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Coming to terms with loss

Explaining how people with amyotrophic lateral sclerosis engage with healthcare services

By

Geraldine Foley

Thesis submitted to the School of Social Work and Social Policy
Trinity College Dublin
In fulfilment of the requirement of the
Degree of Doctor of Philosophy

April 2014

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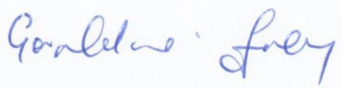

Summary

Amyotrophic lateral sclerosis (ALS), also known as motor neurone disease (MND), is a rapidly progressive and terminal neurological condition. ALS is a highly disabling condition and people with ALS ordinarily engage with healthcare services from symptom onset to end-of-life care. Researchers have already undertaken studies on perceptions and experiences of healthcare services among people with ALS but little was known about how and why people with ALS engage with healthcare services. The overarching aim of the study was to identify key parameters of healthcare experiences among people with ALS and to develop a substantive framework that explains how and why people with ALS engage with healthcare services.

Grounded theory method was used to capture the ALS service user perspective. Thirty-four people with ALS were sampled without predefined geographical location from the Irish ALS population-based register. An in-depth qualitative interview was conducted with each participant about their experiences of healthcare services. All interviews were transcribed. Data were collected and analysed in tandem and emerging findings guided sampling. Data were analysed using open, axial, and selective coding procedures. Reflexive and theoretical memos were compiled to guide sampling and to build substantive theory. NVivo9 was used as a tool to store coded data and memos, and to link memos to codes.

The study identified that loss (including loss of control) is central to the experience of living with ALS. Participants lost control in their lives and did not regain control or normality in their lives. Losing control in their lives led participants to exert control in their interactions with healthcare services. Rendering control to healthcare care professionals on their own terms also engendered feelings of control. Participants perceived control in end-of-life care as having the right to decide about end-of-life care. Few participants viewed life-sustaining interventions in ALS as interventions to alleviate suffering.

Family and life-stage were primary contexts to how participants responded to ALS and engaged with healthcare services. Participants with family made decisions about care in the interest of family, and struggled between needing to reassure family and needing to be reassured by family. Participants without family felt they had more freedom than people with family to follow through with their own preferences for care. Participants

suggested that it was more acceptable to die from ALS in later life and less acceptable to die as a young parent. The majority of participants were resigned to the progression of the disease but participants in later life were somewhat more accepting of death than young and middle-aged participants because they had already reared children who were now self-sufficient. Participants with dependent children were more likely to engage with life-sustaining interventions to continue parenting. All participants with children considered the impact of life-sustaining and supportive interventions on their adolescent and adult children.

Feeling uncertain about healthcare services was central to participants' experiences of healthcare services. Uncertainty about healthcare services comprised uncertainty about how the Irish healthcare system renders services to people with ALS and uncertainty about the future of healthcare services in Ireland. Most participants felt "disconnected" from the Health Service Executive (HSE) and expressed concerns about inequity in the Irish healthcare system. However, participants' uncertainty about the future with ALS was alleviated when they engaged with healthcare professionals who they trusted and who reassured them about their care.

The study reveals that people with ALS engage with healthcare services in line with their life transitions and trajectories and construct their experiences of healthcare services at both family and societal levels. The family roles that people with ALS adopt shape their response to ALS and their decisions about care. Losing control in life and fighting to be in control constitute two central components in how people with ALS interact with healthcare services. However, people with ALS also value reassurance offered by healthcare professionals and in some cases have a preference for rendering control over their care to healthcare professionals. People with ALS negotiate loss by engaging with services on their own terms.

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Chapter One

Introduction

1.1 Introduction

Amyotrophic lateral sclerosis (ALS) is a terminal neurological disorder of the central nervous system. It is a rapidly progressive condition and there are no periods of disease stability or remission in ALS. ALS is also a highly disabling condition. Clinical trials have been ongoing in the quest to develop and test disease-modifying treatments but treatments that have been used during clinical trials have had no real effect on the course of the disease, let alone offered hope of a cure. Healthcare professionals are being continually challenged to adjust their approach away from a model of care focused on medical outcomes to a model of care that seeks to improve (or at the very least) sustain quality of life for people with ALS.

The aggressive nature of ALS means that most people who develop the condition engage with healthcare services. However, despite mounting health services research in the ALS field, few researchers had sought to understand what is important to people with ALS as they engage with healthcare services. Given the resource limitations of healthcare services in Ireland and the shortened lifespan of people with ALS, it is imperative that services fit with service users' expectations of these services, and not merely the expectations of the service provider. What are the things that *really* matter to people with ALS in healthcare? Why do people with ALS engage with healthcare services? What conditions impact on how people with ALS use healthcare services? What shapes how people with ALS make decisions about their care? It is only by understanding the service user perspective that service providers can know whether the criteria used to judge care outcomes are consonant with real-life concerns facing people with ALS.

1.2 Background and rationale: Who *really* decides about healthcare services in ALS?

There is general consensus among policy makers that people who utilise healthcare services have an important role in shaping how services are provided to them (Health Service Executive, 2008). However, service providers do not routinely involve service users in planning services and researchers in health services research do not routinely include service users in research. Moreover, as in the case of ALS care, recommendations for best practice (Andersen et al., 2012; Miller et al., 2009a, b) are based on outcomes that, for the

most part, do not include the service user perspective on healthcare services. Indeed, few studies in the ALS field have focused exclusively on the service user experience of healthcare services (Foley et al., 2012a). This thesis describes and conceptualises the key parameters of healthcare experiences among people ALS from the viewpoint of people with ALS.

From her experience in the field as a clinical specialist occupational therapist at Beaumont Hospital in Dublin, the researcher had long observed (from her position) how people interacted with healthcare services. She observed that people with ALS were generally keen to engage with healthcare professionals but that they also negotiated between accepting, delaying, and declining services that healthcare professionals offered to them. She conducted a systematic review of ALS service users' perceptions of services and preferences for care (Foley et al., 2012a) and found that researchers and healthcare professionals knew little about the processes that underpin *how* or *why* people with ALS engage with healthcare services. She, therefore, set out to identify key parameters of healthcare experiences among people with ALS and to gain a better understanding of what aspects of care matter most to people with ALS.

1.3 Capturing the 'complexity' of the healthcare user perspective

The processes involved in decision making about care are complex (Clark et al., 2007). Healthcare professionals' labelling of illness and of healthcare interventions does not necessarily mirror how people who experience illness and who engage with healthcare services, label illness or healthcare services. Differences between service providers' and service users' understandings and perceptions of care are well documented (Blondeau et al., 1998; Rodriguez & Young, 2006a). Service users' perceptions of care are idiosyncratic, based on personal experiences and everyday knowledge about illness (Ziebland & Herxheimer, 2008). Healthcare professionals might adopt a top-down approach to 'fix' or 'resolve' (Henriksen & Rosenqvist, 2003) and might not necessarily factor patient experience into how to improve and/or render services (Yen et al., 2011).

There are approximately 265 people in the Republic of Ireland living with ALS at any one time and on average, two people are diagnosed with ALS every week in Ireland. Irish experts in the field have already developed guidelines in ALS care (Hardiman et al., 2002;

Phukan & Hardiman, 2009) but researchers had not explored experiences of healthcare services among people with ALS in Ireland.

Clark (2013) suggests that the rendering of healthcare services is complex because not only are interventions in themselves complex, healthcare interventions can vary in impact depending on how they are delivered and received. Healthcare interventions can have meaningful outcomes for people. However, outcomes are shaped by context, and the values and behaviours people have about healthcare interventions vary between contexts (Clark & Thompson, 2010). The focus of the thesis is to capture the complexity of ALS service users' experiences of healthcare services by exploring variation in healthcare experiences among people with ALS, in similar and different contexts (Foley et al., 2012b). Hence, even though this study is focused on healthcare experiences among people with ALS in Ireland, their experiences can be compared to those of people with ALS outside of Ireland.

1.4 Aims, objectives, and scope of the study

The purpose of the thesis is to generate a substantive framework that explains how people with ALS engage with healthcare services. The overarching aim of the study is to identify key parameters of healthcare experiences among people with ALS. The objectives of the study are two-fold: first, to identify key variables that shape how people with ALS engage with healthcare services and second, to unearth key psycho-social processes which explain how and why people with ALS engage with healthcare services.

Substantive theory is theory that is developed for a particular area of concern or for a limited domain of inquiry (in this case, healthcare experiences among people with ALS). It is constructed in the process of identifying differences and similarities in contextualised instances and patterns, relevant to the domain of inquiry, and used to build formal theory in larger conceptual fields (Adelman, 2010). Substantive theory describes *and* explains phenomena (Strauss & Corbin, 1998). This research is conceptually oriented because the findings are not only a basic description of service users' experiences of healthcare services. Experiences are conceptualised, and relationships between concepts are built to form theory that explains how people with ALS engage with healthcare services.

The research is deeply contextualised. Context means "structural conditions that shape the nature of situations, circumstances, or problems to which individuals respond by action/interaction/emotion" (Corbin & Strauss, 2008, p.87). Context might not necessarily determine how people engage with healthcare services but it does incorporate conditions which *shape* how people engage with healthcare services. With this in mind, the researcher set out to explore the ALS service user perspective with the following assumptions. Firstly, experiences of healthcare services for people with ALS are 'grounded' in their everyday lives, and how people with ALS engage with healthcare services is shaped by the social and cultural contexts of their lives. Secondly, multiple contextual factors (micro and macro) shape how people with ALS engage with healthcare services. Contextual factors vary between people with ALS and give rise to both similar and different experiences of healthcare services for people with ALS. Thirdly, healthcare experiences among people with ALS, are best understood by identifying consequences and/or contingencies to their actions.

At a substantive level, the findings enable comparisons between other contexts. The research design was planned so that it could be replicated in other country/systemic contexts, enabling further theoretical development of the findings in relevant substantive areas. Hence, at a substantive level, the findings might be of relevance for service delivery to people with ALS outside of Ireland. In addition, the leverage of the study yields important insights which might be applicable to people with other terminal conditions.

1.5 Defining terms

1.5.1 Amyotrophic lateral sclerosis

Amyotrophic lateral sclerosis (ALS), also known as motor neurone disease (MND) or Lou Gehrig's disease, is a systemic and terminal disorder of the central nervous system (Hardiman et al., 2011a). The condition causes degeneration of motor neurons in the central nervous system and results in paralysis of limb, respiratory, and bulbar (swallowing and articulation) muscles (Wijesekera & Leigh, 2009). Approximately 15% of the ALS population develop frontotemporal dementia and 35% develop mild frontotemporal impairment (Phukan et al., 2012). Rilutek (Riluzole) is a licensed drug in ALS, but has little impact on disease progression (Miller et al., 2007). ALS is insidiously progressive and the average life expectancy for people with ALS is two to four years from symptom onset (Chio et al., 2009). The aetiology of ALS (apart from familial cases) remains unclear but age

and location of symptom onset are primary predictors of survival. Peak onset of ALS is in the mid-60s but people in earlier adulthood can also develop the condition. The term 'amyotrophic lateral sclerosis' is consistently used in the thesis and is not interchanged with other terms by which the condition is also known.

1.5.2 Healthcare services and healthcare system

Healthcare services are services involved in the treatment and management of illness and the preservation of health, provided by medical, pharmaceutical, scientific, nursing, and allied health professionals. In this thesis, the term 'healthcare services' is used to denote the above *and* other services that participants perceive as healthcare services. The World Health Organisation (WHO) defines a healthcare system as that which consists of all the people and actions whose primary purpose is to improve health for those who they serve (WHO, 2007). Healthcare systems commonly comprise a mixture of users, payers, providers, and regulators (McPake et al., 2013). In this thesis, the term [Irish] 'healthcare system' is used to denote facilities, organisations, and personnel engaged in providing healthcare to people with ALS in Ireland.

1.6 Context and the researcher: From 'within' healthcare services

Reflexive research takes account of the researcher's role in the research. The researcher's experience as a healthcare professional in the field prompted her to undertake the study so that she and others might gain a better understanding of what is important to people with ALS. In qualitative research, reflexivity constitutes an awareness of the researcher's contribution to the construction of meaning (Charmaz, 2006a) and an acknowledgement that it is not possible to remain outside of the subject matter (Denzin & Lincoln, 1998). Chapter Three provides a detailed account of the researcher's reflexive processes in the study.

Corbin and Strauss (2008) suggest that reflexivity is shaped by how researchers locate themselves in their research. The researcher subscribes to the notion that qualitative research can never be objective and that she was always relevant to the research process. The researcher 'gave voice' to people with ALS, *and* co-constructed the data (Charmaz, 2006a). However, her experience of ALS is grounded in healthcare services only, and her reasons for conducting the research were not personally emotive. Rather, she undertook this study in the context of being a healthcare professional and a research fellow, and her

experience in the field enabled her to be sensitive to participants and the data. It is in the aforementioned capacities, that the researcher locates herself within the study.

1.7 Locating the research in an Irish context

This study investigates healthcare experiences among people with ALS in the Republic of Ireland. ALS research conducted in the Republic of Ireland is a key contributor to ALS research worldwide and ALS researchers in Ireland collaborate with international colleagues. ALS research conducted in Ireland has focused primarily on the epidemiology, pathogenesis, and cognitive profile of ALS (Hardiman & Greenway, 2007; O'Toole et al., 2008; Phukan et al., 2012). Little was known about how people with ALS in Ireland experience healthcare services.

As described in Section 1.4, people's experiences are shaped by context. It is important to note that this study was conducted in the midst of a severe recession in Ireland, where public financing of healthcare has been curtailed in response to the country's economic plight (Thomas et al., 2012). This context i.e. limitations in resources for healthcare users in Ireland (including people with ALS), is relevant to the study. It is also important to note that people with ALS in Ireland ordinarily engage in research. Researching healthcare experiences among people with terminal illness can pose challenges for research participants and researchers, not encountered in other fields of care (Addington-Hall, 2002; Ewing et al., 2004). Indeed, commentary on the ethical challenges of including people nearing death in research is commonplace in palliative care (Casarett & Karlawish, 2000; Jubb, 2002). In this study, the researcher did not encounter problems recruiting participants and the study was only feasible because of participants' propensity to engage in research.

1.8 Structure of the thesis

The thesis comprises eight chapters. This chapter (Chapter One) has explained the rationale for undertaking the study, outlined the aim and objectives of the study, defined some key terms, and drawn attention to the potential complexity of healthcare experiences among people with ALS. The chapter has also placed the researcher in context.

Chapter Two outlines the literature pertaining to the topic of investigation. Chapter Two provides a detailed review of studies that have investigated experiences and perceptions of healthcare services among people with ALS, and explains how healthcare services are rendered to people with ALS in Ireland. The chapter also reviews literature on adaptation to illness, including terminal illness. The theoretical and sensitising frameworks for the research are made clear to the reader.

Chapter Three outlines the methods used in the study to capture the ALS service user perspective. Here, the researcher explains her ontological viewpoint and describes how her view of reality impacted on the methods she used. The chapter provides a detailed account of the steps taken by the researcher at all stages of the research, and explains how the study procedures enabled her to build substantive theory.

The findings are presented in course of three chapters (Chapters Four, Five, and Six). The central story line begins in Chapter Four that focuses on the 'personal' experiences of participants, and explains how people with ALS respond to ALS and adapt to change in their lives. Chapter Five moves to the 'external reality' (i.e. meso-level) of help and care for people with ALS. Chapter Five explains what type of care is important to people with ALS and why, under certain conditions, people with ALS value the care they receive. Chapter Six focuses on the macro-context of healthcare services for people with ALS in Ireland and describes how macro-level structures impact on their experiences of healthcare services. The central story line is threaded through each of the findings chapters. Chapter Seven outlines how healthcare services might best respond to ALS service users' expectations of healthcare services.

Chapter Eight (conclusions) synthesises the research findings, and outlines key theoretical and conceptual contributions of the research. The concluding chapter also situates the findings within the sensitising framework of the research.

Chapter Two

Literature review

2.1 Introduction

Amyotrophic lateral sclerosis (ALS) is a progressive neurological disorder of the central nervous system which causes paralysis of limb, respiratory, and bulbar (articulation and swallow) muscles. Despite a common perception in the public domain that ALS primarily affects people in middle and later life, ALS is also one of the most common neurodegenerative conditions in young adults. The life time risk of developing ALS is approximately 1:400 (Alonso et al., 2009). Unlike the majority of neurodegenerative conditions (that have a chronic pattern of progression), ALS is hastily progressive. Assisted ventilation can increase life expectancy beyond the natural course of the disease but the care approach for people with ALS is palliative from the point of diagnosis (Oliver et al., 2006).

As stated in Chapter One, no curative treatments exist for people with ALS. Services that address effective symptom management, physical and psychological support, are considered by healthcare professionals necessary to meet the needs of people with ALS (Andersen et al., 2012). The role of healthcare professionals is to improve quality of life for people with ALS and their families. Consensus among service providers is that models of care that are multidisciplinary in nature and responsive to the evolving nature of the condition, are best practice in ALS care (Hardiman et al., 2002).

Despite positive outcomes in ALS multidisciplinary care research on survival and cost (Traynor et al., 2003; Van der Steen et al., 2009), the literature that reports on the ALS service user perspective suggests, for the most part, dissatisfaction with services (Foley et al., 2012a). Few researchers have investigated how people with ALS experience healthcare services and no outcome studies in ALS multidisciplinary care include the service user perspective on care (Foley et al., 2012b). Researchers have explored a broad spectrum of experiences among people living with ALS (e.g., Locock et al., 2009; Young & McNicoll, 1998) but there is a dearth of literature on how people with ALS understand their healthcare services, decide about services, and engage with services.

2.1.1 Engaging the amyotrophic lateral sclerosis (ALS) service user

Williamson (2010) argues that attitudes and beliefs of healthcare users about care are largely based on their 'personal' expectations of services and that key to achieving meaningful outcomes for service users is to identify how phenomena are understood and experienced by them. Service users can offer a unique perspective about care which is grounded in their own personal experiences of care. However, service users' perspective is not always captured by rudimentary survey questionnaires that do not account for these experiences (Entwistle et al., 2012). Understanding the potentially different ways people engage with services can help to illuminate the key parameters of service users' experiences of healthcare services. Identifying key parameters of service users' experiences of services enables researchers to test relationships between domains of care that are important to service users and things that healthcare systems are measured against.

In ALS care, many questions remain over how best to capture the service user viewpoint: which aspects of healthcare services matter most to people with ALS and why; do standardised tools and survey instruments include the domains of care that are important to people with ALS; and if not, how should such tools be developed (Foley et al., 2012b)? People with ALS shift towards values of self-direction as they advance in illness (Fegg et al., 2005) and so might choose interventions based on this orientation. It is possible that they make decisions about care based on what they believe is important to themselves, rather than what is important to service providers. Correspondingly, research needs to explore these diverse experiences in order to identify what aspects of care are meaningful to people with ALS.

This literature review analyses the literature relevant to the topic of investigation: How and why people with ALS engage with healthcare services. The primary aims of the literature review are to draw attention to factors (micro and macro) that impact on ALS service users' experiences of care, and to outline how people with ALS use healthcare services as they negotiate change in their lives. First, the theoretical framework adopted in this study to underpin investigation into how people with ALS engage with healthcare services is outlined (Section 2.2). ALS healthcare services (including the Irish context of ALS healthcare services) are described (Sections 2.3 and 2.4). This is followed by a review of investigations on ALS service users' experiences of services and their preferences for care

(Sections 2.5 to 2.5.7). The findings of the review are then contextualised in terms of how people with ALS might engage with services as they adapt to ALS. Accordingly, literature on adaptation in ALS is discussed (Section 2.6). Attention is given to how people with illness adjust to illness and how they construct their experiences as they adjust (Sections 2.7 to 2.7.4). Awareness of dying in terminal illness might be of particular relevance to people with ALS and contextualise their experience of healthcare services (Section 2.7.5). Finally, the use of the chosen research method is put forward in the context of how this study sought to pinpoint the key parameters of ALS service users' experiences of healthcare services. The chapter concludes by outlining the sensitising framework for the thesis.

2.2 Healthcare services: People with ALS as agents of change

Medical sociology has adopted a number of different theoretical approaches, and is not dominated by any one framework. Post-modernistic perspectives which view the healthcare client/consumer as an agent of change (e.g., interpretive perspective) and models which emphasise the collective social, cultural, political, and economic context of health (e.g., social model of healthcare) are consistent with core principles of healthcare today (Clarke, 2010).

Contemporary theoretical perspectives view healthcare at both the micro-level (individuals), and macro-level in contradistinction to the exclusive focus on the macro-level of earlier functionalist and social constructionist perspectives. Symbolic interactionism (Blumer, 1969) suggests that illness and health are defined within a specific socio-cultural context where the meaning attached to healthcare is determined by the individuals within that context. Symbolic interactionism as a social action theory evolved as a perspective which focused on the intricacies of human interaction and meaning (e.g., within the healthcare context). Applied to healthcare, it views service users as active agents who have the capacity to make decisions about care and so change how services are provided to them.

Context does not necessarily determine experiences and/or set the course of interaction with service providers but it does incorporate a set of conditions which influence how people with ALS respond to healthcare services. The factors that influence understanding of services and decision making among people with ALS about their care have remained

poorly understood. There is both a micro and a macro-context to the experience of care for people with ALS. Micro-conditions are close to each service user (for instance personal resources, family) and macro-conditions consist of the broader political and social contexts which shape how services are provided to them. The situational context of Irish healthcare services for people with ALS shapes their individual and subjective experiences of services which may in turn impact on services and change how services are rendered to them. However, this can only happen when empirical data on their experiences of healthcare services is collected, analysed, understood, and applied.

2.3 Healthcare in amyotrophic lateral sclerosis

International guidelines for best practice in ALS care emphasise the collective importance of physical, social, psychological, and existential care for people with ALS. The American Academy of Neurology (AAN) and the European Federation of Neurological Societies (EFNS) task force on the clinical management of ALS have undertaken extensive reviews of evidence-based care in ALS (Andersen et al., 2012; Miller et al., 2009a, b). For the most part, these reviews outline best practice in domains that service providers (rather than service users) have designated as the key parameters of ALS care.

Hardiman et al. (2011a) state that care for people with ALS is primarily symptomatic and should be delivered by a broad network of healthcare professionals who operate within an interdisciplinary context. Indeed, Van den Berg et al. (2005) reported that multidisciplinary care in ALS had a positive impact on quality of life for people with ALS. Healthcare professionals in ALS care believe that primary components of ALS care include: a timely diagnosis, early use of available neuro-protective treatments, nutritional and respiratory support, symptomatic management, assistive devices, and psychosocial support including advance directives (Phukan & Hardiman, 2009). These services are provided by a broad range of healthcare professionals including neurologists, palliative care specialists, general practitioners, nurse specialists, occupational therapists, physiotherapists, speech and language therapists, clinical nutritionists, social workers, psychologists (including neuropsychologists), respiratory specialists, and gastroenterologists (Andersen et al., 2012).

Radunovic et al. (2007) suggest that people with ALS navigate through different stages of ALS care. These include adapting to the diagnosis and early disability, coping with

progressive disability, and engaging with end-of-life care. Hardiman et al. (2002) state that healthcare services should respond in line with how people with ALS navigate through these different stages. Hardiman et al. (2002) describe a model of care based on a centralised specialist multidisciplinary ALS clinic with strong links between specialised clinic-based healthcare professionals, primary care healthcare professionals, and the voluntary sector. This model emphasises the importance of early access to services at the diagnostic and minimal disability stage, ongoing service provision with evolving disability, and the subsequent extension of services to the pre-terminal phase to maintain quality of life and alleviate burden of care. This model of care has been developed by specialist ALS centres in other countries (e.g., UK, USA) (Leigh et al., 2003; Mayadev et al., 2008).

A number of studies have reported positive outcomes in survival, cost, and service users' quality of life when care is managed by multidisciplinary specialised ALS centres as opposed to non-specialist centres (Chio et al., 2006; Traynor et al., 2003; Van den Berg et al., 2005; Van der Steen et al., 2009). Researchers and service providers have also found that multidisciplinary care is effective in managing end-of-life care for people with ALS (Mandler et al., 2001; Miller at al., 2009c).

McLeod and Clarke (2007) undertook a detailed review of the psychosocial needs of people with ALS and found that psychological and social wellbeing of service users may have a significant impact on their quality of life. Quality of life studies in ALS have shown that physical ability is not a primary determinant of quality of life in ALS and that psychological, existential, and social wellbeing impact on quality of life and satisfaction with life in ALS (Fegg et al., 2010; Neudert at al., 2004). Access to life-sustaining interventions might not necessarily be ALS service users' primary expectation of healthcare services (Borasio, 2001).

There is now international consensus among experts in ALS care that ALS healthcare services are best provided under a palliative care framework (Bede et al., 2011). The palliative care approach seeks to improve the quality of life of service users and their families as they encounter problems associated with life-threatening illness through prevention and/or relief of physical, psychological, and existential suffering (WHO, 2013). ALS is always terminal and in comparison to other terminal conditions (e.g., cancer), there is often more certainty regarding the period of time to death. Over 60% of people with

ALS die within 1,000 days from their first symptom (Chio et al., 2009). Expanding the concept of palliative care into the management of non-malignant conditions is an emerging trend (Veronese et al., 2010). However, the key factors for initiating palliative care intervention, and the types of intervention required for people with ALS remain to be defined (Hardiman et al., 2011b).

2.4 The situational context of Irish ALS healthcare services

In the Republic of Ireland, a national ALS multidisciplinary clinic provides care for up to 80% of the ALS population. Services are primarily co-ordinated by the national clinic with direct referral onto community-based services (Bede et al., 2011; Corr et al., 1998). The remainder of the ALS population access neurology clinics at secondary care centres. Some people with ALS also negotiate their care between the national clinic and other neurology centres. Palliative care services for people with ALS are provided in multiple locations ranging from acute and sub-acute specialist palliative care units to home and community-based support. It is thought that most people with ALS in the Republic of Ireland access community-based care through primary care services at some point in time. However, access to aids and equipment in the Irish healthcare system is means tested. A complex two-tier system of private and public healthcare financing in the Irish healthcare system also limits and/or delays access to multidisciplinary community-based services (Hardiman, 2010).

The Health Service Executive's Quality and Clinical Care Directorate has established a number of national clinical programmes to improve the standard of healthcare in Ireland. The national clinical programme for palliative care (Health Service Executive, 2013) has aimed to establish designated pathways for the delivery of services to people who have life-limiting conditions. Palliative care provision in Ireland has extended since the publication of a national palliative care document (Report of the National Advisory Committee on Palliative Care) in 2001 (Department of Health and Children, 2001). This report defined priorities for palliative care services in the Republic of Ireland but many priorities have not been enacted because of limited resources in the Irish healthcare sector (May et al., 2013).

There is consensus that healthcare services in the Republic of Ireland for people with neurodegenerative disease remain under-developed and under-resourced (Hardiman,

2010). The benefits commonly associated with private healthcare in the Republic of Ireland are of no great advantage to people with ALS beyond the diagnostic phase and ongoing care needs in primary care are met primarily by the public sector and supplemented by the voluntary sector (the Irish Motor Neurone Disease Association). A series of documents published by the Neurological Alliance of Ireland (NAI) on optimum standards in neurological care (Neurological Alliance of Ireland, 2000; 2001; 2002) emphasised the importance of unrestricted and timely access to healthcare services for people with neurological conditions in Ireland, but few of the care standards as set out by the NAI care documents have been achieved (Neurological Alliance of Ireland, 2010). Similar to the situation of other populations with neurodegenerative disease in the Republic of Ireland, fully integrated palliative care is not uniformly available to people with ALS.

2.5 ALS service users' experiences of services and preferences for care: A review

Researchers had already reported on ALS service users' perspectives on healthcare services. In 2011, the researcher undertook a systematic review on ALS service users' experiences of healthcare services and preferences for care. She undertook the review before she embarked on the study. The systematic review included empirical studies published in full between January 1988 and March 2011 on the ALS service user perspective on services (Foley et al., 2012a). Studies published between April 2011 and October 2013[1] (captured by the same search strategy and selected using the same inclusion and exclusion criteria) were analysed during the period of the thesis write-up, and are also included in the thesis. Appendix A is a matrix of all the papers included in the review for the thesis.

2.5.1 Search strategy for systematic review

MEDLINE (including PubMed as a search engine), CINAHL, AMED, PsycINFO, Evidence Based Medicine Reviews (EBMR), and Web of Science were used as databases for the systematic review. Search terms used included; *'amyotrophic lateral sclerosis'* or *'motor neurone disease'* and *'services', 'healthcare', 'experiences', 'expectations', 'satisfaction', 'decision making', 'perceptions', 'perspectives'* and *'preferences'*. Separate electronic manual searches (including early-online articles where available) in the following palliative care journals; *American Journal of Hospice & Palliative Medicine, European Journal of*

[1] Thesis submitted in November 2013 for examination

Palliative Care, Journal of Hospice & Palliative Nursing, International Journal of Palliative Nursing, Journal of Pain & Symptom Management, Journal of Palliative Medicine, Palliative & Supportive Care, Journal of Palliative Care and *Palliative Medicine* were also undertaken using the search terms *'amyotrophic lateral sclerosis'* and *'motor neurone disease'*.

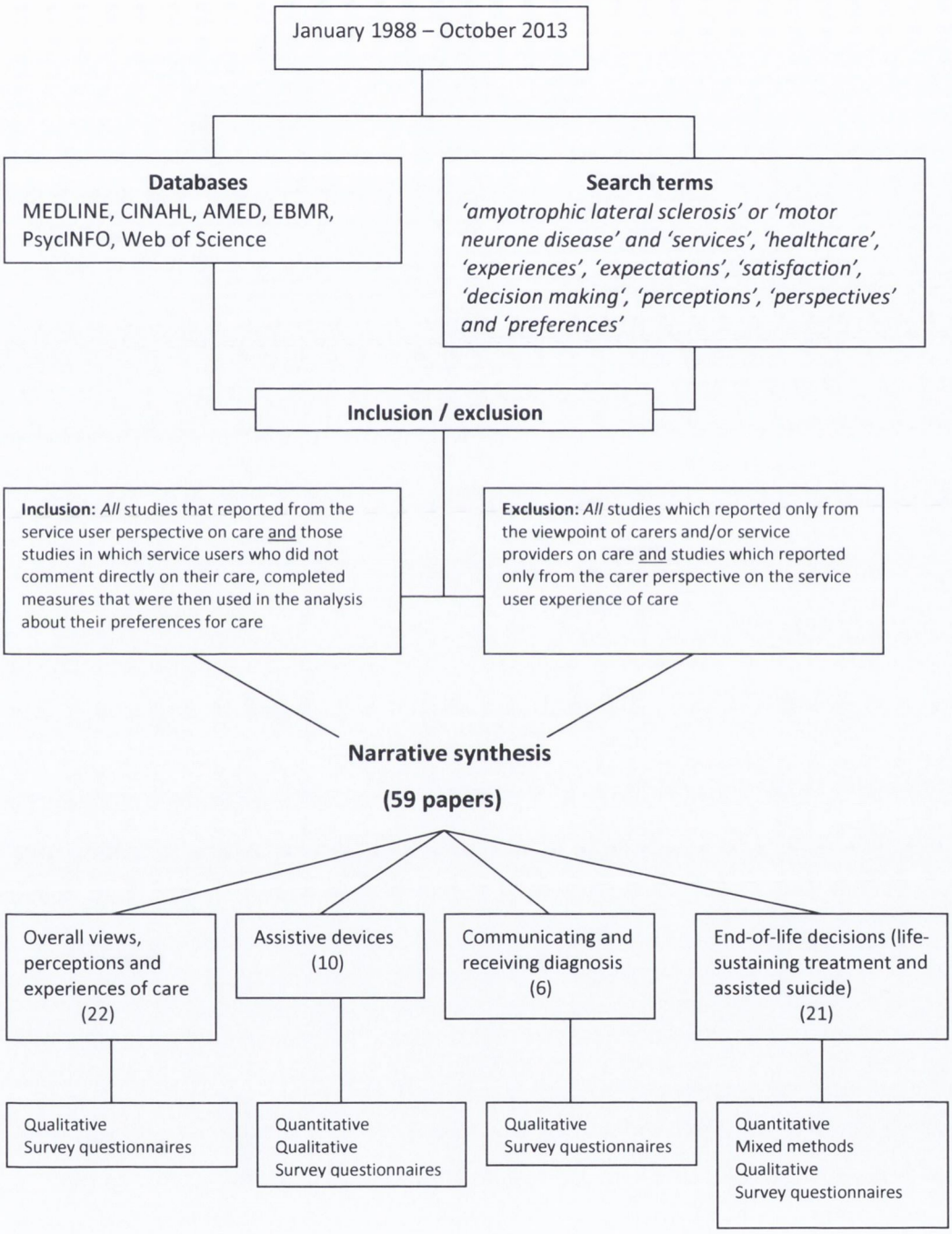
2.5.2. Selection criteria and synthesis of the data

The researcher included studies which reported from the ALS service user perspective on care. She also included studies in which people with ALS who did not report directly on their care, had completed measures which had then been used to investigate factors associated with their preferences for care (Foley et al., 2012a). She sought to focus only on the service user perspective and she excluded studies which reported only from the viewpoint of carers and/or service providers. In addition, she excluded studies not published in the English language. Although structured approaches exist to judge the strength and reliability of quantitative and qualitative research (to determine exclusion criteria for analysis), no criteria for further exclusion was applied owing to the paucity of data found on the topic. Because of the diversity of methods used in the selected studies (as detailed in Sections 2.5.3 to 2.5.7), a statistical synthesis was not appropriate and the researcher undertook a narrative synthesis of the data (Mays et al., 2005). Here, she conducted a preliminary analysis of each selected study, critically appraised each study and then explored similarities and differences between studies. The researcher, her PhD supervisor and her clinical sponsor worked collectively to synthesise and interpret the evidence as it related to the purpose and aims of the review.

Using the above criteria, 47 papers from 43 studies were found (published in the period 1988 to March 2011) which report from the ALS service user perspective on health and social care services (Foley et al, 2012a). An additional ten studies (published between April 2011 and October 2013) are included in the review for the thesis along with two extra papers from one study that had originally been published in the period between January 1988 and October 2013 [i.e. total of 59 papers from 53 studies]. An ALS study recently conducted (and published) by Picker Institute Europe (Cooney at al., 2012) is also included in the thesis. Figure 1 (p.16) illustrates the steps taken by the researcher in the review and summarises the findings of the review. For clarity, studies are categorised under the following headings: Overall views, experiences and expectations of healthcare services;

use of assistive devices; communicating and receiving the diagnosis; and end-of-life decisions.

Figure 1 **Systematic review flow chart**



2.5.3 Overall views, experiences, and expectations of healthcare services

Researchers have found that primary expectations among people with ALS of healthcare services were emotional support, timeliness of care, and access to multidisciplinary care (Beisecker et al., 1988; McCabe et al., 2008; Kristjanson et al., 2006). However, Peters et al. (2013) undertook a large cross-sectional survey of healthcare experiences among service users with neurodegenerative conditions in England (including ALS) and found that the problems most frequently reported by service users were poor planning and integration of care. Wicks and Frost (2008) administered an online questionnaire to 247 people with ALS about their services and found that respondents wished for more information about their services.

Krivickas et al. (1997) conducted a survey on the homecare needs of people with ALS in Ohio, USA, and found that over 50% respondents did not receive services. Ng et al. (2011) conducted a survey on community care needs of ALS service users in Victoria, Australia, and found that 43% of respondents reported deficiencies within services. These findings resonate with a larger-scale survey in Australia on palliative and supportive care for people with neurodegenerative conditions (Kristjanson et al., 2005) where only 35% and 18% of ALS respondents had received homecare services and palliative care services respectively. A study by Van Teijlingen et al. (2001) in Scotland found that only 25% of service users felt that their needs had been met. Problems identified in relation to care included long waiting periods for multidisciplinary care. The majority of respondents also accessed voluntary sector services to alleviate their family carers' burden of care. The findings are consistent with those of an Irish survey on access to community-based services for people with ALS (Hardiman et al., 2003). Hardiman et al. (2003) also found that people with ALS relied heavily on the voluntary sector to supplement their care.

Researchers have conducted qualitative studies on the meaning of care for people with ALS. Participants' primary expectations of care centred on the need to be valued by healthcare professionals, and to be acknowledged independently of their disease (Bolmsjo, 2001; Brown, 2003). People with ALS also complained that the onus to develop and maintain relationships in the clinical encounter lay, for the most part, with service users as opposed to service providers (Brown, 2003; Hocking et al., 2006). However, in an Irish study of perceptions of quality of life among people with ALS (Foley et al., 2007), service users indicated that healthcare services had had a positive impact on their lives.

Budych et al. (2012) undertook a large-scale qualitative study on how people with rare diseases in Germany (including ALS) experience encounters with physicians and found that participants perceived physicians as lacking in expertise. Hogden et al. (2012) conducted a qualitative study at two specialised ALS clinics in Australia to explore factors that influence decision-making in care for people with ALS. Participants expressed satisfaction with specialised multidisciplinary care but expressed dissatisfaction with non-specialised care due to waiting times for services and poorly co-ordinated care rendered by non-specialised services.

A number of UK-based qualitative researchers have explored ALS service users' expectations of care, and compared expectations with care received (Brown et al., 2005; Hughes et al., 2005, O'Brien et al., 2012). They also found that people with ALS were concerned about a lack of knowledge about ALS among healthcare professionals. Participants also felt uncertain about healthcare services and complained that services were deficient in providing timely and co-ordinated care. Problems identified by service users included the absence of care co-ordinators and limited access to specialised equipment and respite care. In contrast, an audit by Bennett et al. (2009) on service users' perceptions of hospice-based clinics co-ordinated through a tertiary ALS centre in Preston, UK, found that participants were satisfied with hospice-based care.

2.5.4 Use of assistive devices

Murphy (2004) undertook a qualitative study in Scotland to investigate perceptions of augmentative and alternative communication (AAC) devices among people with ALS and their communication partners. Service users identified benefits of AAC but they also expressed a strong desire to use their own speech even if it was unintelligible to others. The majority of participants found AAC unsuccessful as it impacted on close personal interaction in real-life settings. Participants also expressed dissatisfaction with respect to the timely provision of AAC and inadequate support and training to use AAC devices.

Surveys of service users' satisfaction with assistive devices at ALS clinics in the United States reported that people with ALS expressed satisfaction with assistive devices (Gruis et al., 2011; Hossler et al., 2011; Trail et al., 2001; Ward et al., 2010). Trail et al. (2001) found that over 75% of their sample (n=42) were highly satisfied with wheelchair mobility because it facilitated greater independence in their lives. Ward et al. (2010) reported

similar levels of satisfaction with powered wheelchair mobility among service users. Gruis et al. (2011) surveyed 68% of their clinic attendees and found that the majority of service users utilised a broad range of low-tech personal care devices and were satisfied with them. Hossler et al. (2011) tested the feasibility of an interactive computer aid for advanced care planning, among 17 ALS clinic attendees. This computer aid assisted participants to translate personal values and goals into an advance directive that best reflected their wishes for end-of-life care. All participants were highly satisfied with the use of this device.

In Italy, Caligari et al. (2013) studied the impact of eye tracking communication devices (ETCD) on disability and quality of life for 35 anarthric people with ALS and reported high levels of satisfaction with ECTD across their sample. At a specialised ALS centre in Brescia, Italy, Vitacca et al. (2010) reported that 75% of participants were highly satisfied with the use of telemedicine for a home-based cough assist programme. Similarly, a preliminary trial on the use of telemedicine in Portugal for home ventilation reported that service users expressed satisfaction with telemedicine (Lopes de Almeida et al., 2010). In the Netherlands, Nijeweme-d'Hollosy et al. (2006) also evaluated the impact of tele-medicine on the care of people with ALS. All four participants expressed high levels of satisfaction with telemedicine but also valued face-to-face contact with healthcare professionals to discuss their concerns about ALS.

2.5.5 Communicating and receiving the diagnosis

Hugel et al. (2006) conducted a qualitative study on ALS service users' experiences of receiving a diagnosis of ALS at a specialist neuroscience centre in Liverpool, UK. They found that participants experienced delays in receiving a diagnosis but were pleased with how the diagnosis was communicated to them. At a large Italian ALS specialised centre in Italy, Chio et al. (2008) undertook a survey to investigate how people with ALS seek information and rate their satisfaction with how physicians communicate the diagnosis of ALS. They found that even though service users experienced negative feelings about their diagnosis, most participants sought information about ALS and were satisfied with how physicians communicated the diagnosis to them. These findings are consistent with an audit on service users' experience of a fast-track system for diagnosing ALS at a specialised centre in Preston, UK (Callagher et al., 2009). However, they are contrary to the findings from a qualitative study at the same centre where people with ALS reported a lack of

sensitivity among physicians when communicating the diagnosis (O'Brien et al., 2011a). They are also contrary to findings from large-scale survey research in the United States where 27% of participants reported at least one misdiagnosis (Belsh & Schiffman, 1996) and 56% of participants rated physician performance in the disclosure as average to poor (McCluskey et al., 2004).

2.5.6 End-of-life decisions

Cooney et al. (2012) conducted a qualitative study in the UK to explore end-of-life issues among people with ALS. They found that self-determination at end-of-life was participants' primary expectation of healthcare services. Participants equated dignity with self-determination at end-of-life and they believed that discussions with healthcare professionals about end-of-life care should include assisted dying in addition to advance directives and palliative care. At two specialised care centres in the United States, Sulmasy et al. (2007) and Nolan et al. (2008) also found that maintaining control in making decisions about end-of-life care was important for people with ALS. Service users preferred to make decisions independently or at least favoured shared decision-making with family and/or service providers. The majority of participants had already issued an advance directive (Nolan et al., 2008).

Quantitative studies of ALS service users' preferences for and decision making in end-of-life treatments (cardio-pulmonary resuscitation, ventilation, and gastrostomy feeding) and physician-assisted suicide have been conducted, for the most part, in the United States (Albert et al., 1999 and 2009; Ganzini et al., 1998; Moss et al., 1993 and 1996; Munroe et al., 2007; Rabkin et al., 2006; Silverstein et al., 1991). Researchers found that people with ALS demonstrated ability to express preferences for life-sustaining interventions and follow through on these preferences (Albert et al., 1999). Desire for information and participation in decision making about care remained stable among people with ALS (Silverstein et al., 1991) but their preferences for life-sustaining treatments changed in some cases (Albert et al., 1999; Silverstein et al., 1991). Some people with ALS who were initially in favour of life-sustaining interventions later opted against them and others changed from declining life-sustaining interventions to opting for them. Service users' care preferences were independent of their functional status and demographic variables (Munroe et al., 2007; Silverstein et al., 1991). Service users' care preferences were determined, in part, by their attitudes to life: those who demonstrated a greater

attachment to life were more likely to find life-sustaining interventions acceptable (Albert et al., 1999).

