disabilities and cannot be siphoned off for other purposes. Thus, DSS can be seen as a generally positive move on behalf of the state to

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support people with disabilities. It can be understood as an agent for promoting "regime change" and a "subversion of established order" (Shore et al. 2011:3) in that it is a policy that addresses the exclusion experienced by many people with disabilities (Beatson 2004, Office of Disability Issues 2002, Moore and Tennant 1997). The disability community in New Zealand has engaged in strategies of biopolitics to challenge the social structures which excluded them. As such the establishment of the DSS shows how one subaltern, dissenting group has engaged with public policy resisting and challenging their social exclusion and influencing change (Pèro 2011:223). But it has the unintended consequence of separating hospitals from other regular parts of life. For young men with DMD hospitals are a recurrent part of normal life.

During my attempts to discover why Anthony could not have his support workers with him in hospital I had a phone conversation with a Relationship Manager, responsible for NASCs, at the Ministry of Health. He explained the rationale for the way the Ministry provides services for disabled people. In line with the New Zealand Disability Strategy: Making a World of Difference – Whakanui Oranga 2001 (Minister of Disability Issues 2001), the justification for the DSS pool of money is to support participation in the community. Any service that is to do with improving health or treatment or rehabilitation must be paid for by another pool of Ministry of Health money, the DHB stream. Any personal care (work undertaken by support workers) that a disabled person under the age of 65 receives is to assist that person to participate in their local community. Once a person requires hospital treatment DSS money cannot be used to support them, all needs must be met by the DHB. For a person to continue to receive care funded by the specific disability appropriation whilst also under the care of a District Health Board is viewed as a duplication of payment ("double dipping"). This is because it is the DHBs responsibility to provide services and they are funded to do so. DHBs, like all other service providers should aim for 'normalisation'. The goal is to remove the barriers to the participation of the disabled person in society. If someone needs a service that service should meet their needs regardless of what those needs are. These beliefs and values that underpin the existing system appear to be positive for people with disabilities – facilitating participation in the community complemented by a barrier-free health system. However, there is another underpinning rationale that is implicit in this model of service provision. The explicit rationales that the Ministry of Health relationship manager offered are part of a complex assemblage and integral to the assemblage are budgetary constraints. Shore et al. (2011:3) note that, "policies reflect the rationality and assumptions prevalent at the time

of their creation." The policies that inform the practice of DSS show the challenges of the disability community to their social exclusion, the assumption (which disability activists helped to inform) that discrimination towards people living with disabilities is structural and institutional and should be addressed. They also show the assumption that capped budgets for population based funding is the best way to provide health services. This rationale of capped budgets was not referred to by the relationships manager I spoke to, perhaps because it is so entrenched in the health system that it has become a taken-taken-granted, common-sense understanding, a self-evident doxa (Bourdieu 1977 (1972):164).

However, as well as being an unquestioned part of the health system the capped budget approach also acts as a policy of concealment. Shore et al. (2011:8) note that policies are often presented as "apolitical and self-evident" but in fact (citing Cruikshank 1999) can be sites where "power is hidden from view and present(s) no visible targets to oppose or resist." This analysis is useful to consider in the context of this particular situation. A great deal of power resides in the way policies of capped budgets are implemented in the health sector. This approach to funding the health system is regularly challenged when individuals or groups of individuals find their own needs are not met by this system (for example the recent campaign to get funding for the skin cancer treatment, keytruda, RNZ 28.6.2016). However in the case of the use of support workers in the hospital, the power of the policy is hidden because in a situation of extreme distress there was no visible target to address concerns to. Those involved in the care of Anthony could not pinpoint who to address their anxieties to. Although they knew about the rules regarding support workers working in the hospital, none of those who were so anxious about the situation knew that the DHB was supposed to provide a barrier free service. I am sure they would have been amazed to discover that their predicament was part of a barrier-free service. These carers (support workers and parents) worked in partnership with the ward nurses to look after Anthony but as they were not next of kin they found it hard to access anyone higher up the hierarchy. Similarly for Ben, he knew the policy about not using paid support workers in the DHB but did not know how to ensure that the DHB service met his needs. He did not know this because the DHB did not provide such a service.