Participants expressed a desire to live on and use services up to the point of death or long-term ventilation (tracheostomy) (Albert et al., 2009). Choosing long-term ventilation was determined by their own perceptions of their health status and satisfaction with life. However, some participants' hope and optimism diminished the longer they remained on long-term ventilation (Albert et al., 2009; Rabkin et al., 2006). People with ALS valued extended life provided by ventilation and were positive about ventilation despite its physical limitations (Moss et al., 1993 and 1996) but they also recognised that assisted ventilation had limitations in maintaining quality of life (Moss et al., 1996) and that it increased the burden of care on their families (Moss et al., 1993). Some people also expressed the desire to stop ventilation on their own request (Moss et al., 1996).

Achille and Ogloff (2003) and Ganzini et al. (1998) found that feelings of hopelessness among people with ALS predicted preferences for physician-assisted suicide. Service users in Oregon, USA, who expressed interest in physician-assisted suicide had greater distress and a lower quality of life than those who did not wish to hasten death (Ganzini et al., 1998). These findings resonate with those of Stutzki et al. (2013) who undertook a prospective questionnaire study of attitudes towards hastened death in ALS at two specialised ALS centres in Germany and Switzerland. Stutzki et al. also found that participants' wish to hasten death was predicted by depression, anxiety and self perceived low quality of life. 50% of their sample indicated that they could envisage requesting physician-assisted suicide or euthanasia.

Young et al. (1994) conducted a mixed-methods study on service users' perceptions of assisted ventilation at a specialised ALS centre in Vancouver, Canada. They found that service users' decisions about assisted ventilation were shaped by their views on the potential for assisted ventilation to improve their quality of life, their ability to control discontinuation of ventilation, and the likelihood of ventilator support to extend their life. Participants expressed concerns about potential adverse effects of assisted ventilation on their quality of life and the impact of burden of care on their family and carers. Some participants did not actively seek medical information about the benefits and/or contra-indications of ventilation. Mixed-methods research undertaken by Hirano and Yamazaki

(2010) and Hirano et al. (2006) in Japan also found that people with ALS feared that assisted ventilation would increase the burden of care on their families. However, contrary to Young et al. (1994), they identified that education about invasive ventilation was a key factor in determining whether or not people regretted choosing invasive ventilation. Participants who received advice and information about invasive ventilation were more likely to have no regret than those who did not receive sufficient information (Hirano & Yamazaki, 2010).

Qualitative investigations undertaken at specialist ALS centres in Montreal, Canada (Lemoignan & Ells, 2010), and in Stockholm, Sweden (Sundling et al., 2009), also found that service users' decisions about ventilation were influenced by their life situation and family support. Lemoignan and Ells (2010) and Sundling et al. (2009) identified that people with ALS had conflicting emotions about assisted ventilation and had fears about adapting to assisted ventilation. These findings resonate with those of Baxter et al. (2013) in Sheffield, UK. Baxter et al. conducted a qualitative study to explore perceptions of non-invasive ventilation among people with ALS and their carers. They also found that assisted ventilation was challenging for people with ALS. Participants reported better sleeping patterns but expressed fears about non-invasive ventilation technology and about the physical limitations that non-invasive ventilation imposed upon them.

Narayanaswami et al. (2000) undertook a survey at a long-term care facility in Tennessee, USA, on assisted ventilation and found that people with ALS were somewhat more ambiguous toward assisted ventilation than people with other neuromuscular diseases. These findings resonate somewhat with those of Cazzolli and Oppenheimer (1996) in the United States. Cazzolli and Oppenheimer conducted qualitative [structured] interviews with 75 ALS service users in Ohio, Pennsylvania, and West Virginia, and found that those who underwent emergency invasive ventilation (tracheostomy) without advance directives in place may not have chosen invasive ventilation had they been aware of the ensuing burden of care. However, McKim et al. (2012) identified that formal education about ventilation at a Canadian respiratory rehabilitation centre predicted real-life choices about ventilation among people with ALS and assisted people with ALS to make decisions about assisted ventilation.

2.5.7 Critique of the literature on the ALS service user experience of care

Researchers have used a number of different methods to investigate perceptions of and preferences for care among people with ALS. Qualitative researchers have explored experiences and expectations of services among people with ALS in general, and in specific domains of care. Experiences for people with ALS have also been captured by a combination of national (Hardiman et al., 2003; Peters et al., 2013; Van Teijlingen et al., 2001) and regional (Kristjanson et al., 2005 and 2006) surveys on health and social care services; regional surveys specific to community-based services (Krivickas et al., 1997; Ng et al., 2011); regional and clinic-based surveys on satisfaction with disclosure (Callagher et al., 2009; Chio et al., 2008; McCluskey et al., 2004); a facility-based survey on the use of assisted ventilation (Narayanaswami et al., 2000); national and clinic-based assessment of satisfaction with assistive technology (Caligari et al., 2013; Gruis et al., 2011; Hossler et al., 2011; Trail et al., 2001; Ward et al., 2010) and telemedicine (Nijeweme-d'Hollosy et al., 2006; Vitacca et al., 2010); online surveys of service users' care preferences (Wicks & Frost, 2008); and by an international survey specific to perspectives on misdiagnosis (Belsh & Schiffman, 1996). Quantitative cross-sectional (Achille & Ogloff, 2003; Ganzini et al., 1998; Moss et al., 1996), quantitative longitudinal (prospective) (Albert et al., 1999 and 2009; Moss et al., 1993; Silverstein et al., 1991; Stutzki et al., 2013; Sulmasy et al., 2007) and retrospective (Munroe et al., 2007) studies, and mixed-methods studies (Hirano & Yamazaki, 2010; Hirano et al., 2006; Nolan et al., 2008; Young et al., 1994) have focussed on the relationship between variables that may influence and/or predict preferences for end-of-life care and treatment choices among people with ALS.

Overall, few studies have explored in depth the *experiences* of healthcare services among people with ALS. Some studies included other diagnostic groups (Budych et al., 2012; Hardiman et al., 2003; Kristjanson et al., 2005 and 2006; McCabe et al., 2008; Narayanaswami et al., 2000; Peters et al., 2013; Sulmasy et al., 2007) and the majority included carers. For the most part, studies are based in Europe, North America and Australia. Some sample sizes are small and in most cases findings are not generalisable to the ALS population. No standardised patient questionnaires on health and social care services have been used. One study (Kristjanson et al., 2006) used a generic patient satisfaction with care measure and only few studies (using Likert scales) (Chio et al., 2008; Gruis et al., 2011; Hossler et al., 2011; Lopes de Almeida et al., 2010; Nijeweme-d'Hollosy et al., 2006; Trail et al., 2001; Ward et al., 2010) have measured service users' satisfaction

with specific aspects of care only. The majority of findings derived from qualitative studies are descriptive and none have developed theory from data. Some findings are contradictory and may be attributed to small sample sizes, purposeful sampling procedures, bias within interpretation, and differences between healthcare systems. There are no control studies and results from qualitative investigations have not been used to construct ALS specific survey instruments for use with randomly selected populations. *The parameters of ALS service users' experiences therefore remain to be more fully mapped* before they can become subjects of large-scale quantitative research focused on the key aspects of their healthcare experiences; a task that this thesis seeks to accomplish.

Despite these shortcomings and variation in design, there are some commonalities between the findings of studies that have sought to capture the ALS service user perspective on healthcare services. Multidisciplinary care is often supplemented by the voluntary sector to compensate for service gaps within public and/or private healthcare. People with ALS have expectations for dignified care and seek multidisciplinary intervention to meet a combination of physical, emotional, and social needs. People with ALS report satisfaction with assistive devices. However, they are often dissatisfied with delays in diagnosis and with how the diagnosis is communicated to them. People with ALS seek information to make choices about care but feel that they do not receive sufficient information. People with ALS have uncertainty in relation to accessing care, feel that healthcare professionals lack knowledge about ALS, and view services as fragmented and inadequate to alleviate their burden of care. Some people with ALS also view service providers as disconnected from the challenges posed by ALS and inattentive to the personal losses incurred by them.

2.6 Decision-making among people with ALS: Adapting to change

The studies that have investigated the ALS service user perspective on care indicate that their decision-making processes about services are complex. People with ALS engage with a broad range of services. They accept *and* decline services, and in some cases, change their preferences for care. A multitude of factors can influence how they make decisions about care. Physical disability alone has little effect on *how* people with ALS make decisions about their care. Instead, their personal values and their desire to maintain control and sustain quality of life are more likely to influence their preferences for care.

Psychological distress and their perceptions surrounding care burden also impact on the decisions they make about their care. Some people with ALS have conflicting feelings about care and their preferences for care change over time in order to accommodate to support systems and their evolving perspective on living with ALS. The literature suggests *that people with ALS adapt to progressive disability within the context of their lives.*

The literature (as discussed in Sections 2.5 to 2.5.7) suggests that people with ALS decide about care in the context of both intrinsic (personal) and extrinsic (external) support systems. People with ALS engage with healthcare services but how they view and make choices about care is primarily grounded in how they interpret their own lives. In a study to identify what people with ALS view as meaningful in their lives, Fegg et al. (2010) found that people with ALS shift their focus from health issues to supportive relationships. They also found that severity of illness and degree of functional impairment might have little impact on what people with ALS find most meaningful in their lives. Fegg et al. (2005) reported that people with ALS shift towards self-transcendence values (benevolence, self-direction, and universalism) in order to cope with progressive illness. Conservation values (safety, harmony, and security) might also influence quality of life for people with ALS (Fegg et al., 2005). These findings suggest that services that focus on providing security and maintaining the status quo have potential to improve subjective wellbeing for people with ALS.

As stated, quality of life studies in ALS suggest that people with ALS alter their life values as they live with ALS. This phenomenon has prompted researchers to investigate (through qualitative research) how people with ALS make decisions in order to sustain what is important to them. King et al. (2009) identified a distinct decision-making process in people with ALS characterised by their ability to appraise change and select strategies in order to adapt to change. King et al. found that adjustment to life changing circumstances fostered a sense of wellbeing among people with ALS in order to cope with further change and progressive loss.

In a study to identify how people with ALS confront the disabling effects of ALS, Brown and Addington-Hall (2008) identified four consistent types of narratives among people with ALS: 'sustaining' life, 'preserving' life, 'enduring' life, and 'fracturing' of life. Brown and Addington-Hall suggest that people with ALS, despite encountering insurmountable

loss (i.e. 'fracturing' of life), have the capacity to 'preserve' and 'sustain' by resisting death and by focusing on what remains. These findings resonate with those of Locock et al. (2009). Locock et al. found that life with ALS was characterised not only by 'disruption' and 'abruption' but also by 'repair' and 'continuity' in order to cope with change. Biographical disruption is characterised by isolation and by alteration in definitions of self. Abruption is denoted by a feeling of life ending with associated loss. Locock et al. found that people with ALS also managed to 'repair' by regaining control, maintaining normality, creating a new normality, finding new meaning and identity, and by negotiating between feelings of acceptance and avoidance. Their findings are consistent with those of Young and McNicoll (1998) who reported that people with ALS reappraised their life situations in order to maintain a positive outlook on life and find meaning in the present. These findings also resonate somewhat with those of Mistry and Simpson (2013) who found that people with ALS experience loss *and* manage to accommodate to loss.

2.7 Understanding adaptation to illness

2.7.1 Adjustment to illness: A psychological approach

Brownlee et al. (2005) state that the process of adjustment to illness constitutes not only standard cognitive processes about cause and effect but also an array of emotional representations which are grounded in people's lives. Brownlee et al. proposed a self regulation model whereby people use their knowledge and life experiences to construct self-representations and these self-representations in turn influence how people view benefit to their actions. The process by which people attempt to maintain their self-representations whilst accommodating what they know about illness i.e. illness representations, involves establishing certain propositional rules which enable people to cope with change imposed by illness. Multiple contextual factors might influence how people construct these rules.

Sprangers and Schwartz (1999) suggest that people with life-limiting illness change their expectations in life in order to cope with illness. In psychological terms, this process is referred to as 'response shifts' which constitute emotional and/or cognitive recalibrations in internal views associated with changing context (Sprangers & Schwartz, 1999). Response-shift studies have found that not only do people who encounter life-limiting illness shift their priorities from one aspect of life to another but they may also scale back the number of their expectations in line with changing circumstances (Carver & Scheier,

2000). This suggests that people with life-limiting illness place more importance on fewer life domains as they adjust to illness.

2.7.2 Finding meaning in illness: A constructivist approach

Sharpe and Curran (2006) suggest that adjustment to illness for people with life-limiting illness requires establishing new and/or altered meanings in life in order to cope with illness. How people construct *chronic* illness and cope with changing health is well studied in the sociological sphere (Bury, 1982; Charmaz, 1991; Frank, 1995) and has been applied to a variety of chronic illnesses (Asbring, 2001; Charmaz, 2006b; Gisquet, 2008; Sanderson et al., 2011; Wilson, 2007). Bury (1982) suggests that illness can alter people's lives but people have the capacity to 'reconstruct' themselves despite disruption in their lives. Frank (1995) identified three different types of narratives in chronic illness: 'restitution' to recover; the 'chaos' associated with loss and despair; and the 'quest' for personal growth.

Charmaz (1995) suggests that people who encounter chronic illness establish altered identities and that a process of repair in chronic illness involves accepting and adapting to an altered identity. Illness can engender loss and people with chronic illness move to an altered identity in order to adapt to loss (Charmaz, 1983). According to Charmaz (1995), some people with chronic illness purse a 'super normal self' attempting to excel but others adopt a 'restored self' attempting to resume their lives as was before. Some people choose to remain in the past ('entrenched self') but many seek to reassess their lives ('developing self') as they live with illness. People's ability to move to an altered state is in turn influenced by time and their degree of disability (Charmaz, 1987).

Contemporary studies which explore how people construct illness have found that encountering illness is not only about 'disruption' and 'repair' but also about normalising the illness trajectory into one's life. Sanderson et al. (2011) describe 'biographical reinstatement' where people with rheumatoid arthritis described returning to a normal life through effective medication. Sanders et al. (2002) describe 'biographical continuity' where people perceived the debilitating effects of osteoarthritis as part of the normal ageing process. Similarly, Faircloth et al. (2004) detected 'biographical flow' in the narratives of people recovering from a cerebrovascular accident with pre-existing co-morbidities.

2.7.3 Terminal illness in amyotrophic lateral sclerosis: Context to experience

Most literature on biographical change in illness has focused primarily on chronic illness. There is a dearth of literature on how people with terminal illness construct meaning in their lives along the lines of their biographical trajectories (Locock et al., 2009). No other studies beyond those undertaken by Brown and Addington-Hall (2008), King et al. (2009), Locock et al. (2009), and Young and McNicoll (1998), describe how people with ALS construct meaning in their lives. This is perhaps surprising as satisfaction-with-life-research in ALS (Fegg et al., 2010) demonstrated that people with ALS can find new meaning as they adjust to the debilitating effects of ALS.

2.7.4 Finding meaning in terminal illness

The meaning of living with terminal illness is a growing area of research in the health and social sciences. McQuellon and Cowlan (2000) suggest that the diagnosis of a terminal illness moves people to an altered state in 'mortal time' from which there is no return. Insights into the adaptive processes for people with chronic illness may not adequately reflect the experiences of people who live with terminal illness. Reeve et al. (2010) studied the experiences of people with terminal cancer. They found that terminally ill people can manage 'disruptive' events to maintain an overall sense of well-being. However, maintaining 'continuity' can be exhausting in the face of ongoing loss and terminally ill people may require considerable external support to maintain continuity.

Wright (2003) suggests that death and the dying person are completely intertwined – death continually shapes the dying person and the dying person continually shapes his/her death. It is a process of constant adaptation, flexible, and open to change. According to Wright (2003), many normative ideas of coping in terminal illness have only served as illusionary in terminal illness. Whether or not acceptance or resolution can enable people to embrace death remains a matter of debate. However, certain factors such as age (Cicirelli, 2001) and culture (Timmermans, 2005) can alter personal meanings in death.

Numerous qualitative studies have investigated terminally ill service users' views on what they value most in their care although the majority of the literature explores the experience of cancer service users as opposed to service users with non-malignant diseases. Similar to the literature in ALS, Carter et al. (2004) found that the need to 'take charge' was of prime importance for terminally ill cancer service users. Karlsson et al.

(2012) found that people with terminal cancer were trusting of service providers but they also feared for the continuity of services to alleviate their suffering. Awareness of impending death can alert people with terminal illness to the need to engage with healthcare services (Fisher & Colyer, 2009). People's fear of suffering in terminal illness might also increase their desire to hasten death (Karlsson et al., 2012).

2.7.5 Awareness of dying: Context to the experience of healthcare services among people with ALS

As stated, life expectancy for the majority people with ALS is comparatively short. Invasive ventilation (i.e. long-term ventilation / tracheostomy) and non-invasive ventilation can improve survival in ALS but these interventions do not alter the neurological course of the disease. Some healthcare systems endorse invasive ventilation in ALS care (Borasio et al., 1998). In the Republic of Ireland, the cost of invasive ventilation extends beyond that which is available through public healthcare services and few people with ALS in Ireland engage with invasive ventilation. However, regardless of what benefits result from life-sustaining interventions, neuro-protective treatments have not yet managed to slow down the neurodegenerative course of the disease. ALS is a terminal disease. Studies of experiences of care from the viewpoint of people with ALS have not reported explicitly on service users' awareness of the terminal nature of the disease. Receiving a diagnosis of ALS and living through the progressive stages of the disease might result, for some people, in an awareness that they will, sooner rather than later, engage with end-of-life care.

Glaser and Strauss (1965) conducted a ground-breaking study on the social process of dying among terminally ill people and in doing so developed a theory about awareness of dying. Glaser and Strauss found that different contexts to care influenced awareness of dying among terminally ill people and different levels of awareness between service users and care providers impacted significantly on their interaction. They described four different types of awareness in dying: closed awareness where the person in unaware that they are dying but the healthcare professional is aware; suspicion awareness where the person suspects they are dying but the healthcare professional counters these suspicions; mutual pretence where both the dying person and the healthcare professional are fully aware but pretend otherwise; and open awareness where both the dying person and healthcare professional acknowledge that death is imminent.

Timmermans (1994) suggests that the way in which people cope emotionally with illness in itself defines the type of open awareness context. Timmermans (1994) describes different levels of 'open awareness context': 'suspended open-awareness context' during which time service users ignore information provided by service providers; 'uncertain open-awareness context' where service users dismiss information about the terminal nature of their condition and hope for positive outcomes; and 'active open-awareness context' when service users accept and prepare for impending death. Mamo (1999) suggests that the 'awareness of dying theory' developed by Glaser and Strauss falls short in explaining how people confront impending death because it does not accommodate the emotional dimensions of the experience of dying. Knowledge about the course of terminal illness and the cognitive processing of this information alone cannot account for the emotional adaptive strategies commonly employed by terminally ill people during end-of-life care (Mamo, 1999). Invariably, multiple factors influence the context of terminally ill people's experience of care. Contextual factors that influence ALS service users' experiences of care remain unclear.

2.8 Conclusion

2.8.1 The complexity of care in amyotrophic lateral sclerosis

Healthcare in ALS is complex not only because people with ALS have a broad range of care needs but also because multiple factors impinge on how people with ALS engage with services to meet these needs. The evidence suggests that ALS healthcare services fall short in meeting the needs of people with ALS.

How people with ALS engage with healthcare services and make decisions about care is central to the care process. Normand (2009) states that care outcomes are often influenced by service users' perceptions of care and not necessarily by care providers' intentions regarding care. Service users' views on services might differ from those of service providers. Service users' understandings of care are based on a combination of personal experiences and 'common sense' knowledge about illness. Greater attention to ALS service users' experiences of and preferences for services is an important component in the quest to achieve meaningful outcomes for people with ALS (Foley, 2011).

A body of interpretative research has emerged on the subjective experience of living with ALS and the decision-making processes in the lives of people with ALS. Living with ALS

poses many challenges for people with ALS but despite physical loss, living with ALS also creates a need to adapt to progressive disability in order to respond to the challenges posed by the disease. However, apart from studies which have investigated decision making about life-sustaining interventions in ALS, little is known about how people with ALS make decisions about their care. How people with ALS adapt to illness might differ from other diagnostic groups. As for non-terminally ill service users, people with ALS move through different stages of care with evolving disability but unlike non-terminally ill service users, people with ALS encounter death at the end point of their disease trajectory. In comparison to other terminal illnesses, there is a greater degree of certainty in ALS in terms of how and when people die. Multiple contextual factors shape how people with ALS experience care. It can be assumed that awareness of the terminal nature of ALS is a factor that shapes healthcare experiences.

People with ALS have expectations around the need to exert control and be autonomous in their decisions about their care as they negotiate between accepting healthcare services and seeking to maintain normality in their lives. Like other terminally ill service users, they make decisions about care as they adapt to change. In this light, further research was required to pinpoint the key parameters of healthcare experiences among people with ALS in order to explain how people with ALS engage with services.

2.8.2 Conceptualising healthcare experiences for people with ALS

The outcomes used in ALS multidisciplinary care studies to date (survival, cost), are conceptually different to service users' experiences of healthcare services. Service users' experiences of care are subjective; cost and survival are objective. Experiences of care are more complex constructs than survival and cost because they are shaped by people's expectations in domains that are important to them. A number of quality of life (QoL) / meaning in life measures (Fegg et al., 2008; Simmons et al., 2006) are used by service providers and researchers to capture service users' views on what is important to them but these measures are rarely used to evaluate service users' perceptions of services or indeed the impact of care from their perspective. This is not surprising as QoL measures are self appraisals of wellbeing and can exist independently of specific experiences of ALS services. The use of generic palliative care outcome tools might not necessarily be sensitive to the needs of people with ALS (Hughes et al., 2004). Apart from a limited number of qualitative studies on perceptions about services and the meaning of care in

ALS, little was known about *how* people with ALS interpret, approach, and experience healthcare services. Further research was required to pinpoint the key parameters of ALS service users' experiences of healthcare services. In the Republic of Ireland, healthcare experiences of people with ALS had not been studied.

Grounded theory is a research method that builds concepts and theory from data with the view to formulating a substantive explanation about phenomena. Identifying the parameters of ALS service users' experiences of healthcare services through grounded theory would in turn facilitate the development of a theoretical framework that explains how people with ALS understand and use their healthcare services. Importantly, grounded theory method saturates concepts in terms of their properties and dimensions and so the framework it builds accounts for variation in experiences. As stated, it is only by exploring variation in context and in meaning for people with ALS about their healthcare services that their experience of healthcare services can be conceptualised and their use of healthcare services understood.

Foley et al. (2012b) argue that inclusion of measures that tap into the ALS service user perspective on healthcare services would yield a different, and fuller, picture of outcomes for people with ALS. Identifying the key parameters of healthcare experiences among people with ALS through qualitative research would first unearth key domains of care that are important to them. These domains can then be translated into a measure that accounts for the ALS service user experience of healthcare services. Such an instrument would enable researchers and service providers to measure ALS service users' satisfaction with services and test the relationship between their satisfaction with services and other outcomes of care. In turn, this would improve quality of care for people with ALS and promote services that are driven by their needs. This qualitative investigation is the first step in *grounding* such a measure in the lived experiences and perceptions of people with ALS.

2.8.3 Sensitising framework for the research

The evidence thus far on ALS service users' decisions about care points to an overall desire to maintain normality and remain in control as they judge what types of services are most appropriate to them. People with ALS expect timely provision of a broad range of services

but they also negotiate between accepting and declining these services. Opting for and/or declining services may well relate to how they construct their own illness trajectory.

It is possible that the experiences of people who encounter terminal illness may differ somewhat from the experiences of people who live for indefinite periods with disability. Unlike many other disabling illnesses, there is more certainty in ALS in terms of how and when people die. People with ALS face difficult decisions as they move along their illness trajectory and inevitably encounter discussions on the end-of-life stage (Oliver & Turner, 2010). How people with ALS approach these discussions is influenced by their awareness, and in turn by the socio-cultural context of their lives. Hence, a deeper understanding of ALS service users' experiences of care, in the context of terminal illness, was required.

Chapter Three

Research methodology and methods

3.1 Choosing qualitative methodology to understand the ALS service user perspective

Much debate exists in the health and social science fields with respect to the role of qualitative research and to the epistemological and ontological positions adopted by qualitative researchers (Bryman, 2012). However, the purpose of this chapter is not to dwell on this debate but rather to outline why qualitative research methods were adopted in order to capture the ALS service user experience of healthcare services.

Unlike quantitative research approaches which excel at testing hypotheses derived from existing theories, the primary purpose and strength of qualitative research lies in generating new theory about human behaviour. The topic of this study, healthcare experiences among people with ALS, can be most fruitfully interrogated with the use of qualitative methods. The fact that this was a poorly understood, novel topic in the literature further added to the suitability of the chosen method because it was essential that the parameters of ALS service users' experiences of healthcare services be mapped out before they can become subjects of further (qualitative and quantitative) study focussed on the key aspects of their experiences. In other words the method, to be outlined in this chapter, was an ideal match to the research question. It is anticipated that the findings generated from this study will facilitate further qualitative and quantitative studies as it pinpoints the most central areas of further scientific enquiry and gives an indication of the key variables that other studies can collect data on.

Pope and Mays (2006) state that qualitative research is important when research questions are not amenable to measurement i.e. when the 'what's', 'why's', or 'how's' of a particular phenomenon need to be identified as opposed to questions of how many, how much, or how often. Identifying how people think about healthcare services and make decisions about care involves understanding how social practices in healthcare are created and what meaning these practices have for people within specific contexts. A basic tenet of sociology is that people act on the basis of their definitions of situations rather than on the 'objective' reality of the situation (Bourgeault et al., 2010). Indeed, focusing on outcomes of care which exclude the service user perspective limits service providers' understanding of how and why service users engage with healthcare services.

Holloway (2005) states that, how service users behave can only be understood when they are asked about their experiences. This study is person-centred because it looks to people with ALS in an effort to understand how *their* perceptions impact on the care process. This study sought to capture the ALS service user experience of services by asking people with ALS about their experiences. A greater appreciation of what is important to people with ALS is necessary in order to shape policies which best fit with their expectations of healthcare services. Identifying what is important to people with ALS also helps service providers understand the emotions and actions of this population group who live with a terminal condition which, in itself, creates the need to engage with healthcare services.

Corbin and Strauss (2008) suggest that researchers are drawn to qualitative research because it allows them to connect to research participants at an individual level. Capturing ALS service users' individual experiences of their healthcare services was necessary in order to understand how they attach meaning to services and decide about care. Using qualitative methods to investigate a particular phenomenon is (and was in this study) an interactive process. The researcher becomes part of the research process and meaning attached to what participants share about their experiences is constructed not only by the participant but also by the researcher (Charmaz, 2006a).

3.2 Critical Realism: An ontological perspective in understanding complexity of experience

Traditionally methodologists have adopted polarised ontological positions between qualitative and quantitative methodologies (Cruickshank, 2012). Some argue that the paradigms which underpin positivism and constructivism are fundamentally different whilst others argue that the dichotomy between the two is largely false. Here, the researcher does not classify her ontological position as that which is constructivist or positivist. Rather, her ontological position is that of a critical realist.

Critical realism, otherwise known as complex realism, is a post-positivist approach that positions itself between positivism and constructivism (Bhasker, 1997). It adheres to the view of one reality and to the existence of a reality independent of mind. However, importantly, critical realists recognise that people have different perspectives on reality and how they engage with phenomena shapes that reality.

Bhaskar (1997) describes a stratified emergent ontology which consists of three realms of reality; the actual, the real, and the empirical. The actual domain refers to events and outcomes that occur in the world. The real refers to the underlying, often latent structures and relationships (which can give rise to tendencies) that change events and outcomes. The empirical represents the human perspective of the actual and the real and so in this light reality is always changing (fallible). A central tenet of critical realism is emancipation (agency) where individuals themselves can effect change because how they act shapes reality.

Clark et al. (2007) state that understanding key factors and processes which impact on health service outcomes is necessary in order to achieve meaningful outcomes for service users. However, the processes that impact on outcomes are complex. Critical realism offers a perspective to help make sense of the complex nature of actions within healthcare and how such actions influence outcomes. It recognises that structural factors and agency have an important role in influencing experiences of phenomena. However, it does not seek to objectify the human perspective or remove the importance of the human perspective on experiences (Clark et al., 2008).

3.2.1 Critical Realism applied to ALS care

Critical realism for qualitative research in ALS care is useful because it seeks to render complexity intelligible by explaining *how* and *why* things happen in ALS care. Critical realism considers the existence of wider knowledge about ALS healthcare services but pays close attention to social meanings for ALS service users. Importantly, critical realists view healthcare services as complex, open, and changing systems in which people with ALS have the capacity to shape their own experiences.

There is *a* reality to healthcare services for people with ALS in the Republic of Ireland because there are 'facts' about and 'variables' within healthcare services which *can be* measured and which differ from other healthcare systems. The purpose of undertaking qualitative research was to illuminate the key parameters of ALS service users' experiences of healthcare services by exploring the different ways in which service users interpret and engage with services. The different ways in which people with ALS interpret and engage with healthcare services shape the reality of healthcare services for people with ALS and so in turn, shape what is measured.

For critical realists, the relationship between agency and structure is complex. Agency is constrained by multiple structural factors but structure can also be influenced by agency. As discussed in Chapter Two, multiple factors might influence how people with ALS engage with healthcare services, and how people with ALS make decisions about services is complex. Both intrinsic (agency) and extrinsic (structure) factors (including the relationship between them) have influenced how people with ALS make decisions about end-of-life care. However, it remains unclear *how* and *why* people with ALS engage with healthcare services. As a critical realist, the researcher acknowledges that how people with ALS interact with healthcare services is influenced by a combination of personal, social, cultural, and political factors. Explaining how and why people with ALS interact with services and make decisions about their care (from their perspective) 'in the real world', is key to understanding complexity of the service user experience. In this study, grounded theory method is used to explain why and how people with ALS engage with healthcare services.

3.3 Grounded Theory: Understanding psycho-social process

Grounded theory is a systematic research approach that builds concepts and theory from qualitative data (Corbin & Strauss, 2008). It is concerned with the basic psycho-social processes of behaviour because it seeks to explain *how* and *why* people behave in certain ways. Originally considered to be inductive in nature, grounded theory methods have evolved to incorporate both deductive and abductive methods (Charmaz, 2006a; Corbin & Strauss, 2008).

3.3.1 Key tenets of Grounded Theory method

Unlike quantitative inquiry, grounded theory does not work from an existing theory but instead generates theory about the phenomenon of interest. Grounded theory method was used in this study because the overall aim of the research was to develop substantive theory about how people with ALS understand and use their healthcare services. Importantly, grounded theory method attempted to identify conditions that might influence ALS service users' actions and interactions with healthcare services. It is assumed that context shapes their actions and so key to explaining their actions, is to capture variation in both context and experience.

Specific characteristics of grounded theory method distinguish it from other qualitative research methods. Grounded theorists not only code data for concepts but also identify relationships between concepts/categories by making theoretical comparisons between them (Corbin & Strauss, 2008). Categories have properties and dimensions. Properties describe categories and dimensions reflect the variation within categories. Participants are sampled based on emerging concepts until categories are exhausted in terms of their properties and dimensions. This form of sampling is known as 'theoretical' sampling. Key to developing substantive theory is to 'saturate' categories in terms of variation and meaning (Corbin & Strauss, 2008).

As for other qualitative research methods, the researcher has a key role in the research process because data is constructed by *both* the researcher and participants (Charmaz, 2006a). In this light, there is a reflexive process to grounded theory method because the researcher's own knowledge and assumptions about the data constitute, in part, theory building.

3.3.2 Symbolic Interactionism: Meaning and action

The epistemology of grounded theory method has evolved largely from symbolic interactionism (Blumer, 1969), a theoretical perspective which emerged from the philosophy of pragmatism. Pragmatism is based on the principle that knowledge is created through action and interaction (Dewey, 1929). Reflexive inquiry arises out of the need to interpret meaning in order to respond to particular situations. Focusing on the consequences and contingencies of actions in turn leads to different actions.

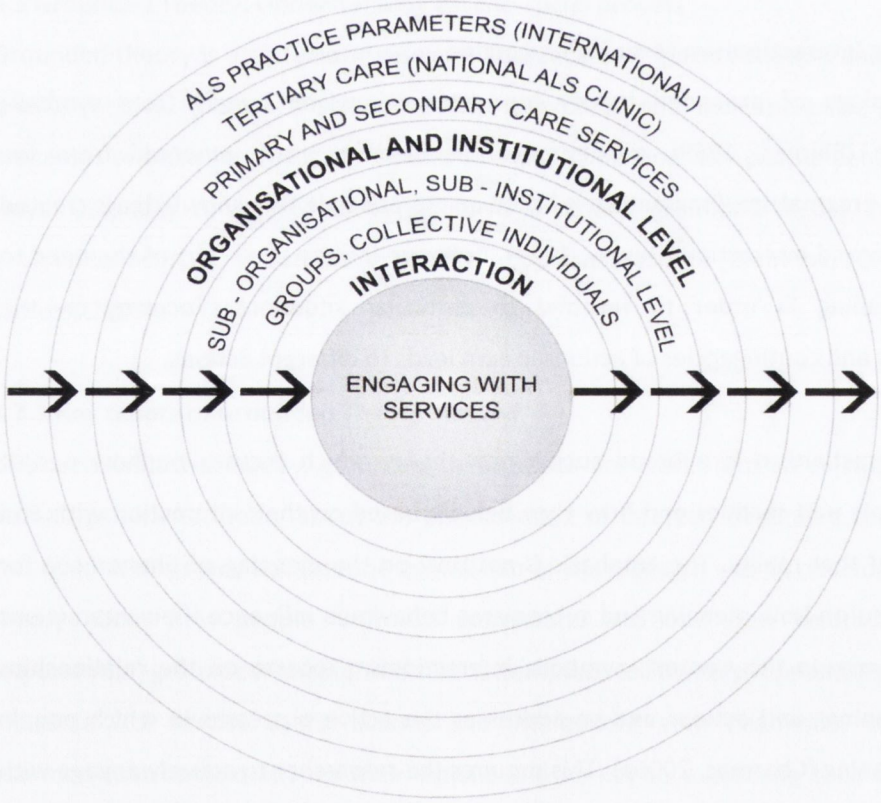
Symbolic interactionism is a broad sociological theory which focuses on how people interpret events and realities and how they behave based on their interaction with and experiences of that reality. The emphasis is not only on the meaning of phenomena for people but also on how meaning and subsequent behaviours influence their interactions with phenomena. In this context, symbolic interactionism focuses on the relationships between meanings and actions and so addresses the active processes in which people construct meaning (Charmaz, 2006a). This requires the researcher to actively engage with the research participant. Adopting a symbolic interactionist perspective was appropriate in this study because the focus was not only on how people with ALS interpret healthcare

services but also on how they interact with services based on the meaning they attach to healthcare services.

3.3.3 Understanding healthcare experiences among people with ALS: The conditional/consequential matrix in Grounded Theory

Corbin and Strauss (2008) provide a conceptual guide labelled the 'Conditional/Consequential Matrix' to depict the complex nature of how and why people act and interact in the empirical world. Many contextual layers surround the individual ranging from individuals and groups i.e. micro-conditions, to broader community, national, and international agendas i.e. macro-conditions. Multiple structural conditions can determine action/interaction and how people respond within specific contexts can also influence outcomes in other contexts. Figure 2 provides an illustration of the 'Conditional/Consequential Matrix' for people with ALS.

Figure 2 Conditional/Consequential Matrix



Adapted from Corbin J, Strauss A (2008). Basics of Qualitative Research (3rd edition)

As described in Chapter Two, how people with ALS interact with healthcare services is complex. There is both a micro- and macro-context to their experience of care. Micro-context refers to the things and individuals close to the ALS service user and macro-context reflects broader agendas and structures (national and international) both within and outside ALS healthcare services. In this study, it is assumed that both micro- and macro-conditions impact on how people with ALS interact with their healthcare services. It is also assumed that decisions people with ALS make about their care shape and/or are shaped by a combination of all contextual layers.

As discussed in Chapter Two, there is a situational context to the experience of ALS healthcare services in the Republic of Ireland. Services are led by a specialised ALS clinic. Models of care have been developed by the clinic and are shaped by international best practice guidelines. Services provided by and organised through the specialised clinic are in turn modulated by the national agenda of healthcare services. National agendas impact both community and institutional levels of care including the individuals within these institutions. It is assumed that a change in structural conditions across all contexts can lead to change in how service users respond to services. The aim of grounded theory in this study is to identify the different ways people with ALS respond to healthcare services across these contexts.

3.4 Some ethical considerations prior to data collection and analysis

It has been argued by some that involving terminally ill people in research is unethical because terminally ill people (by definition, everyone with ALS) invariably don't benefit from the outcomes of research (Duke & Bennett, 2010). However, the researcher argues here that investigating the experiences of care from the terminally ill service user's perspective is no less important than capturing the experiences of services users who do not live with terminal illness. ALS *is* a terminal condition and it is conceivable that ALS service users' concerns and expectations about their healthcare services are shaped, in part, by the context of living with a terminal illness. As stated in Chapters One and Two, it is only by investigating the service user perspective that service providers can know whether the criteria used to judge care outcomes are consonant with the concerns facing people with ALS.

The researcher acknowledges that there are particular research sensitivities in palliative care research (Addington-Hall, 2002; Casarett, 2005) which are not given equal attention in other fields of healthcare research. These can include the effect on people who are asked to participate but who decline, the effect on people who are asked to participate but where certain conditions (e.g., declining health) prevent them from participating, and of course the emotional impact of sharing their experiences which for many includes feelings about death and dying. In some situations researchers also face difficult decisions around sampling vulnerable service users who might offer valuable insights into emerging theory. These were just some of the challenges faced by the researcher in this study. How the researcher dealt with all ethical considerations in data gathering and analysis is described within the relevant sections of this chapter.

3.5 Reflexivity and the researcher

Reflexivity refers to how the researcher's actions and role in the research shape how the research is conducted and how the findings are interpreted (Mason, 2002). The reflexive researcher recognises that all knowledge is affected by the social conditions under which it is produced. In this study, participants' experiences were filtered through the researcher's interpretation and her experience as a clinician in ALS care shaped, to some degree, how she and participants interacted with each other. Reflexivity has an important role in grounded theory. Key to developing theory is to remain open to all possible understandings *and* to remain theoretically sensitive to the data and methods employed to capture the data (Corbin & Strauss, 2008). The reflexive process never ceased for the researcher during the course of the research and the effect she had on all procedures in this study is captured throughout this chapter.

3.6 Sampling ALS service users: Why only the ALS service user perspective?

This study placed people with ALS firmly at the centre of the enquiry. The majority of studies that have explored expectations of services among people with ALS have included carers. This research focuses only on the ALS service user. Living with ALS can have a profound effect on family members (Aoun et al., 2012) and family carers of people with ALS experience increased carer strain when they feel unsupported by healthcare services (Peters et al., 2012). However, the central aim of this study was to generate substantive theory about how *people with ALS* understand and use their services. It was important to bear in mind that, particularly in a qualitative PhD where fieldwork was undertaken by a

single researcher, including caregiver respondents would have reduced the number of people with ALS that could be interviewed. This would run counter to the aim of placing people with ALS firmly at the centre of the enquiry as the most reliable informants regarding their own healthcare experiences. The carer perspective as context to service users' experiences was often captured through participants' narratives and in some cases (as described in Sections 3.7.3.1, 3.7.6.1.1, and 3.7.6.1.2) impacted on sampling and on how interviews were conducted.

Neither did this study seek to capture healthcare professionals' views. Some investigations on the experience of ALS healthcare services have included the service provider perspective (e.g., Brown et al., 2005; Hughes et al., 2005) in an effort to capture a wide range of experiences from different stakeholders. However, as stated, the aim of this study was to identify the key parameters of the ALS *service user* experience of healthcare services so that it could pinpoint further areas of scientific enquiry on aspects of healthcare services that are most important to them.

3.7 Research design

Bryman (2012) suggests that choice of research design reflects the decisions made by the researcher in terms of how to capture data which best fits with the research aims. This research sought to understand the complex nature of ALS service users' interaction with healthcare services and the research was designed to capture this complexity. Capturing complexity in the ALS service user experience of services required the researcher to: explore multiple perspectives of people with ALS on their healthcare services; account for variation within their experiences; and understand how people with ALS engage with healthcare services in response to context.

As stated, the overall aim of the research was to develop substantive theory about how people with ALS understand and use their healthcare services. Corbin and Strauss (2008) state that theories arrange concepts about phenomena in order to define and explain phenomena. They also provide a basis for considering how what is not yet known might be organised (Silverman, 2010). As discussed in Chapter Two, little was known about how people with ALS interpret their healthcare services including key variables that explain why and how they engage with services. In this light, grounded theory method was well

suited to the task of explaining how people with ALS experience services and make decisions about their care.

3.7.1 Sourcing the sample: The Irish ALS population-based register

The researcher sampled participants from the Irish ALS population-based register. The Irish ALS population-based register has been in operation since 1995 and is compiled by an Irish ALS research group based at Trinity College Dublin. The register contains epidemiological and clinical data on people with ALS in the Republic of Ireland and claims over 95% ascertainment of the ALS population. As described in Chapter Two, up to 80% of people with ALS in the Republic of Ireland attend the specialised ALS clinic at Beaumont Hospital in Dublin at some point in their disease progression. These service users are captured by the ALS register when they first attend the clinic. Those who do not access the clinic are ascertained by multiple sources and tracked directly by the ALS register (Yeo et al., 2010). On signing a confidentiality agreement with the Irish ALS research group, the researcher was given permission to sample participants from the register.

3.7.1.1 Using the ALS population-based register to sample participants: Context and benefit

Apart from complex ethical challenges that surround sampling terminally ill people in research, where to *locate* people who are willing to participate in topics that explore sensitive issues can determine who researchers sample and where they go to sample (Stiel et al., 2010). Accessing terminally ill populations may offer additional challenges not encountered in non-palliative care fields. Gatekeeping by different groups can impact on recruitment in palliative care research (Ewing et al., 2004) and potentially limit the researcher's capacity to explore the service user perspective (Wright & Flemons, 2002). Inevitably these obstacles can restrict grounded theory researchers, where developing theory is supposed to guide who they sample and where they go to sample (Corbin & Strauss, 2008).

Sampling from the Irish ALS population-based register removed many but not all of these obstacles. Clear consent by personnel who compiled and monitored the register was required before asking people (who were amenable to the sampling procedure) to participate. A small number of people on the register who the researcher felt were amenable to the aims of sampling (n=2) were highlighted as 'do not contact'. These

people were highlighted as 'do not contact' at the discretion of the Director of the ALS clinic.

As stated, the majority of people on the ALS population-based register attend the national ALS clinic. Permission to access background data on service users through their clinical records was granted by the Director of the clinic. Key to theoretical sampling is that the researcher samples participants based on characteristics and/or experiences that can further develop emerging categories in terms of their properties and dimensions (Corbin & Strauss, 2008). Access to data on service users was an important tool in the research process because it enabled the researcher (in advance) to identify potential participants that could add further variation and meaning to concepts that were emerging from the data. Overall, access to the ALS population-based register minimised delay in sourcing participants who were sampled on the basis of emerging findings and enabled the researcher to secure a sample sufficiently diverse to develop substantive theory about how and why people with ALS engage with healthcare services.