This is an example of the "messiness and complexity" of policies (Shore 2011:8). The establishment of DSS and its reliance on the entrenched capped budget model of service provision does not create a transparent and fair structure for the provision of disability services, despite the explanation of the Relationship Manager from the Ministry of Health.

For those with DMD it creates an opaque system where those whose needs are exceptional continue to be excluded. Frank in his discussion about the incompatibility of restitution and chaos narratives recognises the inability of administrative systems, and in this case the Ministry of Health manager, to take account of suffering "The pedagogy of suffering is my antidote to administrative systems that cannot take suffering into account because they are abstracted from the needs of bodies. When the body's vulnerability and pain are kept in the foreground, a new social ethic is required (1995:146).

Statutory Requirements for Person, Whanau or Patient-Centred Care

These funding boundaries are clearly important to the management of health and disability services but they operate in conjunction with other guidelines and requirements including; The New Zealand Health Strategy 2001, The Disability Action Plan 2014 – 2018, Whaia Te Ao Marama: The Maori Disability Action Plan for Disability Support Services, 2012 to 2017, The Health and Disability Code of Rights and finally, The Health Quality and Safety Framework (Ministry of Health 2011).

All these documents refer to the principle of person, whanau or patient-centred care. The clear implementation policies involving; funding agreements, ministerial, official and community based relationships, and reporting requirements indicate that these documents are mandatory. Government systems should adhere to the principles and goals outlined in them. However, as the ethnographic data in this chapter demonstrates, there is an inconsistency between statutory requirements to provide person-centred care and the current funding model. The following examples reiterate the statutory requirements for patient-centred care:

Table 2, Statutory Requirements for Person-centred Care

Document	Vision
The New Zealand Health Strategy 2001	"… ensure institutional boundaries do not compromise quality of care…" "… Ensure services at all levels of the health sector fully reflect the needs of individuals and communities."
The Disability Action Plan 2014-2018	"Promote disabled people having choice and control over their supports/services, and make more efficient use of disability support funding." "Increase government services' responsiveness to disabled people."
Whaia Te Ao Marama: The Maori Disability Action Plan for Disability	"Enabling Maori Disabled. Greater personal leadership, choice and control

Support Services 2012 to 2017	over disability supports accessed." "Delivering high-quality, effective disability support services. Culturally safe and trustworthy disability support service."
The Health and Disability Code of Rights (Right 4-3)	"the right to have services delivered in a manner consistent with his or her needs"
The Health Quality and Safety Commission	"New Zealand will have a sustainable, world-class, patient-centred health care and disability support system …"

Contested Space

These strategies, codes of rights and visionary statements demonstrate an understanding that health services are most effective when they acknowledge the needs of the person or whanau requiring assistance. Although now accepted as an appropriate way of delivering health (Boon 2012, McCrae 2013, Neuwelt and Matheson 2012), this construction of how health services should be delivered exists within a contested space. Historically, the views of the medical expert tended to dominate ideas about the management of the ward, such as how people with disabilities would be cared for, and decisions were made by senior medical staff. While criticisms of this approach abound (Gibbs et al.1988) it did mean that those senior staff could take into account the needs of the individual in their decision-making. The health reforms of the 1980s and 1990s were driven by the spiralling costs of the health system and changing political ideologies. They introduced a new style of hospital management in which fiscal considerations dominated and budgetary constraints were implemented. Clinicians could no longer make independent decisions about ward management. These decisions now had to be considered within the context of finite budgets applied to each service. This change is evidenced by the attendance of a busy neurologist, at a teleconference call attended by the Deputy Commissioner, Disability, for the Health and Disability Commissioner, to support the request of the MDA to change the policy of not permitting the usual support workers to work in the hospital setting for people with DMD (and others who rely on support workers). The attendance of the neurologist at a telephone meeting instigated by a NFP organisation in an attempt to instigate change at the organisation that employs him demonstrates just how opaque and non-transparent policies of capped budgets have become. This was reinforced when the HDC Deputy Commissioner was unable to advise how the MDA should proceed in

advocating for this change. Her brief only permits her to consider individual complaints about unsatisfactory treatment and not the shortcomings of the health system per se.

Increasingly fixed budgets are applied to smaller and smaller parts of the health system. I discussed the use of support workers by people attending emergency departments (ED) with a nurse who had experience working as a district health nurse (a community-based nurse) working in the residential part of the Wilson Centre (a centre for children with disabilities) and in paediatrics before her work in ED. She had knowledge about DMD both for children and adults and was aware of the high levels of support that accompany advanced DMD. She was not aware that officially people with disabilities are not meant to bring their support workers into the hospital. She also explained that it would be impossible for an ED nurse to provide the sort of support that she knew was required. The job of the ED is to triage patients and to save life and limb. The ED has to ensure that the most urgent cases are seen first. This nurse was aware how much time the support and monitoring DMD required and knew that the nurses just could not do it. Their job was to keep people alive and that always took precedence over other duties. She noted that there was a six patients to one nurse ratio unless it was a resuscitation situation, in which case there was a 1:1 ratio. The nurse I spoke to said, "We love it when patients come with support workers. There is no way we could provide for the activities of daily life." The doctor referred to the support plans that people with conditions like DMD often have on their records and how the ED staff endeavour to follow these plans and have patients admitted to the ward quickly. He was very familiar with the policies and procedures of the emergency department but he too was surprised to hear that the people who came with support workers were not actually meant to do so.

The increasing fracturing, or segmenting, of the health system by the allocation of budgets was highlighted when one ED staff member mentioned that recently the ED they worked in had stopped doing blood tests. Previously if a patient was to be admitted to a ward the ED would do the necessary tests so that the results were ready in a timely fashion for the ward doctor to facilitate treatment. Recently the ED had stopped doing these tests as they were charged to the ED budget. If the doctor on the ward wanted to know the results of blood tests the ward would have to organise and pay for the test out of their own budget. This was an ED management decision and shows how the system of fixed budgets applies on a service by service basis, which causes managers to make decisions that prioritise the boundaries of the budget rather than the overall care of the patient. These concerns, about stretching the money allocated to each fragmented area of medical service, apply throughout the health system.

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Despite the requirements that DHBs should provide services that meet the needs of all patients, this system of fragmented funding does not facilitate patient-centred care when the patient has additional needs that fall outside the boundary of the particular budget. The question of which budget, or how a budget, will cover the activities of daily life becomes a difficult issue. It is an unusual and unexpected cost. In the case of Anthony, in addition to not being informed that the DHB had a duty to provide services to meet the need of the individual, the support workers and parents knew that people had to be properly trained to do this work. The type of support that Anthony needed was not something that just anyone could do. The situation of those involved in this policy network was complex and included "incompatible managerial demands and expectations" (Shore et al. 2011:20). In the case of advanced DMD it is not possible for the DHB to provide appropriate, barrier-free care without an individual's usual carers.

DHBs negotiate their funding requirements with the Ministry of Health and receive an equitable share of the Vote: Health money to implement agreed health services. DHBs must also adhere to the policies established by the Ministry in the implementation of service. The current notion that person/ patient/whanau-centred care is the optimal way to provide health services exists in a contested space. Currently people with DMD, their whanau/family and specialist clinicians all agree that the use of usual community support workers will have the best health outcomes for the patient, but the constraints of Ministry of Health funding policies prohibit this. This is despite the fact that Ministry policies aim for a barrier-free health system.

People living with severe impairments find themselves in an unfortunate position of being caught between conflicting ideologies and bureaucratic imperatives in a dynamic and changing health system. This contested space suggests a shift is occurring in the ideological landscape. The funding model for service provision described here has been designed to enhance the lives of disabled people. The underlying rationale is to have a pool of money that can be used to support only people with disabilities to live independently and to participate in society. For people with DMD it is something of a false dichotomy to distinguish between their everyday life and their hospital admissions. Their everyday life involves managing their increasingly fragile health and the possibility of hospital admissions.

The situation is further complicated by the requirement for each DHB to establish its own policies and procedures to manage the way care will be provided. Again the language of these

official policies is inclusive and based on notions of person or whanau-centred care. The terminology used includes; family, whanau and patient centred care, collaborative care planning, Health and Disability Commissioner (HDC) Health Passport, trendcare system for hospitalised patients, multi-disciplinary team (involving medical, nursing, OT, physiotherapy, social workers, hospice and Hunga Manaaki staff), Clinical Nurse Mangers – ensure patients are cared for, and a comprehensive care plan.