3.7.1.2 National ALS clinic: Context to sampling and participant consent

It is important to note that sampling non-ALS clinic service users from the population-based register offered more challenges than sampling service users who attended the national ALS clinic. In these cases, the researcher had less information available to her about potential participants. Nonetheless, data captured by the ALS population-based register on non-ALS clinic service users and general background information on them known to the register personnel was also used to inform the sampling procedure.

Pre-existing relationships between participants and the ALS clinic more than likely influenced some participants' decisions to participate. ALS research in the Republic of Ireland is primarily conducted by (or in collaboration with) the Irish ALS research group based at the clinic. Approximately 65% of people with ALS in the Republic of Ireland participate in research (Elamin et al., 2011). ALS service users' propensity to engage in research based at the clinic was in itself context to service users' participation in this study. Although subsequent sections of this chapter discuss the reflexive role of the researcher in interviewing and data analysis, her association with the ALS clinic was also context to obtaining participant consent. Prior to conducting the research, she worked at Beaumont Hospital as a clinical specialist occupational therapist and had been a long-

standing member of the multidisciplinary ALS clinic team. This research was carried out in collaboration with the Irish ALS research group based at the national ALS clinic. In addition, the Director of the ALS clinic and of the research group was clinical sponsor to the researcher for the duration of the study.

3.7.2 Ethical approval to conduct the research

Ethical approval to conduct this research was granted by the Beaumont Hospital Ethics (Medical Research) Committee, Dublin on the 27th September 2010 and by the Research Ethical Approval Committee at the School of Social Work and Social Policy, Trinity College Dublin, on the 18th April 2011.

3.7.3 Obtaining participant consent

The process of obtaining consent involved a number of steps. Firstly, each selected individual was contacted via a phone call during which the researcher explained the study in full. In the case of people who were anarthric or severely dysarthric, first and/or subsequent contact was invariably with their significant other or paid carer. Three people (two of whom were anarthric) were contacted by email because they ordinarily communicated by email. A participant information leaflet (Appendix B) was posted to each individual who expressed interest in participating. Each individual was given at least five to seven days to read the participation information leaflet and to consider their involvement. The researcher then followed up with each individual (or where appropriate with their significant other and/or carer) by phone or as appropriate by email. For those who agreed to participate in the study, each participant signed a consent form (Appendix C) or if physically unable to do so, permitted their significant other / carer to sign the consent form on their behalf.

As stated, the researcher was aware of the potential for some participants to give consent to participate based on the fact that many people with ALS in the Republic of Ireland (the majority of who attend the tertiary ALS clinic at Beaumont Hospital, Dublin) engage with research. The participant information leaflet clearly stated that participation was entirely voluntary and that choosing not to participate would have no effect on the care they received. However, it can be assumed that the above context influenced, to some degree, the decision to participate.

3.7.3.1 Carer role in negotiating participant consent

Although the researcher set out to capture the *service user* perspective, variation among participants in terms of their physical abilities meant that for some participants, their significant other or paid carer was a gatekeeper at first and subsequent contact and had an active role in negotiating participant consent. A number of participants had already developed severe dysarthria (n=6) or anarthria (n=2) and were therefore less likely to communicate by phone. It is worth noting that more service users with high level disability (including deteriorating or loss of motor speech function) and whose significant other was *the* point of contact, declined to participate than did service users with high disability who were themselves the first point of contact. In these situations the decision not to participate was communicated to the researcher by their significant other and so it was impossible to ascertain whether or not these service users were informed about the invitation to participate in the research. Service users without speech difficulties were more likely to be the first point of contact.

There is no doubt that for some participants, their significant other was to all intents and purposes, their gatekeeper. In truth, this bothered the researcher somewhat because for some service users who were severely dysarthric and/or who had already encountered significant disability, the researcher sensed that their carers made the decision (whether or not to participate) on their behalf. The researcher recognises that family carers have an integral role in the lives of people with ALS and as service users' needs increase, the burden of care falls predominantly on their carer. However, people with ALS and their carers are likely to differ in their opinions about care.

Guidelines for best practice in ALS care (Andersen et al., 2012) highlight the need to include the carer and/or significant other in decisions about care but there is little debate on how carers should, if at all, mediate between service users and researchers. It is important to note that all participants who still lived at home wished to participate in the study from their home. It is possible that the decision to capture the service user perspective in the naturalistic setting required in some cases that carers be open to research being conducted in their homes. In this context, it was perhaps inevitable that some carers negotiated participant consent. It is also likely that some carers were understandably protective of their partner/family member and had reasonable concerns about the potential effects of participation on their partner/family member.

3.7.4 Ethics and sampling

Access to the ALS population-based register removed many of the logistical problems often associated with sourcing terminally ill participants but asking people with ALS to talk about their experiences was still complex. The emotional impact of participation on participants or indeed the emotional impact of being asked to participate, was for some, significant. Not all people who were asked to participate agreed to participate. Some people declined on the grounds of deteriorating health. Two people were upset that they were approached about the study. Both indicated that living with ALS entailed high levels of anxiety, and both expressed despondency about their future with ALS. Their response was relayed by the researcher to personnel who compiled the register. Three people who had agreed to participate were subsequently unable to do so due to rapid deterioration in their condition.

3.7.5 Recruitment: Sampling for variation and theoretical sampling

Theoretical sampling in grounded theory is a sampling procedure whereby the sample size is determined by the data gathered (Corbin & Strauss, 2008). At the onset of this study, it was impossible to determine *ex ante* exactly how many participants would be selected in total. This is because theoretical sampling is driven by concepts / categories that emerge from the data and the need for further elaboration to develop theory that is grounded in the data (Corbin & Strauss, 2008).

Some debate exists on what exactly constitutes theoretical sampling (Coyne, 1997) and how other forms of sampling, in particular purposive sampling, is required in the preliminary stages of most grounded theory studies (Morse, 2007). Consistent with grounded theory method, the researcher collected and analysed data in tandem in order to generate concepts for further sampling. During the early stages of the study, she purposively sampled participants to look for variation in experiences of healthcare services among people with ALS in an attempt to capture diversity of experience. From early analysis of the data emerged concepts. She then theoretically sampled participants based on emerging concepts in order to 'exhaust' these concepts and categories (larger concepts) for both meaning and variation (Corbin & Strauss, 2008). For example, when she identified that ageing and parenthood were emerging as key categories that shaped how participants made decisions about their care, she continued to sample participants for variation in these contexts (e.g. people with ALS at different life stages, and those who

had dependents and those who had no dependents). Ongoing comparisons between data in the theory building process enabled the researcher to identify relationships between concepts as she sampled. Sampling ended when the data were 'saturated' for variation and meaning i.e. when no conceptually or theoretically significant data were emerging from additional interviews (Corbin & Strauss, 2008). Data collection and data analysis are detailed separately in the sections that follow.

In total 34 people were sampled from the Irish ALS population-based register between September 2011 and August 2012 using purposeful and theoretical sampling procedures. The researcher sampled participants without pre-defined geographical location and so she captured experiences of participants who lived in different regions across the Republic of Ireland. Section 3.8 describes the sample. The sample captured variation along a number of dimensions including age, geographical location, ALS type, duration of disease, level of disability, utilisation of healthcare services, and life stage. In the interest of participants' anonymity, specific locations cannot be disclosed and geographical locations for all participants are denoted by region in Table 1 (pp. 60-62).

3.7.6 Data gathering

3.7.6.1 Qualitative interviews

In this study, it was decided that data collection would comprise qualitative interviews with participants. Although other forms of data collection such as observation are common in grounded theory (Timmermans & Tavory, 2007), choosing qualitative interviews as the primary source of data collection was appropriate in the context of both inevitable intrusion into participants' time and the time-frame of the research as a PhD study. However, qualitative interviews did not alone constitute the data. Consistent with grounded theory method, analysing qualitative interview data generated both reflexive and theoretical memos on the data (Corbin & Strauss, 2008) which guided theoretical sampling and in turn helped when generating theory. Section 3.7.8 describes how a memo was used as a tool for analysis.

3.7.6.1.1 Ethics and the qualitative interview

There were potential risks in conducting qualitative interviews with people with ALS including the emotional impact of the interview on participants who were living with such a rapidly progressive disease. The researcher was also attuned to how participation in

qualitative interviews can result in fatigue, in particular for those participants in advanced stages of the disease and/or those who were close to death. However, seeking variation in concepts that emerged from the data inevitably guided the researcher to sample participants at various stages of the disease and the researcher did not shy away from interviewing participants in advanced stages of the disease when the grounds for sampling them fit with the sampling procedure. The researcher was sensitive to participants' needs in the context of her professional expertise in ALS care and the extent to which she probed was modulated by her in accordance with how she judged the capabilities and awareness of each participant. All participants were aware that they could stop the interview at any time or take as many breaks as they wished during the course of the interview. As expected, this was necessary for some participants with dysarthria because the physical effort to speak for sustained periods was exhausting for them.

Decisions to include (or not to exclude) people with dysarthria and anarthria when such people were amenable to the aims of sampling, were complex. In total, two participants were anarthric and six participants were severely dysarthric and so inevitably shaped both how these interviews were conducted and the content of the data which was subsequently analysed. Anarthric participants used high-tech-switch-activated and/or touch-screen alternative and assistive communication (AAC) devices with synthetic speech output to communicate their responses. The majority of severely dysarthric participants used low-tech AAC devices (e.g., communication boards, writing pads) to assist them. All but one of the participants who had severe dysarthria (i.e. five) were assisted by their significant other who was attuned to participants' worsening intelligibility. As the researcher interviewed anarthric and severely dysarthric participants, momentarily, she decided to sample this group nearer the point of data saturation and include their perspective to test out categories. She considered potential limitations in their data for unearthing new categories. However, on reflection, she decided against this because the emphasis was not on the amount of data that she captured but on the meaning of what participants communicated. Dysarthria is a common feature of ALS and many people with ALS develop dysarthria at some point along their disease trajectory. To have excluded the perspectives of severely dysarthric and/or anarthric people with ALS whilst building theory would have rendered theory building less meaningful. The main purpose of conducting the research was to capture the service user perspective, not to exclude it.

It is important to note that nine of the 34 participants, prior to the research study, had encountered the researcher in a clinical setting at Beaumont Hospital. The researcher had worked at Beaumont Hospital until the point of data collection. Of these nine participants, she had encountered five on one occasion only and one on two occasions only at the ALS clinic. She had had more frequent contact with two other participants (at the clinic) and with one participant in the inpatient neurology ward. Sampling was guided by emerging findings (not by pre-determined stratification of potential participants) and so in effect, this resulted in selecting some participants who were already known to the researcher. It is difficult to ascertain how much, if at all, previous clinical encounters between the researcher and participants shaped subsequent interviews. It is possible that encounters with the researcher prior to the interview might have negated, for some, feelings of apprehension about participating in in-depth qualitative interviews that would more than likely raise sensitive issues. However, the majority of participants were relaxed and all participants were open to sharing their experiences.

Although this study did not seek to capture the carer viewpoint, seven participants were interviewed with their significant other present for the entire interview and two participants were interviewed with their significant other present for a portion of the interview. All other participants were interviewed on their own. Of those interviews conducted with the significant other present, three interviews were conducted with an additional family member present (in all cases, an adult daughter) for a portion of the interview. The researcher's preference was to interview participants on their own knowing that the presence of another would inevitably influence how participants shared their experiences. During early sampling, there was an oversight on behalf of the researcher in that she did not consistently explain to participants that it was her preference that interviews be conducted without their significant other present. Notwithstanding the expression of this preference for later interviews, some participants' significant others wished to be present for the interview. It was anticipated that the carer role as context to the service user experience would come through participants' narratives but the study's aim was not to explore the carer viewpoint. However, it would have been unethical to exclude participants (who had already been asked to participate) because of their preference that their significant other be present or because their significant other wished to be present.

It can be assumed that participating in in-depth qualitative interviews about their experiences of healthcare services had an emotional impact on participants. However, this study was designed to give people with ALS the opportunity to share their viewpoint and all those who agreed to participate were keen to share their experiences. People with terminal illness often want to participate in research provided it is conducted sensitively (Kendall et al., 2007). The researcher always adhered to the study's ethics protocol and she drew on her experience as a healthcare professional in seeking at all times to be sensitive to the needs of participants.

3.7.6.1.2 Conducting the qualitative interviews

Qualitative research methods favour the 'real life' i.e. naturalistic, setting. Choice of location for the interview was at the discretion of the participant, and all participants chose their residence as the interview setting. In-depth qualitative interviews were conducted with participants in their homes. Two participants (participant #17 and participant #21) were interviewed in care settings where they resided. Each participant was interviewed on one occasion. Duration of interviews ranged from 40 minutes to 2 hours and 10 minutes; the average duration of interviews was I hour and 20 minutes. All interviews were digitally-audio recorded and transcribed by a professional transcriber within days of each interview.

Variation exists among grounded theory researchers in how they conduct qualitative interviews, but often how participants respond to the interviewer determines how and under what conditions interviews progress (Charmaz & Belgrave, 2012). All participants wished to talk about their experiences but some participants reached a more comfortable mode of expression than others. In some cases, the preferred interviewing style of the researcher as an 'active' interviewer (Holstein & Gubrium, 1995) was helpful. As an active interviewer, she lent her own voice to participants' experiences to help participants expand on their viewpoint. Overall, each interview was calibrated to how best data might be co-constructed by both the participant and researcher (Charmaz, 2006a).

Interviews in grounded theory can be unstructured where the interview is conducted without a pre-established interview guide (Corbin & Strauss, 2008) or semi-structured where each participant is asked a set of opening, central, and closing questions (typically no more than 10 questions in total) that are intended to structure the interview (Charmaz,

2006a). Unstructured interviews are suited to enquiry that embarks on a very poorly understood topic, and/or intends to extract the basic parameters of phenomena with the view to maximum openness to what might be the aspects of it that matter most to the participant. The researcher took the unstructured approach in this study because she had established (by conducting a systematic review [Foley et al., 2012a]), that little was known about how and why people with ALS engage with services and because she was open to the possibility that parameters of ALS care as agreed by service providers might be *very* different to what people with ALS value in healthcare services.

Most interviews began by inviting participants to talk about their experiences of healthcare services "since ALS came into their lives". Where necessary and fruitful, participants were 'prompted' when they struggled with phrasing a particular experience. Additional information on issues that were particularly pertinent to individual participants was pursued spontaneously (in the course of the interview) by adding questions that elicited this additional information ('probing'). Furthermore, as data analysis (that proceeded in parallel with data collection) progressed and began to yield a conceptual and theoretical framework to explain the ALS healthcare experience, some new questions were asked of subsequent research participants in order to be able to refine the concepts (that had already emerged from earlier interviews) and theory. Participant #31 interview [Martin] (Appendix D) is an illustration of the above. Martin was sampled primarily to saturate the concepts ageing, acceptance, parenthood, and family in ALS care, for meaning and variation, and to consolidate relationships already identified between them. During the course of the interview, Martin spoke spontaneously about losing control over his life and the researcher asked questions that elicited more data on loss and control in ALS. Loss and control had already been identified as primary categories from the data prior to the interview. Analysis of the data from Martin's interview (in combination with earlier data) identified the relationship between loss and control in ALS care. Overall, each interview contained a mixture of open-ended, probing, prompting and clarifying questions.

Interviews were informal and conversational but variation in participants' physical abilities inevitably impacted on how the participant and researcher interacted with each other. Prior to data collection, the researcher had worked in the ALS clinical field for 11 years and so she anticipated that she would encounter participants with high levels of physical

disability. How interviews were conducted with anarthric and severely dysarthric participants differed from how interviews were conducted with participants whose speech was unaffected or who were only mildly or moderately dysarthric. Interviewing severely dysarthric and anarthric people with ALS often meant that the researcher spoke more because she had to verify more (paraphrase what she understood the participant to be articulating, and seek confirmation / correction to her understanding). However, emphasis in these interviews was on the meaning of what participants communicated and not necessarily on the amount of data that would be captured.

Conducting qualitative interviews with anarthric and severely dysarthric participants who were keen to share their perspective required that the researcher be attuned to participants' deteriorating intelligibility and for those who did not use an AAC device, being able to decipher what participants sought to articulate. In this, she drew from her clinical experience. The researcher also anticipated that the professional transcriber would have difficulty in deciphering what severely dysarthric participants communicated in the course of the interview and so in parts where she judged participants' speech unintelligible for the transcriber, she repeated verbatim or summarised close to verbatim what participants communicated to her. This strategy inevitably shaped how these interviews were constructed but never seemed to diminish participants' desire to share and expand on their viewpoint. To ensure all interview data was captured by the transcriber, the researcher checked all transcripts against the digital-audio recording.

As stated, a minority of the sample were interviewed with their significant other present. In these cases, the interview was also co-constructed by participants' significant others. However, the main focus of the interview was to capture the service user viewpoint and questions were primarily directed to the participant as opposed to their significant other. One participant (P#33) (anarthric) offered additional data (i.e. data beyond the interview) to the researcher and gave consent that the data be used as data for analysis. This comprised an article she compiled for the Irish Motor Neurone Disease Association newsletter in which she shared her experiences of healthcare services.

From her clinical experience, the researcher was aware that she would invariably sample participants with some degree of cognitive impairment. Cognitive and/or behavioural impairment is a feature of ALS. As stated in Chapter Two, approximately 15% of the ALS

population develop fronto-temporal dementia and a mild form of executive impairment occurs for up to 35% of the population (Phukan et al., 2012). Behavioural change can also occur in the absence of cognitive impairment. This study did not seek to exclude people with ALS with cognitive impairment although conducting qualitative interviews with people with *severe* fronto-temporal dementia was not possible (within the design of this research). As she sampled, the researcher sampled participants who she knew in advance to have a mild form of fronto-temporal dementia (n=1) or a somewhat more advanced form of fronto-temporal dementia (n=1). She also sampled participants who she judged (through the course of the interview) to have mild to moderate cognitive impairment (n=2). A small number of participants (n=7) (which included the majority of participants who were known or suspected to have cognitive impairment) were mildly disinhibited during their interview (which manifested itself e.g. in the use of profanity, mild agitation, excessive preoccupation with the interview set-up) which might have been suggestive of behavioural change. However, all participants actively engaged with the researcher.

3.7.7 Member checking

All transcripts were thoroughly checked against the audio-digital recording to ensure that data was captured in full. Each participant was invited to check the accuracy of the encounter by offering them a copy of the typed-up interview and a brief summary of the interview. The purpose of providing each participant with a brief summary of their interview was to give them the opportunity to assess whether the researcher's early analysis of the interview was in their view an accurate reflection of the conversation. This was done before the researcher's interpretation was abstracted onto a conceptual level. None of the participants requested that the interview summary or their transcript be modified. Variation exists in grounded theory in terms of when and how researchers use member checking as a tool to verify participants' responses and to validate their own analysis. Some choose to return to participants and validate the accuracy of codes, categories, and developing theory (Charmaz, 2006a). In this study, the researcher did not conduct a second interview with participants largely on the grounds of rapid progression of the disease. Instead, she invited each participant to review their transcript and the researcher validated emerging concepts between interviews. Data collection ceased soon after the point at which no new findings added meaning to the data. At this point in time, 10 of the 34 participants were deceased.

3.7.8 Data analysis

As stated, the researcher gathered and analysed the data in tandem because emerging findings guided sampling. Analysis of the data involved a process of open, axial, and selective coding procedures (Strauss & Corbin, 1998). The researcher commenced coding after the first two interviews as early data served as the foundation for ongoing data collection and analysis. In open coding, the data was broken down into discrete parts and formed concepts that represented segments of raw data. These segments (otherwise known as indicators) comprised words, phrases or indeed large blocks of data that were abstracted under conceptual headings (e.g. "this segment is about the participant being trustful of his physician at the ALS clinic; I will code this as 'trusting clinic physician'"). She then began to code for similarities and differences in the data which involved constantly comparing indicators and concepts with new data which in turn led to new concepts (e.g. "several subsequent participants disclosed being trustful of healthcare professionals at the clinic – I have decided to label this as "trusting ALS clinic"'). Key to all coding was to code the data in terms of basic psycho-social process. This was done by looking closely at the data in terms of what participants described themselves as doing, feeling, thinking, and being. To this effect many lower level concepts were labelled using gerunds e.g. trusting (Charmaz, 2006a).

The researcher not only coded comparatively between blocks of data within the interview, she also coded data comparatively between significant incidents or situations both explicit and implicit in the data. In this light, the researcher always coded for *process* (Corbin & Strauss, 2008) which means how participants acted, thought, and felt about things in response to different contexts. For example, based on the above analysis, she sampled participants who had never accessed services at the ALS clinic). As stated, key to analysis in grounded theory is to look for variation in both context and experience. Identifying conditions which shaped participants' experiences and attempting to capture different and/or similar contexts that could add variation to concepts and categories (larger concepts), enabled the researcher to delineate and eventually exhaust concepts and categories (e.g. trust) in terms of their properties (i.e. characteristics), and dimensions (i.e. variation) (Corbin & Strauss, 2008).

Theory building began when the researcher began to make tentative propositions about how concepts and/or developing categories might relate to each other and about how

variation in context might shape participants' experiences. In grounded theory, this phase of analysis is known as axial coding. This required the researcher to think abductively about the data and move back and forth between concepts and different contexts in order to identify primary psycho-social processes. Here, she drew somewhat on her prior experience as a clinician in ALS and neurology care but always sought to remain open to all possible theoretical understandings. Occasionally, she found that her context i.e. *being* a healthcare professional, helped identify potential gaps in the data. As theory building became more solid, data collection became more focused on saturating core categories for meaning and variation. In the early phases of axial coding, the researcher looked for relationships between concepts and between concepts and categories. By exploring tentative relationships, some concepts became characteristics i.e. properties of categories, which served to describe categories in more detail. Looking for relationships between concepts and between concepts and categories inevitably built up categories and so in real terms, open coding flowed into axial coding. Axial coding progressed as the researcher built relationships between categories.

As she coded, the researcher wrote both reflexive and theoretical memos about the data. In her reflexive memos, compiled within days of each interview, she recorded in-depth observations, methodological insights, and comparisons between data (see Appendix E for an example of a reflexive memo). During open and axial coding phases, these reflexive memos informed and/or became part of theoretical memos in which the researcher recorded (on an ongoing basis) insights about possible connections between concepts and between concepts and categories i.e. theory building. Memoing is an important component of grounded theory analysis (Corbin & Strauss, 2008). In her theoretical memos, the researcher recorded theoretical comparisons between data which guided sampling and theory building. For example, in an early memo entitled 'making decisions in the context of family', she made comparisons between how different family contexts were impacting on participants' decisions about care and then sampled participants who had varying degrees of family support available to them. As she continued to sample participants and analyse the data, it emerged that family context also encompassed how participants themselves sought to provide support to their family and that their parenting roles at different life stages influenced how much support they sought to provide to their family.

The final coding phase, known as selective coding (Holton, 2007), involved identifying a core category which could incorporate large categories and supersede all other categories in explanatory importance. The relationship between categories constitutes substantive theory about how people with ALS engage with healthcare services. The core category did not emerge on the basis of how *many* participants or how *much* data contributed to the core category but rather by how the main categories had theoretically informed and/or were theoretically informed by the core category. Selective coding began towards the end phase of interviewing without the need to search for large amounts of additional data that had little relevance to the developing theory. During selective coding, the researcher developed and repeatedly revised an integrative diagram which helped to establish relationships between categories. The purpose of developing and refining the integrative diagram was to provide a graphic description of the substantive theory and illustrate the relationships between categories (including the core category).

Refinement of the main categories (including the core category) and the relationships between categories continued after interviewing had ceased. This was because the scope for new findings had already been exhausted and so no new data were required to support the substantive theory. Here, the researcher expanded on insights from existing theoretical memos stored on NVivo9 to compile additional theory building memos about the data. This final stage of theory building helped synthesise the relationships between categories that explained how and why participants engaged with healthcare services. For example, loss emerged as the core category in the data and the researcher identified the relationship between loss and other categories (e.g. control): participants felt they had no control over loss in their lives and exerted control in healthcare in response to loss of control in their lives. The computer software package NVivo9 (QSR International, 2010) was used as a tool to: store all memos; record coding procedures; illustrate how and what concepts formed categories; and to link concepts and categories to the memos that described and informed how the concepts and categories developed.

3.7.9 Ethics in data management

The safe storage of the data was ensured by the following procedures. All signed consent forms were stored securely at Trinity College Dublin. All data was kept securely with the researcher with password access on a secure drive in Trinity College Dublin. Only the researcher, her academic supervisor, and the clinical advisor to the researcher had access

to the data. The interviews were recorded by an MP3 digital-audio-recorder. Audio recordings were then transferred to the secure computer drive and subsequently deleted from the MP3 player. The audio recordings will be deleted from the computer drive one year after acceptance of the PhD thesis. All interviews were transcribed by a professional transcriber and were then anonymised whereby any detail in the data that could potentially make participants identifiable was removed and replaced with descriptions that reflected the significance of the original text within the context of the transcript.

A list of participants including their personal data and the link to their code name (i.e. P#1, P#2), was stored separately on the password secure computer drive. Any data or extract of quotes that are used in presenting the findings in the thesis are delineated from any characteristics that could potentially make participants identifiable. All names of participants in this thesis and related publications are pseudonames. The transcribed (and anonymised) interviews remain on the password secure computer drive in Trinity College Dublin. The researcher intends to deposit the data with the Irish Qualitative Data Archive (www.iqda.ie). The Irish Qualitative Data Archive is the central access point for qualitative social science data generated in Ireland.

3.8 Characteristics of the sample

Table 1 (pp.60-62) provides a detailed summary of the sample. As stated, sampling of participants from the population-based register resulted in sourcing participants from locations across the Republic of Ireland. Seventeen men and 17 women participated in the study. Fifteen participants lived in urban areas (urban was defined as cities and towns, including suburbia) and 19 participants lived in non-urban areas. Twenty-six participants had spinal onset ALS (limb onset), six participants had bulbar onset ALS (speech and swallow), and the remaining two participants had respiratory onset ALS. Duration of disease (i.e. since onset of symptoms) for participants ranged from four months to 169 months and the average duration of disease for the sample was 31 months. The majority of participants (n=21) were diagnosed within one year of their symptoms. Age of participants ranged from 37 to 81 years and the mean sample age was 60 years. Two participants were anarthric and six participants were severely dysarthric. A further two participants were moderately dysarthric and three other participants were mildly dysarthric.

Participants varied in degree of physical disability but the majority of participants required assistance for everyday activities. In Table 1, low level disability denotes where participants did not yet require physical assistance from another for activities of daily. Medium level disability denotes where participants required physical assistance from another for everyday activities but were not fully dependent on another. Participants who had high levels of disability were those who where fully dependent on others to engage with activities of daily living. At the time of the interview, all but five participants were using one or more assistive devices. However, participants' use of assistive devices was not always an indicator of the degree of disability among the sample as participants who had predominantly bulbar involvement (i.e. dysarthria and/or dysphagia) were also restricted in terms of limited physical endurance. (It important to note that people with spinal onset ALS may also develop bulbar dysfunction as their condition worsens and people with bulbar onset ALS invariably develop weakness in their limbs).

As depicted in Table 1, eight participants had already engaged with non-invasive ventilation and/or gastrostomy feeding. Five participants were using non-invasive positive pressure ventilation (NIPPV) only, and two participants had engaged with both NIPPV and gastrostomy feeding. One participant had engaged with gastrostomy feeding only. At the time of the interview, eight of the 34 participants had not attended the ALS clinic at Beaumont Hospital, Dublin. For those who had accessed tertiary care services at the ALS clinic, there was variation in the extent to which participants had engaged with services at the clinic. The majority attended the clinic periodically but a small number of participants (n=4) had either stopped attending the clinic or only attended sporadically. At the time of interview, some participants (n=3) were new attendees at the clinic.

The majority of participants were married and lived with their spouse who for most participants was also their primary carer. All but one of the participants who were married had dependants. Participants who had not married had no dependants and participants who had not married and/or who were not in a relationship either lived alone, in a care facility, or with a sibling. Four participants were widows: two of them lived alone, one lived in nursing home, and one lived with family. All participants whose spouse was deceased had dependants. All participants under 50 years old with dependants lived with their dependants, and some older participants with dependants also lived with their dependants. Nearly one third of the sample (n=10) had already retired from paid or self

employment prior to the onset of ALS. The majority of participants (n=19) were in paid or self employment at the time of onset of the condition but only three of these participants were still working at the time of the interview. In Table 1, the term 'retired' is used to denote those participants who had retired prior to the onset of ALS. The term 'early retirement' denotes those participants who retired because they were no longer able to continue in their work role because of the disabling effects of ALS. All participants had accessed tertiary and/or secondary care services and all but two participants had engaged, to some degree, with primary care services.

Table 1 **Sample characteristics**

		Age	ALS type	Status	Region	Interview Date	Diagnosed	Duration of Disease	Severity of Disability
P#1	Mona	50	Spinal onset	Married Living with spouse and son Home-maker	North East (non-urban)	12/09/2011	October 2010	24 months	Medium level disability (NIPPV)
P#2	Ita	68	Bulbar onset	Widow Living alone Retired Pensioner	Dublin (urban)	13/09/2011	July 2011	9 months	Medium level disability Mildly dysarthric
P#3	Callum	67	Bulbar onset	Married Living with spouse Retired Pensioner	West (non-urban)	19/09/2011	January 2009	44 months	High level disability Severely dysarthric
P#4	Peter	69	Spinal onset	Married Living with spouse Healthcare professional Lecturer	South West (non-urban)	27/09/2011	December 2010	11 months	Low level disability
*P#5	Vincent	62	Spinal onset	Married Living with spouse Labourer Early retirement	Mid West (urban)	06/10/2011	December 2010	24 months	High level disability
*P#6	Andrew	64	Respiratory onset	Married Living with spouse Fisherman Early retirement	South East (non-urban)	10/10/2011	August 2011	19 months	Medium level disability (NIPPV)
P#7	Eve	45	Spinal onset	Married Living with spouse and children Software engineer Early retirement	South (non-urban)	19/10/2011	June 2010	24 months	Medium level disability
P#8	Richard	52	Spinal onset	Single Living alone (no dependants) Farmer Early retirement	Midlands (non-urban)	27/10/2011	April 2010	25 months	High level disability
P#9	Danielle	45	Spinal onset	Single Living with sibling and sibling's family (no dependants) Software engineer	Mid West (non-urban)	08/11/2011	August 2009	45 months	High level disability
*P#10	Morris	67	Bulbar onset	Married Living with spouse Retired Pensioner	South East (urban)	15/11/2011	September 2011	11 months	Medium level disability Severely dysarthric (PEG)

Table 1 continued

P#11	Terry	45	Spinal onset	Married Living with spouse and children Civil servant Early retirement	Dublin (urban)	13/12/2012	June 2010	29 months	Medium level disability
P#12	Helen	56	Spinal onset	Widow Living alone Public servant Early retirement	South East (urban)	04/01/2012	December 2009	46 months	Medium level disability
P#13	Paul	80	Spinal onset	Married Living with spouse and daughter Retired Pensioner	Midlands (urban)	09/01/2012	December 2011	4 months	Medium level disability
*P#14	Edward	81	Spinal onset	Married Living with spouse Retired Pensioner	South West (urban)	18/01/2012	July 2011	13 months	High level disability Severely dysarthric (NIPPV) (PEG)
*P#15	Philip	74	Bulbar onset	Married Living with spouse Famer Early retirement	West (non-urban)	30/01/2012	July 2011	12 months	Medium level disability Severely dysarthric
P#16	Tim	47	Spinal onset	Married Living with spouse and children Community worker Early retirement	Dublin (urban)	13/02/2012	May 2011	19 months	High level disability
*P#17	Gregory	81	Spinal onset	Widower Nursing home Retired Pensioner	North East (non-urban)	20/02/2012	October 2010	81 months	High level disability Moderately dysarthric
P#18	Mary	57	Spinal onset	Single Living alone (no dependents) Civil servant Early retirement	Dublin (urban)	29/02/2012	November 2010	21 months	Medium level disability
P#19	Eilish	51	Spinal onset	Married Living with spouse and children Home-maker	South (non-urban)	12/03/2012	May 2011	25 months	High level disability
P#20	Sally	62	Spinal onset	Married Living with spouse Home-maker	North West (non-urban)	21/03/2012	November 2011	11 months	Medium level disability (NIPPV)
P#21	Aine	39	Spinal onset	Single Hospice (no dependants) Student (prior to ALS)	North East (urban)	30/03/2012	May 1999	169 months	High level disability Anarthric
P#22	Samantha	58	Spinal onset	Married Living with spouse Healthcare professional Early retirement	South East (non-urban)	10/04/2012	January 2011	22 months	High level disability Mildly dysarthric
P#23	Pascal	41	Spinal onset	Married Living with spouse (no dependants) Commercial banker	North East (non-urban)	16/04/2012	April 2011	18 months	Low level disability
P#24	Ann	67	Spinal onset	Married Living with spouse Home-maker Previously healthcare professional	Dublin (urban)	25/04/2012	June 2011	57 months	High level disability Severely dysarthric (NIPPV) (RIG)
*P#25	Jennifer	73	Spinal onset	Single Living alone (no dependants) Retired Pensioner	South (non-urban)	04/05/2012	December 2011	16 months	Medium level disability

Table 1 continued

P#26	Jack	61	Respiratory onset	Married Living with spouse and children Industrial worker Early retirement	North East (urban)	10/05/2012	September 2011	15 months	Medium level disability (NIPPV)
*P#27	Melanie	77	Bulbar onset	Widow Living with son and daughter-in-law Retired Pensioner	South (non-urban)	29/05/2012	August 2011	17 months	High level disability Moderately dysarthric
P#28	Cathal	37	Spinal onset	Single Living with sibling and sibling's family (no dependants) Labourer Early retirement	West (non-urban)	04/06/2012	June 2011	27 months	High level disability (NIPPV)
P#29	Melisa	60	Spinal onset	Married Living with spouse Care assistant Early retirement	North East (non-urban)	26/06/2012	April 2011	20 months	Medium level disability
P#30	David	51	Spinal onset	Married Living with spouse and children Engineer Early retirement	Mid West (non-urban)	03/07/2012	April 2012	39 months	Medium level disability
P#31	Martin	73	Spinal onset	Married Living with spouse Retired Pensioner	North East (urban)	13/07/2012	November 2011	18 months	Medium level disability
P#32	Cara	64	Spinal onset	Married Living with spouse Home-maker	South East (urban)	20/07/2012	December 2011	37 months	Medium level disability Mildly dysarthric
P#33	Denise	55	Spinal onset	Not married Living with partner (no dependants) Literature & communications Early retirement	South (non-urban)	01/08/2012	May 2011	91 months	High level disability Anarthric
P#34	Maureen	62	Bulbar onset	Married Living with spouse and grandson Retired	Dublin (urban)	09/08/2012	June 2011	17 months	Medium level disability Severely dysarthric

* denotes non-ALS clinic service users
NIPPV (non-invasive positive pressure ventilation)
PEG (percutaneous endoscopic gastrostomy)
RIG (radiologically inserted gastrostomy)

3.9 The researcher and the research design: A critical review

The researcher was not an experienced researcher prior to conducting the research. She had undertaken qualitative research in ALS care in the past at Masters' level, but her experience was primarily in the clinical field. Undertaking a PhD as a research fellow was in itself a learning process.

There is debate among qualitative researchers as to the extent coded data should, if at all, be checked by other researchers. Some grounded theorists (Charmaz, 2006a) reject the need for agreement among coders because how and when data is analysed and by whom inevitably shapes the analysis. Data collection and data analysis was monitored closely by

the researcher's supervisor and her supervisor interrogated the analysis. However, the data was gathered by the researcher and no data was coded by another. It can be assumed that a larger qualitative study on the topic undertaken by a team of researchers would have unearthed similar *and* different findings which would have, in turn, guided sampling somewhat differently. Nonetheless, careful documentation of procedures in this study enables other researchers to replicate the method. Although replication of the method would not necessarily yield the exact same analysis, there should be commonalities in the main contours of experiences identified by all researchers in the same or very similar contexts.

As stated, the researcher did not undertake a second visit with participants largely based on the grounds of rapid progression of the disease. Longitudinal studies in ALS are conducted but to allow for the rapid attrition through death, they have to date been quantitative (large-N) in design and recruited participants at diagnosis or in the early stages of the disease. The aim of this study was to capture as much diversity as possible and theoretical sampling invariably guided the researcher toward service users at different stages of the disease. Grounded theory studies seek to capture *process* (over time). Capturing diversity in experience in itself captured process over time because participants spoke about their healthcare services since symptom onset. A single interview encounter with participants in a sample that comprised variation in context and experience also captured (albeit to a limited degree, subject to recall bias) variation in how participants had responded over time.

Sampling participants through sources other than the ALS population-based register could have resulted in a very different sample. It is possible that without a population-based register to sample from, it would have been more difficult to locate service users in advanced stages of the disease and/or who were engaging with end-of-life care. Sometimes uncertainty surrounding people's prognosis or uncertainty surrounding their awareness of the terminal nature of their condition renders it difficult to recruit research participants who have a limited life-expectancy. In this study, the researcher did not encounter these problems because for the majority of participants, she was aware of their prognosis and all participants were aware that ALS is terminal.

Why and how this research was conducted illustrates the complex relationship between agency and structure. The researcher sought to capture the viewpoint of people with ALS so that people with ALS can inform how healthcare services might best be delivered to them. However, multiple structural contexts to ALS healthcare services in the Republic of Ireland influenced how participants were sampled and the conditions under which participants agreed to participate. The Irish ALS research group and the national ALS clinic were, in themselves, gatekeepers. Structural factors closer to people with ALS also impacted on how the research was conducted. Those who had not yet encountered high levels of disability and/or significant deterioration in their speech were more likely to be the first and only point of contact for the study but for many people with ALS with high level disability and/or severe dysarthria, their significant other was a gatekeeper. The researcher was often dependent on the significant other to relay information about the study to the service user and in some cases carers made the decision about participation on behalf of people with ALS. It is possible that as people with ALS engage with healthcare services (with increasing disability) and become dependent on others to care for them, complex structural factors restrain (some forms of) agency. In this light, focusing on the service user perspective alone to the fullest possible extent was necessary in order to identify what is most important to them.

3.10 Conclusion

This study set out to capture the ALS service user perspective on healthcare services. As a critical realist, the researcher sought to understand the complexity of healthcare experiences among people with ALS and placed people with ALS at the centre of the enquiry as the most reliable informants about *their* experiences of healthcare services. Capturing variation in participants' experiences in the 'real world' enabled the researcher to show (as will be described in the chapters that follow) that people with ALS construct healthcare experiences i.e. give meaning to their healthcare services, in the context of their lives.

This study is the first to map out the key parameters of healthcare experiences among people with ALS from their perspective. It is also the first study to identify key psycho-social processes which underpin how people with ALS engage with healthcare services. Knowing *how* and *why* people with ALS in the Republic of Ireland engage with their

healthcare services is important because how people with ALS engage healthcare services, in turn, shapes the reality of services for people with ALS in the Irish healthcare system.

The findings as described in the chapters that follow are not representative of people with ALS. However, the findings are substantive because they reflect a broad range of experiences among a diverse group of people with ALS who were sampled from a population-based register. At a substantive level, the findings pinpoint central areas of further scientific inquiry and identify key variables that other studies (both within and outside ALS) might usefully collect data on. A scale has been developed from the data (Appendix F) and is being used in a study aimed at mapping out optimal care for people with ALS.

Chapter Four

Loss, control, and awareness in ALS: The personal experience of people with ALS

"I read a very interesting book some years ago by Stephanie Meyer and she was talking about, it was an alien species came down to earth (....) and it took over your brain, took over your body. Well, I often think that MND is like that. I feel like I've been taken over by a sort of an alien presence and it's, it's in my brain and I think it's like an alien that has nuclear weapons, that it can do all sorts of things to me and I'm combating it with bows and arrows (....) So I find myself in a constant battle, day after day because this, this MND alien that has taken over my body and that I have not too much defence against. Normally in, in a war you win some battles, I'm losing all the battles and I know for a fact I'm going to lose the war. The war, the war is lost already because MND is going to take me anyhow, but I'm not winning any battles because I feel that I don't have, the arms that I'm provided with in no way, can in any way defeat or even hold back this alien that is taking me over." (Martin, participant #31).

4.1 Introduction: Loss of control and adapting to insurmountable loss

In this study, all participants spoke openly about the uncontrollable nature of ALS. Indeed, all participants felt unable to control the losses they had encountered since the onset of ALS. For the most part, participants resisted loss by attempting to exert control in their lives and over ALS. However, the experience of unremitting loss fuelled awareness among participants that it was impossible for them to control ALS or indeed arrest the loss they encountered in ALS. Participants' personal accounts of their experiences suggest that there is a time dimension to how people with ALS lose control and "suffer" loss: People with ALS lose control and bear more loss as they advance in the disease. Nonetheless (as described in the sections of this chapter), the majority of participants managed to somehow "get through" their journey with ALS as they adapted to the uncontrollable nature of ALS and to insurmountable loss. This chapter describes, at a substantive level, the meaning of loss for people with ALS and how people with ALS adapt to loss. Understanding how people with ALS respond to (and in most cases) adjust to insurmountable loss is important because (as described in Chapter Five), participants' lived experience of ALS shaped how they thought about and interacted with the external reality of care in ALS.