These DHB documents do not refer to budgetary constraints regarding the provision of personal care and this is an important issue. Some of the responses from the DHBs were couched in terms of progressive verbs such as "embedding" and "developing". The HDC Health Passport is a new initiative. Whilst these new developments may eventually benefit those with severe impairments in the health system, participants in this research lack faith in them. Participants referred to constant changes in the health system and changing rhetoric, but no real change in service. If a truly patient-centred approach to the provision of care is to be implemented then either there needs to be flexibility between budgetary boundaries or additional capped budgets to facilitate person-centred care must be provided.

Chapter Conclusion

In the previous chapters of this section of my thesis, which address the bureaucratic entanglements confronting the DMD community, I have reiterated Foucault's analysis of biopower and governmentality that is based on the premise that the rationality of government in contemporary capitalist societies, like New Zealand, increasingly concerns managing populations as a whole rather than addressing the requirements of individuals. Here we see the start of a social shift, not just in advocating that the needs of the individual be better met in particular circumstances, but with the institutional and social acceptance of person-centred care. In the case that I discuss in this chapter, the ability of support workers to work in the hospital, this change could occur without additional cost to the health system as the funding for support workers has already been allocated. However, the wider application of this approach requires a shift in the way services are provided. Arthur Frank's advocacy of a new social ethic offers an idea about the type of shift required. He argues for a pedagogy of suffering as a moral change in which multiple voices are recognised and respected. "Ultimately moral values require a complementary politics with attention to inequalities of power" (1995:152). In such a recalibrated system the (currently disempowered) voices of those who were aware of the risks of the present, rigid funding system would be recognised

and respected. Furthermore these voices could offer a moral orientation that shows how suffering can be accepted (not just cured) but listened to, acknowledged, and alleviated as far as possible. The allocation of fixed budgets onto increasingly fragmented aspects of the health system entrenches the fragmentation of service provision. Improvements to person-centred care require a change that allows for provision to transcend rigid, inflexible, bounded entities. The role of policy is central as policies currently create and reinforce the rigidity of the system. The policies in this thesis determine what money can and cannot be spent on. They are about the control of smaller and smaller levels of governance. This allocation to increasingly smaller regimes of authority conceals the complexity of human life, particularly lives that are affected by this single gene mutation that has such systemic impact on the human body. This chapter demonstrates that the pervasive dominance of this way of organising the health sector creates an insurmountable health risk, an impenetrable barrier, for some people living with DMD. Despite the ideological belief that this system provides transparency, that it is clear how services for people with DMD (and other similar severe disabilities) are provided, the system is opaque for those immersed in it. Here I refer to the meeting I attended as a doctoral candidate who could provide relevant research data. This meeting was also attended by the CEO of the MDA, a neurologist; Miriam as a key liaison between the DHB and the MDA; and the Disability Commissioner for the HDC. These were all highly knowledgeable experts and even for us it was not clear how best to address these concerns. We came to understand there was no one point of reference, one person or office within the Ministry of Health, where these concerns could be addressed in a straight forward fashion. So if the system seems opaque to a group of concerned experts how much more difficult must it be for those in situations of crisis dealing with grave illness. The pervasive dominance of financial transparency does not create clarity for those receiving services within the system. The message that the rigid boundary between DHB funding and DSS funding gives to the DMD community is that the policies which control spending are more important than the care that directly supports their survival. This chapter, following Fassin (2012:112) is an analysis which addresses the politics of life and the worth of lives. While not an intention of the ring –fenced funding of DSS, the outcome, in this case, is a life-threatening, invisible barrier that cannot legitimately be traversed.

Section 3 Conclusion: Bureaucratic Entanglements

In Section Three I aimed to create a sense of the repeated and ongoing frustrations that the DMD community faces as people attempt to negotiate a path through multiple bureaucracies and service organisations. Not only were such negotiations a recurrent experience of daily life but they were accompanied by an increasing sense of injustice as again and again individuals and families found themselves falling outside the parameters of service provision, not able to access life preserving or enhancing equipment or sufficient support to maintain health and independence. Their experiences fell short of the inspirational vision statements of government and other agencies that reiterate social values of acceptance, inclusion and participation for people with disabilities.

People with disabilities form an enormously diverse group and their requirements are also diverse. The needs of people with DMD are high and even within the wider disability community they form an anomalous group. Foucault's analysis of biopower, where state assistance for unexpected circumstances provides a general level of security, not one specific to individual need, helps explain the repeated exclusion of people with DMD from appropriate service provision. Their exclusion is reinforced by the particular structure of the New Zealand health system which is based on a hybrid neoliberal philosophy. Some parts of the system are classic free market, private businesses. Other parts of the system are based on internal markets where one arm of government purchases services from another government agency. In other arrangements NGOs fulfil the role of service provision. These diverse arrangements by which the state now fulfils its duty of care for citizens means that people with DMD, who need comprehensive assistance, engage with numerous agencies and organisations. Whichever type of agency people are seeking services from, policies act as gatekeepers to service. They are key parts of the service assemblage.

As Shore and Wright note, although policies are presented and written with impersonal language, as if they are "pragmatic, functional and geared to efficiency" (1997:10) in fact they are morally loaded and can conceal and obscure political agendas. Shore and Wright point out that in the shift from the social democracy of the welfare state (the model of governance through which New Zealand's healthcare was provided from 1938 until 1984) to the current neoliberal era of individual responsibility, policies have not reduced regulation, as was intended, so much as made "the regulatory power of the state more diffuse and less visible" (1997:29). Foucault argues that one aspect of governmentality is that people are

inculcated with dispositions so that they will be responsible for themselves. Policies have a part to play in this, by not fully meeting the needs of some people with disabilities (including those with DMD) there may be an expectation that people will find ways to manage themselves. In some cases this may be effective but this is generally not the case for those with DMD. The examples of entanglements with bureaucracies considered in this section of my thesis show that people with DMD, particularly those whose families are not wealthy, are not induced to govern themselves. Partly because they cannot and partly because they refuse to. Their disabilities prevent them from participating in the workforce and their lack of income prohibits people from providing services autonomously, or acting as the type of responsible, self-governing individual that Foucault identified in his lectures regarding technologies of self. People with DMD refuse to self-govern in this way and instead repeatedly contest and challenge bureaucratic decisions, instigating and participating in assemblages that talk back to power. This section of my thesis shows the tactics that members of this community use to respond to their exclusion, to talk back to policies and power. In terms of technologies of self, people with DMD are not docile bodies that acquiesce to the technologies of governance, the policies that marginalise them. The rapid degeneration of muscle function, which increasingly impacts those muscles associated with vital organs such as lungs and heart, invests these contestations with a sense of urgency and immediacy.

The cases I consider in this section of my thesis demonstrate that care and concern for the health system takes priority over the care of people with very high levels of disability. The lack of guaranteed access to either to an effective system of respiratory care or a cough assist machine is clear evidence of an inadequate system that, as Foucault (1997:247) suggests, lets people die. The prioritisation tool described in Chapter Nine is an acknowledgement that the current system is inadequate. The difficulties that families have in challenging the exclusions of existing policies and systems reiterates that these technologies are not transparent, rational documents but rather operate within complex assemblages where they can serve to conceal political agendas. The current situation does not provide the additional support that people with DMD need when they are in hospital. In order to fulfil the requirement to be; person-centred, responsive, collaborate across agencies and ensure that services reflect individual needs, the delivery of support services to people living with severe impairments requires some flexibility. Failure of the Ministry of Health to recognise that people with severe impairments may need their community support workers with them in hospital, at times, reiterates a repeated experience of the DMD community that policies and funding models are

currently more important than the health of individuals. In New Zealand the rigid structure of different funding channels outweighs the views of the DMD community, neurologists, ward staff and respiratory specialists, all of whom acknowledge that community support workers provide the best care for people with DMD when their families cannot be with them in hospital. The needs of people with DMD are an anomaly in a system designed to address average needs.