4.2 The meaning of loss from the ALS service user perspective

4.2.1 Feeling "shocked" and living with a "death sentence"

Receiving a diagnosis of ALS was a significant event in the lives of all participants. Most participants suspected something amiss prior to their diagnosis in the light of their symptoms but the majority of participants did not suspect ALS before their diagnosis. For the most part, participants were overwhelmed by their diagnosis and their reaction to the diagnosis was one of shock and dismay:

> *"I hadn't suspected, I didn't know it was fatal (....) so that was like somebody literally just punched me in the stomach and just, I could hardly breathe, it was like you know this physical reaction I had when I heard it (....) I was numb, I was stunned."* (Danielle, participant #9)

> *"When I came out of there [ALS clinic] it was like someone had you know kicked the chair from underneath me and I just fell flat on my back and was just completely deflated."* (Pascal, participant #23)

> *"Shock, when they told me that I had motor neurone, I said oh good god, I couldn't believe I had it. Because I thought it was [just] something that was wrong with my hands."* (Mary, participant #18)

> *"I'm still finding it hard to take in that I actually [have ALS] because I felt I was doing everything right in exercising all my life. Doing things right, never smoking, never even trying it, taking drugs or anything like a drug (....) then of course this [ALS] came like a bolt from the blue."* (Jack, participant #26)

Some participants suggested that a diagnosis of ALS is "shocking" even when suspecting the diagnosis and participants questioned whether one can ever be prepared for such a diagnosis. For some, the "shock" subsided with time, for others the "shock" still remained:

> *"Yes I'm still so shocked because I mean it had never crossed my mind."* (Melanie, participant #27, end-stage ALS)

Regardless of suspecting or not suspecting ALS, all participants had hoped that the diagnosis would not be confirmed because nearly all participants had understood the diagnosis of ALS to be a "death sentence". Nonetheless, no participant questioned the diagnosis. All participants were aware of the terminal nature of ALS at the time of the interview and all were resigned to the irreversible nature of ALS:

> *"It's like someone getting a death sentence. You know there's no jury to come back on this. There's no cure (....) It's like a contract over your head. Someone told you that you are going to be whacked. You don't know when, but there's a contract on your head."* (Tim, participant #16)

4.2.2 Loss and feeling "absorbed" into a world of ALS

Living with a "death sentence" provoked feelings of profound loss for participants that comprised both physical and psychological dimensions. Not only was physical loss devastating for participants but their deteriorating physical ability also engendered loss for them in other domains. The meaning of loss for participants consisted of loss of normality, loss of participation, loss of independence, loss of identity, loss of the future, loss of parenthood, loss of hope, loss of expectations and loss of control. Participants' perceptions of loss were formed by their awareness of physical change *and* by their awareness of how their ability to engage in important aspects of life (e.g., family, parenthood, leisure) was disintegrating and/or had already disintegrated:

> *"I can't hug my daughter, enormous loss, it's incredible."* (Tim, participant #16)

> *"I missed being able to tell a story, or a joke, have a proper argument or just being able to explain myself."* (Denise, participant #33)

> *"I'm not able to cook, light the oven or I couldn't do any, I can't do any of those things (....) and I can't, I can't drive. Oh I miss [driving] the car, I really miss it, I really do."* (Mary, participant #18)

Central to participants' experience of loss was emotional loss. Participants indicated that people with ALS "suffer emotional loss" which includes the loss of participation and most importantly, loss of what they "love" to do:

"You know I can look at a magazine because I used to like knitting and I looked at a knitting pattern and then say oh shite!" (Jennifer, participant #25)

"Well I've opted out of a lot of things that I loved to do. I'm a writer. I'm no longer writing (....) that's a big thing, I would say it's THE big thing (....) very important, so important but I've lost them." (Melanie, participant #27, end-stage ALS)

"You lose everything, absolutely everything (....) I used to love in the evening to sit down and have a glass of wine, bit of cheese, no real desire now. No, no feeling now." (Martin, participant #31)

The majority of participants were deeply distressed by the devastating impact ALS had on their lives and all of them suggested that their lives had been utterly transformed since the onset of ALS. Gregory (participant #17) and Ann (participant #24, end-stage ALS) disclosed:

"I felt terrible [when diagnosed] because I knew I'd never be the same again (....) [now] I'm just lying here, I've no future." (Gregory)

"You see MND is the worst, MND is the worst neurological condition one could get (....) This for me has taken away so much. It's going in the direction of taking all my faculties, my independence, my speech, my mobility, concentration, eating (....) The whole thing is a nightmare, well the whole, the whole concept [of MND] is a nightmare (....) Everything I was doing is now gone (....) I just don't want to be absorbed into it. I just don't want to talk about it apart from this. I just don't want to fall into that hole." (Ann)

A number of participants struggled in their attempt to resist ALS. Participants revealed a strong desire to live their life as they had lived their life prior to the onset of ALS and participants were keen not to be defined by ALS. Of note, were participants' personal accounts of their relationship with assistive devices and in particular for those who could no longer ambulate, their perceptions of self as a wheelchair user. Striking was how Danielle (participant #9) described her relationship with her wheelchair. She believed her powered wheelchair had impacted positively on her quality of life but she tried to forget

that she was a wheelchair user. She stored her wheelchair outside her bedroom so that it would not be the first thing she woke up to i.e. a reminder of being "absorbed" into a world of ALS:

> *"You know it's not a part of you, so you don't want to get into it (....) When I'm in bed at night like we take the wheelchair out of the room because I don't want to be looking at it (....) I don't want it to be the first thing I see in the morning. You have to trick yourself into it because as soon as you see it it's like oh here we go again. You know, you are straight back in it."*

However, all participants perceived themselves to be powerless in the face of insurmountable loss. The majority of participants were overwhelmed by the aggressive nature of ALS and felt "robbed" of the present and of the future. Richard (participant #8) disclosed:

> *"It [ALS] turned me into an old man overnight because suddenly you're going with a walking stick and then you're with a rollator, now you're in this wheelchair and you've really, it's just a robbery of everything (....) I was interested in having my own tools and doing me bit of work on the farm and now there's not a single one of them any use to me because you can't take out your lawnmower and cut the lawn. You can't take out your chainsaw and cut a few branches off a tree or you can't get up on a tractor, you can't drive a car, you haven't got the freedom to go to a pub or any place because it's, the awkwardness of it outweighs the whole lot (....) I dread the future."*

Losing hope was also central to participants' experience of loss. Hope for many participants encapsulated their quest for a cure or at least treatments to slow down the progression of the disease. However, participants' rapidly deteriorating physical abilities reinforced for them the likelihood of further disease progression:

> *"This thing has been a downhill from the very first, it's been downhill all the time and it's accelerated now."* (Richard, participant #8)

Hope diminished for participants as their losses "accelerated". Most participants indicated that there is less hope in ALS compared to other disabling and/or life-limiting conditions

and some participants even suggested that there is no hope in ALS in the context of the incurable nature of the condition:

> *"Nobody with this illness does really well (....) It's a load of bunkum (....) There are no heroes in motor neurone. If there are, they are lying to you (....) You see Geraldine, motor neurone is without hope. You can't treat it and you can't get well (....) So cancer you can treat and get well (....) With motor neurone there is no way out of your situation, can't be improved. It's a funny disease in that way that it's without hope and that's the worst feature of it."* (Morris, participant #10)

Comparing self to other people with ALS helped some participants adapt to the progression of the disease but seeing other people with ALS also eliminated hope of overcoming ALS. Participants' growing awareness of the limitations imposed on them diminished hope of a future:

> *"I mean you're sitting there [in ALS clinic] as close as I am now to you with another [more disabled] patient across from you (....) that was the reality of the situation in that I was facing myself."* (Richard, participant #8)

> *"People say, ah sure we're all going to die. But I know I am going to die you know."* (Cara, participant #32)

A minority of participants (n=3) indicated that they were hopeful either for the sake of their children (Terry, participant #11) or because they hoped they would survive long enough for a cure for ALS (Tim [participant #16] and Áine [participant #21]). However, for the most part, participants' hope diminished in line with mounting loss. Remaining hopeful was beyond most participants because their losses never ceased. Instead, their losses accelerated with disease progression. The clinical course of ALS offered no periods of remission or stability to participants and all participants in this study suspected they would encounter more loss as they advanced in the disease. Participants shared the perception that ALS was an insurmountable disease:

> *"It's very difficult in this whole fucking disease, it seems to be one step forwards and two steps backwards, the whole time (....) You always feel you are losing a little bit*

which is mentally very, very difficult. So I said fuck it, I'm not going to put my mind through that any more." (David, participant #30)

4.2.3 Failing to control the uncontrollable

Participants' growing realisation that they were unable to control ALS or indeed the losses they encountered because of ALS compounded their feelings of loss. Losing control (and/or fighting to remain in control) was central to how participants acted and interacted across different contexts. For participants, losing control constituted "having no choice". Indeed, the majority of interviews centred on how participants felt they were *forced* to adapt (to ALS and related healthcare services), *forced* to accept (ALS and related healthcare services) and *forced* to plan for the future in order to cope with mounting loss. Participants' deteriorating physical abilities forced many participants to adapt to their ever-increasing physical limitations:

"I mean I know I can't walk so I have to use a wheelchair. I know I can't stand up so I have to be lifted up and I have to be hoisted into bed and hoisted onto the toilet. You see so like I mean you have to accept it if you like (....) You don't have a choice. I mean if you are sitting there and you can't get up, what choice do you have only you have to be lifted up off it?" (Eilish, participant #19)

Regardless of whether participants felt they accepted ALS or had not accepted ALS, all accepted the need to engage with healthcare services. However, participants' perceptions surrounding "having no choice" were not necessarily about the decisions they made about their care. Instead, "having no choice" reinforced for participants a reality in which they had no control over the effect of ALS on their daily lives:

"I still try and do whatever I can myself, but I'm, I'm beginning to realise that you know, I'm not going to be able to control all these things and I really don't like the fact that I will depend so totally on people to do things for me." (Martin, participant #31)

"I loved gardening, spent my whole time gardening. I loved cooking, entertaining, playing golf. I was always active and I feel I want to be active but I can't and I find that depressing (....) not to be able to do anything for myself. I'm a very private person and to think that people would have to do everything for me." (Cara, participant #32)

Beneath participants' growing awareness of the uncontrollable nature of ALS was, for some, a strong resentment towards ALS and towards the need to use assistive devices in order to live through insurmountable loss. Participants recognised the benefits of using assistive devices but also spoke of their contempt towards assistive devices because they had no choice but to use them. Melanie (participant #27) described her reaction when she received a bed lever that would assist her to move in bed without assistance from another:

> *"I mean looking at that the first day I felt like throwing it out the window."*

Indeed, many participants continued to struggle with the concept of having to use assistive devices because assistive devices reminded them of how much they had already lost:

> *"The thought of a wheelchair, you know, and it's just that you're getting it into your head that you're losing your independence altogether (....) I just hate it and I still hate the wheelchair."* (Sally, participant #20)

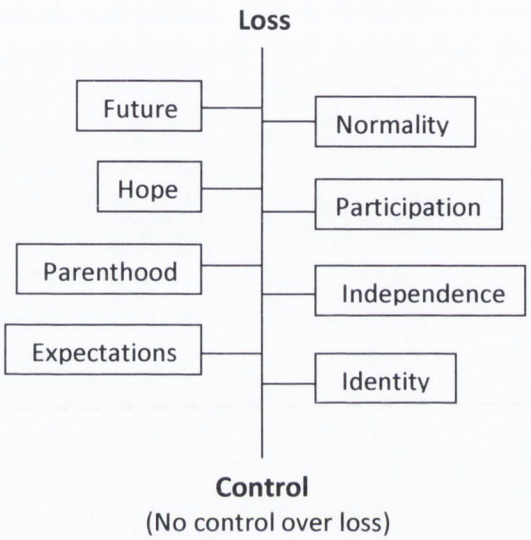
Losing independence accentuated participants' fears about the prospect of losing more control over their lives. Disease progression forced participants to accept assistance for their daily needs but some participants who had not yet needed full assistance hoped to die before they would become fully dependent on others. Melisa (participant #29) and Martin (participant #31) explained:

> *"I'm hoping that I don't live to be too, that I'm going to have to depend on someone you know what I mean at the back of my mind. You know the way you can get very invalided and you could be in say a nursing home and you could, now lets be honest you could be at the mercy of people isn't it."* (Melisa)

> *"The thought of like sitting in a wheelchair paralysed and depending on other people to do for me, that to me that's, that's a total nightmare, that is my worst nightmare. I would hope before I got to that condition that I would, I would die."* (Martin)

Overall, participants' experience of unremitting loss alerted them to the uncontrollable nature of ALS and most importantly, to the fact that they were unable to control the impact of ALS on their lives. Figure 3 below summarises how participants interpreted loss including loss of control in their lives.

Figure 3 The meaning of loss for people with ALS



4.2.4 Time frame and feeling uncertain

Participants' experience of disease progression prompted them to think about the future. A small minority of participants (n=4) anticipated death within a specific time frame but most participants were somewhat uncertain about this. Participants' uncertainty about time frame subsequently evoked more feelings of loss for them because uncertainty surrounding prognosis reinforced that they were unable to exert control in their lives:

> *"I wish somebody could turn around and say right, 2014 probably in around June or something like that, you'll be gone, that's when it'll be all over I can take that, you know what I mean, I wish someone could say that to me (laughing)."* (Terry, participant #11)

> *"You never know exactly what is coming (....) you don't know what ability you will have at any time so it's hard to plan."* (Denise, participant #33)

Uncertainty about ALS in turn fostered feelings of anxiety among participants. Feeling uncertain ranged from being uncertain about how people die from ALS to uncertainty about what was yet to come with ALS. Participants were dismayed by what was not yet known about ALS:

> *"I don't know what happens when people die, I don't know what progression, how far it goes or what it stops, does it stop the heart or does it stop the lungs or will it stop vital parts of the body (....) I don't know what finishes people in the end."* (Paul, participant #13)

> *"This sounds kind of harsh, sometimes I think they [researchers / healthcare professionals] just don't have a clue about it (....) They don't know really how to start to fix it because they just can't figure it out. Like I know at one stage they were thinking that maybe the dementia side of it was maybe a separate illness, stuff like that and this is all relatively new like, and then you think what the fuck."* (Terry, participant #11)

Some participants seemed realistic about time frame (in the context of what the researcher knew of and/or expected of their disease progression) but others appeared somewhat optimistic about their time frame. It is possible that some participants held a somewhat more positive outlook on time frame in order to cope with ALS. However, despite participants' awareness of their progression, the vast majority were shocked that they had acquired such high levels of disability and had not suspected ALS to be as aggressive as they experienced it to be. Sally (participant #20) shared:

> *"It's something unreal really, you know, it's kind of hitting every bit of you, you know what I mean, every bit of your body is kind of hit with it, whereas if you had something else, it might be only a part of your body, but like from your hands and your neck and your legs and your swallow."*

It is possible that despite participants' awareness of the progression of their disease, some were not attuned to the pace of their respiratory decline and/or to how prognosis in ALS is affected by how and when people with ALS encounter deterioration in their respiratory function. It is also possible that participants who believed they had a slower progression of the disease (compared to other people with ALS) suspected they would live for some time

before they encountered respiratory problems. In addition, the majority of participants who searched for more certainty surrounding their prognosis failed to secure assurance of a more certain time frame from service providers:

> *"I have found from the very start with MND that it's kind of smoke and mirrors, no one is prepared to say to you, like for instance where are you in the disease, they all, everyone says, well everybody's different (....) I said well how do I gauge where I am if everybody's different, well you gauge yourself against yourself. Well how do you gauge against yourself, well how are you, how were you this time last month. Well, yeah but you know that still doesn't tell me at this moment in time that's where I am."* (Martin, participant #31)

4.2.5 Becoming more aware of loss

Central to participants' experiences of living with ALS was their growing awareness of how much they were losing out on living their lives on their own terms. Participants felt restricted by ALS because mounting physical and psychological loss rendered them less able to participate in activities that they had previously enjoyed:

> *"I wouldn't like not being able to walk (....)" [participant #15 writing] G: "I'm going to read it out again for the transcriber @@Philip##. You also 'like to go about work around the house and farm a bit'. So what's important to you is that you continue to do what you were doing before?" P#15: "Ah well, I wouldn't have as much energy (....) I try a bit (....) [but] it [ALS] has progressed."* (Philip, participant #15)

> *"Well I think I try to do and go as many places as I can (....) with my friends (....) but maybe that's coming to an end (....) I can't, I can't stay up longer than an hour. I am so tired."* (Ann, participant #24, end-stage ALS)

As described in Chapter Three, all participants in this study were aware that ALS is a progressive and ultimately terminal condition. On receiving a diagnosis of ALS, participants moved from suspecting to open awareness or from unsuspecting to open awareness. Most participants spoke openly about the terminal nature of ALS and for the majority of participants, the reality of living with ALS never left their consciousness:

"I'm aware that it is getting worse (....) because I can't walk as well as I could (....) general weakness (....) I'm aware, not suspecting." (Melanie, participant #27, end-stage ALS)

"There's not a single place you go that this thing never leaves the back of your mind." (Richard, participant #8)

"It's always there (....) you'd never forget, you know like you'd never, ever forget." (Terry, participant #11)

"There's no getting away from, from the illness because as soon as you wake up in the morning it's there you know. It's with you all day because you're going to do something and you can't." (Helen, participant #12)

As described, the majority of participants were unsure about time frame but all were very aware of the loss they had encountered thus far. Participants' awareness of how their bodies physically changed stands out within the data. The pace of disease progression raised participants' awareness of what they could no longer do and all participants were very much attuned to the mounting restrictions ALS imposed on them. As participants advanced in the disease, the pace of disease progression also reminded them that they were moving hastily towards death:

"Up until the end of the summer, I could still get into my car by myself, drive to wherever I wanted to drive, I could even push the trolley around the supermarket and do light shopping, whereas I couldn't possibly think of that now." (Eve, participant #7)

"If I first had the symptoms in January 2011 but like once I was diagnosed from when I thought I had it to the diagnosis, I was pretty right in where I thought I'd be in my head. So that we arrived at May and I'm having speech problems and that was what I calculated. I mean I didn't sit down with paper and calculate it (laugh) (....) It's something people don't like hearing but I think I'll go [die] between October and December." (Jennifer, participant #25)

Comparing self to other people with ALS and comparing ALS to other disabling and/or life-limiting conditions also alerted participants to their loss. Participants' accounts suggest a genuine interest in other people with ALS and a curiosity as to how other people with ALS cope with advancing illness. However, seeing other people with ALS was distressing for many participants because it reminded them of the progressive nature of ALS. Participants spoke frequently about how "tragic" it was to see other people with ALS who had already advanced along the disease trajectory:

> *"Looking at the patients, when I was diagnosed first you know the way there would be people sitting there [in ALS clinic]. And maybe they would be in different kind of, different stages. And I used to say god is that the way now I'm going to end up."* (Melisa, participant #29]

Comparing ALS to other conditions was also a red flag to "realising" the terminal nature of ALS. Participants frequently compared ALS to cancer and to other disabling neurological conditions including Parkinson's disease (PD) and Multiple Sclerosis (MS). Some felt despondent about the absence of effective neuro-protective treatments for people with ALS when compared to those available for people with MS:

> *"I know a woman that has MS and she said she got it about ten years ago, ten or eleven years ago. I think there's an injection she's to give herself three times in a week I think and it's made an awful difference to her."* (Eilish, participant #19)

Some participants suggested that whilst people with cancer might "suffer" more pain than people with ALS, living with cancer might also offer people with cancer the opportunity to fight a somewhat more uncertain prognosis and for those who live with terminal cancer, the possibility of being more physically independent in the latter stages of their illness:

> *"Now I know there's levels of cancer where you've no chance but I do, the cancer I think you might be able to fight physically, like right I'm going to try the chemo and this might work for me or whatever, I'm going to stay positive and all this, but with the motor neurone the one thing I've learned is that you can't really fight the physical, like I can't make this arm work, there's nothing I can do to make this arm work again."* (Terry, participant #11)

"Well, you know, I'd probably die from cancer a lot quicker but at least I'd be up walking, I'd be out fishing, I'd be doing me few bits and pieces." (Cathal, participant #28)

Overall, unremitting physical and psycho-social losses encountered by participants in their lives, and their attention to other people's (with ALS) losses, sensitised participants to the progressive and terminal nature of the disease.

4.3 Adapting to insurmountable loss

As described, losses encountered in ALS were, from the participants' perspective, insurmountable. Participants spoke about ongoing loss and about never regaining what they had already lost. For the most part, participants developed strategies in order to adapt to insurmountable loss, including exerting control over and engaging with healthcare services. The following sections of this chapter describe how participants adapted to insurmountable loss. How participants exerted control over and engaged with healthcare services to adapt to loss (i.e. the meso-level) is described in Chapter Five.

4.3.1 Striving to maintain normality but losing out on normality

Participants' desire or indeed their struggle to maintain normality was evident throughout all interviews. Many attempted to resist change in order to cope with unremitting loss. Participants' personal accounts of their experiences of living with ALS and of engaging with healthcare services suggest that there is a time dimension to "normality" in ALS. Receiving a diagnosis of ALS was a ticket to an "abnormal" world and a reminder to participants that they were no longer living in a "normal" world. In response, participants strove to maintain normality and the strategies they employed in their struggle for normality became for them valuable coping strategies in the face of mounting loss.

Participants' desire to live in the present was a key strategy which enabled them to resist (if not overcome) the life-altering effects of ALS. Participants were almost always aware of ALS in their lives but many participants tried to "live for the moment" instead of confronting ALS all the time:

"I get up in the morning and think for today and that's it, you know. It's like Garth Brook's song, 'If Tomorrow Never Comes', tomorrow never does come because when I go to bed tonight and get up in the morning, it's another day." (Cathal, participant #28)

"It's like giving people more possibilities, it just makes life more rewarding because you are not focused on the end, you are focused on living the day and that's really important for your quality of life." (Danielle, participant #9)

Participants attempted to maintain the status quo by striving to maintain some sense of normality in their lives. Their inclination to carry on as before (i.e. "getting on with it"), and their reluctance to look towards and/or plan for the future permeates many interviews. Some participants preferred not to think too far into the future, and purposely avoided timelines to deal with the condition on a day-to-day basis. Jack (participant #26) and Philip (participant #15) commented:

"I mean getting to bridge ten before you cross bridge one. You don't want to be really thinking about things like that, even though you know you have to do them. But you try and put it to the back of your mind all the time." (Jack)

"I'm dealing with things as they come (....)" [participant #15 writing] G: "I'm just going to read this out again for the transcriber, you 'try not to think too much about it. I try to go on with life'. Are you dealing with the present then?" P#15: "Yeah, yeah for the most part." (Philip)

Downplaying their own physical limitations and paying attention to what they had not yet lost (e.g., remaining abilities, family) energised some participants in their struggle to maintain normality. Participants' attention to their remaining abilities was an important coping strategy because it cushioned the otherwise devastating impact the experience of loss had on them:

"I'm speaking fine, if I didn't have to get on to my feet and move around you would never think that there was a thing wrong with me." (Eve, participant #7)

"You see my hand was getting very thin, this one is you see I'm lucky enough I am a ciotóg [left-handed person], left handed and I do everything with my left hand." (Melisa, participant #29)

Nevertheless, despite participants' fight to maintain or regain normality, no participant suggested that he or she managed to maintain or regain normality. Rather, participants indicated in the course of interviews that their lives were, for the most part, defined by "unbelievable" loss. Morris (participant #10) disclosed:

"What was important to me was always the family, then food, drink and conversation." P#10 spouse: "All the things that he loved, yeah." P#10: "Yeah." P#10 spouse: "Yes, the dining room table was a great feature of our house and so, and @@Morris## at the top of it." (....) P#10: "I can't do anything of it now (....) huge, unbelievable loss."

A minority of participants (n=4) [Danielle (participant #9), Áine (participant #21), Pascal (participant #23) and Denise (participant #33)] were keen to live life to the full. Living life to the full for these participants meant making the best of every day and (where physically possible) pursuing life-long goals which they had not yet achieved. However, all participants still suggested that "normality" was passing them by:

"I think there's more happening than people can see or realise. Personally I think it's, I think it's my, my own, my own body is withdrawing from my old life into this new situation and I'm, I'm discarding all the things that, that I used to do, used to enjoy. I'm not doing it deliberately. My body is saying you know pull back from that and that's what's happening." (Martin, participant #31)

4.3.2 Struggling between the present and the future: Needing time to process

Participants' ability to move between the present and the future also aided them to cope with insurmountable loss. The majority of participants were moving at varying speeds towards acceptance of ALS and the process of acceptance for participants was complex. In this study, much of the data depicts how participants wished to live on "in the present" but also how they were resigning themselves to death from ALS:

"I try not to think about it, however last night was the first time that I found it difficult to breathe (....) I tend to get a plug of mucus in my throat that I can't clear. And when I was breathing it was causing like wheezing but it was all in the throat. But it panicked me and I know that's what's going to kill me. Em and that is going to be hard to get through that." (Samantha, participant #22)

Participants' struggle between the present and the future comprised a number of dimensions. Participants struggled hard to deal with everyday challenges imposed on them by ALS but as loss mounted in their lives, they also recognised the importance of considering the future in order to somehow plan (if not fully prepare) for the challenges that were yet to come. Participants struggled between the "here and now" and the inevitable progression of their condition, but they were also resigned to (if not always fully accepting of) ALS. Helen (participant #12) explained:

"It's not that I'm totally laid back and accepting my lot and of course I get angry, you know. I have to because that's always going to be there but I mean I've never thought why me, I thought why not me. I'm not a fatalist, em but I have it, that's it, I have it, you know, so where do you go from here, well you do the best you can in the meantime."

Nonetheless, participants feared death from ALS and even though some participants suggested they had accepted ALS, they stationed themselves closer to the "here and now" than to "the future with ALS". Indeed, participants struggled between attending to information about the future with ALS and not wanting to know about the future with ALS:

"I know you have to plan ahead and that you have to be organised, I'm an organised type of person, you have to be organised for what's coming down the road and like I'm not oblivious to that, but for the moment it definitely helps me to stay where I am." (Terry, participant #11)

"I want to take one day at a time but I'm not too sure, in me heart I suppose I'd want to know. I suppose you'd be more prepared if you could know." (Sally, participant #20)

Moving back and forth between the present and the future enabled the majority of participants to negotiate the challenges of living with ALS. Many participants were attuned to the fact that they themselves wanted to accept loss. Nevertheless, they judged carefully when and how to shift their focus of attention back and forth between the losses encountered thus far and the losses they had not yet encountered. Needing time to process was central to how participants managed to cope as they moved between focusing on the present and thinking about the future. Needing time to "digest" rapidly changing circumstances permeates all interviews. Participants suggested they coped better when they had had time to adjust:

> *"Overall I'd say I'm doing okay because I have the time to adjust to it, you know, yeah definitely."* (Helen, participant #12)

Participants who had experienced rapid disease progression struggled more (than those who had a somewhat slower disease progression) to process change. Participants who believed they had encountered rapid deterioration in their function suggested they had had little time to adjust to loss:

> *"I get very emotional when I think about what I have lost (....) When I was diagnosed I was walking with a limp but no aids. Within six months I was needing to use a frame because I kept falling over. Within nine months I needed people to walk with me. Within a year I could hardly do two steps (....) there is so little time to take it on board, it's [loss] incessant (....) The future is so grim because of all the loss."* (Samantha, participant #22)

Participants felt less in control when they felt they had little time to process loss. Cara (participant #32) endured a "nervous sanity" in the period between her diagnosis and her son's wedding because preparing for the event gave her little time to come to terms with her diagnosis:

> *"I couldn't even think of the wedding. I wished it wasn't on because I was so panicky (....) it was nervous sanity I had, I wanted more time to think, I wanted to digest different things myself but a lot of time I wasn't getting the chance."*

Nevertheless, having at least some amount of time to process was necessary for all participants to cope with the adverse effects of ALS. Participants hoped for time to "digest" loss because as Helen (participant #12) remarked, simply living with ALS might force people with ALS to adapt to loss:

> *"You're constantly adapting because it's changing (....) your movements through the day are, are affected, they have to be affected because constantly the nerve is being weakened on a constant basis."*

4.3.3 Balancing between fighting and accepting ALS

Finding a balance between fighting and accepting ALS also assisted participants to adapt to insurmountable loss. Indeed, fighting ALS was, for most participants, central to how they attempted to resist loss. Not only did fighting ALS prompt participants to engage with healthcare services in an attempt to exert some control (as described in Chapter Five), fighting ALS also unearthed feelings of resolution for participants that they would somehow manage to minimise the effects of ALS in their lives. Pascal (participant #23) described how he felt resolute in the face of ALS:

> *"When I got out into the car park and my wife was in floods of tears and that you kind of get this, I don't know, feeling of determination and resolution and as I say you got to be strong. Not so much for yourself but for the people and family around you. You can go two ways, you can either fall into em, a sorrowful state and kind of woe is me kind of scenario or you can be resolute about it and say I'm going to do whatever I can do to minimise the impact and you get on with life and enjoy what's left."*

Central to fighting ALS was time and participants varied in how and when they chose to fight ALS. Some participants suggested that it was important to continue fighting ALS regardless of how much loss they encountered. Vincent (participant #5) had already encountered severe limitations in his mobility but he still struggled to move without assistance despite the restrictions ALS imposed upon him:

> *"I fell in the bathroom and I couldn't get up, @@Tara## [spouse] was trying to lift me, I said don't, leave me, leave me, I'm dead weight now like, just pull my leg out, my leg went in under me, the bad one, I was there for about ten minutes. And I started crying,*

she said what are you crying for, I can't help myself I said, if I could help myself like it'd be grand but I couldn't, so the next thing I said no, don't lift me, so I kind of made my way to the toilet and I caught the bar on the wall, I was kind of frightened after the fall."

However, Áine (participant #21) who was anarthric and quadriplegic, was adamant that she would only continue to fight ALS up until the point where she would require assisted ventilation to stay alive:

G: "@@Áine## I'm getting the sense that you are fighting MND?" P#21: (P#21 indicating yes). G: "And yet you have made certain decisions such as not getting the feeding tube, opting against assisted ventilation should you need it which on one level might suggest giving into MND. Do you think there's any conflict there or?" P#21: [using switch-activated AAC device] "When you can't breathe on your own it's game over."

For the majority of participants, early to mid-stages of the disease (as perceived by them) were characterised somewhat more by fighting than accepting ALS. Participants' inclination to move was strong and their openness to consider "any remedy" reinforced how few were "waiting" for death. Fighting ALS also engendered feelings of hope for a minority of participants [Danielle (participant #9), Tim (participant #16) and Áine (participant #21)]. Feeling hopeful about the future with ALS enabled them to remain resolute in the face of insurmountable loss:

"I've always had that mentality where I'll fight this and I'll beat it (....) I don't know how I'm going to beat it, I haven't got a clue what that is (....) but it's a better day when you feel that way than it is when you are down." (Tim, participant #16)

Fighting ALS comprised both physical and psychological dimensions but participants suggested that fighting ALS (in the early to middle stages of the disease) was perhaps somewhat more physical in nature. Indeed, participants' resistance to physical change permeates all interviews and resistance to physical loss centred primarily on their desire to maintain normality:

"I feel I don't have energy (....) but it doesn't stop me trying. I still go up to the shops. As soon as you are gone I'll be up to the shops." (Maureen, participant #34)

However, despite participants' reluctance to submit to ALS, the data strongly suggests that the majority of participants were at least attempting to find a balance between fighting and accepting ALS and were perhaps, with increasing awareness of their loss, moving towards acceptance. Cathal (participant #28), who died four months after his interview, remarked:

"Like there's nothing I can do about it. I have battled this as much as I could, I fought with this thing to try and stop it from beating me, to keep me walking, I just, I wasn't able (....) I'm still fighting it a small bit, the little bit of fight I have left in me I am."

The relationship between fighting and accepting ALS was complex because most participants felt that they could not accept ALS unconditionally. As described in Section 4.2.3, participants' feelings of "having no choice" permeate all interviews whereby participants felt forced to adapt and in some cases, forced to accept. As Cathal explained:

"I can't turn around and say to myself, I'm getting out of the wheelchair and going for a walk, you know, I'd love to be able to do that. But it's not by choice I'm in this fucking thing you know, it's not my choice to be in this but I'm in it and nothing I can do about it, just accept it."

Moving towards acceptance of ALS was complex for participants. All participants resigned themselves to the difficulties posed by ALS and the majority of participants like Jennifer (participant #25) felt the need to accept ALS. However, some participants such as Tim (participant #16) were adamant that they would never accept the condition *per se*:

"I've had a lot of trouble getting my head around losing physical independence and thought it would [involved being] dressed like a child, or undressed you know. But I have to get over it." (Jennifer)

"I don't accept the disease. I accept the difficulty but I will not accept the disease. Or I will not take ownership of it, it's not mine. It doesn't belong in my body. I'll never accept it." (Tim)

Overall, participants struggled between fighting and accepting ALS. Denise (participant #33) believed that life-sustaining treatments are inevitable for most people with ALS but she would abstain from such interventions up until the point where she could no longer survive without them:

"I would put if off to the last minute but would do it."

The *meaning* of acceptance for participants was complex and varied across the sample. For the majority of participants, acceptance meant accepting ALS and/or the need to stop fighting ALS in order to accept ALS. However, for a minority of the sample, acceptance meant coming to terms with life-altering events associated with ALS but not necessarily ALS *per se*. Indeed, in the minds, of some participants, fighting ALS equated to accepting loss because failure to accept loss meant one was fighting oneself:

"By accepting loss you are fighting it [ALS] because if you don't accept it [loss] you are fighting yourself. I believe our body is equipped to fight any illness, any disease, but don't use that energy on a useless fight." (Tim, participant #16)

Participants' personal accounts of their experiences suggest that there is also a time dimension to acceptance of ALS for people with ALS. In this study, the majority of participants were moving towards acceptance as they encountered more loss. However, similar to how participants moved back and forth between the present and the future, participants also moved back and forth between fighting ALS and accepting ALS. Indeed, striking was how some participants suggested that they had accepted loss but not necessarily the loss they had yet to encounter:

"I've a big challenge at the moment with eating because it's getting harder and harder to actually feed myself sort of thing, I'm like I am not having someone feed me. I'm just not going, you know, I know eventually it's going to happen but like not for a while." (Danielle, participant #9)

Participants' negotiation of mounting loss in ALS also comprised "bargaining" for no more loss. Participants negotiated mounting loss by accepting the loss they had already encountered in return for no more loss. The majority of participants "bargained" for no more loss. Danielle shared:

> *"I don't think you can go back and I don't think they can make me better but if you said it's never going to get any worse than this that would be like brilliant, fantastic I'd be thrilled to bits."*

In this context, participants' acceptance of ALS was not only shaped by their experiences of mounting loss. Moving towards acceptance of ALS (or simply the challenges posed by ALS) was also shaped by their ability to prioritise what they had not yet lost:

> *"He [other person with ALS] just said to me don't be worrying about what you're losing, concentrate on what you have and so that I suppose is what I am doing."* (Helen, participant #12)

Overall, the relationship between acceptance and loss was complex because regardless of whether or not participants accepted or struggled to accept ALS, all participants encountered loss. Moreover, participants' inclination to bargain for no more loss suggests that whilst many (not all) participants were moving towards acceptance at varying degrees, *how* they moved towards acceptance was shaped by their anticipation of more loss.

4.3.3.1 Acceptance: Shaped by age and life stage

As described in Section 4.3.3, participants struggled between fighting and accepting ALS and/or the losses they encountered because of ALS. This study found that age and life stage shaped how accepting participants were of ALS. Participants were more accepting of ALS when they had already fulfilled their life-long ambitions and had had the opportunity to live a long life. Edward (participant #14, age 81) commented to the researcher:

> *"You [younger] people haven't lived"*

The relationship between age and acceptance in ALS was complex along a number of dimensions. Overall, participants indicated that it is more acceptable for people in later life to die from ALS than it is for "young" people to die from ALS. Indeed, participants (regardless of their age or life stage), thought it was "tragic" to witness "young" people with ALS struggle with such a life-limiting condition. Some middle-aged and older participants suggested that they would give their life in return for a "young" person with ALS to survive:

> *"There's a young boy, he got it at fifteen he said, he's twenty-four now, struck down with it. I said to the wife, I'd prefer if I was taken now and left that boy, young boy like, you know he was young, I had a good auld spin [good life so far] at sixty-two, he was only a young fella [man], young boy, so young."* (Vincent, participant #5)

Young participants, middle-aged participants, *and* those in later life suggested that it is more acceptable to die when one has reached old age. Indeed, all participants aged 80 years and over indicated that they were more than ready to die. Apt was Paul's (participant #13, age 81) wish for a "perfect" death in the context that he had already accomplished what he set out to achieve in life:

> *"I'd be jealous of fellas [men] I went to school with. There's one fella now died having his porridge, oh man what a way to go [die] (laughing). I don't mind death at all but I feel sorry for people that would get it young you know, but like thinking about it, god I would hate to have it younger. I've my kids [children] raised and everything."*

Middle-aged and older participants also revealed that even though they themselves were accepting of their own impending death, they still remained hopeful that "young" people with ALS might live long enough for a cure:

> *"I think with younger people they might find a cure and there is hope for them, who are younger now (....) I'm going to die because there's no cure for me."* (Cara, participant #32, age 64)

Seven participants in this study were under 50 years old. The researcher classified these participants as "young" because they considered themselves young in the context of

having a condition they associated with middle and later life. Young participants suggested that older people are perhaps more vulnerable in the fight against ALS and are more likely to resign themselves to ALS because they are less equipped to resist ALS. In addition, many young and middle-aged participants implied that ALS is somewhat more acceptable in later life as people in later life are more likely to have already acquired morbidity:

> *"I think if you are a young healthy man you are going to have a better chance than being an old man, who has probably other things [morbidity] going on, the whole thing is more acceptable. It's like a car, if the car is old and starts getting trouble it's going to the scrap yard because the car has done its bit."* (Tim, participant #16, age 47)

Some participants in later life were more accepting of ALS and of their impending death because they no longer felt worried about the prospect of living to "old age". Indeed Jennifer (participant #25, age 73) suspected that her quality of life would be less than she would wish it to be if she were to live to "old age". She lamented the loss of living through her 70s and early 80s but not necessarily the life she might otherwise have encountered beyond that point:

> *"I come from a long-lived family and I was liable to live into my mid-eighties and I didn't want to get to ninety. But eighties is like ten years away and I was going to get a lot of retirement living into that time (....) [but] it would be much harder to maintain your independence in your nineties."*

Participants spoke of *becoming old* with ALS in the context of experiencing physical decline which they ordinarily associated with ageing. For some participants, ALS had the capacity to "age" them which they suggested, in turn, forced them to accept ALS:

> *"I see people over eighty and I'd say god I'm never going to get old and then this thing [ALS] happened to me (....) I never felt old until this thing started (....) It's going to come to a stage that I probably just can't go on any longer."* (Cara, participant #32, age 64)

As already described, a number of participants struggled between resisting and accepting ALS and the meanings participants attached to ageing prompted feeling of acceptance *and* resolution among participants. Andrew (participant #6, age 64) was glad of the

opportunity to have lived a worthwhile life but his desire to live on was strong and he was not "waiting" for death:

> *"I mean at my age I'm sixty-three or whatever, sixty-four, whatever it is now, you know, I suppose I haven't done too bad but if there's any, if you can get a few years longer out of it, you'd like that."*

Participants' perceptions of ageing were closely linked to how they perceived their roles at different life stages. Indeed, for the most part (as described in Chapter Five), participants spoke about their concerns in the context of life-course trajectories. Achieving one's personal milestones (which centred primarily on the opportunity to have parented and raised one's family) fostered feelings of acceptance among participants. However, young participants who felt denied the opportunity to raise their children were less accepting of ALS. Eve (participant #7, age 45) disclosed with visible emotion:

> *"It hits a very raw nerve (crying), not for me, but for my children (crying). I don't think it would be as devastating if my children were reared, that is the hardest (crying)."*

4.3.4 Becoming more certain: Coping with the prospect of future loss

Moving from uncertainty (pre-diagnosis) to a *somewhat* more certain world of ALS also assisted participants to adapt to insurmountable loss. With increasing awareness of the progression of the disease, feeling more certain about disease progression enabled some participants to prepare for future loss and come to terms with loss. Many participants indicated that they would cope better if they knew how or why they developed ALS but the certainty of a diagnosis of ALS in itself assisted participants to cope with loss. As Mona (participant #1) remarked:

> *"It's good to know where you stand, to know what's going on."*

Seeking certainty in ALS centred primarily on time frame so that participants could somehow plan (if not fully prepare) for the future. Participants felt they were losing control over their lives as they advanced in the disease but they still possessed the ability to plan how they might at best deal with and in time, come to terms with ALS and mounting loss. Feeling more certain about ALS had the potential, for some participants, to

render life more "bearable" in the context that they had already lost so much control over their lives. Martin (participant #31) described his position as he desperately searched for some certainty in an attempt to regain control in his life:

> *"I felt I was floating on my own and this thing [ALS] is progressing (....) I've a high tolerance to pain but I'm terrified of being, of my life being in everybody else's hands except my own (....) I said well I feel I'm in trouble (....) I wanted to know."*

The majority of participants feared what was yet to come with ALS (e.g., respiratory failure, deterioration in and/or loss of speech). However, becoming more certain about the progression of ALS through bearing the loss they encountered in ALS, alleviated (but did not remove) their fears about the future:

> *"I was delighted, she [consultant neurologist] said when you have this, there's no more spinal taps, there's no more fucking blood tests, there's no more MRIs which I hate. So that's fine and then I don't have to have treatment, and I'm not sick. So when I'm talking to the kids [children] I'm me. Okay physically I'm fucked but mentally I'm the same."* (David, participant #30)

As described in Section 4.2.5, comparing self to other people with ALS and comparing ALS to other disabling and/or life-limiting disorders reinforced for participants the extent of their own loss. However, on the other hand, comparing self to other people with ALS and ALS to other disorders also assisted participants to come to terms with ALS. Participants perceived the disease trajectory of ALS to be more certain than that of other diseases (e.g., cancer) and the "certainty" of ALS helped participants to adapt to loss. Jennifer (participant #25) explained:

> *"You have to accept that you are not going to get better (....) I mean I would have thought having cancer is ten times as hard. I mean you put yourself through ferocious treatments. And then you get better for a bit, if you are lucky and then it comes back." G: "Are you suggesting then @@Jennifer## that perhaps the predictability of MND even though there is variation..." P#25: "Well it's predictable." G: "Do you think that the predictability of MND versus the unpredictability of cancer or other conditions where*

there are periods of remission (....) makes it easier to cope with MND?" P#25: "Yes I think so."

Indeed, a minority of participants had already encountered previous illnesses and they compared ALS favourably to these conditions. Callum (participant #3) who had moderate fronto-temporal dementia (FTD) and who was fully dependent for all his care needs spoke of how living with ALS was less painful than living with depression:

"I had depression before (....) I handled this better."

Comparing themselves to other people with ALS also assisted participants to cope with the prospect of future loss. Indeed, some participants were encouraged by the longevity of other people with ALS in the context that perhaps the losses they had not yet encountered would somehow not be as severe as they had expected them to be:

"I met a woman above in the clinic, her speech was affected, her walk was affected you know a little like, she was able to walk, she was able to speak but she had we'll say symptoms all over. But like in three months like she hadn't gone back anything [had not gotten worse]." (Eilish, participant #19)

How participants responded to other people with ALS in order to cope with impending loss was complex. Participants were curious about how other people with ALS managed to confront loss and compared their own coping strategies to those of others. The question that Melanie (participant #27, end-stage ALS) posed was striking:

"It's been at the back of my mind, can't quite put it into words or form a pattern of thought. Eh all these people you've spoken to with motor neurone, I don't quite know how to put it, are they generally depressed and looking into a dark tunnel?"

However, some participants also sought to remain at a distance to other people with ALS because meeting other people with ALS accentuated their own fears about future loss. Jack (participant #26) explained:

"I just don't want to be with somebody else who has it as well and sit and talk to them about it, maybe it's because he might say something that I don't want to hear or I might say something to him he doesn't want to hear."

Overall, most participants were simply keen "to get on with it". Making the best of the remainder of their lives assisted participants to adapt to loss:

"There's days I think oh fuck and then I give myself a kick (....) I can either sit in a corner and whinge and moan (....) or I can get on and do what I'm able to do and that's what I try to do (....) I need to have something ahead to look forward to." (Helen, participant #12)

4.3.5 Thinking about "holding on" and thinking about "letting go"

Participants' abilities to navigate between wanting to live on and not wanting to live on also assisted them in adapting to insurmountable loss. Participants' experiences of unremitting loss inevitably pressed them to think about death from ALS. Some participants were unacquainted with how people die from ALS (i.e. respiratory failure) but all were very much attuned to the fact that they would sooner rather than later encounter the final stages of the disease. Few wished to live on in a state which would be, in their view, unacceptable to them. Apt was the response of Paul (participant #13) to how he might approach the end stage of ALS:

"Jesus lets be practical here. Wouldn't it be lovely just lying in this chair and I woke up dead (laughing) you know to see what you are going to through (....) Look I'm chicken, I don't want to suffer, I just don't want to suffer you know. So that's why I say if you woke up dead yeah that would be nice."