As I note in Chapter Ten, the hospital system is also designed to fulfil a restitution narrative. The care provided in hospital is primarily for recovery. The terrible situation that Anthony and support workers and friends encountered reveals even more of the complexities of the situation. The bureaucracies (and the epistemologies entangled in them) and the moral orientations of the support workers to 'go the extra mile' with their caring are focussed on contradictory notions of what good care is. These different notions reflect Fassin's (2012) analysis of the value of life. Hospital care is a about the biological value of life, it is about compartmentalising the systems within the body and fixing parts of the system in need of repair. The support workers care is about the political worth of lives, it is about acknowledging and responding to suffering and accepting that people will not be returned to full health. This consideration of bureaucratic entanglements reveals the complex ways in which lives with DMD are valued.

Chapter 11: Thesis Conclusions

At the end of 2011 I made my first foray into the world of DMD. In September I enrolled as a doctoral candidate and in November I attended an inaugural Disabilities Studies conference where I met some of the MDA fieldworkers and National Office staff. By mid-2012 I began volunteering at the MDA office, and had set out, with some trepidation, to conduct my first interviews. I met fieldworkers from across New Zealand and as a result attended the MDA children's camp in October 2012. I continued with fieldwork, attending camps, social events and conducting interviews for two more years. People generously shared their stories and time with me and some in the Duchenne community have become long term friends. Here I share some reflections on my personal involvement with DMD, summarise my findings and reflect on the theoretical frames I used and the implications of this research.

I have used the staple tools of social anthropology, participant observation and key informant interviews, to understand the lived experiences of people living with DMD in Aoteraroa New Zealand. In addition I examined legislation, policies and the historical antecedents that contribute to the particular and distinctive context within which people with DMD live. The theoretical perspectives I adopted assisted in the final analysis. Actor-network theory allowed me to see how all actants were enmeshed in the technologies of governmentality, the often unacknowledged beliefs and structures that frame policy and resource allocation. So much of the disability literature is about struggle, difficulty and limits that reading about hope usefully matched the data. The hopes I have discussed are both the small hopes that each day will be a good day, hopes with short, immediate timeframes and hopes with much longer trajectories. Local biologies allowed me to see the diversities and commonalities of lives lived with DMD. Each theoretical approach added a particular facet to this study and contributes to knowledge generally. Actor-network theory offered an analytic frame particularly for policy within the disability sector. The discourse of hope allowed me to identify a distinctive relationship between time and hope. My use of local biologies emphasises the importance of sociocultural factors even within the biomedical considerations of this predictable, genetic condition.

Personal Reflections

From my first tentative steps in 2011 to a kind of 'baptism by fire' at the children's camp in 2012 and ensuing interactions I gradually came to appreciate some of what it means to live

with DMD. The thesis that I present here focuses on the two contradictory discourses that emerged during this research. The frustrations with bureaucracies that individuals with DMD and their families repeatedly endured was unmistakable. People would get cross and upset again when recalling such experiences. These were difficult and emotional parts of interviews and for this reason they stood out, they were an unmissable common thread of this research. The enthusiasm with which people spoke about the way they created meaningful and purposeful lives provided a distinct contrast. People spoke eagerly about the things they enjoyed doing. I recall leaving one interview and feeling as though I had been at an introductory workshop for computer coding. I came to share with the research participants their sense of injustice at the intractable systems they encountered and their sense of relief when things went well.

The special and sad nature of this research added an extra dimension to the research process. I interviewed recently bereaved, grieving parents and young men in very fragile health who talked about not wanting any more medical interventions, who had asked for DNR (Do Not Resuscitate) to be added to their medical notes. This made that task of analysis difficult. Some days I felt like the grief was unending although, of course, I was getting an unusually concentrated dose of it. I was asking people to share sad memories – diagnosis, experiences of service provision, and memories of much loved family members. I interacted with the grief of those interviews many times, initially recording the interviews and then transcribing them, coding data, analysing and writing up. Although people had agreed to be interviewed, I had to think carefully about how I used the information they gave me. My intense experience of grief was partly to do with the interview process. I realised that people were not constantly experiencing the grief that was expressed in some of my interviews. Some of this grief was recalled, which is not to diminish it, but when a parent or sibling sits in an interview and describes the circumstances surrounding the death of their much loved relative that action recreates specific memories and emotions of a very sad event. The sadness that family members live with on a daily basis seemed different. The daily grief is about missing their son or brother and remembering him in many ways and not necessarily recalling the specific details of his death. Interviews with people who had a recent diagnosis or a recent death were inevitably fraught but in some of the other interviews people were describing experiences that were historic and the interview process was the catalyst for sad recollections. I have tried to balance the sadness inherent in my research findings with the fulfilling activities that people talked about and which I saw during fieldwork. I find writing about Kerry especially hard