Indeed, participants' desire not to "suffer" at the end stage of the disease permeates most interviews:

"If you want an honest opinion the only thing that I am afraid of is that I wouldn't strangle to death or something, you know what I mean, that you don't get, you're not going to end up half strangled and dying that way. I know that's bloody morbid to be talking about but that would have crossed me mind." (Andrew, participant #6)

A minority of participants suggested that they would fight (or indeed were in the process of fighting) ALS to the end but many suggested in the course of their interview that they were beginning to plan for (if not fully embrace) death. However, planning for end-of-life was complex because not only did participants welcome death because they felt they had accepted ALS, some also welcomed death because life had become unacceptable to them amidst the experience of unremitting loss:

> *"If you get up every day with the tube [gastrostomy feed] in that's okay, if you are quite happy with that, to be still alive on that basis, that's okay but there are other people like me who don't want the bloody tube to keep them alive."* (Morris, participant #10)

Adaptation to loss in ALS not only comprised the struggle between fighting and accepting ALS; participants also adapted to insurmountable loss by losing the desire to live on with ALS. Indeed, freedom to choose not to live on with ALS in a state of "suffering" was central to the hopes and expectations of participants as encapsulated by the following remark by Richard (participant #8):

> *"I mean why go down that road (....) you drink out of a straw and that, sure I don't want to live forever with this bloody thing if it gets worse you know."*

As participants encountered more loss and endured the restrictions ALS imposed on them, many deliberated on what could be tolerable to live with and what might well be unacceptable to them. Participants placed more importance on quality of life than on quantity of life and for many the struggle between choosing quality of time over quantity of time had moral dimensions. Apt was the account that Martin (participant #31) gave of his struggle between wanting to die through the natural course of ALS and feeling obliged to live on with a gastrostomy feed:

> *"I'm not a religious person as such, although I am you know, I am a moral person. I wouldn't want to put myself in a situation where I thought myself that I were committing suicide by not taking the tube. But having said that, I would recognise that if, if the natural way to nutrition is to swallow food and you can't do that, well then the moral argument about whether I should use a feeding tube or not has a different dimension."*

Participants struggled to find a balance between enduring ALS and "letting go". Balancing between wanting to live on with ALS and wanting to die soon comprised the ability to judge when life no longer offered meaning. Indeed, participants indicated ALS would be at some point in the future, unacceptable to them:

> *"I thought if I am so bad that, obviously if I can't swallow, I can't talk, who wants to go on? That's my feeling. Now because I don't know anybody in that condition I don't know what kind of quality of life they have. But I feel it wouldn't be much."* (Jennifer, participant #25)

> *"When I can't do anything I enjoy it's time to stop."* (Denise, participant #33)

Indeed, for some participants, the difference between living with ALS with some quality of life and living with ALS with no quality of life was diminutive. Cara (participant #32) explained:

> *"There's a very thin edge, or a thin line (....) I feel if my diaphragm is going to go, my speech, my hands, how am I going to communicate you know (....) I'm going to die so why prolong it."*

Nonetheless, many participants (including those who wished to die) struggled between wanting to live on and "letting go". Some participants hoped that death would come soon but many still deliberated between sustaining life and ending life:

> *"I mean I don't want to be brought back from the brink. But I mean I don't want to say no resuss [resuscitation] at all, no treatment, if you say no to them it's too drastic. I mean one gets over pneumonia."* (Jennifer, participant #25)

> *"I've obviously had patients [as a medical doctor] in this kind of situation, obviously not with motor neurone but where you'd, would have wondered about the wisdom of treating but on the other hand I suppose one is reluctant to let go of that tenuous chord of life kind of you know."* (Peter, participant #4)

Indeed, despite mounting loss, very few participants seemed ready to die:

"Sometimes I think I have accepted and then other times I think maybe I haven't accepted it at all (....) I really don't want to think about it any much further. I know it's going to happen but it's not going to happen next week." (David, participant #30)

4.4 Conclusion

This chapter has described the meaning of loss for people with ALS and how people with ALS adapt to insurmountable loss. Participants were overwhelmed by loss but how they responded to loss, suggests that people with ALS might indeed adapt to insurmountable loss.

The findings suggest that adaptation in ALS is bidirectional. Many participants were moving towards some level of acceptance in line with increasing loss but adaptation was shaped by how participants navigated back and forth between resisting and accepting change. The findings reveal that negotiating loss in ALS comprises complex appraisal strategies which enable people with ALS to move between accepting ALS and resisting ALS. However, regardless of whether people with ALS accept or resist ALS, fundamental to the experience of living with ALS is loss. Some people with ALS fight ALS in an attempt to arrest loss but people with ALS also move from fighting to accepting in the face of unremitting loss.

How participants in this study responded to living with a terminal illness is contrary to much of the literature on how people with chronic illness respond to change. Charmaz (1995) suggests that people with chronic illness move to altered identities in order to adapt to loss. The findings in this study suggest that participants struggled to find a meaningful identity in the face of *constant* loss. ALS is a rapidly progressive condition and the clinical course of ALS differs greatly from the disease trajectories in chronic illness. There are no periods of disease stability or remission in ALS. It is possible that as people with ALS encounter *rapidly* changing circumstances, they may not have sufficient time to move to a meaningful identity. Although some participants accepted insurmountable loss, none indicated that they felt better for the experience of such loss.

The findings also differ from findings of more contemporary research on how people with chronic illness adapt to change. Older participants in this study approached death as it were an inevitable life event i.e. part of biographical flow, but there is little evidence to

suggest that participants continued as before i.e. biographical continuity (Sanders et al., 2002) or returned to a "normal life" i.e. biographical reinstatement (Sanderson et al., 2011). Instead, the diagnosis of ALS moved participants to mortal time (McQuellon & Cowlan, 2000) i.e. a "death sentence" characterised by intractable loss. Importantly, a diagnosis of ALS eradicated a future in which loss would at some point cease and allow participants to regain what they had already lost.

The findings resonate with and are contradictory to studies that have investigated how people with terminal cancer adapt to illness. As described in Chapter Two, Reeve et al. (2010) found that people with terminal cancer manage 'disruptive' events to maintain an overall sense of wellbeing. Maintaining 'continuity' is exhausting in the face of ongoing loss but people with terminal cancer might still manage to 'restore' stability in their lives (Reeve et al., 2010). In this study, the evidence suggests that people with ALS manage disruption in their life. However, no findings suggest that participants actually managed to restore normality because their losses never ceased at any particular point in time. Literature on how people with terminal illness adapt to illness focuses primarily on cancer service users. The disease trajectories in different forms of terminal cancer are hugely diverse. In comparison to cancer, ALS is in most cases relentless from outset, with little effective treatment available to service users. In addition, people with terminal cancer can encounter a somewhat more uncertain disease trajectory where periods of remission or stability may offer hope of survival (Astrow et al., 2008). Although many participants in this study were unsure about their prognosis, all were acutely aware that they were living with a terminal disease which would offer no periods of remission. The findings suggest that people with ALS fail to maintain normality because they enter into a state of *constant* flux characterised by *relentless* loss. In this study, failing to maintain or restore normality in ALS was a product of having to adapt to unremitting loss.

The findings resonate with and differ from those of other researchers who have studied how people with ALS adapt to change. The data suggest that participants encountered excessive loss i.e. 'fracturing' of life, and adapted to ALS by 'enduring' loss (Brown & Addington-Hall, 2008). The study also identified biographical abruption (i.e. "death sentence") and disruption in the lives of people with ALS (Locock et al., 2009). However, Locock et al. also detected biographical repair in ALS. They found that biographical repair comprised balancing avoidance and acceptance, regaining control, restoring normality,

keeping hold of the old normality, creating a new normality, living life to the full i.e. heightened normality, and finding new meaning and identity. Brown and Addington-Hall (2008) reported that people with ALS might also 'preserve' and 'sustain' despite disease progression. In this study, some participants resisted being "absorbed" into a world of ALS and there is variation in terms of how accepting participants were of ALS. However, no data suggest that participants regained control, held onto normality, or indeed found a new normality. Participants in many cases attempted to remain positive by focusing on what was still important in their lives but did so *knowing that they would eventually lose it in the end*. A minority of participants were keen to live life to the full (i.e. heightened normality) but all participants were resigned to more immediate loss. In this light, it is possible that people with ALS fail to regain, sustain, or preserve because they live with unremitting loss that is beyond their control. The data suggest a *forced* 'continuity' in ALS. Despite losing out on normality, participants continued to live through ALS and associated loss. Theoretically, living with ALS might well force people with ALS to adapt simply because the disease trajectory in ALS is one of constant neurodegenerative change. No data suggest participants stopped "living" and many participants felt they had "no choice" but to live through unremitting loss.

Locock at al. (2009) and Brown and Addington-Hall (2008) conducted biographical and narrative-based studies. Their aim was not to develop a theory. Sampling by Brown and Addington-Hall (n=13) was restricted to four National Health Services (NHS) primary care trusts in the UK. Locock et al. purposively sampled for variation in experience from different sources in the UK (ALS centres, local and regional voluntary associations, ALS specialist nurses, an on-line forum) but it is difficult to ascertain whether their sample captured variation across a population. As described in Chapter Three, Grounded Theory method enabled the researcher to saturate key concepts and categories (e.g., loss) in terms of their properties and dimensions. It is possible that by sampling participants from a population-based register to saturate concepts for variation, the researcher managed to capture the magnitude of loss for people with ALS. Locock et al. and Brown and Addington-Hall sampled participants with primary muscular atrophy (PMA) and participants with primary lateral sclerosis (PLS). PMA and PLS are highly disabling motor neuron disorders but they differ in disease trajectory to ALS[1]. All participants in this study

[1] Primary Muscular Atrophy (PMA) and Primary Lateral Sclerosis (PLS) are variants of motor neurone disease (MND). In the United Kingdom and the Commonwealth, *MND* refers both to ALS and to the broader spectrum of related motor neurone disorders that include PMA and PLS. In the

had a diagnosis of ALS. Variation in disease trajectories contextualises the difference in findings between studies.

This study set out to understand how people with ALS engage with healthcare services and what factors shape the decisions they make about their care. As described in this chapter, loss was insurmountable for participants and all participants felt they lost control over their lives. The next chapter explains, at a substantive level, how people with ALS interact with healthcare services in response to losing control and "suffering" insurmountable loss. The inner-most experience of the ALS service user shaped how participants interacted with the external reality of help and support available to them.

United States and continental Europe, the term *ALS* is used for the specific disorder ALS and as a blanket term for the entire spectrum of MND.

Chapter Five

How and why people with ALS make decisions about care: Understanding interaction at the meso-level

"In relation to MND I have to be very realistic about this. The healthcare professionals do very little for me in relation to the disease because they can't. They've done as much as they can, they have me on the Rilutek and they have me on high protein and high fat diet, after that they react to whatever problems I run into and make the disease more bearable. I mean I see @@the consultant neurologist at the national ALS clinic##[1] *and her team as like a group of shepherds who, who are shepherding me towards you know, towards my, towards my death really, you know. And what they're doing is they're, they will in so far as they can make it more bearable for me (....) like my shepherds to move me as gently and as bearably possible towards where you know, when I die. And I know they'll do that with compassion and efficiency and indeed the local services would be, will be the same but that's about it really."* (Martin, participant #31)

5.1 Introduction: Needing healthcare services and family to "get through" loss

As described in Chapter Four, people with ALS live through insurmountable loss. In this study, all participants engaged with healthcare services as they lived through loss. For participants, the world of ALS included healthcare services because ALS forced participants to engage with healthcare services in order to cope with mounting loss. As illustrated in Chapter Four, participants failed to control ALS or the losses they encountered because of ALS. Few participants wanted to engage with services because the need to engage with services was a reminder to them of what they had already lost and of what loss was yet to come. However, as Martin (participant #31) described, healthcare professionals are important to people with ALS because they have the potential to make life "more bearable" for people with ALS. Participants felt they had "no choice" but to engage with healthcare services but they sought to exert control over healthcare services. Indeed, failure to control the effects ALS on their lives prompted participants to exert control over when and how services were rendered to them. Engaging with healthcare services was in itself an adaptive strategy for participants because engaging with services assisted participants to cope with loss.

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[1] *@@the consultant neurologist at the national ALS clinic##* denotes that there is only one consultant neurologist at the ALS clinic. This de facto enables identification. Consent has been given not to anonymise.

As outlined in Chapter Three, a key tenet of critical realism is that people interact with phenomena in the 'real' world. This chapter explains how people with ALS engage with healthcare services i.e. where participants were in contact with, thought about and received care from service providers (at the meso-level, between the micro level of individual lives and macro level of policies and systems). Importantly, this chapter also explains how the external reality of other supports (i.e. family) shaped how participants interacted with healthcare services. The findings in this chapter draw on those of Chapter Four because the personal experience of participants shaped how they responded to healthcare services and to support from those close to them.

5.2 Thinking about and engaging with healthcare services

The majority of participants *took action* in healthcare services prior to the diagnostic phase of ALS. Searching for services to investigate their worsening symptoms stands out in participants' accounts of their early experiences of healthcare services:

> *"I attended my GP about March 2009 and, he didn't think there was anything really wrong with me and he said hold over until the end of the summer before I would seek an appointment with a neurologist, because my symptoms were so slight. But I said no, I'd like to be referred to a neurologist for no other reason to rule out things."* (Eve, participant #7)

Taking action not only included participants' request to engage with physicians. Engaging with services to cope with mounting loss also included requesting services from a broad range of healthcare professionals, and in time accepting physical assistance (e.g., home care) and psychological support (e.g., counselling) from healthcare services. Samantha (participant #22) explained:

> *"I think as the disease has progressed we've had to address the disability, for instance I noticed it was harder to get me up out of the chair so I just rang the OT [occupational therapist] and said I needed a standing hoist (....) We went and saw the palliative care consultant on Friday for the first time and we had a long chat about the services available down this end of the country (....) The palliative or home care team come out as well, she's [palliative care nurse] has been out regularly (....) and we've been seeing a social worker within that team."*

Participants valued services rendered by allied healthcare professionals (e.g., ALS specialist nurses, occupational therapists, physiotherapists, speech and language therapists, nutritionists, and counsellors) because they felt these services "made a difference" in their lives. Such services offered little hope of a "remedy" but participants relied on these services to cope with the physical and psychological effects of mounting disability:

> *"I know it will progress and it will be worse as time goes on but I still think with the support out there I'll be fine you know. I know now that all these people that are there I can get the help or whatever I need and that's good to know."* (Ita, participant #2)

A minority of participants (n=4) disclosed in the course of their interview that they had availed of alternative treatments (e.g., acupuncture, herbal medicine). Indeed, despite participants' growing awareness of the uncontrollable nature of ALS, many struggled between accepting the progression of the disease and seeking a cure for ALS. Samantha (participant #22) disclosed:

> *"We did go to the Chopra centre in California for Ayurvedic because [it] is meant to help heal, it's Indian medicine (....) and we were going to go to China for Chinese traditional, Chinese medicine. There are some write ups about motor neurone disease and how they've helped people. So we tried acupuncture and traditional Chinese medicine but we did it from here because we found an acupuncturist that trained in Beijing."*

As depicted in Chapter Four, participants moved back and forth between accepting and resisting ALS. Some participants felt they accepted ALS but many participants wanted to participate in research to find a cure for ALS or at least treatments to slow down the progression of ALS. Danielle (participant #9) spoke of her disappointment when she did not fulfil the clinical criteria to participate in a clinical trial that had commenced prior to her interview:

> *"I was two years diagnosed and it wasn't for people two years diagnosed (....) Oh I was so disappointed but you see you have to appreciate where I am coming from. I am looking for a cure, other people try to understand the illness so it depends what the objective of the trial is and a lot of the other trials that are going on at the moment are*

about studying what causes the illness. And I just think, the time frame for that to deliver results is too long for me. So I'm really interested in the trials that are trying to put forward new treatments to slow it down."

Participants' interpretation of a "cure for ALS" varied. Some participants perceived a cure as any disease-modifying agent that might slow the progression of the disease and enable them to maintain at least some independence in their lives. However, for other participants, a cure constituted a treatment that could "get rid of" ALS and enable them to regain what they had already lost. Apt was how Paul (participant #13) explained what he understood a cure to be:

"Well you see it's either, it's either a nice death or be perfect. That's it." G: "So no in between?" P#13: "No, no life to be perfect, it's like what's your man JR, I'm perfect but in every way, you know (laugh). Oh yeah, perfect in every way. I don't want to live with physical disability."

Participants' desire to engage with medical research extended beyond the potential benefit for them. Coming to terms with loss also included the need to be of help to those who may encounter ALS in the future. Ita (participant #2) explained:

"I think for the future, I feel that the fact that there is only one medication there for people with motor neurone and there's no, no cure and no sign of a cure. I feel that something has to be done for those people."

Participants engaged with healthcare services to fight ALS (i.e. fighting to feel in control) but participants also engaged with services as they moved towards acceptance. Jennifer (participant #25) shared her feelings about the prospect of increasing support from healthcare services:

"It's inevitable but she [primary care occupational therapist] only recommends things as we go along. She never told me all the things because it would be scary when you didn't need them (....) I mean the thought of a hoist would have horrified me in the beginning. But as I have trouble moving around, I can see that I could come to that."

As depicted in Chapter Four, adaptation in ALS is complex and involves both resistance and acceptance. In this study, it was not the case that participants only engaged with healthcare services when they resigned themselves to ALS or only engaged with services when they fought ALS. Instead participants engaged with services to fight *and* accept ALS as they adapted to insurmountable loss.

5.2.1 Thinking about and engaging with end-of-life care

As described in Chapter Four, participants' awareness of disease progression heightened their anticipation of the likelihood of further disability. The experience of rapid neurodegenerative change forced participants to consider (if not fully plan) how they would engage with healthcare services in line with mounting loss. Participants' decisions about care ranged from decisions for the "here and now" to decisions about end-of-life care. Helen (participant #12) disclosed:

> *"I would like a DNR [Do Not Resuscitate order] because like I know it's pneumonia that'll kill me. Well it stands to reason if my lungs fail it'll probably be pneumonia (....) to my mind that's probably the way it'll be."*

As described in Chapter Three, a number of participants (n=8) had already engaged with life-sustaining interventions (i.e. non-invasive ventilation and gastrostomy feeding[2]). No participant had engaged with invasive ventilation (long-term ventilation / tracheostomy). In general, participants felt that life-sustaining treatments were acceptable in ALS care but only prior to a state of complete physical dependence. Some participants were unsure about the function of life-sustaining treatments at the end stage of ALS:

> *"In relation to the invasive ventilation (....) does the airways just close that much and then there's no more breathing unless it's invasive treatment? Or does it never get that bad that the likes of a NIPPV [non-invasive positive pressure ventilation] won't solve?"* (Jack, participant #26)

[2] Invasive ventilation and non-invasive ventilation prolong survival in ALS. There is some debate as to whether gastrostomy prolongs survival in ALS (Andersen et al., 2012; Miller et al., 2009a). Both non-invasive ventilation and gastrostomy are palliative by definition.

Few participants in this study spoke specifically about invasive ventilation. This is perhaps not surprising as people with ALS in the Republic of Ireland ordinarily do not engage with invasive ventilation. The cost of invasive ventilation extends beyond the resources of the Irish healthcare system and it is possible that few participants were advised about invasive ventilation. Among those who spoke about invasive ventilation, few considered invasive ventilation a viable option because they believed that invasive ventilation would increase the likelihood that they would live on in a state of dependence and render them less able to communicate their preferences for care:

> *G: "One thing you mentioned there @@Helen## as well was about the gentleman who has written a few times in the [Irish] Times and [that] he's on mechanical ventilation, long-term ventilation. Do you have any views on long-term ventilation?" P#12: "Well if you've lost your speech and all that how can you tell people when you've had enough?"* (Helen, participant #12)

Participants questioned the role of life-sustaining treatments in ALS care in the context of the rapidly progressive and incurable nature of the disease. Losing independence rendered their lives less satisfactory and some participants suggested that life-sustaining interventions or even supportive care prolonged their lives "unnecessarily". Of note, participants construed gastrostomy feeding as a life-sustaining intervention. Some participants also perceived non-invasive ventilation and gastrostomy as interventions that would sustain life beyond the time frame in which they ordinarily sustain life for people with ALS:

> *"I felt I was rushed into it [gastrostomy feed]. I would have liked more time to have thought about it (....) When you get to that stage, what's the point (....) It's [gastrostomy feed] just a life line for the medics to put in food and drugs (....) to sustain life beyond what it should be."* (Morris, participant #10)

> *"I do wonder a lot, I'm saying why are they [healthcare professionals] keeping me alive for longer, why don't they just let me fade away, you know the end is going to be the same when it comes, so why prolong the suffering?"* (Cara, participant #32)

"I think I'd be dead [without gastrostomy feed and non-invasive ventilation]. I don't eat anything now, nothing by mouth (....)" G: "You haven't really engaged with community palliative care services?" P#24: "Well it's not (cough), it's not for me at the moment (....) I know it's [death] a while away." (Ann, participant #24, died 7 weeks after interview)

Indeed, engaging with life-sustaining treatments that might extend duration of their distress was unacceptable to most participants. Melanie (participant #27) who was in the end stage of ALS disclosed:

"I mean there are days when I don't feel like living (....) hopefully there won't be too much future [left] (....)" G: "Do you have any particular feelings about life-sustaining treatment?" P#27: "Dragging it out. Well I don't see the point (....) I think people might incorrectly believe that a life-sustaining treatment might lead to a general improvement (....) the feeling of having something [like this] dragging on and on. I don't want that."

Six participants spoke about physician-assisted suicide and all of these participants indicated that assisted suicide was a reasonable care option for people with ALS in the context of mounting loss. They also believed that people with ALS have a right to engage with assisted suicide. No participant (at time of interview) disclosed that they intended to travel abroad to avail of physician-assisted suicide but some had considered physician-assisted suicide and/or implied they would not rule out physician-assisted suicide at some stage in the future:

"I think you know, it gets to the stage where somebody's life where you know, there's just nothing going on for them, you know, pain all around (....) I would defy anyone to say they wouldn't think of it." (Terry, participant #11)

The freedom to decide about place of care at the end stage of ALS was extremely important for participants and for the most part, participants wished to die at home in familiar surroundings:

"I'm staying here and I'm not going to hospital (....) I spoke to the consultant in @@regional hospital## on Friday (....) I can have that care here [at home] through palliative care services." (Samantha, participant #22)

Indeed, freedom to decide about care permeates most interviews with participants and participants exerted control over how they engaged with services and made decisions about their care. The following section describes *why* and *how* participants exerted control as they took action and engaged with healthcare services.

5.3 Exerting control over healthcare services to feel in control of care of care

Although participants felt they had "no choice" but to engage with services, they exerted control over how they engaged with services. For the most part, participants used services in line with how *they* perceived their disease progression to be and only when they themselves deemed these services necessary. *How* and *why* participants exerted control in their interactions with healthcare services was complex along a number of dimensions. As described in Chapter Four, losing control over their lives was central to participants' experiences of living with ALS. This study found that losing control over their lives prompted participants to exert control in one of the very few domains that enabled participants to exert control i.e. healthcare services. Overall, participants engaged with services to feel in control of care. As Maureen (participant #34) explained:

"I think as I say that when you have a pain you say I need a pain killer. My breathing is changing but I don't want to lose control so if you need to breathe, you have to say I need that [NIPPV] to be in control."

The right to choose how and when to engage with healthcare interventions (e.g., life-sustaining treatments, allied healthcare services, symptomatic pharmacological agents, and homecare services) was extremely important to participants, and feeling in control of healthcare services enabled participants to accept or refuse services. Samantha (participant #22) [former public health nurse] explained how engaging with services might foster feelings of control for people with ALS:

"I'm hoping it [speech] will stay as it is (sigh) but if I do lose it, yes I would be looking for the devices to communicate. I have to be in control because otherwise if I weren't in

control I'd have let the public health nurse stick a blooming catheter in and look at all the issues you have with them."

Participants expressed a strong desire to follow through with their own preferences, and making decisions about their care enabled participants to feel in control of their care. As described in Chapter Four, participants resented the limitations that ALS imposed on them. In response, participants believed it was important for them to decide when to engage with services. Participants needed to be in control over *when* they would engage with services and needed time to "process" the life-altering impact of ALS before they willingly accepted assistance from healthcare services. Danielle (participant #9) explained why she had previously declined homecare services even though she had needed assistance for personal care activities:

"because it takes a long time to get used to carers, like to have somebody come into your house and get you up in the morning and give you a shower and all that kind of stuff, it takes a lot to get used to that."

As described in Section 5.2, participants engaged with services in line with their own perceptions of mounting disability and when participants themselves judged that they needed services. Subsequently, some participants exerted control by delaying specific interventions:

"I'd listen to all what I'd be told you know and I'd be taking it in and I'd be inclined to say I need that, it would be great, but I wouldn't think of getting anything until the time comes." (Sally, participant #20)

"Well I suppose if I was working outside the home I would be using my voice more whereas I'm here a lot on my own, my only social is playing golf, so I feel that I'm doing ok (....) I do find it difficult talking but I don't have to talk, you see that's why I feel that I don't need to see a speech therapist [locally] yet." (Cara, participant #32)

As described in Chapter Four, participants resented having to use assistive devices because assistive devices reminded them of what they had already lost. In this light, participants declined assistive devices offered by healthcare professionals up until the

very point at which such devices became essential for them to function. Declining assistive devices until essential to function was emblematic of how participants negotiated loss:

> *"Three or four months ago the OT [occupational therapist] was here in the kitchen and I says how do you see this going and she says I see a wheelchair. I said fucking no way, absolutely no way says I. At the time I was on a crutch (....) now that this leg is gone and this leg is just beginning to go, so I'm hoping that when I go into the wheelchair I'll have use of the wheelchair, manual wheelchair for years. I'm hoping I will. And I'll be able to get in and out of the car because I still have plenty of upper body strength. Now, you know if I end up at Christmas having to get an electric wheelchair, that'll be hard to take."* (David, participant #30)

Of huge importance to participants was their need to be *the* central figure within *their* world of healthcare services. Participants indicated they would complain if they felt the care they received failed to meet with their expectations. Some participants also believed that they would disengage from healthcare professionals if they felt their preferences for care were ignored by healthcare professionals. Eilish (participant #19) remarked:

> *"Well I wouldn't go back to them (laugh). I definitely wouldn't go back to them (laugh). I'd say no I'm not coming back and that's it."*

Exerting control in healthcare services also assisted participants in their struggle to remain in the present and/or move at their own pace towards acceptance. As described in Chapter Four, some participants attempted to station themselves in the present as they struggled (unsuccessfully) to maintain normality. Correspondingly, participants exerted control in their dealings with healthcare professionals so that they might somehow "live for the moment". Maureen (participant #34) who was increasingly dysphagic disclosed why she opted for the moment against a gastrostomy feed offered by her consultant neurologist:

> *"I told her I lost weight and she [consultant neurologist] asked me did I want a..." (P#34 pointing to her abdomen) G: "A feed through your tummy." P#34: "Yeah and I said no (....) Oh I don't want one until I feel the need. I don't know want to know an awful lot*

about motor neurone (....) I take each day as it comes. I don't want to let my mind run away."

Participants expressed a strong desire to be in control of end-of-life care and the fear of losing their ability to communicate their preferences during end-of-life care encouraged participants to make their preferences for end-of-life care known. The majority of participants (n=27) spoke openly about dying from ALS and all of these participants felt that people with ALS have the right to sustain or end their life on their own terms. Pascal (participant #23) remarked:

"When you get to a stage where the only thing you can move is your eyelid you wonder what, whether you are kind of just existing. I know your brain is alert and you feel, you know you've got your sense of touch (....) [but] if I got to the stage where I was just existing I would be leaning towards going to Switzerland [for assisted suicide]."

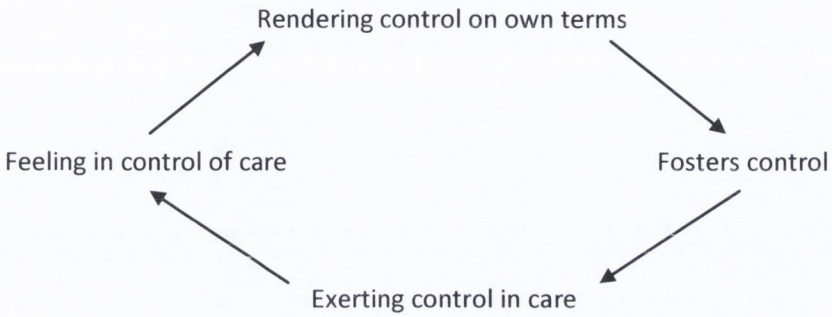
Participants sensed that control inevitably shifted away from them and towards service providers because service users rendered more control to service providers as they advanced in the condition. However, having the time to "process" loss also fostered feelings of control among participants. As Eve (participant #7) shared:

"I suppose I'm good at, you know I'm processing it [ALS], I know its there and when I'm ready I'll have no problem with it (....) I think we're all a bit like that, you process stuff internally for quite a while."

This study found that rendering control to service providers also enabled participants to feel in control of their care because exerting control in healthcare services included the freedom to relinquish control on their terms. Figure 4 (p.112) illustrates how rendering control to healthcare professionals on service users' own terms fosters control among people with ALS. Danielle (participant #9) described how she exerted control in her interactions with service providers *and* remained in control of her care by accepting help on her terms:

"I could probably go on indefinitely and not go back to the [ALS] clinic (....) I'm going to avoid going back for as long as I can, at some point I probably do need to go back (....) that is my choice."

Figure 4 Exerting and rendering control over healthcare services



Most of all, participants were uncertain about how healthcare services would meet their needs as they encountered more loss. In turn, participants exerted control in their interactions with service providers to alleviate their uncertainty about what healthcare services could offer them. Martin (participant #31) explained how exerting control in his interactions with service providers made him feel more reassured about his future with ALS:

"I said to @@the consultant neurologist at the national ALS clinic##, look I said, I know that you pride yourself on the service that you personally and your team give to MND people. But to be perfectly honest, I'm not seeing it. I said last time, I didn't see you, I was here three months ago, I didn't see you I said, I didn't have a chance to really to discuss my condition, I'm kind of apprehensive about a lot of things (....) so she said three months was too long (....) she said we will organise a conference with the people up in the primary care team in @@local town## (....) So @@ALS clinic specialist nurse## and @@IMNDA specialist nurse## from the Motor Neurone Association arrived here and they met the occupational therapist, the physiotherapist and the local nurses here. And they discussed me and my condition and then they brought @@daughter## and myself in and they discussed with me what they had been saying about me (....) I'm very happy with all that now."

The following sections of this chapter describe why participants felt uncertain about healthcare services and how they turned towards service providers and family to (as described in Chapter Four) *become more certain* and cope with the prospect of future loss.

5.4 Feeling uncertain about healthcare services

In addition to feeling uncertain about ALS (as described in Chapter Four), participants were also unsure about their healthcare services. Most participants felt unsure about how to access healthcare services and few participants knew much about their entitlements to healthcare services. In the early stages of their disease, some participants also felt disconnected from healthcare services as they encountered more loss:

> *"Well I think probably the biggest thing is not knowing what I need and what I'm entitled to (....) As the progression gets worse obviously not realising or not knowing where to go to get things, not realising what you are entitled to, what you need, em it's quite traumatic you know."* (Pascal, participant #23)

> *"I was starting to get a feeling of like I couldn't lift, this arm was coming to the point I couldn't lift it and I couldn't lift the kettle and my breathing was getting slower, not as good, in that three month period I felt a complete disconnect. I hadn't got, the support services were doing nothing for me really (....) I didn't know what would happen if I, say for instance if I became unwell."* (Martin, participant #31)

Participants' uncertainty about healthcare services ranged from uncertainty about how to access services beyond the diagnostic phase to uncertainty about what services are available at different stages of the disease, to uncertainty about what services are rendered by different healthcare professions:

> *"I'm not sure who does what, like I know the physio[therapist] is physio, the OT [occupational therapist] is for the house, the public health nurse introduced herself but I don't know whether maybe there's something they could help me with that I'm not aware of."* (Helen, participant #12)

Some participants were also perplexed why some service providers prescribed specific treatments to them which others had not recommended. Cara (participant #32)

questioned why she had been prescribed Riluzole (Rilutek) in the context that Riluzole would more than likely have no tangible impact on her life:

> *"He [consultant neurologist] said I could put you on Rilutek but he says it's no good, I'm not going to put you on it, and then I went to @@the consultant neurologist at the national ALS clinic## and she put me on it, that confused me (....) From reading the leaflet, it says it, what they think there's too much glutamate being released from the brain and it [Rilutek] slows down the release of it but I don't know, its not clear. It can make you live what, three months longer, is that not prolonging the agony (humour)?"*

Some participants were also deeply frustrated by some healthcare professionals' lack of expertise in ALS. Healthcare professionals' lack of expertise raised participants' anxiety about their future and diminished participants' confidence in healthcare professionals:

> *"Well they [neuro-medical team] took out a nerve to analyse it to see was my body attacking the nerves, to see was it an autoimmune type problem and the results were negative, first test was negative, the second test and then the third test was negative (....) At this stage we were about a year and a half into it and I went back to @@consultant neurologist##. He said look, I don't know what's wrong with you but I will recommend somebody else but at that stage I'd lost confidence in the whole fucking thing."* (David, participant #30)

However, as participants encountered mounting loss, they *searched for certainty through healthcare services* by engaging with healthcare professionals to ease their fears about their mounting losses. The following sections describe how participants engaged with external supports to adapt to loss.

5.5 Searching for certainty to adapt to insurmountable loss

As described in Chapter Four, uncertainty about the future with ALS engendered feelings of loss (including loss of control) for participants. In response, participants interacted with two primary external supports: healthcare services and family, in order to alleviate their fears about the future and adapt to loss. Participants' uncertainty about healthcare services was a dimension of the concept *uncertainty*. However, participants' uncertainty

about healthcare services and about ALS led them to "connect" with healthcare professionals in their quest to become more certain about their future.

Six primary categories (and the relationships between them) shaped how certain or how uncertain participants felt as they encountered unremitting loss: trust, reassurance, being cared for, healthcare services, family, and parenthood. Having *trust* in service providers *reassured* participants. Feeling trusting of healthcare professionals and feeling reassured by healthcare professionals and by family alleviated participants' fears about the future. Placing trust in services and feeling reassured by services and by family also engendered confidence among participants that they would *be cared for* as they advanced in their condition. *Being a parent* was primary context to how and why participants felt reassured by family and/or in some cases, why they felt concerned about family. Feeling reassured as a parent alleviated participants' uncertainty about the future but their concern for their children also heightened their own anxiety about the future with ALS.

5.5.1 Trust: An essential component of care for people with ALS as they endure loss

Participants indicated that they needed to trust in healthcare professionals to "get through" ALS. Participants had a strong desire to place trust in healthcare professionals at all stages of the disease but varied in how much they trusted healthcare professionals. In some cases, participants spoke of mistrust. David (participant #30) disclosed:

> *"I was going down to [private consultant neurophysiologist] in [regional hospital] and he wasn't saying anything to me, he was making a pile of money (....) In fairness I thought this is fucking bullshit, nobody knows what's wrong with me and it's going on and on (....) The whole thing was disjointed and the worst of it, I was being kept in the dark."*

Participants' trust in healthcare services centred primarily on trusting that their diagnosis was correct, trusting that services would continue to be available to them, and trusting that the terminal phase of their care would be managed with dignity. Eve (participant #7) shared:

> *"@@regional hospice## is our big huge hospice here and quite recently they moved from their old premises to state of the art premises (....) I've a very good friend and she*

happens to be in the hospice, she's actually in her final stages of cancer (....) I went to see the hospice and it was a very good experience from my point of view knowing down the line that I will be using this service and it was good for me to see the brand new premises, to experience people who are, you know in a caring role looking after the patients that were there."

Participants placed trust in healthcare professionals who demonstrated expertise in ALS care. Indeed, as participants advanced in the disease and encountered more loss, they sought to engage with healthcare professionals who specialised in the field of ALS care. Pascal (participant #23) remarked:

"When you read and see what @@the consultant neurologist at the national ALS clinic## is doing, you very quickly realise that she's at the top end of her field so why would you need to go somewhere else."

However, professionals' expertise in ALS care did not alone fortify participants' trust in service providers. Participants were also trusting of healthcare professionals who were attuned to how participants exerted control in healthcare services to adapt to loss. Richard (participant #8) disclosed, with visible emotion:

"I was in a lot of trouble [difficulty mobilising] until she [primary care occupational therapist] brought me the wheelchair even though I rejected it at the start because I felt I didn't need that kind of thing (....) of course I hid it in the shed outside for a while (....) I just hated the thought of being wheeled in that (crying) but I've no bother now (....) It's a process to get to that, that's the process you have to do yourself. I thought that end of it was very professional the way it was left to me and it wasn't forced on me (....) I trusted her then."

Participants trusted service providers who engaged openly with them and conveyed empathy in the clinical encounter. Danielle (participant #9) described those service providers who she most trusted:

"People who are interested in me, people that see the individual and not the disease. Some people see the disease first and don't get beyond that. I think what makes, what works for me is somebody who sees the individual first."

Many participants expected healthcare professionals to be trustworthy. However, even though participants developed trusting relationships with healthcare professionals as they advanced in their disease, some participants also questioned the motives of some service providers. Participants' mistrust of service providers featured mostly in their accounts of engaging with private healthcare practitioners:

"I went to see this private physiotherapist and he couldn't detect anything, he gave me a few sessions, the usual, they'll all treat you for three weeks and send you off then for a month when they can't do anything for you so that you won't go with anyone else in the meantime." (Richard, participant #8)

"It's gone from a time when now they [consultants in private practice] see four patients in a hour where as before it's used to be one. That's six hundred euros an hour (....) You go to your GP now and the first thing is they take a pad out and prescribe and you pay for it." (Morris, participant #10)

In particular, participants were mistrusting of pharmaceutical companies. Participants questioned the sincerity of the pharmaceutical industry's efforts in the quest for a cure. Some participants suggested that given the low prevalence of ALS in comparison to other life-limiting and/or disabling diseases, the costs incurred by the pharmaceutical industry in the development of neuro-protective treatments for people with ALS might extend beyond what they would recoup by supplying disease-modifying treatments to healthcare. Eve (participant #7) disclosed with anger:

"I suppose I can be cross [angry], the fact that it's down to money. The big pharmaceutical companies won't look into the disease because when you've got only a very small percentage of the population getting motor neurone disease, it doesn't pay them to investigate and research it."

Feeling trusting of healthcare services enabled participants to develop lasting relationships with healthcare professionals. Placing trust in service providers also enabled participants to feel more reassured about their future care than they might otherwise have felt had they not trusted service providers. Terry (participant #11) shared:

> *"I found from the last trip or two up to the clinic that I have begun to trust them. I expressed that my legs were gone bad and I went in waiting for them to put a plan of action in for walking aids or stuff like that, and they said no, you don't need that yet (....) that they feel that it's not, we're not there yet."*

Overall, feeling trusting of service providers reassured participants but when participants mistrusted healthcare professionals, healthcare professionals failed to reassure them. The next section explains why feeling reassured and/or being reassured is important for people with ALS as they adapt to insurmountable loss.

5.5.2 Reassurance in ALS: Needing reassurance to live through loss

Fear about the future with ALS permeates the vast majority of interviews with participants in this study and participants' assessment of ALS as a "feared" disease surfaced early in the data. As described in Chapter Six, participants feared for the future of the Irish healthcare system. However, participants' fears extended beyond their concerns about the future of the healthcare system. Participants also feared what was to come with ALS, feared the prospect of future suffering, and perhaps most of all, feared that they would not be in control of their services as they advanced in the disease. In their effort to ease their fears about the future with ALS, participants sought reassurance from healthcare services and from family.

5.5.2.1 Seeking reassurance from and feeling reassured by service providers

Feeling reassured by healthcare services alleviated (if not removed) participants' fears about the future with ALS. For some participants, feeling reassured by healthcare services enabled them to find at least some relief despite mounting loss. As Jack (participant #26) shared:

> *"But eh it's the time I accept it [ALS] most now is when I go to bed at night and I'm lying awake before I eventually fall off to sleep, that's when I say to myself you are on your*

own this is yours, it's me that has this motor neurons (....) Accepting it is hard but then what makes it an awful lot easier is every time you talk to somebody let it be in Beaumont[3] *or wherever they seem to always have an answer (....) you'd probably be stuck in an asylum [without support]."*

Reassurance as a category comprised multiple properties and dimensions and there was variation across the sample in terms of how reassured participants felt about their healthcare services. For some participants, reassurance was an intangible concept because of the uncertainty in ALS:

"Eh but this [reassurance] is not a tangible thing because you can't tell me how long I'll survive with this, so these are questions nobody can answer." (Melanie, participant #27, end-stage ALS)

Nonetheless, participants sought reassurance from and felt reassured by service providers along a number of dimensions. Participants felt reassured when they received information on request, when they were guaranteed that services would be available to them on request, and when they in fact secured the services they requested. Most of all participants wanted healthcare professionals to "reach out" to them. Participants needed to feel "connected" with services as they encountered more loss:

"They probably need to sort of reach out and go okay you actually do need to come back, as good and all as you are dealing with this, we actually do want to see how you are doing and see if there's anything else we can do to help." (Danielle, participant #9)

"At the moment I am, I am reassured (....) I felt before I wasn't part of the whole. I felt I was kind of like floating on my own whereas now, I am connected and it's a team effort." (Martin, participant #31)

Despite participants' uncertainty about the future of the healthcare system, the majority of participants felt reassured that they would be cared for by service providers as they lived through insurmountable loss. Feeling reassured by healthcare professionals despite

[3] The national ALS clinic at Beaumont Hospital is not anonymised. The majority of people with ALS in the Republic of Ireland (i.e. 80%) attend the national ALS clinic.

feeling concerned about the future of the healthcare system is an important finding in this study. Participants expected a "human" response from service providers and the degree to which participants trusted and felt reassured by service providers was shaped more by their "personal" interaction with service providers than by (as described in Chapter Six) broader (i.e. macro) contexts to their services. Participants felt reassured when they felt listened to by healthcare professionals and when healthcare professionals themselves cared to understand the service user perspective. Participants engaged comfortably with service providers when they felt service providers engaged at a personal level with them. Helen (participant #12) explained how a "personal" interaction with a healthcare professional enabled her to cope with mounting loss:

> *"She [consultant neurologist] gave me the time. She didn't rush me (....) She'll chat, how are the kids [children], how are they getting on with it. I come out of there feeling better because she reassures me that yes you are doing well. Now three days later I might go ooooh (sigh), but I'd wait just for that."*

Participants also shared experiences about engaging with healthcare professionals who were insensitive to their needs. Engaging with insensitive healthcare professionals offered little reassurance to participants that they could rely on such people to assist them as they encountered more loss. Cathal (participant #28) spoke about his experience of a physician who was insensitive to his needs:

> *"He [gastroenterologist] wasn't a very nice man, he hadn't a very nice bedside manner (....) For this guy to sit down with one leg across the other and start on about motor neurone and asking what was I doing in the hospice, how come, is there no disability places for young people in @@residing county##, you know to me it came across as a person that hadn't got a clue."*

Participants felt reassured by healthcare services' support of research to find a cure for ALS if not during their lifetime, then for future generations. Terry (participant #11) recognised that his participation in a clinical trial offered him little hope of survival but he felt reassured that his participation would in the end be of benefit to others:

"It'll never be a waste of time because at least at the end of the eighteen months or whatever they'll turn around and say well that didn't work, but at least now they know it didn't work, they can try and find another fix."