because he became a good friend and he facilitated my research, taking me along to ice skating and powerchair soccer and introducing me to some in the community. Initially I followed the anthropological tradition of giving Kerry a pseudonym but it just felt wrong. Also, as he was the only person with a neuromuscular condition working at the National Office when I did my voluntary work, it was unrealistic to think that a pseudonym would offer him much in the way of anonymity. Miriam suggested that if I was going to name her, perhaps I might consider using Kerry's real name too. Kerry had passed away by this time so I was unable to ask his permission. As my thesis developed into its final form I realised just how difficult the ethics of anonymity are in a small community. For Kerry I consulted with family and for others sent parts of chapters so people could see how I had used their stories and changed identifying details.

Loss and grief are an inherent part of the Duchenne condition. People talked about the difficulties of losing ability as well as the particular grief associated with death. There are many different types of sadness for DMD families but as well as grief I learned about commitment. For some families the troubles that DMD brought to them also brought them close together. The devotion within families was noticeable as each family devised ways of providing care for the person with DMD. The focus on ensuring that the person with DMD received the best possible care shaped much of family life as parents and siblings worked out how to meet their own needs as well as the needs of the person with DMD. The families tended to play this down saying they really had no choice but it struck me as quite remarkable. Through my observation of some DMD families I became aware of the very close bonds that exist in the families that participated in the research. Clearly those families where things were going reasonably well were more likely to participate in research, and there is debate about a higher rate of marital separation or divorce when parenting children with disabilities (Sobsey 2004). However the desire to provide the best possible life for the person(s) with DMD characterised relationships in families (both single parent families and two parent families). These families displayed unusual selflessness where constantly putting the needs of those with DMD first was central to the family dynamic. As well these families talked about how DMD had changed their ideas about what mattered. Whereas at one time they would have wanted their children to do well at school, perhaps to go to university or follow some other career path now they were much more focussed on their children being happy, finding things they were good at and supporting them to do those things, because they gave them pleasure, self-confidence and had immediate benefits.

Research Findings

This brings me to a consideration of my research findings. Fundamentally this thesis addresses contradictions in the way lives of people with DMD are valued in Aotearoa New Zealand. Within the immediate community of family and friends these lives are cherished. I discuss this through an anthropological discourse of hope. Considering hope also facilitated an analysis of the advances in care and treatment including pharmaceutical developments. What is shown by this analysis is the value placed on each individual life. Immediate family and friends love and cherish the child or young man with DMD with special attentiveness as people with DMD need highly responsive care. As the network widens to include those whose relationships are professional other ideas about the way lives are valued become apparent. Fassin's (2012) ideas about the value of life apply as each individual life is still considered important. However as Novas (2006) shows the involvement of biomedicine and in particular the development of pharmaceuticals involves a political economy of hope. The desire of the immediate members of the community for an effective treatment that will reduce suffering is counterposed with economic considerations associated with drug development.

The immediate network of family and friends is intimately acquainted with the physical and emotional suffering associated with the condition and this knowledge contributes to the devoted care that many boys and young men receive. It is knowledge of this suffering and the hope that it can be alleviated that creates an assemblage in which the value of lives lived with DMD changes. The focus on each individual life remains but an economic calculus and financial assessment is introduced. This financial consideration contrasts starkly with the considerations that those families who discussed their reproductive decisions addressed. The way in which couples highly valued the lives of a son or brother with DMD was of overriding significance to the decisions they made. The different decisions that couples made about the use of reproductive technologies and the interplay of hope and political economy illustrates the complexities of ideas that inform the way lives with DMD are valued. The value that members of the DMD community place on each individual person, the desire to live a meaningful and purposeful life, the importance of alleviating or managing suffering and the hope for future effective treatments – all these ideas inform the way these lives are valued.