Participants' fear about the end stage of ALS was tangible in interviews and their desire "not to suffer" at the end stage of life permeates most interviews. Participants sought reassurance from healthcare professionals that all efforts would be made by them to relieve any suffering that participants would encounter during the dying phase. Primary neuropathic pain is not a feature of ALS but participants shared concerns about the prospect of pain in the very final stages of ALS. Participants' primary expectation of healthcare professionals was that they would alleviate suffering at the end stage of ALS:

"I'd have a fear about, you know will this hurt me when I'm dying, you know the final stage of the illness, people do die from it, that I just want to make sure that the services are going to be there and that any pain that's to be had, that that's all looked after." (Eve, participant #7)

As participants engaged with service providers to feel reassured, they also sought reassurance from family about the future. However, seeking reassurance from family was perhaps somewhat more complex than seeking reassurance from service providers because (as described in the following section), in order to feel reassured by family, participants also has a need to reassure their family.

5.5.2.2 Seeking reassurance through family: Feeling reassured by family and needing to reassure family

Family as a category emerged from how participants attached meaning to life as a parent, grandparent, spouse, sibling, and partner. Family (or the absence of family) was primary context to what types of decisions participants made about their care and why they made the decisions they made about their care. This study explored variation in family context for ALS service users and saturated the category 'family' in terms of its properties and dimensions. In doing so, this study found that people with ALS feel reassured when family reassure them *and when they feel they reassure family*. In this light, participants' interaction with healthcare services was shaped not only by the role of family members in their lives but also by *their* role in the lives of their family.

The majority of participants (n=24) lived at home with their spouse (partner) and all but two of these participants required assistance from either their spouse (partner) or other family members. Some participants who had never married and who had no dependants lived with their sibling (n=2) or resided in care (n=1). The majority of participants who lived alone and who had no dependants suspected they might move to residential care if they failed to secure sufficient homecare services. Those who had never married and who had no dependants felt they were more reliant on healthcare services than those who had a significant other and/or adult dependants. Overall, participants relied on family to supplement the care they received from public healthcare services. Danielle (participant #9) disclosed:

> *"I have two carers that come in the morning, helping me out of bed and to shower but I depend on my sister at night time who helps me get to bed at night before I wait till somebody gets me up the following morning."*

As discussed in Chapter Three, for some participants, their significant other was to all intents and purposes their gatekeeper. In this context, it was perhaps not surprising to find that *family* shaped how participants engaged with healthcare services and made decisions about care. Participants were keen to know how ALS would progress for the "sake of the family" and the majority of participants included their family in decisions about care. In particular, those who had developed dysarthria or anarthria, relied on family members to voice their concerns on their behalf. Morris (participant #10) and his spouse explained why it had become so important that she (his carer) advocate on his behalf:

> *P#10 Spouse: "I would say I did most of the sourcing and finding out and all of that." P#10: "Self-protection" (laughing). P#10 Spouse: "Oh self-protection for me, yes self protection for me (laughing). Well I just feel that somebody who is sick, @@Morris## can't make phone calls and he can't do that sort of thing so somebody has to do it, somebody has to be his advocate." P#10: "I would have done the same but I'm the one losing speech."*

Family was context to how and why participants opted for or declined services. Those who had no spouse (or partner) and/or adult children required more physical assistance from

service providers than those who had a spouse (or partner) and/or adult children. Participants who had immediate family also felt less reliant on service providers than those with no immediate family. Participants felt reassured when they made decisions about care with their family and when service providers sought to include family members in decision making about care:

> *"She [ALS specialist nurse] asked me a couple of months ago did I feel I should get what do you call it, a homecare assistant but I haven't done that (....) I don't want one at the moment because my husband does what I can't do (....) He is here and I don't think he gets anxious."* (Ann, participant #24, end-stage ALS)

> *"I am more relieved for my husband, because he came in with me to her [consultant neurologist] and he was absolutely a different person after that visit because obviously he would worry, probably more than I would. I always think it's the other person who has to put up with an ill person, goes through more."* (Cara, participant #32)

It was important for participants that their family members endorsed the decisions participants made about care as they and their family encountered more loss. Some participants paced services in line with how accepting their significant other was of change. Samantha (participant #22) spoke about how she delayed homecare services until her spouse had come to terms with the need for such services:

> *"I felt I needed to wait for him [spouse]. He was very reluctant to have anybody into our home and he's been doing all the housework and the garden because we had always done it together you see (....) but he is getting there slowly and so now we have home help which started last week."*

Attention to the wellbeing of loved ones often restricted participants in making the decisions they themselves wished to make about their care. Martin's (participant #31) preference was to move to a nursing home so as to alleviate the burden of care on family. However, such a move would have devastated his ailing spouse who had always relied on him:

"I thought it may have been the right decision for me. If I were living on my own I would have said yes, it would have made a lot of sense to me and the very fact that @@Emily## [daughter] will have to look after both of us and @@Pam## [spouse] is not hugely mobile either (....) But I, I can't do this, if I said that to @@Pam## it'd kill her."

Indeed, participants who had no spouse or dependants suggested that they had more freedom to make decisions about end-of-life care than those who had family. Jennifer (participant #25) explained why she refused Rilutek and (despite onset of dysphagia) opted against a gastrostomy feed:

"You see I don't feel I have the need to hang on at any cost. If I had a husband, wife, children, it might be different so I'm free to make the choice."

Needing family to endorse decisions about care extended beyond decisions about homecare services or about place of future care. Participants were also eager to involve family in making decisions about advance directives and some were keen to know whether or not their family would support them should they at some point in the future, wish to end their life. Terry (participant #11) shared:

"My biggest fear you know [is] around being totally debilitated (....) I constantly crave to be, to be told by the people that matter most to me that you won't be left like this for years and years and years you know (....) I suppose I'm talking euthanasia stuff like that."

However, Morris (participant #10) who expressed the desire to take his own life, and Helen (participant #12) who had considered travelling to Switzerland for assisted suicide soon after her diagnosis, indicated that they remained deterred from ending their life because they believed such action would devastate their family. Helen explained:

"I had to consider my family and the implications, it's like a suicide, it's like a suicide that you know, the pebble in the water, it spreads out, it affects so many people and I suppose a lot of family like with, with suicide, you know, they get angry that you were selfish, you know, why did you do it (....) there are serious implications for them because

they want you for just another day, and of course you want them but you know I mean if push came to shove now I really don't think I could do it to be honest with you."

Participants' decisions about life-sustaining treatments were, for the most part, shaped by how participants themselves struggled to find a balance between drawing support from and providing support to loved ones. As described in Chapter Three, some participants had already engaged with gastrostomy (i.e. radiologically inserted gastrostomy or percutaneous endoscopic gastrostomy) and non-invasive positive pressure ventilation (NIPPV). Other participants were likely to engage with these interventions in the near future. Maureen (participant #34) admitted that non-invasive ventilation provoked anxiety for her. However, she felt obliged to engage with non-invasive ventilation so that *she* could be of support to her spouse:

"I want to be here for as long as I can yeah." G: "So then @@Maureen## you might choose them [life-sustaining interventions] to help you stay alive?" P#34: "Oh yeah but without being too much of a burden on @@Fred## [spouse]." G: "So is it nearly a balance then. You want to continue to live on with MND but you also don't want to be a burden on your family. I'm just trying to figure out, I suppose my question is what is the point where you think it becomes too much of a burden?" P#34: "Well I say this to @@Fred## but at the moment he tells me that he wants to hold onto me! (humour)"

As described in Section 5.2.1, few participants thought that invasive (i.e. long-term) ventilation was a viable option in ALS care. Participants indicated that invasive ventilation would increase burden of care on family. Sally (participant #20) explained why she would never opt for invasive ventilation:

"I just saw my mother suffer, she was told at the time that she had creeping paralysis and there was nothing that could be done, just go home that was all (....) So I'd love to just go [die] quick. I would hate to have to depend on everyone having to do everything for me (....) all the pain and anguish and the anger and everything that I went through, and I'd hate to inflict that on them [family]. It would be unfair to them really."

Indeed, feeling a burden on family featured strongly throughout participants' personal accounts of their experiences and in this study, "burden" of care fell predominantly on

spouse or partner, on adult children, and on siblings. For participants, "burden" of care represented the ever-increasing physical and psychological strain placed upon family as participants encountered more loss. Participants were reluctant to place burden on their family but were also resigned to the fact that they would become dependent on their family:

> *"My daughter is saying that she'll give up work and she'll move in as a carer. But then she's trying to get pregnant, so, and I keep saying to live your own life, the time will come when I do need you, really need you yeah it'll all kick off you know."* (Helen, participant #12)

However, participants' families did not always reassure them and in some cases, troubled relationships between participants and their significant other prior to the onset of ALS, deteriorated even further. David (participant #30) spoke about how conflict between him and his spouse constrained him in the decisions he wished to make about his care:

> *"I'm finding it very difficult at home. My wife doesn't seem to have any sympathy for me, empathy or whatever and we're looking at building, obviously we don't have the money but we could build just a toilet maybe you know, facilities for maybe eight, nine, ten grand [thousand]. I'm thinking, you know if we had a bedroom as well, you know, it would be, might cost twenty grand but at least we'll have, I'll have my own little bit of independence and what have you. So that's trouble now at the minute."*

Analysis of participants' personal accounts of their experiences suggests that participants required more assistance from family as they (participants) encountered more loss. Having family who were willing to provide physical care reassured participants and for some participants, having family increased the likelihood that they would remain at home during the dying phase:

> *"I accept that we must all die. I have to accept what is coming (....) I prefer to be at home and to have someone near me at night, my family, @@Annette## [spouse], my son and daughters (....) I don't want to be alone."* (Edward, participant #14)

Although many participants spoke about their fears of becoming a burden or indeed being a burden on their family, the data also suggest that participants struggled to live on in order to "be there" for their family. As described in Chapter Four, participants struggled between wanting to live on with ALS and resigning to the inevitability of ALS. Conflicting feelings between "letting go" of family and wanting to remain with family was a dimension to the struggle between "holding on" and "letting go". As described in the next section of this chapter, participants' struggle between holding onto family and letting go of family was complex.

5.5.2.2.1 Feeling reassured and struggling to be reassured: Extending the concept of reassurance to parenthood

As described in Chapter Four, older participants were more accepting of ALS than were young and middle-aged participants because older participants felt they had fulfilled their ambitions and had the opportunity to live a long life. Living a long life to fulfil one's ambitions centred primarily on the ability to raise a family and the extent to which participants were reassured by family (or felt concerned about their family) was influenced predominantly by *being a parent* at different life stages. Indeed, *being a parent* was principal context to how the majority of participants who had children made decisions about their care and adapted to insurmountable loss.

This study found that older participants were more accepting of death than young and middle-aged participants because they had already reared children to a point where they were now self-sufficient. Having children who were no longer dependent on them alleviated participants' concerns about the future:

> *"I retired when I was fifty-five and if I had been diagnosed at fifty-five it would have been a totally different ball game. It would have killed me, it would have destroyed me because like as you see @@Emily## [daughter] is forty-two, her sister's forty-four, they have their families. I have no kind of responsibilities towards them. If I had young children, this would be the most devastating thing that would have ever happened, the fact that I may not have been there for them."* (Martin, participant #31, age 73)

Indeed, some participants without children suggested that ALS would be more devastating for people with ALS who have young children and all participants with young children suggested that ALS might have been less devastating for them had they not had children:

> *"People hear that I have MND and they say how awful and I think well I've had a pretty healthy life, I'm getting old. So I mean it's tough but there are a lot of worse things. I don't have young children. It would be quite different for someone with young children".* (Jennifer, participant #25, age 73)

> *"I do think it would be easier to stomach if you were like a single person with no children that have to go through this bloody thing with you. I think it'd have to be easier, yeah without a doubt, without a doubt. It's more so because they're so young, you know."* (Terry, participant #11, age 45)

Being a parent was a dominating feature in the lives of those who had children. Participants suggested that parenthood had the potential to foster feelings of hope and energised some participants to resist ALS "for the sake of the children". Tim (participant #16, age 47) explained with visible emotion why he was adamant to engage with available life-sustaining and supportive care to enable him to parent:

> *"I have a wonderful relationship with my children, we're very close. If they were adults, they would probably deal with it better, so I'm kind of hanging on for that if I can. It's all for them and that's what brings the tears."*

The meaning participants attached to parenting with ALS differed depending on participants' life stage. Overall, participants with young children were more devastated by ALS in comparison to older participants who had already raised their family. Losing out on parenthood had a profound effect on participants with young dependants and all participants who had young dependants felt overwhelmed by the prospect that they would lose out on raising their children. Apt was Eve's (participant #7, age 45) response to the prospect of no cure for ALS in her life time:

"At the beginning obviously I've cried every day for months (crying). Look my response to that is I'm young, as a parent, as a mother, you worry about your children and that's my worry (crying)."

The prospect of "leaving children behind" was also distressing for middle-aged participants with dependants because many of their children still relied heavily on them. Andrew (participant #6, age 64) remarked with humour:

"You see I don't want to die anyway, because there's @@Kara## the daughter, she has four [children], she's four, my son has, he has two [children], but I mean @@Kara## can't stay away from [parental] home, you know and we see them as children even sure bloody hell, they are thirty, nearly forty odd years of age now."

Not only did participants' duty towards their children (at different life stages) shape how they engaged with their children, parenting at different life stages also shaped the decisions they made about care. Young and middle-aged participants' desire to live on for their children was strong and many felt obliged to avail of life-sustaining treatments so that they could continue to "be there" for their children. Maureen (participant #34, age 62) explained why she felt she had no choice but to accept non-invasive ventilation:

"I have a grandson living with me, he is twelve and his mam [mother] has a drug addiction. We have an agreement between her and me that we [participant #34 and spouse] are sort of guardians and he [grandson] keeps me going (....) I have a son, he's an alcoholic and he has a condition called @@congenital disorder##. His legs and one of his arms is kind of paralysed. I am afraid of him drinking and falling. So I need to be around for as long as I can be for them."

Participants with young children were very much attuned to the "devastating" impact ALS had on their children and all participants with young and adolescent children opted for, declined, and paced services in line with how their children responded to the presence of healthcare services in their lives:

"During this summer just gone by I didn't want to be making calls to physio[therapy], occupational therapy, district nurse, I wanted a holiday from the disease myself. Well

you know I didn't need it and I just felt that once the kids [children] are back at school sure I can make a phone call again, I'll wait until the holidays are over." G: "So as you said wanting a break from the whole thing nearly." P#7: "Yeah." G: "Is that to do with the kids [children] do you think?" P#7: "Definitely." (Eve, participant #7, age 45)

As described in Section 5.2, participants revealed a strong desire to engage with research for the benefit of others. Participants' decision to engage in medical research (including research on the genetics of ALS) was also for the benefit of their children:

"Down the line all the information that's gathered it may be helpful to somebody. Now it mightn't be in my time but I would hope that, you know, later on (tearful) (....) I'd like to give any help you could you know, because maybe your children might be affected or your grandchildren, you know what I mean because as I say my mother had, had a neurological disease and there was nothing that could be done." (Sally, participant #20)

The majority of young and middle-aged participants wished to live on to support their children but living on with ALS also prompted participants to consider the impact of life-sustaining treatments and support services on their children. As described in Section 5.5.2.2, participants were, for the most part, reliant on their family. However, participants with children were also keen to minimise the painful impact that ALS had on their children. Peter (participant #4, age 69) explained why he wished to "expire in solitude" without any form of life-sustaining intervention:

"I think you know if you try to do something that you can't do and you fall on your face in philosophical terms, I think it's much more distressing for those around you. I suspect it is anyway, because then they're looking out to see if you're going to tumble over or whatever so it puts an awful lot of unnecessary distress on them really."

Overall, participants with children wished to live on with ALS but they were also mindful of the "devastation" ALS had already inflicted on their children. In this context, many participants with children struggled between living on with ALS and hoping for death prior to losing their independence:

"[I] just want to get them [children] across the line (....) I've three of them, my eldest is eighteen, my youngest is, he's nearly ten, there's a bit of rearing on him left to be done (....) I also know I don't want them being so young seeing me quite ill you know (....) so it would be peace of mind for me to think that when this thing goes so far that it'll be stopped." (Terry, participant #11, age 45)

Sections 5.5.2.2 and 5.5.2.2.1 have explained how and why participants searched for reassurance through family and felt reassured by family as they engaged with healthcare services. The following section describes how feeling *cared for* by family and by healthcare services rendered life "more bearable" for participants.

5.6 Coming to terms with loss: "Being cared for" by family and by healthcare services

Feeling trusting of service providers and feeling reassured by service providers and by family, instilled confidence in participants that they would be cared for as they advanced in the disease. As Jack (participant #26) explained, "being cared for" by healthcare services made the experience of engaging with services better than had he not felt cared for by healthcare services:

"Sometimes I think before people really knew what this disease was and when there was no one around to tell people what they had, what to do or how to go about doing things, sure it would drive you mental."

As already described, the experience of living with ALS is primarily one of loss and people with ALS exert control over service providers in the face of mounting loss. However, this study also found that participants' need to feel cared for by healthcare services and by family increased as they (participants) encountered more loss:

"I have now got to the stage you know the steps of being independent are further away (....) [but] I mean one is very lucky if one gets the back-up that I've got from the services (....) As my mobility declines I can ask them what to do and they come up with answers." (Jennifer, participant #25)

"I mean there are days when I don't feel like living (....) So important is the quality of the people who are doing the caring, my daughter-in-law and the various doctors and the local people [healthcare professionals]." (Melanie, participant #27, end-stage ALS)

Indeed, even participants who valued efficiency most sensed that their need to be cared for would increase in the face of mounting loss. Morris (participant #10) disclosed:

"Caring alone is not enough (....) they've [healthcare professionals] to be caring and efficient (....) a carer who's not efficient, shouldn't be a carer unless you are a lost cause, maybe efficiency is not so important then (....) I think efficiency and care, they are probably intertwined so much you can't separate them (....) but it comes to the stage I'd say where caring predominates everything."

The meaning of *being cared for* centred primarily on how participants "connected" with family and with healthcare services. Participants' relationships with family were different to participants' relationships with service providers. Relationships with family were long-standing and most participants "connected" with family. As Tim (participant #16) shared:

"I have a wonderful wife and wonderful family support around me, wonderful. That makes my life somehow okay (....) I could probably see where you could die quickly without it (....) I'm blessed with the support I have."

Participants' relationships with service providers were, for the most part, new relationships because the majority of participants had not had prior need to engage with a broad range of healthcare services. As described in Section 5.4, some participants felt disconnected from healthcare services. However, participants also endorsed the presence of service providers in their lives when they "connected" with service providers and when they felt that service providers genuinely cared for them. Ultimately, "being cared for" by healthcare services enabled participants to endure loss. Cathal (participant #28) explained:

"If I'm up at night in the hospice, if I can't sleep I'll go down to the smoking room. One of the members of staff will come up and sit with me and ask me what's wrong. Like I'd an awful bout of depression the last time I went in there. But I was up one night, about

two o'clock in the morning and one of the nurses came over and sat down with me, she asked me what was wrong. She made me comfortable and she reassured me."

Participants valued healthcare services when service providers were accessible to them and when they rendered services to them in a timely manner. However, participants felt most cared for when healthcare professionals demonstrated empathy in the clinical encounter. Helen (participant #12) described how her consultant neurologist helped her to come to terms with her diagnosis:

"She had time for me you know, she doesn't send her minions out to call you in, that's the way I looked at it. She comes out herself and she doesn't rush you, even if you ask her to repeat something again (....) I kind of blamed her for giving it [diagnosis] to me but she was honest with me (....) I know some people think the healthcare service is a thankless job but for me certainly it wouldn't be thankless, that despite all the crap that's going on, when somebody can still put a smile on their face and come out and just totally focus on you. That to me is very important."

Most of all, healthcare professionals and family rendered life "more bearable" for participants as they moved closer to death. Feeling looked after by family and by healthcare services enabled the majority of participants to adapt to insurmountable loss:

"For a long, long time I didn't accept it, I didn't because I was of the opinion that there would never be anything wrong with me. I thought I had everything covered in so much as you could (....) [but] I have had nothing but the best of help from everybody." (....) G: "That somehow perhaps makes you feel more accepting of having MND?" P#26: "Yeah, yeah." G: "But do you still think you are in the process of accepting or have you got there?" P#26: "I think I'm there already. I think I'm there now (tearful)." (Jack, participant #26)

5.7 Conclusion

This chapter has described how people with ALS interact with the external reality of care and support available to them as they adapt to insurmountable loss. All participants engaged with healthcare services as they adapted to loss. How participants engaged with

healthcare services to adapt to loss was shaped by how they engaged with and sought support from family.

The findings from this chapter both resonate with and differ from those of other studies that have explored perceptions of healthcare services among people with ALS. As described in Chapter Two, surveys on satisfaction with healthcare services among people with ALS found that people with ALS expressed dissatisfaction with healthcare services (e.g., Kristjanson et al., 2006; Ng et al., 2011; Van Teijlingen et al., 2001). Studies that have explored service users' perceptions of services in ALS care with the help of qualitative research methods (e.g., Brown et al., 2005; Hughes et al., 2005; O'Brien et al., 2012) also found that people with ALS were generally dissatisfied with healthcare services. Participants in this study were uncertain about what services were available to them (Hughes et al., 2005; O'Brien et al., 2012), and expressed dissatisfaction with delays in services and with service providers' lack of expertise in ALS care (e.g., Brown et al. 2005; Hughes et al., 2005; O'Brien et al., 2012). However, even though participants felt disconnected from healthcare services when they did not receive adequate information about ALS, similar to findings from Brown et al. (2005), the thesis also reports positive perceptions among people with ALS about healthcare professionals.

The findings are somewhat inconsistent with studies that have captured ALS service users' satisfaction with specific domains of care. Studies on ALS service users' satisfaction with assistive devices found that people with ALS expressed high levels of satisfaction with a broad range of assistive devices (e.g., Gruis et al., 2011; Ward et al., 2010). In this study, participants recognised the benefit in assistive devices but they felt despondent about having to use such devices because assistive devices reminded them of their loss. Using grounded theory method to saturate the meaning of loss for participants revealed that people with ALS might resist assistive devices even if they report satisfaction with assistive devices. The findings resonate with and differ from findings of studies which captured ALS service users' experiences of receiving a diagnosis (Belsh & Schiffman, 1996; McCluskey et al., 2004; O'Brien et al., 2011a). A number of participants in this study also expressed deep frustration with non-ALS specialist physicians' slowness to recognise ALS symptoms (e.g., general practitioners, consultant physicians who specialise in fields outside of neurology care, and consultant neurologists who do not specialise in ALS care). However, contrary to findings from the aforementioned studies, few participants in this study disclosed negative

experiences about the disclosure *per se* and no participant had been misdiagnosed by a consultant neurologist.

Overall, participants in this study conveyed more positive perceptions of healthcare professionals than participants in other studies have done. Even though participants felt uncertain about healthcare services, the majority trusted healthcare professionals. The majority of participants also felt reassured by healthcare professionals as they encountered mounting loss. As described in Chapter Six, the specific macro-contexts to ALS healthcare services in the Republic of Ireland might explain the difference in findings between studies. However, use of Grounded Theory method to identify *how* and *why* people with ALS engage with services might also explain the difference in findings. Grounded Theory method not only identified participants' overall views of healthcare services. Grounded Theory method enabled the researcher to unearth key concepts (e.g., trusting healthcare professionals, feeling reassured by healthcare professionals) which *explain* how participants adapted to loss. Participants' need to trust service providers, and their need to feel reassured by service providers resonates with findings from other studies that have investigated perceptions of healthcare services from the service user perspective. Nanton et al. (2009) found that empathy from healthcare professionals in the clinical encounter reassured people with cancer and alleviated their uncertainty about care. Woodward et al. (2004) found that service users' trust in service providers helped maintain continuity of care.

Similar to other terminally ill service users (e.g., cancer patients) (Sand et al., 2009; Volker & Wu, 2011) participants in this study exerted control over a broad range of decisions about their care. Participants' expectations surrounding control resonate with the findings of other studies in the ALS field. Sulmasy et al. (2007) also identified that remaining in control of care is important to people with ALS and O'Brien et al. (2012) found that service users' desire to maintain control delayed their use of services. However, not only was being in control of care important to participants in this study, feeling in control of healthcare services enabled participants to adapt to insurmountable loss. Exerting control over healthcare services *and* rendering control to service providers on their own terms fostered feelings of control among participants and assisted participants to come to terms with loss. *Feeling in control of healthcare services by rendering control to service providers on their own terms* is a key finding of this thesis.

As described, participants involved family in decisions about care and in some cases relied on family members to assist them in making decisions about care. As explained in Chapter Three, this study focused exclusively on people with ALS in order to identify the domains of care that are important to them. Numerous qualitative studies in ALS care have explored the carers' perspectives in ALS care (e.g., Aoun et al., 2012; Ray & Street, 2006) or sampled carers as well as service users in studies about service users' experiences of healthcare services (Brown et al., 2005; Hughes et al., 2005; O'Brien et al., 2012). However, few qualitative studies have identified how people with ALS (from their perspective) interact with family as they engage with services. This study found that participants felt more reassured about their future care than they might have felt had they not had family to support them in making decisions about care. The findings resonate with and differ from findings of other studies that report on the role of family in terminal illness (including ALS). Sulmasy et al. (2007) and Nolan et al. (2008) found that even though people with ALS opted for shared decision-making with family, some tended to make decisions about care independent of family. Hauke et al. (2011) found that family members of people with terminal cancer did not participate in decision-making about end-of-life care.

Participants' perceptions of being a burden on family resonate with that of other people with ALS (Hirano et al., 2006; Lemoignan & Ells, 2010; Young et al., 1994). Hirano et al. (2006) found that feeling reassured by family is important for people with ALS but people with ALS also consider the impact of life-sustaining interventions on their family. Feeling a burden on family (due to advancing disability) and seeking family involvement in care are not unique to people with ALS. Aldred et al. (2005) reported that people with advanced heart failure also expressed concern about how their increasing care needs impacted on family wellbeing. Janssen and MacLeod (2010) found that people with terminal cancer felt more reassured about their care when healthcare professionals included family in their care.

Researchers have reported on how people with life-limiting conditions experience illness in the context of parenthood (Bell & Ristovski-Slijepcevic, 2011; Duvdevany et al., 2008) but few studies have illuminated how people with terminal illness make decisions about their healthcare services in the context of being a parent. Hirano and Yamazaki (2010) found that people with ALS who chose long-term ventilation had a desire see their

children and grandchildren grow up. In this study, all young and middle-aged participants wanted to live on to support their adolescent and adult children and considered the potential impact of life-sustaining treatments (including the support services needed to sustain life) on their children. Parents of young children paced services in line with how their children responded to the presence of healthcare services in their lives and few participants expressed a desire to live on with ALS should ALS have an adverse effect on their children.

This chapter has explained how people with ALS engage with healthcare services and with family to adapt to insurmountable loss. Family (including the experience of parenthood) was primary context to how participants engaged with healthcare services. Chapter Six describes how the macro-context of health and social care services for people with ALS in the Republic of Ireland shaped how participants responded to their healthcare services and adapted to loss.

Chapter Six

The macro-context of healthcare experiences among people with ALS

"The thing is I think the service that you get when you go through the process of application and it's granted the service you get is really good. Anyone that I've dealt with, the physio[therapists], the OTs [occupational therapists] et cetera (....) they've come out to the house here, the CRC [Central Remedial Clinic] has been out to the house. I've been to see my physio a couple of times, em the OTs, I've spoke to the OTs from the district nurse that's in here, really nice people and very obliging and very helpful. That piece of it is not a problem it's the administration end of it (....) The part that I find frustrating is like understanding what you need and where you need to get it and how long is it going to take before you get it (....) A lot of stuff on the internet on the HSE website et cetera, it's pretty vague, you are reading it and you are going I'm still no clearer on what I actually need to do (....) And especially with MND you know as I said two months in the lifespan of someone who's got a fast progression rate is an eternity. You know some people last eight, nine, ten months, so from diagnosis to death so two months in that time frame is a long time." (Pascal, Participant #23)

6.1 Introduction: The national context of healthcare services for people with ALS

How people engage with healthcare services is shaped by the macro-context (i.e. systems) of healthcare services (Harwood & Clark, 2012). All participants in this research engaged with healthcare services and the majority (n=26) engaged with national-level services (i.e. the national ALS clinic). National services impacted on the delivery of local health and social care services to participants. However, shortcomings within the Irish healthcare *system* (e.g., diminishing resources, inequalities in access to services) meant that participants did not always receive care in line with their own preferences for care.

This chapter describes how the macro-context (i.e. local, national, and international policies and services) of health and social care services shaped the participants' experiences. The chapter highlights the ambiguities (and in some cases, inequity) in the Irish healthcare system which fostered uncertainty among participants. It pays close attention to the relationship between agency and structure (as explained in Chapter Three) and explains how local, national, and international contexts of ALS healthcare services both enabled and hindered participants' adaptation to loss.

6.2 The Health Service Executive (HSE): An "uncertain" place

For the most part, participants' perception of the Health Service Executive (HSE) was that the HSE was difficult to negotiate as a service user. Not only were participants uncertain about what healthcare professionals do and what they offer to people with ALS at different stages of the disease (as described in Chapter Five), participants also encountered difficulty sourcing information from the HSE about what services might (or might not) be available to them. Maureen (participant #34) commented:

> *"I don't know how I will go about [getting] a bus pass. I've tried to find out up there [local HSE office] if I'm entitled but still it's unclear."*

Participants' uncertainty about the HSE stands out in the data. The majority of participants were confused about why some services were provided directly by the HSE and why other services that were primarily funded by the HSE were not rendered by the HSE:

> *"I don't understand the whole, between MND support [voluntary sector] and between the health board [HSE] and where the money is coming from or who gives money for what (....) I need a wheelchair, I've been measured for a wheelchair (....) Is it the CRC [Central Remedial Clinic] or whatever? I said, well they're [Central Remedial Clinic] are kind of semiprivate as well, they are getting funding [from HSE] as well?"* (David, participant #30)

Participants' uncertainty about the HSE also comprised not knowing how long it would take the HSE to process their application for different services (e.g., home care services) or how the HSE might notify them if such services were available to them. As described in Chapter Five, the majority of participants sought services when they felt they needed them. However, some participants felt they needed to "chase" the HSE when they chose to engage with services:

> *"How to navigate the health services like and you know that can be a challenge actually. To actually get somebody to answer a phone is like, its must be the hardest thing in the world (....) What I find is the patient is always chasing (....) I'm supposed to be chasing them [health services] (....) I'm the one that has to keep tabs on it all (....) The*

problem is you can't get your question into the system because it's very hard to track people down." (Danielle, participant #9)

Some participants encountered difficulty in contacting primary care healthcare professionals when they required services in between scheduled contact and frequent change in front-line personnel gave rise to feelings of uncertainty among participants. Some participants also encountered delays or stoppages in their services because the HSE did not replace personnel who left their position or personnel who were on temporary leave from their position. Helen (participant #12, former HSE clerical officer) remarked:

"I only maybe saw her [public health nurse] once or twice then as, as typical in the health service, you ring up, she's gone and there's another one and then she's gone on maternity leave, and then there's no one else to replace. So it would be hard to pin down the one person. But having worked in the health service I know the way it works so I'm not complaining about it, it's just the way it is."

Participants expressed concern about the future of public healthcare services. As described in Chapter Five, participants were reassured by service providers when service providers guaranteed them that services would be provided to them and when service providers rendered services that were amenable to them. However, participants' attention to the downturn in the Irish economy also raised concerns among participants about the public healthcare sector's ability to deliver services for them. Apt were the remarks of Sally (participant #20) and David (participant #30) about the impact of the recession on the Irish healthcare sector:

"Well I hope that we all survive (laughing) but you'd be a bit worried definitely you know about the future of the health services. I could see an awful lot of changes but just I hope it keeps going somewhat (....) It's [recession] not looking good at the moment." (Sally)

"It's [recession] definitely going to affect it [services] and I'm going to need home help and definitely the State is gone. Obviously the country is broke, we're living away as we always did, people are saying fuck the banks, we're not going to pay off the banks, but we have to, like somebody has to pay otherwise we're going to go broke." (David)

A minority of participants (n=2) reported that they were subsidising home care services through private means at the time of the interview, but many participants suspected that they would soon have to use whatever private means they had to supplement public-sector home care services. Jennifer (participant #25) conceded:

> *"Well the HSE told me about private packages because they told me they couldn't fund much help." G: "How do you think you would fare, if you were totally dependent on the State?" P#25: "Ah! (laughing)." G: "You think you'd be struggling more?" P#25: "I would."*

Participants were sympathetic to the challenges encountered by front-line staff. However, they criticised the higher echelons of the HSE for creating a "maze" that participants struggled to negotiate their way through. Indeed, the majority of participants suggested that healthcare professionals did the best they could for people with ALS but were constrained by a lack of funding in the HSE:

> *"At the beginning I was at a bit of a loss to eh why there's not more being done and why people don't know more than they should (....) I don't think that's anyone's [healthcare professionals'] fault, I think it's the climate we live in, funding, I think everyone is doing their absolute best and people have great ideas. But I just think it's a little bit weak."* (Tim, participant #16)

Participants admitted that prior to ALS they had not given much thought to how the HSE operated or indeed to how healthcare services (public, private, or not-for-profit) rendered services to people with progressive illness. The majority of participants had no pre-existing illness or disability and having to engage with services with advancing disability was challenging for them. The following sections of this chapter explain why participants found the Irish healthcare sector difficult to negotiate.

6.3 Public and private healthcare services: Do they stand alone or are they one of the same?

All participants in this study accessed public healthcare services and the majority had engaged with private healthcare services from onset of symptoms until and during the diagnostic phase of care. Despite recent reductions in the number of people in the

Republic of Ireland who have private health insurance, 46% of the population still hold private health insurance (Health Insurance Authority, 2013; Wall, 2013). In the Republic of Ireland, private *and* public hospitals deliver inpatient care to private healthcare users. Private care is in part, funded by the public sector. Consultant physicians in the public sector run private practices with public practices and the physical infrastructure of public hospitals is often used to facilitate consultations and care of private service users (Tussing & Wren, 2006).

In the majority of cases (n=23), participants' first point of contact with a consultant neurologist was in a private clinic and all but three participants received a diagnosis (or provisional diagnosis) of ALS before they attended the public-based ALS clinic at Beaumont Hospital Dublin. Participants' path to a consultant neurologist varied across the sample. Most participants were referred to a consultant neurologist (in most cases, a private consultant neurologist) by a general practitioner. However, some participants were referred by a general practitioner to private consultant physicians in other fields of medicine who subsequently referred participants to either a public or private consultant neurologist. The majority of participants were referred to the national ALS clinic to confirm the diagnosis and to access multidisciplinary tertiary care. Jack (participant #26) explained:

> *"When @@private consultant neurologist## done all his tests with his wee needles and eh he said that he wasn't in a position to diagnose what it was." G: "So he wasn't too sure do you think?" P#26: "Well when I pushed him on it he says it could be this, it could be that (....) he says it could be a case of motor neurone but I'm not qualified enough to diagnose it so I want you to see @@the consultant neurologist at the national ALS clinic## in Beaumont."*

Most participants questioned the role of private healthcare services for people with ALS in the Republic of Ireland in the context that they would have to engage primarily with public healthcare services as they advanced in the disease. As Danielle (participant #9) and Pascal (participant #23) explained:

> *"When I came back from the US, it was like yeah you have to go to the clinic at Beaumont, it's the only clinic and that's a public clinic you know so like private*

healthcare wasn't going to do anything for me. It wasn't going to get me seen any faster you know and the community services that you need you are not going to get them through the private system either." (Danielle)

"Even if I won the euro millions in the morning and I wanted some of the stuff that I need I probably wouldn't be able to get it [through the private sector]. I can only get it through the HSE, the type of care that I need. I can't get it anywhere else." (Pascal)

Participants engaged with public *and* private healthcare services but most participants relied on public sector services as they adapted to insurmountable loss. Prior to the onset of ALS, the majority of participants had had little reason to engage with a broad range of public healthcare services. However, as participants encountered insurmountable loss, they felt their needs were best met by public healthcare services. Sections 6.3.1 and 6.3.2 describe how participants came to realise that private and public healthcare services for people with ALS in the Republic of Ireland are very different.

6.3.1 Private healthcare services: Few benefits and many limitations for people with ALS

As stated, the majority of participants in this study engaged with private healthcare services during the diagnostic phase of care. Participants who had attended a private consultant neurologist and who had been admitted as an inpatient to a private hospital for further investigation of their symptoms, encountered little delay in accessing diagnostic services:

"I saw him [private consultant neurologist] in @@private hospital## and he brought me in two days later to the @@private hospital##, that's where he could get a bed quicker." (Cara, participant #32)

"My GP said he was more or less looking at a neurological thing so he contacted @@public consultant neurologist##, he's in @@regional hospital##, he'd be the neurologist there, and there'd be a waiting list, I think it was at least six months or more (....) then my GP he said oh I can get you somebody maybe from @@private clinic## and it'd be much quicker, so my GP got me @@private consultant neurologist## from @@private clinic##." (Sally, participant #20)

The national ALS clinic endeavours to see service users with suspected ALS within four weeks from date of referral. However, some participants reported that their general practitioner referred them to a private consultant neurologist because their general practitioner assumed that the waiting period to see a consultant neurologist in the private sector would be much shorter than that which people encounter in the public sector. As Melisa (participant #29) disclosed:

> *"She [general practitioner] said now with the health service the way it is you could be a long time before you see them [consultant neurologists]. And I said to her well look I'm in the VHI [private healthcare insurance] which I was when I was working and she said then I know this good man. So she recommended him, so she sent me to @@private consultant neurologist## in the @@private hospital##. So I was called within a month to go there."*

Participants had thought that attending a private consultant neurologist had enabled them to access diagnostic services which they assumed service users who did not use private services at the diagnostic phase, encountered difficulty accessing:

> *"I was lucky in that I had Aviva [private health insurance] and I was able to get to see the specialist quickly. I was able to write a cheque for a couple of hundred euros and claim it back, no problem for me. I wouldn't like to have been at the bottom of a queue waiting for a scan for six months or whatever."* (David, participant #30)

However, a number of participants felt that the cost of attending a private consultant neurologist (even with private health insurance) was expensive and some found themselves less able to afford private health insurance and/or the cost of various diagnostic tests in the private sector. Terry (participant #11) and Eve (participant #7) recounted:

> *"When I first made the appointment to see @@private consultant neurologist##, I asked the secretary, I went how much will that be and she said three hundred euros, you know and I had to ask again no, no, how much did you say, three hundred euros, sorry would you say that again please (laughing) you know like, I just think that's off the Richter scale!"* (Terry)

"As he [private consultant neurologist] was looking for more and more tests to be done, my health care cover only covered me for, oh it didn't cover me for a lot of things. I had reduced my cover just before I got my last policy (....) like the EMG [electromyography], had it been done in the @@private hospital## it would have cost me an awful lot of money." (Eve)

Participants indicated that private health insurance was of no great benefit to them as they advanced in the condition. Although some participants encountered delays in accessing community-based services in the public sector, private health insurance did not cover the cost of (or only contributed a fraction of the cost towards) private services from allied healthcare professionals. Eve explained once more:

"Once the diagnosis was made I do know that he [private consultant neurologist] referred me to the private physio[therapist] and of course that was going to cost me something like forty euros or fifty euros a trip and the same with the occupational therapy but my insurance wouldn't cover much of that."

Participants reported that mounting physical disability alerted them to the likelihood that they would become more dependent on allied healthcare services (e.g., physiotherapy, occupational therapy). However, of those who engaged with private allied healthcare services in the earlier phase of their condition, none felt that private allied healthcare professionals were sufficiently competent in the specialist field of ALS care. Richard (participant #8) remarked:

"I was disappointed with the private physios [therapists] that they couldn't actually spot that well there's something different here, wrong, we can't say what it is, but I'd advise you to be going such a way. I'm disappointed with that."

Some participants believed that in comparison to public healthcare services, post-operative care and continuity of care between different medical specialties in the private sector was unsatisfactory:

"He [private consultant neurologist] had no back up services in the private. There is no back up services in the private (....) There is no advantage for me in the private sector

(....) like the surgery post op [surgery post gastrostomy feed insertion] in the @@private hospital## was an accident waiting to happen." (Morris, participant #10)

"I was going down to @@private consultant neurophysiologist## and he wasn't saying anything to me, he was making a pile of money. I mean you could see he was doing his half hour and then he kind of got interested (....) but he wouldn't say anything to me because his job was to report back to @@private consultant neurologist##. Then I had to wait weeks or months to get back to @@private consultant neurologist##." (David, participant #30)

A number of participants felt that they would soon have to pay for private homecare services to supplement public-sector homecare services. However, private health insurance did not cover the cost of private homecare services. The HSE did not provide night-time care assistance to participants and should participants have felt the need to access night-time services, they had to pay for these services. Edward (participant #14) and his spouse explained:

"We have VHI [private health insurance] plan E (....) We can pay separate for night-time." P#14 Spouse: "We are thinking of that soon, if we need it (....) I don't think there's any for that [from VHI], no that's extra."

Due to the limitations of the private healthcare sector in meeting participants' broad range of care needs, some participants felt they had "no choice" but to engage with public healthcare services. All participants were grateful to public-sector services for providing services to them but some participants felt they should at least have had a choice to access either or both sectors at all stages of the disease. However, those who wished for choice also resigned themselves to the prospect that private healthcare services might never provide the broad range of services required by people with ALS. The reason voiced by participants as to why private healthcare services could never meet all their care needs was that it was not financially feasible for private health insurance to cover all the services that they required:

"I would have to say it would be just too expensive, simple as that, because there's a whole team involved, it's not just the neurologist (....) there's a whole team like, there's the occupational therapist, the physiotherapist, the dietician. I don't think you could do

that on a private basis (....) I would just say it's just not economically viable to give private care for that." (Terry, participant #11)

In this context, participants who engaged with private healthcare services before and during the diagnostic phase of care, moved from the private sector to the public sector in their attempt to secure the broad range of allied healthcare and support services required by them.

6.3.2 Public healthcare services: Does "public" mean public?

Participants suggested that healthcare professionals do their best for people with ALS regardless of which sector (i.e. public or private) they provide services, but the majority of participants in this study felt that public healthcare services met their needs best. For the most part, participants felt more reassured by public healthcare services and more cared for by healthcare professionals in the public sector. Martin (participant #31) remarked:

"I find in my dealings with the public sector there's a whole lot more sympathy going on, there's a whole lot more sympathy (....) there is a different attitude, it's more of a, a person to person rather than I'm doing a job."

Nonetheless, engaging with public healthcare services also posed challenges that participants had not previously encountered. Prior to the onset of ALS, the majority of participants knew little about service users' entitlements to public healthcare services. Indeed, as already described, some participants admitted that they did not understand how the HSE functioned even when they accessed services through the HSE. In the Republic of Ireland, people who are granted a so-called medical card by the HSE have an entitlement to access public healthcare services free of charge (Tussing & Wren, 2006). The allocation of the medical card to services users is means-tested although diagnosis and/or severity of illness can also play a role when eligibility is determined. The majority of participants in this study had assumed (prior to ALS), that the HSE automatically granted medical cards to terminally ill service users. In addition, most participants had also been unaware (prior to ALS) that they required a medical card in order to access in full a broad range of primary care services in the HSE:

"I never knew, absolutely not, I had VHI [private health insurance] through work, company plan plus so any time I ever went to a doctor or whatever, either the doctor I

paid for myself and claimed it back or if I went into hospital I was always on the VHI. So I had no experience of the health care system [public system] at all." (Pascal, participant #23)

"We are not entitled to see the public health nurse unless we have a medical card. I thought public health was public health (....) I thought it would have happened the minute I was diagnosed." (Morris, participant #10)

Prior to the onset of ALS, the majority of participants did not have a medical card and the experience of obtaining one varied amongst the sample. Some participants encountered great difficulty obtaining a medical card but others obtained a medical card without difficulty:

"There was a bit of a battle, a long battle to get that [medical card]." (Jack, participant #26)

"Ah it was alright actually, I found them okay. I suppose I got it in November, six weeks afterwards [after diagnosis]." (Eilish, participant #19)

Indeed, even after obtaining a medical card, one participant (Eve, participant #7) still encountered difficulty accessing general practitioner (GP) services because many GPs in her local area refused to provide services to medical card holders:

"By the time I got my medical card then at the end of August or something I had go and find myself a GP and believe it or not the GPs in the area aren't taking on medical card holders, you know it's very hard, the first few phone calls that I made booked up, my books are full, not taking on any more medical card patients."

All but six participants had a medical card at time of participation in the study but the majority of participants still encountered delays in accessing primary care services. As described in Chapter Five, some participants felt "disconnected" from healthcare services in the early phase of their illness. However, not only did participants experience difficulty *getting into the system*, they also found it difficult to secure services when they managed to "get into the system". Danielle (participant #9) shared with frustration:

"So I got the medical card and I feel really lucky to have it (....) and so the first thing I think was making contact with them but then yeah the waiting lists are ridiculous in the system."

For some participants, (with or without a medical card) their income and/or assets limited the amount of public sector homecare services they received:

"I don't count myself as wealthy, but I have to spend my income (....) I have to spend my savings which is paying for the care (....) I don't qualify for much." (Jennifer, participant #25)

As described in Section 6.2, some participants feared for the future of public healthcare services, in the context of the downturn in the Irish economy. This study revealed that cuts in public healthcare expenditure impacted directly on participants' experiences of healthcare services. For some participants, access to services (even with a medical card) was fraught with difficulty because the services they required had suffered reductions in personnel who provided these services. Participants were also resigned to the prospect of further cutbacks in public healthcare services given the reality of public service cutbacks:

"I think it was the visit in September to the motor neurone clinic, I went to see the occupational therapist and physiotherapist and they at that stage said they were going to contact is it the local occupational therapist and physio[therapist] which they did (....) [but] I've heard nothing from them [primary care] since." (Terry, participant #11)

"I'm sure at some stage someone is going to turn around and say sorry we haven't got anyone to do that at the moment. So that's probably going to happen, you know, I'm realistic about that in that sense (....) I'm sure as it gets worse, when I start having to be passively incubated with oxygen or whatever and those sorts of things am I going to come a cropper [fail to secure services] at some stage." (Pascal, participant #23)

A number of participants spoke about their universal right to healthcare regardless of their means. As described thus far, access to public healthcare services was not universal and some participants encountered significant delays even when they had been granted, in principle, full access to public healthcare services. Those who spoke about universal

access to healthcare were annoyed about having to engage with a healthcare system that denied some service users a basic human right:

> *It SHOULD be good (....) services SHOULD be available to everyone as a human right."* (Melanie, participant #27)

> *"Philosophically I think we should have the best public health system. I don't think we should need a private system. I think we should all be public, and I don't mind paying taxes for that, so philosophically I think everybody should be treated equally. When you are sick it doesn't matter."* (Danielle, participant #9)

However, not all participants experienced delays in public healthcare services and some participants were surprised by the fact that the public sector rendered community-based services. The majority of participants felt public healthcare services were more than sufficient to cater for their needs once they managed to "get into the system" *and* engage with services when they wished to do so. Indeed, most participants were more than willing to access public healthcare services for all their care needs should they manage to secure access to these services. Morris (participant #10) and Melisa (participant #29) remarked:

> *"Well I can tell you there is no comparison between the public service and the private provided you can access the [public] service."* (Morris)

> *"I think when you get to the doctor that you want to see in the public you have very good services, it's just getting these appointments to get into the system."* (Melisa)

Despite the challenges encountered by participants in either public or private healthcare services, some participants believed that healthcare services for people with ALS were more than likely better than services provided to other diagnostic groups. Indeed, of the few participants who had already engaged with services in other fields of care, all were more satisfied with ALS services than they had been with services in other fields of care. Ita (participant #2) had engaged twice with cancer services and compared ALS services favourably to cancer services:

"I thought it would be like a lot of other illnesses you just go and get your whatever treatment you are supposed to have and then you go home and there's nothing else (....) any time I've had the operations or whatever and there was no real aftercare I found (....) I'm quite surprised now that there's so many people there you know to help."

As explained in Chapter Three, a key tenet of critical realism is that reality is formed in part by structure and structure shapes both experience and outcome. This study found that from within the macro-level of ALS care, two primary structures alleviated participants' fears about the public healthcare sector's capacity to meet their needs. These included the national ALS clinic at Beaumont Hospital Dublin and the Irish Motor Neurone Disease Association (i.e. voluntary sector). Sections 6.4, 6.5, and 6.5.1 explain how the national ALS clinic and the ALS voluntary sector shaped participants' experiences of healthcare services and how participants adapted to insurmountable loss.

6.4 The voluntary sector: Supplementing care for people with ALS

The Irish Motor Neurone Disease Association (IMNDA) is the primary support organisation in the Republic of Ireland providing care for people with ALS and their families. Participants revealed that the IMNDA rendered valuable support to them and all participants who had engaged with the IMNDA were deeply grateful for their services. All participants had received information about ALS from the IMNDA and many participants had already received practical assistance from the IMNDA:

"I rang the association [IMNDA] and they sent out this wonderful folder here with all the information I could need really is in that." (Ita, participant #2)

"I mean I'm amazed at the amount of equipment that they [IMNDA] are prepared to lend me." (Jennifer, participant #25)

The IMNDA provided services to participants when participants were unable to secure services from the HSE. Participants' need for specialised equipment and homecare services rose in line with mounting loss but, in some cases, the HSE was unable to fund additional home care services and/or high-tech assistive devices for service users. For this

reason, the IMNDA funded a broad range of assistive devices for participants and provided financial assistance to them toward the cost of private homecare services:

> *"They [IMNDA] got a hoist for me and they have a bed waiting for me now once when I'm, when I have my new room done."* (Eilish, participant #9)

> *"They [IMNDA] have been very useful. They put in the equipment. They put in the stair lift. They have given me a motorised wheelchair (....) and I've got a bed from them and now the hoist."* (Samantha, participant #22)

> *"They're [IMNDA] helping me too with home help." G: "So you're getting home help from the HSE as well as the IMNDA?" P#12: "They're just giving me a few bob [money] towards it. But obviously you don't tell the HSE, don't tell the HSE! (both laughing), yeah so that is good."* (Helen, participant #12)

Overall, participants engaged openly with the IMNDA. Participants drew on the voluntary sector to subsidise services rendered by the HSE (e.g., assistive devices) and to finance services they acquired through the private sector (e.g., home care).

6.5 The national ALS clinic: Reassuring people with ALS and engendering loss for people with ALS

As described in Chapter Two, up to 80% of people with ALS in the Republic of Ireland attend the public, national ALS clinic at Beaumont Hospital Dublin. Population-based comparative studies between specialised ALS clinics and general neurology clinics in Italy have shown that between 43% (Chio et al., 2006) to 66% (Zoccolella et al., 2007) of people with ALS access specialised clinics. It is thought that almost all people with ALS in the Netherlands attend specialised ALS clinics (of which there are three).

As illustrated in Chapter Three, a large proportion of the sample (n=26) had already attended the national clinic at Beaumont Hospital. In most cases, participants were referred to the clinic because it is a national clinic. Of those who had not attended the clinic, three participants suggested that they were too frail to travel to the clinic, three participants were sufficiently satisfied with local / regional services not to warrant travel,

and two participants had been advised by their neurologist that the clinic would offer no more than what was offered to them by community and/or secondary care services.

The majority of participants continued to attend the clinic on a scheduled basis. Indeed, all participants who had continued to attend the clinic and those who were new attendees to the clinic felt that they would more than likely continue to access clinic-based services as they encountered more loss. The clinic was for some participants a "sad" place. However, most participants who accessed services at the clinic suggested that they would have encountered greater difficulty in adapting to loss had they not had healthcare professionals at the ALS clinic to support them:

> *"When I started having breathing problems first em I went to my doctor [general practitioner] and he suggested I took a cough bottle. But I didn't have a cough you know what I mean and then I was sent to @@regional hospital## and I didn't find them very helpful at all. They just didn't know what they were dealing with (....) so then they transferred me to the Beaumont and the whole team is there and they just know what they are doing, they understood straight away how to help me deal with this."* (Mona, participant #1)

As described in Chapter Five, healthcare professionals' lack of expertise diminished participants' confidence in them. However, this study found that healthcare professionals at the national ALS clinic engendered confidence in participants because of their ability to anticipate participants' care needs. Ann (participant #24, end-stage ALS) shared:

> *"Eh the services are primarily in Beaumont, very attentive (....) In fairness the RIG [gastrostomy feed] was put in before I needed it (....) I got this (pointing to NIPPV) before I needed it (....) everything was anticipated." G: "If you didn't have the service based at Beaumont Hospital clinic, how do you think things would be?" P#24: "I think I'd be dead. I don't eat anything now, nothing by mouth."*

Most participants who had attended the clinic were impressed by the clinic's ability to link them directly with primary care healthcare professionals. As described in Section 6.2, participants were confused about how the HSE functioned and found it challenging to

source services on their own. In this context, the role of clinic personnel was not only to provide care directly to participants but also to link them directly to primary care services:

> *"I've met a physiotherapist, I got a letter from her, obviously from the communication in Beaumont, she just wanted to meet me (....) so that's a good system (....) and then about four weeks ago I got a phone call from the local clinic, from the occupational therapist (....) also the speech therapist in Beaumont wrote to maybe the clinic here to see where I would be going to when I did have to or felt like meeting with a speech therapist (....) I thought that was very impressive."* (Cara, participant #32)

As described in Chapter Five, exerting control included the need for participants to decide about how and when they would engage with healthcare services. Participants who attended the national clinic found that the clinic accommodated their need to be in control of their care. Denise (participant #33) remarked:

> *"They [ALS clinic healthcare professionals] try what I want and don't argue and if something doesn't work there is no hassle about stopping it."*

Participants favoured the flexibility of the ALS clinic compared to other services they had encountered. Some participants who attended the clinic on a frequent basis contacted personnel at the clinic between visits. Participants were reassured by the fact that they could, if necessary, hasten their next appointment at the clinic to deal with new challenges they encountered between visits. Samantha, (participant #22) shared:

> *"The physio[therapist] has been good. My left shoulder was giving me quite a bit of pain and @@Beaumont clinic researcher2## had been down here to do the research and he suggested that I had an injection. Well to be fair between him, @@the consultant neurologist at the national ALS clinic##, and the physio[therapist], they sorted it the day I went up. Yeah so I got the injection when I was up there."*

Participants who had either stopped attending the clinic and/or only attended sporadically (n=4) also felt reassured by the fact that they still remained in contact with the clinic from a distance:

"My first interaction was with Beaumont itself. I got an appointment there very fast and what is important for me is that subsequent appointments are really down to me, just to schedule and stuff like that, so that was a positive experience." (Danielle, participant #9)

As described in Section 6.3.2, most participants felt that healthcare services for people with ALS are best delivered by public healthcare services. However, some participants perceived the ALS clinic as separate from the HSE. Pascal (participant #23) disclosed that the ALS clinic surpassed his expectations of public healthcare services because the clinic provided specialist services which he felt were not ordinarily associated with public healthcare services:

"I mean you go through this grey box which is the HSE (....) I never had any experience of the HSE at all em nor did I really want to you know (....) I wouldn't associate the clinic with the HSE at all."

However, participants also complained about long waiting periods at the clinic and some felt the national clinic was under-resourced to cater sufficiently for the needs of all clinic attendees. Maureen (participant #34) disclosed:

G: "What are your impressions of the clinic?" P#34: "Well (laugh), well last time we were there I saw the doctor and had to wait for physiotherapy but I didn't see her so I got fed up waiting (....) I saw the speech and language girl only and I was to see the dietician and the physio[therapist] (....) all I saw was the speech and language (....) we were there at twelve o'clock and we were sitting there at four o'clock (....) I think when you see the doctor that is fine but when you have to see the other people, it's too long of a wait, it's not rolling over (....) because they [clinic-based healthcare professionals] are probably looking after somebody else. I think there are not enough people [healthcare professionals] there."

Waiting at the clinic was upsetting for some participants, in particular for those who struggled most to come to terms with their loss. Waiting for long periods among other people with ALS at the clinic reinforced the sense of losses they had yet to encounter:

"The problem there [ALS clinic] is the long waits in the waiting room and you're really, you're meeting people far worse than yourself in the waiting room and that's horrendous, they've [people with ALS] to spend in that waiting room especially at the beginning because any other surgery I was in up until that I wasn't meeting people with the complaint you know, and now I was subject to a big waiting room of maybe ten or twelve or more people waiting there, some people in very, far worse condition with the upper part affected than myself." G: "Was that, was that distressing for you?" P#8: "That was very distressing yes, it is, it is." G: "Were you shocked by that?" P#8: "Well I knew before we went there that day that we'd be meeting people but of course I didn't know I'd be meeting so many people you know and say being a few hours waiting there, sometimes a good few hours and that. It's pretty devastating." (Richard, participant #8)

Participants' perceptions of the national ALS clinic were also shaped by a medical science documentary which aired on national television during the period in which interviews were conducted with participants. The documentary *MND - The Inside Track* (2012) chronicles the story of Colm Murray (a well-known Irish sports broadcaster) who participated in ALS research including a clinical trial at Trinity College Dublin and the National Neuroscience Centre, Beaumont Hospital Dublin. Colm Murray had also spoken about his experience of the ALS clinic on national television (The Late Late Show, 2010) prior to the documentary. His story drew attention to the role of the clinic as a tertiary care centre and to research undertaken from the clinic. As Edward (participant #14, non-ALS clinic attendee) and his family questioned:

P#14 spouse: "And how is the research going under @@the consultant neurologist at the national ALS clinic##?" P#14 daughter: "But they have some new one drug that they developed or is being developed?" G: "It's a clinical trial, have you heard anything about the clinical trial @@Edward##?" P#14: "I did, I know about it. Colm Murray, isn't it (....) Colm Murray has done a lot (....) they are developing a new pill."

The participants who attended the clinic already had a diagnosis and/or provisional diagnosis of ALS (which was confirmed later). Some participants were drawn to the ALS clinic consultant's profile through the aforementioned publicity and no participant indicated that they wished to transfer their care to another neurologist after they

encountered the consultant neurologist at the ALS clinic. Samantha (participant #22) shared:

> *"Well she's got the reputation so you'd expect to get the better care and the better treatments, so I would choose to go up to see her." G: "Based on her reputation?" P#22: "Yes, but I only knew about that reputation after I was diagnosed*[1] *because I was diagnosed on the Tuesday, had ten thousand texts to watch the Late Late Show on the Friday and the girls said to me, my daughter, well at least you are under the same people as the stars because Colm [Colm Murray] had been."*

However, as the researcher continued to saturate *BH clinic* in terms of its properties and dimensions, it became clear that it was not only the reputation of the clinic's consultant neurologist that reassured participants. As discussed in Chapter Five, participants were most reassured by healthcare professionals who "connected" with them and who were sensitive to the service user perspective. Most participants "connected" with healthcare professionals at the ALS clinic as they (healthcare professionals at the clinic) moved between the micro- and macro-contexts of ALS care to reassure participants. Cara (participant #32) recounted:

> *"I met @@the consultant neurologist at the national ALS clinic## I think for the first time in, I think it was mid February. Obviously I was very apprehensive, very nervous and that and I got a fright when I saw four or five people [research fellows and ALS specialist nurse] sitting in front of me when I went in, that was a shock but she was extremely nice and I felt human, you know I felt it [interaction] was human, she reassured me."*

As described in Chapters Four and Five, participants felt they coped better with ALS when they felt they had time to "digest" loss and when service providers responded to how participants themselves were coming to terms with loss. Despite long waiting periods at the clinic, the majority of participants felt that they were afforded time by healthcare professionals at the ALS clinic when they engaged with healthcare professionals:

[1] Participant #22 was diagnosed with ALS prior to attending the national ALS clinic.

"I think I was in [clinic room] for about an hour you know and I'm after taking up an hour of their time and there's other people waiting, that's the type of clinic that it is. If I wanted to stay in for two hours and talk about something I probably would have been allowed you know. There's no pushing you out or anything. If you have concerns they'll not push you out." (Jack, participant #26)

A minority of participants (n=3) suggested that (in the absence of a cure for ALS) a national clinic was of no tangible benefit to them. However, most participants suggested that healthcare professionals at the clinic reassured them even though participants encountered insurmountable loss. As Samantha (participant #22) disclosed:

G: "So I'm sensing despite the waiting times, you get something out of the clinic?" P#22: "Personally I think I do, if you asked @@Seth## [spouse] he'd say waste of bloody time what they do up there." G: "When you say you think you do, what is that something?" P#22: "I feel, I don't know it gives me that bit of reassurance although physically there's probably nothing, well I know deep down there's fuck all they can do about it, you know they can't stop it."

As illustrated in Chapter Five, feeling trusting of and reassured by healthcare professionals assisted participants to adapt to insurmountable loss. Overall, most participants who engaged with the clinic trusted and felt reassured by the national ALS clinic because (as explained in Chapter Five) what participants searched for most (i.e. to be cared for in accordance with their own wishes), most participants found at the ALS clinic:

"I don't worry so much about that [waiting times] because I feel I'm being cared for." (Cara, participant #32)

6.6 The international context to the national ALS clinic: Shaping experience and loss for people with ALS

Services rendered to participants by the national ALS clinic were shaped by the international context to ALS care. The national ALS clinic at Beaumont Hospital Dublin is a care *and* research centre for ALS. The clinic provides tertiary care in line with practice parameters in ALS care and the ALS clinic's consultant neurologist is co-author of the European Federation of Neurological Societies (EFNS) guidelines on the clinical

management of ALS (Andersen et al., 2012). Researchers at the clinic work collaboratively with researchers in other countries on the pathogenesis of ALS. During the period in which interviews were conducted with participants in this study, the national ALS clinic was one of 81 centres in 11 countries that participated in a phase III ALS clinical trial (Corcia & Gordon, 2012).

As described in Section 6.5, participants who attended the national clinic shared, for the most part, positive perceptions of the clinic because they valued how healthcare professionals at the clinic interacted with them. Participants' perceptions of the clinic were also shaped by the clinic's international standing in ALS research. Indeed, some participants who attended the clinic suggested that given the clinic's connections within the international ALS community, the clinic offered more to them than services elsewhere:

> *"I don't want to disrespect any of the other neurologists not knowing whether they are better or worse than @@the consultant neurologist at the ALS clinic## but you know she's probably the more vocal and one of the most visible in the area of MND so I wouldn't have any cause to question anything she says."* (Pascal, participant #23)

As described in Chapter Two, international guidelines on the clinical management of ALS (Andersen et al., 2012) recommend that care in ALS should be led by specialised multidisciplinary clinics. The national ALS clinic is a multidisciplinary clinic and is staffed by a range of healthcare professions including medicine (neurology), specialist nursing, physiotherapy, occupational therapy, dietetics, and speech and language therapy. The clinic also has close links with neuropsychology services and with palliative care, respiratory and gastroenterology specialists based at Beaumont Hospital. As described in Chapter Five, the experience of mounting disability alerted participants to the fact that they relied on a broad range of services. However, participants felt that healthcare professionals in community-based HSE services (i.e. primary care) and in private healthcare services were, for the most part, unfamiliar with ALS. In this context, some participants' primary reason for attending the clinic was to access specialised multidisciplinary care. Eve (participant #7) explained:

> *"I suppose my reason for continuing to go to Dublin, it is the centre of excellence (....) What I'm finding when I meet people in general, obviously up in Beaumont everybody*

knows about motor neurone disease, out in the community for an awful lot of them, for the district nurse, for the physios [therapists], I'm their first motor neurone disease patient (....) they all compare the disease instantly with MS (....) in fact my GP has never had a motor neurone disease patient (....) you know for health professionals who have never come across motor neurone, they're only learning from you [service user]."

As depicted in Chapter Four, participants moved back and forth between accepting and resisting ALS as they adapted to insurmountable loss. Although the majority were becoming somewhat more accepting of ALS with mounting loss, the findings also suggest that few participants would have declined a cure for ALS or treatments to slow down the progression of ALS. A small minority of participants suggested that they would prefer not to participate in a drug trial. However, a significant proportion of the sample (n=7) were (at the time of their interview[2]) participating in the trial and more participants wished they had been selected for the trial. Being selected for the trial was a positive moment in the lives of participants because the trial offered them hope of a treatment for ALS:

"Without being stupid about it that [selection for trial] was probably the biggest boost, one of the biggest boosts I've got since the start of this thing, it was fantastic, it was brilliant." (Terry, participant #11)

Many participants who did not participate in the trial viewed it positively because a successful outcome to the trial raised the possibility that they might at some point in time, receive the drug tested in the trial:

"@@the consultant neurologist at the ALS clinic## is saying you know there'll be new drugs on the market, maybe you know, something like that will come out of it [trial], maybe able to do more for you, so we'll see (....) She's not talking about clinical trials now, she's talking about actual, the results of the clinical trials. So yeah, sure because since I went on the other thing [Riluzole] I feel I've fallen off a cliff [experienced rapid decline] you know." (David, participant #30)

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[2] Participant #23 had participated in the clinical trial *prior* to his participation in this study. His participation in the clinical trial ceased due to medical complications arising from participating in the trial.

However, the experience of mounting disability whilst on the trial engendered more feelings of loss. Some participants' optimism was quickly replaced by disappointment because they continued to advance in the disease even though they were participating in the clinical trial. As Terry explained:

> *"My health has deteriorated [since start of trial], like you know things have moved on and I tend to think maybe I'm on the placebo and not the real thing or if I'm on the real thing, I do know it's a trial drug, it's probably not working, that type of thing, that the drug itself mightn't be up to scratch (....) or else maybe the drug is working and I am [still] getting worse than I am."*

Indeed, Richard (participant #8) disclosed that the experience of disease progression whilst participating in the trial, evoked feelings of despair. He indicated that he had been informed by the consultant neurologist at the clinic that participation in the trial could potentially be of benefit to him. However, he found little benefit to participating in the trial:

> *"The trial, I've mixed feelings about it, it was sold to us as say being the greatest thing that, the saviour of you (....) whereas now I just, I go through the procedure of it and I'm taking the tablets and getting the tests done but my arms are weakening considerably under it so I don't think it's doing anything for me (....) After a while it becomes more negative you know, the enthusiasm goes out of it after a while when you find out it's not delivering for you."*

Overall, the national *and* international context to the ALS clinic shaped participants' experiences of services and how they adapted to loss. The ALS clinic's reputation reassured service users and participants were, for the most part, satisfied with multidisciplinary care at the clinic. The clinic's participation in a large clinical trial engendered optimism among some of the participants. However, the clinical trial also engendered loss among participants because their experience of mounting disability whilst on the trial diminished their hope of overcoming ALS.

6.7 Conclusion

This chapter has described how the macro-context to ALS healthcare in the Republic of Ireland shaped how participants engaged with healthcare services and adapted to loss. Multiple macro-level factors (local, national, and international) shaped participants' experiences of healthcare services.

The findings indicate that people with ALS require co-ordinated multidisciplinary care in line with their perceptions of their needs. As described, the disease trajectory of ALS is one of constant change and people with ALS rely on healthcare services as they advance in the disease. For the most part, participants believed that the national ALS clinic responded to their needs. These findings are consistent with those from other countries on service users' perceptions of and satisfaction with ALS specialised clinics (Hogden et al., 2012; Miller et al., 2000 and 2009c; O'Brien et al., 2011b).

Guidelines on the clinical management of ALS (Andersen et al., 2012) do not refer to barriers often encountered by people with ALS in accessing services. Healthcare systems vary and different systems impact on how services are rendered to people with ALS (Borasio et al., 2001). In the Republic of Ireland, the healthcare system is fragmented and complex (Ryan et al., 2009; Smith, 2010). A large proportion of the population has private health insurance cover but the range of services required by people with physically disabling and life-limiting conditions (including ALS) are not rendered by the private sector. The cost of private health insurance in Ireland continues to rise but private insurance cover is of little benefit to people with ALS beyond (and in some cases, during) the diagnostic phase of care.

The findings resonate with studies that investigated access to and financial burden of services for people with other life-limiting conditions in Ireland. Hardiman et al. (2003) found that people with multiple sclerosis also encountered difficulty accessing community-based services in the private sector. Timmons et al. (2013) found that private health insurance did little to offset a wide range of additional cancer-related medical and non-medical expenses. Cancer service users also encountered difficulties in obtaining medical cards which would otherwise have entitled them to access public-sector primary care and community services free of charge (Timmons et al., 2013). Overall, the mix of

public/private provision and financing of services in the Irish healthcare system is of little benefit to people with ALS.

Participants' uncertainty about their healthcare services is not unique to people with ALS. People with other life-limiting conditions (e.g., Parkinson's disease, multiple sclerosis) also feel uncertain about their health and social care services (Edmonds et al., 2007; Giles & Miyasaki, 2009). Timmons et al. (2013) reported that cancer service users also complained that service providers fail to inform them sufficiently about their entitlements to services. Participants' uncertainty about healthcare services resonates with people with ALS in other healthcare system contexts (Hughes et al., 2005; O'Brien et al., 2012). Indeed, Whitehead et al. (2012) found that anxiety heightened among people with ALS during end-of-life care because they were unsure about what healthcare services could offer them. Similarly, in this study, feeling uncertain about healthcare services was distressing for participants.

Fallon and Foley (2012) argue that managing the spectrum of palliative care needs for people with life-limiting non-malignant disease (including ALS) is a challenge for healthcare services. It is thought that people with ALS in the Republic of Ireland engage with specialist palliative care services but no data is available on how many people with ALS in the Republic of Ireland access palliative care services. O'Leary and Tiernan (2008) found that barriers to services for people with non-malignant terminal disease in Ireland included a lack of expertise among healthcare professionals about non-malignant terminal disease. In the UK, evidence suggests that terminally ill people with non-malignant disease may not necessarily access specialist services (Elkington et al., 2005). Studies on access to palliative care services in Ireland for people with non-malignant terminal illness have shown that less than 10% of terminally ill people access specialist palliative care services (Igoe et al., 1997; O'Leary & Tiernan, 2008). These findings are similar to those of other countries which have investigated palliative care services for non-cancer service users (Aoun et al., 2007; Harrison et al., 2012).

In the UK, experiential knowledge of service users is recognised as a key factor that shapes people's experiences of cancer care (Attree et al., 2011). Involving the service user in deciding how services should be delivered is meaningful to healthcare users (Cotterell et al., 2011). However, public participation in the delivery of care Ireland is unknown and

there have been few if any tangible outcomes from a national strategy for service user involvement (Health Service Executive, 2008). This study found that from the viewpoint of people with ALS, the Irish healthcare *system* offered little opportunity to shape how their services were delivered to them.

This chapter has explained how the macro-context of ALS healthcare services in Ireland shaped how participants engaged with healthcare services. Multiple macro-contextual layers (as depicted in figure 2, p.39) shaped participants' experiences of healthcare services, and influenced outcomes for participants in other contexts. International consensus on practice parameters in ALS care shapes how tertiary services are rendered to people with ALS in Ireland. Tertiary (national) services shape how local services are rendered to people with ALS but local and national policies in the Irish healthcare system also impact on how community and tertiary services render care. The next chapter (Chapter Seven) outlines a number of practice recommendations for ALS care based on the study findings i.e. how service providers might best support people with ALS as they adapt to loss. Chapter Seven also provides specific recommendations for healthcare services for people with ALS in the Republic of Ireland.

Chapter Seven

Practice recommendations

7.1 Responding to insurmountable loss in ALS: Recommendations for practice in ALS care

As described in Chapters Four, Five, and Six, loss is central to the experience of ALS. Chapter Four explained how people with ALS construct loss and outlined internal domain resources that enable people with ALS to adapt to loss. Chapter Five explained how people with ALS interact with external domains (i.e. healthcare services and family) as they adapt to loss. Chapter Six described how broader contextual factors (i.e. macro-context) shape how people with ALS engage with healthcare services to cope with loss.

As outlined in Chapters One and Two, the purpose of undertaking this study was to identify the key parameters of healthcare experiences among people with ALS. As stated in Chapter Three, the relationship between agency and structure is complex. The situational context of Irish healthcare services for people with ALS shapes their individual experiences of services but their experience of services can also impact on how services are provided to them. Mapping out the key parameters of the ALS service user experience of services was important because mapping out key parameters of healthcare experiences among people with ALS identified domains that are important to people with ALS and reasons why these domains are important to them. Accordingly, the following sections discuss the implications of the findings for ALS care practice, and outline how structures in the Irish healthcare system need to change to help people with ALS adapt to loss.

7.2 Alleviate uncertainty for people with ALS

A core finding in this study is that people with ALS live with uncertainty as they encounter loss. As described in the findings chapters, uncertainty extends from uncertainty about the future with ALS, to uncertainty about how to access healthcare services, to uncertainty about the future of Irish healthcare services. Sections 7.2.1 to 7.2.7 outline how service providers can help to alleviate uncertainty among people with ALS.

7.2.1 Provide information about ALS to service users

Providing information about ALS to service users on their request would help to alleviate their uncertainty. People with ALS move back and forth between accepting and resisting

ALS but they also have a need to be informed about ALS. In this study, participants were uncertain about ALS and were disappointed that healthcare professionals in primary care failed to provide sufficient information about the disease to them. ALS service users receive information about ALS at specialised clinics (Chio et al., 2008; Hogden et al., 2012; O'Brien et al., 2011b) but they might not necessarily receive sufficient information about the disease from primary care services (Brown et al., 2005; Hughes et al., 2005; Peters et al., 2013). Primary care healthcare professionals have a responsibility to ensure that people with ALS (should they wish to be informed) are informed about the disease.

7.2.2 Explain the role of healthcare professionals in ALS care to service users

As described, people with ALS engage with a broad range of healthcare professionals as they advance in the disease. In comparison to other life-limiting conditions, the pace of disease progression is more rapid and in most cases people with ALS require services from different healthcare specialties simultaneously. However, engaging with a broad range of healthcare professionals can also foster ambiguity among service users about the role of different healthcare professionals in ALS care. This study found that participants were somewhat uncertain about the specific roles of different healthcare professionals and what healthcare professionals were likely to offer to them as they advanced in the disease. The skill sets of different professions overlap in multidisciplinary care. However, healthcare professionals should take time to explain their role to servicer users *and* how their role might in some cases complement the role of other healthcare professions.

Some participants in this study were unsure about whether services (e.g., specialist nursing, assistive devices) were rendered to them by the voluntary sector (i.e. IMNDA) or by public-sector healthcare services. In the Republic of Ireland, the ALS voluntary sector ordinarily provides assistive devices to people with ALS on recommendation by its own specialist nurse or by relevant healthcare professionals in the public and/or private sector. Voluntary ALS associations need to explain to people with ALS *how* their services complement services rendered by public and private-sector healthcare professionals. Public and private-sector healthcare professionals should also take time to explain to people with ALS *how* they interlink with the voluntary sector.

7.2.3 Inform service users about their entitlements to healthcare services

This study found that people with ALS were uncertain about their entitlements to health-

care services. As outlined in Chapter Six, ALS service users' early interactions with healthcare services (i.e. at the diagnostic phase) in the Irish healthcare system occur, for the most part, in the private sector. People with ALS move from the private sector to the public sector in order to access allied healthcare and homecare services. However, access to some components of care (e.g., aids and equipment) in the public sector is means-tested and people with ALS encounter delays in accessing community-based services.

In order to minimise delays, service providers at first point of contact (regardless of which sector they provide services from) need to inform ALS service users that they require a medical card to access services in public community-based care. Education about entitlements to services has traditionally fallen to social workers but ALS service users' first point of contact with healthcare services is ordinarily with a physician (i.e. general practitioner / family physician). Healthcare professionals (regardless of profession) should ensure that they are sufficiently able to advise people with ALS about their entitlements to healthcare services.

In the Republic of Ireland, the Department of Health and Children and the Health Service Executive (HSE) have an important role to play in educating service users about how the Irish healthcare system functions and how the HSE delivers services. As described in Chapter Two, the HSE has established a number of national clinical programmes to improve standards of healthcare in Ireland. A national strategy for service user involvement (Health Service Executive, 2008) has also been developed. However, participants in this study were, for the most part, perplexed by the HSE. The HSE needs to be explicit about service users' entitlements to care and explain to people with ALS *why* services rendered by community-based public sector healthcare professionals are, in some cases, restricted to those who have a medical card.

7.2.4 Be attuned to ALS service users' understandings of life-sustaining interventions

Participants spoke openly (with and without prompting) about their experience of life-sustaining interventions and/or the possibility that they might engage with life-sustaining interventions at some point in the future. Participants understood life-sustaining interventions as interventions to extend life but some participants were vague about the life sustaining properties of gastrostomy feeding (if any) and of non-invasive ventilation at the very end-stage of ALS. Moreover, some participants perceived that these interventions

would extend life beyond the time-frame that these interventions typically extend life for people with ALS.

How participants perceived non-invasive ventilation and gastrostomy feeding at end-of-life care has implications for ALS care practice. Non-invasive ventilation and gastrostomy feeding are recommended by physicians in ALS care (Andersen et al., 2012) and people with ALS in Ireland engage with these interventions. Non-invasive ventilation prolongs survival in ALS but its function is also to alleviate respiratory distress (Andersen et al., 2012; Miller et al., 2009a, b). Gastrostomy feeding might not necessarily prolong survival in ALS (Andersen et al., 2012). As illustrated in Chapters Four and Five, people with ALS seek more certainty about time-frame as they advance in the condition and their struggle between wanting to live on with ALS and "letting go" is complex. Some people with ALS desire to live on but others wish to die before they become totally dependent on others. Service providers need to pay particular attention to how people with ALS construe life-sustaining interventions when they recommend (or advise against) life-sustaining interventions for people with ALS.

7.2.5 Be attuned to family members' roles in decision making about care

In this study, family was an integral feature in the lives of participants and their relationships with their family shaped how they adapted to loss. As described in Chapter Five, family as a category emerged from participants' meanings attached to life as a spouse, partner, parent, grandparent, and sibling. Participants made decisions about their care with family members and in some cases family was a key factor in determining how much assistance participants sought from service providers. The majority of dysarthric and anarthric participants relied on family members to voice their concerns. In some situations, involving family in discussions about care might to all intents and purposes be necessary because some people with ALS depend on family members to communicate with service providers.

These findings have implications for ALS care practice. People with ALS depend on family for support and, in the Irish context, become dependent on their family for physical care. The majority of participants wished to be cared for at home and they were, for the most part, reassured by close family members who provided care to them at home. Burden of care for families of ALS service users is high (Peters et al., 2012) but people with ALS rely

on family members for care (Hirano et al., 2006). A study focused on family dynamics in ALS care would be of benefit and could help formulate guidelines on how to involve family members in decisions about care.

7.2.6 Support the family in ALS care

Families of people with ALS experience emotional strain in their caring role (Ray & Street, 2007). They also encounter loss of social support which can impact on their wellbeing and on the wellbeing of people they care for (Love et al., 2005). This study did not seek to capture the carer perspective. However, the findings indicate that people with ALS themselves have concerns about being and/or becoming a burden on their family. *Being a parent* heightens their anxieties about placing burden of care on family.

The findings highlight the need for service providers to support family as they care for people with ALS *and* to support people with ALS as they seek to reassure their family. *The family in ALS includes people with ALS*. The amount of assistance people with ALS request from services is often shaped by how they and others, including service providers and authorities, judge their families' capacities to cope with their care needs. People with ALS request more physical care from healthcare services as they advance in the disease and homecare services become essential to support family who care for people with ALS at home. However, service providers in ALS care need to judge carefully *when* and *how* they support the family. The findings reveal that people with ALS seek to minimise disruption in the lives of their children and regulate how services are delivered to them to accommodate for this. Service providers need to pay close attention to the wellbeing of service users' dependants and provide psychological support to their dependants. Supporting the children of people with ALS might reassure people with ALS.

7.2.7 Reassure service users about end-of-life care

Mitsumoto and Rabkin (2007) suggest that end-of-life discussions in ALS are often triggered by service users' request for these discussions. McCarthy et al. (2010) found that people in Ireland might not be sufficiently informed about the processes surrounding death and dying. This thesis demonstrates that people with ALS need to be reassured about end-of-life care because feeling uncertain about the end-of-life stage of ALS provokes anxiety for them. The fear of losing speech also hastens people with ALS to

decide about their future care. Having confidence in healthcare professionals to alleviate potential pain at end-of-life, reassures people with ALS.

Reassuring people with ALS about the dying phase should include (as appropriate) discussions on: where service users wish to die (e.g., home, hospice); who will render end-of-life care to them (e.g., hospice service, community-based palliative care services); how and who will administer symptomatic relief; how can or should end-of-life care services be supplemented (e.g., family, ALS voluntary sector); how to make a DNR (Do Not Resuscitate order); who has authority to render and/or withhold services; and how people with ALS ordinarily engage with physician-assisted suicide. People with ALS seek information and assistance from healthcare services in line with how they themselves are coming to terms with loss and they have a need to be reassured about the dying phase as they move towards death. Service providers in ALS care have a responsibility to reassure people with ALS about the dying phase in ALS.

7.3 Enable service users to feel in control

This study found that people with ALS need to be in control of their care. Enabling people with ALS to be in control of care is important because people with ALS might disengage from service providers if they feel they have no control over their care. Enabling people with ALS to be in control of care is complex because for people with ALS, feeling in control also includes the freedom to relinquish control to service providers. Sections 7.3.1 to 7.3.3 outline how service providers can enable people with ALS to feel in control of their care.

7.3.1 Engage with service users when they are ready to engage

A key finding in this study is that people with ALS need to be in control of *when* they engage with services. People with ALS seek information about ALS and about what services are available to them but they engage with services in line with their own perceptions of disability and not those of service providers. People with ALS might also resist assistance (e.g., assistive devices) because as they struggle to come to terms with their loss, accepting assistance reminds them of what they have already lost.

These findings have implications for care practice in ALS. International guidelines in ALS care (Andersen et al., 2012; Miller et al., 2009a, c) refer to timing of interventions based on disease progression. Regular multidisciplinary care is advised and so, too, is early

initiation (where appropriate) of life-sustaining interventions (i.e. non-invasive ventilation, gastrostomy feeding) to improve survival. However, this study revealed that people with ALS vary in terms of how they come to terms with loss and they move at different rates towards acceptance of ALS. Participants exerted control over healthcare services by deciding *when* to engage with services and some participants delayed services until they felt ready to engage. Service providers need to pay greater attention to factors beyond disease progression (e.g., control, acceptance) when they recommend care to people with ALS at different stages of the disease. Simply asking people with ALS about how they are coping with loss is useful to ascertain if they are ready to engage with services.

7.3.2 Afford choice to service users about care

For the most part, participants felt in control of their care *when they were given choices about their care* and *when they rendered control to service providers on their own terms*. The findings indicate that people with ALS need to feel in control of their care (i.e. have choices about care) before they readily accept help from service providers.

These findings have important implications for care practice. In this study, having the freedom to opt for *and* decline services (e.g., home care services, assistive devices, NIPPV, RIG/PEG) and follow through with the decisions they made about services, enabled participants to feel in control of their care. However, *not having a choice* about services (e.g., community care services, patient-assisted suicide[1]) even if they were unlikely to engage with services was disconcerting for participants. People with ALS want "options" in care so they can make choices about their care.

7.3.3 Involve service users in research

As described in Chapter Three, ALS research in the Republic of Ireland is conducted primarily by (or in collaboration with) the Irish ALS research group based at the national ALS clinic and Trinity College Dublin. Research participants are sampled from the Irish ALS population-based register and a large percentage of people with ALS in the Republic of Ireland participate in research. In this study, participants expressed a strong desire to participate in research and it stands to reason that the above context influenced participants' propensity to participate in this study. Nonetheless, participants indicated that research participation was meaningful to them even though they were losing control

[1] Some people with ALS choose physician-assisted suicide (Veldink et al., 2002). Physician-assisted suicide is illegal in Ireland.

(and/or had already lost control) over most other aspects of their life. Indeed, simply *being asked by service providers to participate in research* was important for most participants.

Practice parameters in ALS care (Andersen et al., 2012; Miller et al., 2009a, b) do not include guidelines on how people with ALS are approached about research. Research in ALS is conducted by researchers but service providers may also have dual roles as service providers and researchers. Indeed, most clinical and scientific research in ALS is only feasible because researchers sample participants from the clinical setting. Protocols on conducting research with healthcare users are commonplace but no guidelines exist on how service providers and/or researchers might ask people with ALS to participate in research. This study found that people with ALS expressed a strong desire to participate in research. The evidence suggests that actively engaging ALS service users in research or simply offering people with ALS the choice to participate in research fosters control among people with ALS. Research as a component of ALS care is a new concept. The findings support the concept of dual care and research centres for people with ALS.

7.4 Responding to ALS service users' needs: Challenges ahead for the Irish healthcare system

This study found that people with ALS expect service providers to care for them. Indeed, most participants trusted healthcare professionals and relied heavily on healthcare professionals to guide them. The following sections outline how the Irish healthcare *system* might best support people with ALS as they live through insurmountable loss.

7.4.1 Invest in the pubic sector to render healthcare services to people with ALS

A key finding in this study is that people with ALS in the Republic of Ireland believe that the services they require are best delivered by public healthcare services. The range of services required by people with ALS is not routinely available to them in the private sector (Hardiman et al., 2003). A proportion of medical and allied healthcare professionals in the Republic of Ireland work in the private sector but private-based services are rarely multidisciplinary in nature. Private health insurance reimburses professional services only in part, and rarely (if at all) specialised equipment.

Overall, this study revealed that private-sector healthcare services were of little benefit to people with ALS. A large proportion of the sample accessed care through the private sector during the diagnostic phase of care but all moved willingly and/or without choice to public-sector healthcare services as they advanced in the disease. The following sections outline how the Health Service Executive can alleviate uncertainty for people with ALS and enable them to adapt to loss.