However, as this thesis has demonstrated, there is a contrasting view about the way these lives are valued. As I demonstrated in Section Three the lives of those with DMD often fall outside the state's capacity to care. My application of a Foucauldian analysis has allowed me

to show how this happens. Rather than a deliberate social exclusion of this group, people with DMD are an anomalous group whose needs do not align with the needs of the rest of the population. As a result services do not meet their needs. People with DMD effectively fall outside the economic rationale that justifies resource allocations in the health system: and their care therefore becomes a matter of bureaucratic indifference. These unmet needs and the challenges and contestations that people with DMD go through in attempts to access services create an additional level of suffering for lives which are inevitably imbued with pain and difficulty. The bureaucracies that characterise the neoliberal state and in particular the policies that legitimate the activities of bureaucracies (Shore and Wright 1997:11) authorise social exclusion. The technologies which structure the health system – diverse types of organisations managed by fixed budgets, service specifications, reporting schedules and policies that determine eligibility and funding streams – take the provision of care out of the moral sphere and make it a technical phenomenon. Hence the technologies discussed in detail in Section Three that address: access to cough assist machines, the availability of power chairs, the limit to support worker hours and the eligibility of support workers to work in hospitals, renders compassion a technical concern of utilitarian budgetary provision.

Ideas about the moral value of each life lived with DMD (ensuring people's lives have purpose and meaning, that they are included in society and participate in and contribute to their communities) are replaced by calculations about the pecuniary worth of lives (Fassin 2012). Technical considerations about service delivery and what level of provision is affordable replace moral decisions about how to improve life. Engaging with the theories of policy, governmentality and Fassin's ideas about the ethics of government has enabled me to explain the dominance of a technical concern for a utilitarian application of budgets in which services provide the greatest benefit for the greatest number. This rationale for the provision of services reinforces a sense of social exclusion for people with DMD. The message that the community receives is that their lives are not fully valued, but are in fact somewhat inconsequential.

Next Steps: Implications of the research

Participants in this research illustrate ways the indifference to their suffering can be addressed through various assemblages of contestation: the cough assist machine assemblage is one such example. The documents outlining international standards of care (Bushby et al 2010a and 2010b) also address the ways some of these exclusions can be mitigated. These

documents highlight the need for multi-disciplinary teams to provide "anticipatory and preventative measures as well as active interventions" (2010b:177). Condition-specific national services are already provided for some other anomalous biosocial groups in Aotearoa New Zealand. Haemophilia is an example of such a condition. Through the 'taxing' of DHBs and the involvement of all interested parties, a National Haemophilia Management Group and comprehensive treatment centres that meet the needs of people with haemophilia in New Zealand have been established since 2008 (Park, 2013). This provides a model of care for a similar nationwide service for people with DMD. A dedicated organisation that recognises the specificity of DMD is required for adequate care.

For people with DMD much of their lives is structured by numerous medical appointments and struggles with diverse bureaucratic services. A nationwide system designed to ensure a comprehensive service and national standards of care for all people with DMD would alleviate some of the social suffering that individuals and families endure. This structural change would remove the burden of constant struggle from each individual family. A comprehensive national DMD-specific service could provide the biomedical expertise that people require, the disability support and a responsive and accessible administration system. Such a service would contribute to a local biology in which New Zealand was able to offer optimal care for people with DMD, relieving individuals with the condition and their families of the enormous effort of endlessly battling bureaucracies. This is an overwhelming task given that these bureaucracies are not set up to meet the anomalous needs of this group.

The provision of such a comprehensive and specific service fulfils the description of the interactionist approach to disability issues mooted by Tom Shakespeare (2013) and addressed in the introduction. This approach would address both individual medical concerns and structural barriers. The goal of a national DMD service should be the provision of a multi-disciplinary approach that addresses issues to do with the body (that demonstrates the value of the individual life) and issues to do with the elimination of barriers to social participation, the elimination of discrimination and marginalisation (issues that address the political worth of lives). Such provision would provide the comprehensive governance proposed by Arthur Frank (1995) and Didier Fassin (2012), which could be deemed ethical care.

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