7.4.2 Educate healthcare professionals about ALS

A common complaint among participants in this study was healthcare professionals' lack of expertise in ALS care. Indeed, some participants believed that healthcare professionals had little if any knowledge of ALS. In some cases, healthcare professionals disclosed to participants that they knew little about ALS. These findings are consonant with experiences of services among people with ALS outside of Ireland (Hughes et al., 2005; O'Brien et al., 2012). ALS is a rare disease relative to other life-limiting conditions. However, ALS is a highly disabling condition and its disease trajectory is more certain than most other terminal conditions. People with ALS have care needs and become dependent on service providers. *Health care professionals in primary and community care services need to be informed about ALS.*

The Primary, Community and Clinical Care Directorate of the HSE needs to work closely with national ALS services to up-skill primary and community care healthcare professionals who provide services to people with ALS. The national ALS clinic co-ordinates care at a tertiary care level but it also has potential to render expertise to community-based healthcare professionals. The national clinic, the ALS voluntary sector (i.e. IMNDA) and Research Motor Neurone[2] have in the past co-ordinated and delivered study days for healthcare professionals in primary care. However, these initiatives require investment support from the HSE. Section 7.4.3 outlines how the national ALS clinic (with investment from the HSE) can expand its role in both clinical and educational fields.

7.4.3 Expand the national ALS clinic service

As discussed in Chapter Two, people with ALS who access specialised ALS clinics have better outcomes in survival without increased costs to services. However, people with ALS also believe they benefit from specialised tertiary services (Hogden et al., 2012; O'Brien et

[2] *Research Motor Neurone* is the research group who conduct research from the national ALS clinic.

al., 2011b). This study found that people with ALS valued specialised services provided to them by the national ALS clinic but most participants who accessed services at the national ALS clinic also felt that the national ALS clinic was under-resourced.

The findings highlight the need for the HSE to support and expand on existing services at the national ALS clinic. As already described, the national clinic is staffed by a range of healthcare professionals but most allied healthcare professions are staffed by just one healthcare professional in each field. Participants' primary complaint of the clinic was waiting times at the clinic. Allocation of additional personnel to the clinic would improve services at the clinic and reduce delays often encountered by service users.

As stated, the national clinic has an important role to play in educating healthcare professionals about care practice. Healthcare professionals at the national ALS clinic link closely with primary and community care healthcare professionals (Hardiman et al., 2002). However, community-based healthcare professionals encounter ALS service users less frequently than clinic-based healthcare professionals and the linking of care between the clinic and community services is primarily needs-based. In addition, people with ALS in Ireland ordinarily access community-based services prior to attending the clinic. Investing in the national clinic to render formal and structured clinical support to community-based healthcare professionals has the potential to improve the provision of services to people with ALS in community and primary care.

7.4.4 Support and co-ordinate care within Primary, Community and Continuing Care

There is general consensus that the Irish healthcare system lacks a properly resourced and integrated framework for the management of neurological conditions in the community setting (Hardiman, 2010). This study found that participants encountered delays in accessing community-based services and some had to fight for the right to access these services. The findings support the need for the HSE to develop and sustain primary and community care services. People with ALS rely heavily on a range of community-based services as they advance in the disease. People with ALS also *expect* integrated community-based healthcare services.

As described in Section 6.3.2, full access to multidisciplinary community care in the HSE is means-tested. Terminally ill service users are not automatically granted a medical card by

the HSE and the majority of people with ALS (whose means are above the threshold for automatic qualification) advance further along the disease trajectory before they receive a medical card on compassionate grounds. The findings support the need for people with ALS to have unrestricted access to public-sector healthcare services because the range of services they require is not available to them in the private sector. An adjustment in budget allocation within the healthcare system, so that funding 'follows the service user', would be of significant benefit to people with ALS.

7.4.5 Develop a clinical pathway in ALS from diagnosis to end-of-life care

People with ALS in this study trusted and depended on healthcare professionals but negotiating through the Irish healthcare *system* was challenging for most. The findings add weight to the rationale for developing an integrated framework for clinical care in ALS. An ALS health services research programme funded by the Health Research Board of Ireland is underway (Academic Unit of Neurology, 2013). The research programme will develop a framework for ALS care. The framework will account for estimated and actual costs of ALS care in the Republic of Ireland. The costing of care for people with ALS is an important component of the framework because it will map out how resources should be allocated to 'follow the service user' as they encounter mounting loss.

The framework will also incorporate a multidisciplinary-*palliative* approach to care. Living with ALS means living with insurmountable loss and never regaining what is already lost. The evidence suggests that people with ALS (in the absence of disease-modifying treatments) move to an altered state in 'mortal time' (McQuellon & Cowlan, 2000). People with ALS have a desire to be in control of when and how they engage with services but they also have a need to be reassured about palliative care. The evidence-based framework seeks to integrate palliative care into the management of ALS. Fully integrated palliative care services are not uniformly available to people with ALS in the Republic of Ireland. A framework for integrating palliative care into the management of ALS will be of benefit to both services users and service providers.

7.4.6 Support the ALS voluntary sector

The voluntary sector is an important advocate for people with neurological conditions in Ireland. This study found that people with ALS in the Republic of Ireland rely on the ALS voluntary sector to supplement their care. A number of participants received specialised

equipment and financial assistance toward home help services from the IMNDA. Home visits from a specialist nurse also reassured participants. These findings substantiate survey-based research on access to services for people with ALS in the Republic of Ireland (Hardiman et al., 2003).

The ALS population in Ireland remains stable (ca. n=265 April 2013) and the IMNDA continues to support people with ALS in Ireland. The IMNDA has the capacity to respond to service users' needs without delay because it does not impose restrictions on people's entitlement to access its services. There is a need for continued State investment in the ALS voluntary sector. The IMNDA is *primarily* dependent on fundraising by its own members. The State accounts for less than 25% of the Association's income and the economic crisis in the Republic of Ireland has impacted on the ability of its members and supporters to raise funds (House of the Oireachtas, 2012). The IMNDA had a positive impact on the lives of participants. Continued State investment in the IMNDA is warranted.

7.5 Conclusion

Chapter Seven has provided recommendations on how healthcare services might best render services to people with ALS and accordingly, assist people with ALS to come to terms with loss. The next and final chapter, the thesis conclusions, integrates and synthesises the research findings. The final chapter also provides direction for future healthcare services research in the ALS field.

Chapter Eight

Conclusions

8.1 Introduction

This study set out to identify key parameters of healthcare experiences among people with amyotrophic lateral sclerosis. Psycho-social processes that underpin how people with ALS (in Ireland, and in other health care systems) engage with services were, for the most part, unknown. Understanding how and why people with ALS engage with healthcare services is important because a greater understanding of the service user perspective enables healthcare professionals and policy makers to deliver services that best fit with service users' needs. Attention to the micro- and macro-context of participants' lives enabled the researcher to identify a broad range of conditions and consequences which shaped how participants interacted with healthcare services. Drawing from an analytical strategy that integrates structure and process (Corbin & Strauss, 2008), the researcher identified categories (i.e. variables) and relationships between them that shed light on key processes that underpin ALS service users' decision making in care.

The next two sections of this chapter debate key strengths and weaknesses of the study and summarise the key reflexive components of the study. The remainder of the chapter synthesises the research findings, outlines key theoretical and conceptual contributions of the research, and provides direction for future healthcare services research in the ALS field.

8.2 Strengths and weaknesses of the study

The findings in the thesis arise from 34 qualitative interviews with 34 people with ALS in one country / healthcare system. The findings are not representative of the ALS population but neither are they intended to be. The rationale for conducting the study was to explore in depth, the experiences of healthcare services among people with ALS and to develop theory about how people with ALS engage with services. This required using qualitative methods.

Corbin and Strauss (2008) suggest that making judgments about the 'quality' of qualitative research is difficult because so much depends on who conducts the research and the conditions under which the research was undertaken. The role of pre-existing knowledge

in grounded theory has long been debated. 'Classic' grounded theorists (Glaser, 1992) encourage minimum engagement with the literature prior to data collection and analysis because they feel that engagement with the literature limits the scope for developing new theory. It is worth noting that the researcher compiled multiple drafts of the literature review chapter during the course of the study. She conducted her review of the ALS service user experience of and preferences for care, *before* she embarked on the study. However, she argues that people invariably come to their research with preconceived notions and some degree of knowledge about their topic (Charmaz, 2006a; Corbin & Strauss, 2008). The researcher is an experienced clinician in ALS care and she was already familiar with ALS healthcare literature (including some of the literature on experiences of healthcare services among people with ALS prior to conducting the review). Familiarity with the literature was a prerequisite to secure funding for the research.

Each participant was interviewed on one occasion only and so the thesis fails to capture (potential) change in participants' perspectives on care over time. Grounded theory method necessitated obtaining a range of healthcare experiences among people with ALS and variation in contexts to their experiences of services. This meant sampling some participants who were at the end stages of their life. Returning to participants (who were still alive) to capture change or indeed to refine and verify theory (Charmaz, 2006a) might have been fruitful. However, this study was limited to the scope of a PhD. As stated in Chapter One, the purpose of undertaking the PhD was to develop substantive theory that explains how people with ALS engage with healthcare services. Developing substantive theory is a lengthy process because it involves ongoing sampling from multiple contexts relevant to the inquiry in order to 'saturate' the data. Conducting a second interview with participants was beyond the scope and time-frame of this study.

The researcher argues that focusing primarily on the service user experience of care, is a strength of this study. As stated in Chapters One and Three, the aim of the study was to identify the key parameters of healthcare experiences among people with ALS from *their* perspective. As described in Chapter Five (and Section 8.7 of this chapter), family emerged as primary context to the healthcare experiences of participants and so the findings are contextualised within participants' experiences of family. Section 8.7 emphasises the central role of family in the lives of people with ALS in Ireland. However, preferences for care often differ between people with ALS and their family. As discussed in Chapter Three,

the relationship between agency and structure is complex and multiple factors impact on agency for people with ALS including family. This study placed people with ALS at the centre of the inquiry in order to identify what is most important to them.

There are numerous sets of guidelines for judging qualitative research (e.g. Corbin & Strauss, 2008; Lincoln & Guba, 1985; Malterud, 2001; Morse et al., 2002). Criteria for evaluating qualitative research are also debated (Charmaz, 2006a; Mays & Pope, 2000). Overall, 'reliability' and 'validity' hold somewhat different meanings in qualitative research than they do in quantitative research. The 'dependability' (reliability) of the study refers to the extent to which the findings of the thesis are an account of participants' perceptions and experiences of their care. Extracts from the data in Chapters Four, Five and Six, are verbatim, and dense in substance and description. The 'credibility' (validity) of this study is a measure of how the procedures of the study did in fact tap into the phenomenon under investigation (healthcare experiences among people with ALS from the perspective of people with ALS). Investigation of participants' experiences of healthcare services from the 'ground up' identified key domains of care that are important to them and psycho-social processes underpinning their engagement with services not previously identified by researchers in the field (Sections 8.3 to 8.11). The credibility of the findings is also judged by the documented methodological procedures in Chapter Three that fit with the qualitative approach (i.e. Grounded Theory) taken by the researcher. Here, she sought to provide a detailed account of all the steps taken in the study (i.e. audit trail) so that the research could be *replicated* by other researchers as needed.

Qualitative researchers often choose to 'triangulate' their data. This typically involves collecting data from different groups/sources that might have different perspectives on the topic or in some cases, where a number of investigators work together on data collection and analysis (investigator triangulation). Some researchers might also use different methods in data collection to compare data (methodological triangulation). However, as stated, this PhD study was limited to exploring the ALS service user perspective on care and other methods for data collection in grounded theory (e.g. observation) were beyond the scope of the study.

More specifically, the quality of grounded theory research is judged by how well the data has been contextualised and 'saturated' for variation and meaning (Corbin & Strauss,

2008). Contextualised and 'saturated' data means that the data captures the complexity of the phenomenon under study and is therefore likely to be highly 'applicable' (i.e. relevant) to the practice setting. As described in Chapter Three, the researcher sampled for variation based on the emerging findings (i.e. theoretical sampling) and stopped sampling when no new data added meaning to the categories (i.e. ageing, trust, reassurance, etc) that emerged in the data. The relevance of the key findings to practice has been summarised in the conclusion sections of publications arising from the thesis and in the dissemination of the findings to clinicians in the practice setting.

8.3 Reflexivity

As described in Chapters One and Three, reflexivity refers to *how* researchers locate themselves in their research. Reflexivity constitutes awareness among researchers of how their own values and philosophical orientation impact on the data and on the decisions they make in the course of the study (Corbin & Strauss, 2008). As stated, the researcher subscribes to the view that researchers invariably enter into a study with certain opinions about what the research is about and how they might best conduct the research. She had worked 11 years as a clinician in ALS care before she embarked on the study, and her clinical expertise as an occupational therapist is primarily in neurodegenerative disease. She argues that it is not possible to 'bracket' her views and experiences from how she approached the study and responded to participants. The 'credibility' and the 'dependability' of this study are also judged by her role in the research.

First, as described in Chapter Three, the researcher views reality from a critical-realist perspective. Critical realists argue that reality is constantly shaped by structure and agency (emancipation), and agency matters to her. In this study, she acknowledges the central role of family in the lives of people with ALS but she is also attuned to the fact that family might also limit agency for people with ALS. In addition, the researcher conducted the study in a healthcare *system* which in her view assigns little importance to 'agency' for healthcare users. Hence, she sought to focus on the service user perspective. Her decision not to exclude participants with severe dysarthria or anarthria or indeed cognitive impairment (when such participants fitted with theoretical sampling) was strongly influenced by the importance she attaches to agency. Some people with ALS develop cognitive impairment but the researcher believes that their experiences are no less meaningful to them than are experiences to people with ALS who do not have cognitive

impairment. How people with cognitive impairment act/interact in the world also shapes reality.

Second, as stated in Chapter One, the researcher's motivation to conduct the study was to learn from the ALS service user perspective so that healthcare professionals might focus on what is *really* important to people with ALS in healthcare. She conducted the study in the context of being a research fellow *and* a healthcare professional. Conducting qualitative interviews with people who have a terminal illness can be emotionally challenging for researchers. However, the researcher was not emotionally challenged by conducting the qualitative interviews with participants in this study. Although she had already undergone training in qualitative interviewing, she also brought valuable skills to the interview setting. She had already identified from her experience in the field that people with ALS valued empathy over sympathy, and that they engaged meaningfully with her when she was empathetic to them. She always sought to be empathetic to participants over the course of the study.

Third, the researcher worked closely with the data in her effort to remain 'sensitive' to the data. As described in Chapter Three, she wrote reflexive memos following each interview which documented her observations and insights on the data. She also sought to remain in tune with how data inferred meaning (Corbin & Strauss, 2008). Here, she endeavoured to delineate between how much of the data arose through analysis (i.e. induction) and to what extent might some of the data have been based on preconceived ideas or existing knowledge about the data (i.e. deduction). As described in Chapter Three, grounded theory analysis is primarily inductive i.e. taking seriously the exhortation to seek to understand phenomena 'from the ground up', but some analysis and extrapolation in grounded theory research can be deductive in nature.

Overall, the findings of the study have been co-constructed (Finlay, 2002). Each interview was shaped by how the researcher responded to participants and by how the participants responded to her. The researcher captured the data from the viewpoint of participants and she then conceptualised their experiences to build theory about how they engage with healthcare services.

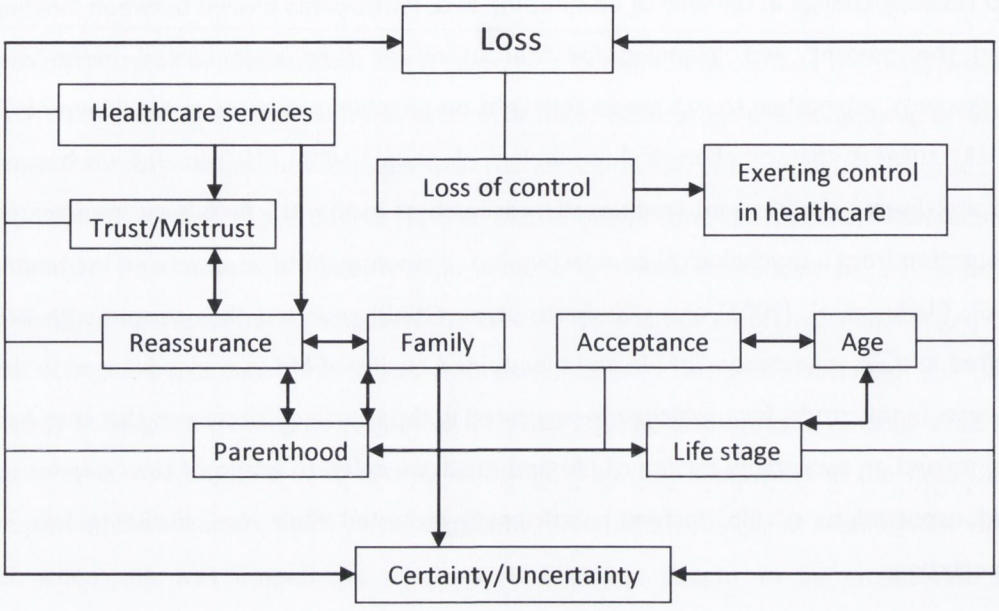
8.4 Adaptation in ALS: Understanding the socio-cultural context of people's lives

As described in Chapter Four, participants navigated back and forth between accepting and resisting change in the face of unremitting loss. Participants moved between 'holding onto the present' and 'planning for the future' as they encountered more loss. Participants' adaptation to ALS sheds new light on adaptive processes in ALS. Cupp et al. (2011) suggest that psychological wellbeing of people with ALS remains unchanged despite disease progression. Traditionally, researchers in the ALS field have investigated adaptation from a psychological perspective (i.e. 'response shifts' as discussed in Chapter Two). Clarke et al. (2001) and O'Doherty et al. (2010) reported that people with ALS shifted in their expectations of life and maintained quality of life as they advanced in the disease. In this study, few participants suggested in the course of interviews that they had maintained an acceptable quality of life and most struggled to let go of their previously held expectations of life. Instead, participants mourned their loss, including loss of expectations.

Drawing on qualitative social science methods (i.e. interpretive research) to capture the service user perspective, the findings reveal that people with ALS might not necessarily maintain an acceptable quality of life because *unremitting* loss is central to the experience of living with ALS. The researcher paid close attention to participants' social contexts (Corbin & Strauss, 2008). In doing so, she identified that adaptation in ALS is intertwined with other complex social processes (e.g., parenting, ageing) that shape how people with ALS respond to ALS and how they move at varying rates between resisting and accepting change. Adaptation in ALS is not simply a process of emotional or cognitive recalibration (Sprangers & Schwartz, 1999). Rather, people with ALS *construct* their trajectories as they engage with others. Understanding adaptation in ALS requires attention to psychological *and* socio-cultural contexts of people's lives.

Figure 5 (p.183) illustrates the primary categories (including the core category i.e. loss) that emerged in the data, and the relationships between these categories. The following sections of this chapter explain, in substantive terms, the relationships between the categories.

Figure 5 Coming to terms with loss



8.5 The central story line: Loss and control

In this study, loss was central to the experience of participants. As discussed in Chapter Four, people with ALS can accommodate to loss in their lives but contrary to previous literature in the field, participants encountered unremitting loss and did not regain normality or control in their lives (Foley et al., 2014a).

Loss is a universal concept (Murray, 2001) and feelings of loss (including loss of control) are common among people with terminal illness (Kutner et al., 1999). The meaning of loss for participants expanded across multiple dimensions including loss of control, loss of identity, loss of the future, loss of parenthood, loss of hope, loss of participation, and loss of independence. The terms 'perceived control' and 'locus of control' are already documented in social psychology research in ALS (Goldstein et al., 2003; Plahuta et al., 2002) but there had been a dearth of literature in social science fields that illuminate *how* people with ALS perceive control or indeed *why* they feel they lose control. A recent systematic review of qualitative research in ALS identified that people with ALS feel they do not have any control over the disease but exert control by making choices in life and in healthcare (Sakellariou et al., 2013). Sakellariou et al. (2013) called for further exploration of how people with ALS make decisions as they lose control over ALS and attempt to exert control in their lives.

The research yields new insights into how people with ALS experience loss (including loss of control) and how they exert control in order to adapt to loss. For participants, loss was unremitting, insurmountable, and accelerated with disease progression. Loss of control in ALS was central to the experience of loss. People with ALS lose control over their life and feel unable to control the losses they encounter in ALS. Adaptation for people with ALS involves adapting to loss without the ability to control loss. As described in Chapter Two, people with ALS have a strong desire for self-determination (Cooney et al., 2012). In this study, participants' need to feel in control of healthcare services arose from their experiences of loss and they exerted control over healthcare services as they searched for control. Exerting control in care enabled people with ALS to adapt to loss without feeling in control of their lives.

As stated, the findings pertaining to control and loss differ, in part, to those of Locock et al. (2009) who suggest that people with ALS can adapt to loss by restoring normality and control. Participants in this study did not restore normality or regain control in their lives. Some participants found solace in focusing on the present and by paying attention to what they had not *yet* lost but all were resigned to the likelihood of more loss. No participant suggested they managed to maintain or regain normality because they perceived ALS and the losses they encountered because of ALS (e.g., identity, hope, and expectations) as beyond their control. The data suggests that regaining normality or control (other than control in healthcare) are not components of the adaptive process in ALS.

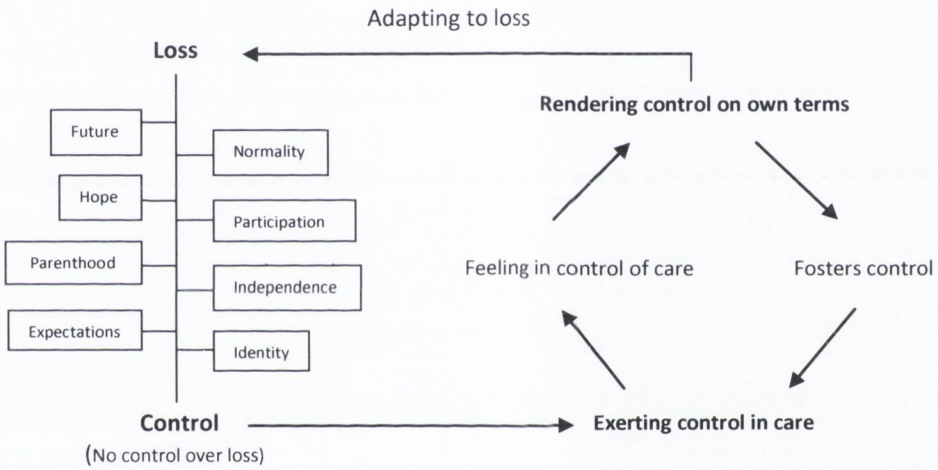
The findings suggest a clear distinction between people with ALS and other population groups who incur loss over a prolonged period of time and regain some of what they have already lost through periods of disease remission or (partial) recovery [e.g., multiple sclerosis, HIV] (Boeije et al., 2002; Russell & Seeley, 2010). ALS is rapidly progressive and has no periods of remission. As described in Chapters Two and Four, Charmaz (1995) suggests that people with chronic illness establish altered identities to adapt to loss. The clinical course of ALS differs greatly from the disease trajectories of chronic conditions. Most participants in this study indicated that ALS offered little time to adjust to their illness. Instead, they struggled to find a satisfactory existence because of the unrelenting nature of ALS and the subsequent unremitting loss they encountered in ALS.

The findings pertaining to loss and control resonate somewhat with those of other researchers who have investigated the experiences of people in palliative care. People with advanced cancer also encounter multiple losses and grieve current and future loss (Cheng et al., 2010). Broom and Cavenagh (2011) found that living with terminal illness comprised a liminal state, bereft of parts which people had previously recognised as components of their life. However, the researcher found no palliative care studies which illustrate the *unremitting* nature of loss as depicted in this study. Instead, Reeve et al. (2010) reported that people with terminal cancer managed to maintain an overall sense of wellbeing in the face of loss and Ranchor et al. (2010) found that people with cancer can regain some control as they live with illness. As described in Chapter Four, the disease trajectories in malignant disease are diverse. In comparison to ALS, people with terminal cancer can encounter phases of remission or stability that could offer hope of survival. In this study, participants did not maintain an overall sense of wellbeing because their losses never ceased at any point in time.

Anderson and Asnani (2013) identified that people with life-threatening illness seek to re-establish control in response to loss but little was known about the relationship between losing control in illness and exerting control in healthcare services. The findings shed light on how and why people with ALS engage with healthcare services. People with ALS engage with services to exert control over healthcare services and to feel in control of care because they perceive that they lose control over other aspects of their lives. Few researchers had reported on psycho-social processes surrounding control for people with ALS as they make decisions about their care. This thesis found that being in control of care was of huge importance to participants. Participants felt in control of care when they made decisions about their care. Feeling in control of care included the right to accept *and* decline services as participants pleased. Indeed, of central importance to participants was the freedom to decide *when* to engage with services. Rendering control to service providers also fostered feelings of control for participants because exerting control in healthcare services included the freedom to relinquish control on their own terms. Participants wanted to engage with healthcare services on their own terms and were likely to disengage from healthcare services if they did not feel in control of decision making about care. Similar to Cooney et al. (2012), this thesis found that feeling in control of care in ALS means having the right to decide about care.

Sakellariou et al. (2013) suggest that people with ALS seek to maintain control in their lives by making active choices about care. However, the thesis findings suggest that people with ALS construe control in life differently to how they construe control in healthcare services. Participants engaged with healthcare services on their own terms to be in control of care but equated living with ALS to a life of unremitting loss which included loss of control. Indeed, participants perceived they never regained control in their lives. In this study, people with ALS engaged with healthcare services on their own terms to adapt to loss. Figure 6 below illustrates how people with ALS adapt to loss by exerting control over healthcare services.

Figure 6 **Exerting control and adapting to loss in ALS**



The findings have implications for clinical practice in the field. The researcher paid close attention to how participants constructed their experiences of healthcare services in the context of social structures (Corbin & Strauss, 2008) but she did not identify structures beyond healthcare services that fostered feelings of control among participants. She postulates therefore that service providers are primary enablers (or at least have the potential to be primary enablers) for people with ALS to exert control. Second, losing control and fighting to remain in control comprised two central components in how participants interacted with service providers. Participants negotiated loss via engaging with services on their own terms. Hence, people with ALS are more likely to accept assistance from healthcare services when they perceive assistance to be on their own terms.

The findings demonstrate that control and loss are key variables that shape how people with ALS engage with healthcare services and that exerting control in healthcare services in an adaptive response to loss in ALS. The finding shed light on the complex relationship between loss and control in ALS. The relationship between loss and control in terminal illness is amenable to further theoretical development and testing. People with ALS experience unremitting loss. The magnitude of loss in ALS as perceived by people with ALS might correlate to disease progression in ALS. The findings are pertinent for research that is attuned to how people with conditions of similar disease trajectories construe loss and exert control in healthcare services.

8.6 Awareness of dying in ALS: Loss and open awareness

Awareness of dying in ALS (or what this might entail for people with ALS) was the sensitising framework for the research. The researcher held the assumption that the context of living with a rapidly progressive and terminal illness that does not offer periods of remission or even disease stability, might in itself be context to how and why people with ALS engage with healthcare services.

As discussed in Chapter Two, researchers have developed theories about awareness of dying (e.g., Glaser & Strauss, 1965; Mamo, 1999; Timmermans, 1994) and grounded theory methods have indeed evolved from a study on awareness of dying (Glaser & Strauss, 1965). Glaser and Strauss suggested that the context to care can influence people's awareness of dying and that varying levels of awareness among terminally ill people shape how they engage with healthcare professionals. Timmerman's (1994) suggestions that people with terminal illness might disbelieve a physician (i.e. suspended open-awareness), or dismiss information and hope for a better outcome (i.e. uncertain open-awareness), or accept death and prepare for it (i.e. active open-awareness), offer a perspective on contexts that might shape how terminally ill people confront mortality.

All participants in this study had received a diagnosis of ALS. All participants were "aware" they had ALS. Indeed, participants spoke *openly about being aware* of ALS and about the impact ALS had had on their lives. Moreover, the majority spoke about how they were experiencing and/or envisaged end-of-life care. Some participants were uncertain about time-frame but none dismissed the terminal nature of ALS. The majority of participants were resigned to further progression of the disease as they encountered more loss. No

participants ignored information provided by service providers. Even participants who felt they could never accept ALS engaged with service providers in order to cope with loss.

Seale et al. (1997) advised that open awareness of dying is desirable in the clinical setting because open awareness enables service users to plan end-of-life care and to have control over their care. In this study, not all participants were actively planning end-of-life care but they exerted control in terms of when and how they engaged with services to adapt to loss. Awareness of dying among participants arose from their experience of loss and from how they sought to accommodate to loss. Awareness of dying in ALS centers on awareness of loss.

8.7 Acceptance and decision making in ALS: Life-course trajectories and family roles

Participants' interactions with healthcare services were shaped by family roles and life stage (Foley et al., 2014b). Participants interpreted loss of independence in ALS as analogous to dependency in later life and indicated that later life was a more acceptable life stage to develop ALS. Acceptance of ALS was closely associated with participants' family roles and life stage. Participants were more accepting of ALS when they had reached later life, had fulfilled their ambitions and for those who had children, had already raised their children.

Young participants (and in some cases, middle-aged participants) were far less accepting of ALS than participants in later life. Young and some middle-aged participants wanted to live on to support their family (which included in some cases, dependent children) and engage with supportive care and life-sustaining interventions for the sake of family. However, participants in later life, regardless of their parenthood status, wanted to die soon. Those who had no children or a significant other felt they had more freedom than people with family to make decisions about their care. Participants with family drew support from family but also provided support to loved ones and expressed a strong desire to "be there" for family. Overall, participants suggested that it was more acceptable for people to die in "old age" and not acceptable to die as a young parent. Participants shared these views regardless of their age-differentiated roles, life stage, or parenthood status.

The life-course perspective is a sociological framework for analysing how social contexts and interactions shape people's lives over time (Hendricks, 2012). Alwin (2012) suggests that people's lives comprise a set of events, transitions, and trajectories and as people age, life transitions and trajectories are shaped by age-differentiated roles. Ageing is more than a biological process or chronological marker. Rather, people attach different meanings to different life stages (e.g., childhood, adulthood, and later life) and different stages of life are characterised by change in people's roles (Hendricks, 2012). People are also linked across family generations and people's trajectories and transitions are shaped by family (Macmillan & Copher, 2005). This study reveals how people with ALS make decisions about care in the context of life stage and family roles.

There are many definitions of age (e.g., chronological, biological, psychological) but Timonen (2008) argues that most attributes of age are in fact culturally determined. This study found that people with ALS perceive dependency in illness as analogous to later life experiences. Sanders et al. (2002) and Pound et al. (1998) noted how people in later life contextualised illness in the context of their age, and Gambling and Long (2012) described how people in earlier stages of adulthood struggled with physical disability they had associated with later life. In this study, participants, regardless of age or life stage, constructed later life as a period of resignation and acceptance.

Researchers have investigated the impact of ALS on family (e.g., Aoun et al., 2012; Ray & Street, 2007) but as stated in Chapter Five, few had explored how people with ALS (from their perspective) interact with their family as they engage with healthcare services. Participants sought to reassure family. They also felt reassured about their future care when they engaged family members in their care and when family members involved themselves in their care. The majority of participants in the study lived with their significant other and were becoming increasingly reliant on family to provide care. It is possible that this context (living with and becoming reliant on family) shaped participants' preference to involve family in decisions about care. It is also possible that the wider (i.e. macro) context of formal health and social care services in Ireland also impacts on how people with ALS in Ireland engage family in their care. Homecare services in the Republic of Ireland are under-developed (Timonen et al., 2012) and the level of formal service provision in the Republic of Ireland is low compared to other developed welfare states

(Tussing & Wren, 2006). This means that people in Ireland are more reliant on their family members than people in welfare states with more extensive formal care provision.

Drawing on the life-course perspective, researchers have argued that parenthood is a pivotal life-course transition, and that people's experience of parenting has a significant impact on their lives and shapes their trajectories (Umberson et al., 2010). Parenting young children exposes parents to life stressors (Evenson & Simon, 2005) but relationships with children are also salient to parents (Knoester, 2003; Mandemakers & Dykstra, 2008). Yellen and Cella (1995) found that having dependent children predicted people's decision to engage with aggressive cancer treatments. Nilsson et al. (2009) found that patients with advanced cancer who had dependent children were more distressed and more likely to choose aggressive treatments than patients with no dependent children. Hirano and Yamazaki (2010) reported that people with ALS who chose long-term ventilation had a desire to see their children and/or grandchildren grow up but little was known about *how* people with ALS engage with healthcare services in the context of parenthood. This study found that parenting in ALS was important for all parents but young and in some cases middle-aged participants, were far more distressed than participants in later life about their children's imminent loss of a parent. Indeed, participants' contradictory emotions surrounding the desire to live on with ALS and the desire to die soon from ALS were, for the most part, related to their perceived responsibilities as parents (or to their freedom from such responsibilities). Having children who were (seen to be) self-sufficient (or indeed having no children at all), relieved participants' anxieties about the future and fostered acceptance.

The findings show that family care-giving in ALS is reciprocal: people with ALS receive care from family but also seek to reassure their family and make decisions about care in the interest of their family. Macmillan and Copher (2005) argue that the life course is not simply defined by transitions and trajectories, but also by how multiple role trajectories within families interlock to configure roles and pathways within families. This study reveals diversity in generational relationships among people with ALS. Participants of different age and life stage were dependent (or were becoming dependent) on their family but they also struggled to maintain their care-giving role to their spouse and to generations below them (i.e. children, grandchildren).

In biographical accounts among people with terminal illness (Reeve et al., 2010), researchers have tended to focus on individual and personal biography without explicit reference to social contextual factors. Hendricks (2012) states that social structures (e.g., family) have both implicit and explicit temporal dimensions, that impose pathways and sequence individual biographies. In this study, the researcher paid close attention to how people constructed their experience in the context of social structures (Corbin & Strauss, 2008) and in doing so, she found that family (including participants' roles within their families) impacted on the decisions participants made about their care. Elder (2003) has argued that people construct experiences interdependently and that their socio-historical roots (e.g., family) shape their transitions and trajectories. The findings demonstrate that family is *the* primary context to how and why people with ALS in Ireland make decisions about their care.

The findings have implications for research and clinical practice in the field. International guidelines in ALS care (Andersen et al., 2012; Miller et al., 2009a, b) refer to timing of interventions based on disease progression. Clinicians advocate regular multidisciplinary healthcare intervention for people with ALS and when clinically appropriate, encourage early initiation of life-sustaining interventions to improve survival. In this study, people with ALS engaged with healthcare services in line with their own life transitions and trajectories and constructed their experience of healthcare services at both family and societal levels. Disease progression on its own does not determine how and why people with ALS engage with healthcare services.

The researcher argues that service providers need to pay close attention to life-course trajectories and family relations of people with ALS by asking people with ALS about their roles within family. The family roles that people with ALS adopt shape their response to ALS and their decisions about care. Traditionally, clinicians and researchers in the ALS field have conceptualised family and family care burden without sufficient attention to the roles people with ALS adopt within their families. People with ALS have roles within family (e.g., as parent, spouse/partner) even though they become physically dependent on family. The findings are relevant for future research that is attuned to the impact of family relations on decision making in care.

8.8 Trust and reassurance in ALS care: The micro-level of healthcare experiences

Participants engaged with healthcare professionals on their own terms but they also placed trust in healthcare professionals and had a need to be reassured by them. Participants also trusted and felt reassured by service providers who enabled them to be in control of their care. Participants' primary expectation of healthcare services was to secure services when they wanted them and they sought reassurances from healthcare professionals about end-of-life care. Moreover, participants valued meaningful relationships with healthcare professionals and trusted healthcare professionals who were attuned to how participants were coming to terms with loss.

Most of the findings pertaining to reassurance and trust are new to the ALS field. Brown (2003) and Bolmjso (2001) identified that existential issues are important to people with ALS and Beisecker et al. (1988) found that people with ALS expected honesty and sensitivity from healthcare professionals. However, this thesis is the first study to identify that reassurance and trust are key variables that shape how people with ALS engage with healthcare services. People with ALS feel reassured about their care when they feel in control of their care and have a need to trust healthcare professionals as they plan for and/or engage with end-of-life care. Most importantly, people with ALS trust healthcare professionals who demonstrate empathy in the clinical encounter and trusting relationships with healthcare professionals also reassure them. However, mistrust of healthcare professionals in the clinical encounter can foster feelings of uncertainty among people with ALS and diminish their confidence in healthcare services

The findings resonate to some extent with those of other studies in palliative care. Steinhauser et al. (2000) also found that terminally ill people seek reassurances from healthcare professionals about end-of-life care. Terminally ill people in other population groups (e.g., cancer, chronic obstructive pulmonary disease, congested heart failure) also have a need to trust healthcare professionals as they approach death (Heyland et al., 2006), expect healthcare professionals to communicate openly with them (MacPherson et al., 2013), and value honesty and sensitive sharing of information (Janssen & Macleod, 2010). Evans et al. (2012) also found that service users' trust (or mistrust) in healthcare professionals is calibrated by how they perceive healthcare professionals' approach towards them. Empathy and genuine concern from healthcare professionals engenders

trust in the service user but a lack of sensitivity on behalf of service providers diminishes service users' trust in service providers.

Carr (2001) suggests that negotiating trust in service providers is an important process among people with life-limiting illness. Thorne and Robinson (1988) argue that having trust in service providers can be personally supportive for service users and trusting relationships between service users and service providers are likely to increase service users' satisfaction with healthcare services. Rowe and Calnan (2006) argue that trust in healthcare services and satisfaction with healthcare services, are conceptually different. Service user satisfaction with healthcare services is an evaluation of distinct healthcare dimensions but trust reflects a *personal* commitment that service users and service providers demonstrate in the relationship between them.

The findings point to the importance of trust and reassurance in ALS care. People with ALS exert control in their interactions with healthcare professionals but also value meaningful relationships with healthcare professionals. As described, the researcher was attuned to both micro- and macro-contexts of participants' experiences of services. The findings reveal that people with ALS construct trust and reassurance differently in their interactions with healthcare professionals (i.e. meso-level) than they do when they think about their healthcare system (i.e. macro-level). As described in Chapter One, this study was conducted in the midst of a severe recession (Thomas et al., 2012), and some participants expressed concerns about the Irish healthcare system in the context of restraints imposed on healthcare expenditure (Keegan et al., 2013). However, feeling trusting of and feeling reassured by healthcare professionals alleviated their concerns and anxieties about future care. The findings indicate that 'trust' and 'reassurance' for people with ALS operate predominantly at the micro-level (i.e. clinical encounter) of healthcare experiences (Foley et al., 2014c).

8.9 Perceptions of end-of-life care in ALS: Life prolonging versus life ending

As discussed in Chapter Five, participants resigned themselves to the likelihood of becoming more dependent on healthcare professionals as they advanced in the disease and actively engaged with services to cope with loss. Resigning to the inevitability of disease progression prompted some participants to plan for end-of-life care, and planning end-of-life care lessened anxiety. As described in Chapter Two, palliative care is defined as

care that alleviates physical, psychological, and existential distress. Key opinion leaders in ALS care (Andersen et al., 2012; Bede et al., 2011) contend that service users should be approached from a palliative perspective and that specialist palliative care services should be available to service users from the time of diagnosis (if not always directly involved). In the Republic of Ireland, specialist palliative care services are not routinely available to people with ALS but service providers estimate that a significant portion of people with ALS in the Republic of Ireland engage with palliative care services at end of life.

Participants' primary expectation of healthcare professionals during end-of-life care was that they would alleviate "suffering" that participants felt they were likely to encounter at end of life. Few participants associated life-sustaining interventions in ALS with a palliative care approach even though non-invasive ventilation and gastrostomy feeding alleviate symptoms in ALS (Andersen et al., 2012; Miller et al., 2009a). People with ALS in Ireland engage with non-invasive ventilation and gastrostomy feeding[1] but participants did not construe these interventions as interventions to ease potential "suffering". Moreover, some participants understood non-invasive ventilation and gastrostomy feeding, to sustain life beyond the time-frame that they ordinarily sustain life for people with ALS. Participants also construed gastrostomy as a life-sustaining intervention even though gastrostomy might not necessarily improve survival in ALS (Andersen et al., 2012).

A key finding of this study is that life-sustaining interventions can provoke anxiety and uncertainty for people with ALS if their function is to prolong life for those who anticipate (or experience) "suffering". Participants struggled between wanting to live on with ALS and resigning to death in the face of inevitable disease progression, but engaging with life-sustaining interventions that had the potential to prolong the duration of distress was unacceptable to most. The findings resonate to some extent with those of other studies on service users' preference for end-of-life care and their expectations of healthcare professionals in palliative care. Cooney at al. (2012) also found that people with ALS wanted to avoid interventions that had the potential to sustain life beyond the natural course of the disease. Heyland et al. (2006) found that terminally ill people with cancer, heart failure, and obstructive lung disease sought to avoid life-sustaining support. Rodriguez and Young (2006b) also found that service users questioned the benefit of life-

[1] Unpublished data from Irish ALS population-based register

sustaining interventions in situations where they perceived their quality of life as unacceptable.

Participants' perceptions of life-sustaining interventions are contextualised within ALS service users' experiences of services in Ireland. Invasive ventilation is not readily available to people with ALS in Ireland and people with ALS in Ireland are rarely intubated. The findings differ from those of Lemoignan and Ells (2010), in Canada, who reported that people with ALS made a clear distinction between non-invasive ventilation and invasive ventilation and understood non-invasive ventilation to be symptomatic care rather than life-sustaining care. As noted in Chapter Five, few participants made explicit reference to invasive ventilation. People with ALS in other healthcare systems where invasive ventilation is more readily available (e.g., Japan, United States) were glad they chose invasive ventilation (Moss et al., 1993), expressed mixed feelings about invasive ventilation (Hirano & Yamazaki, 2010; Moss et al., 1996) and regretted choosing invasive ventilation (Cazzolli & Oppenheimer, 1996; Hirano & Yamazaki, 2010). It is possible that people with ALS in other healthcare systems that do not routinely offer invasive ventilation have similar perceptions about invasive and non-invasive ventilation to participants of this study.

The findings in this thesis shed light on perceptions of life-sustaining treatments and of palliative care among people with ALS in Ireland. Service users' perceptions and understandings of 'palliative' and of 'life-sustaining' interventions in ALS might differ from those of healthcare professionals who render services to them. Clinicians regularly advocate early initiation of non-invasive ventilation and gastrostomy feeding to improve survival and/or to alleviate symptoms. However, many participants were indifferent to these interventions because they did not perceive them as interventions to alleviate "suffering". Participants were averse to the notion of prolonging life in ALS for the sake of longevity alone and most participants nearing death wanted to die soon. Participants did not associate palliative care with care before the dying phase even though the care approach in ALS is deemed by healthcare professionals in the field to be palliative from the point of diagnosis (Andersen et al., 2012; Oliver et al., 2006). Moreover, people with ALS might not necessarily equate alleviation of symptoms with alleviation of suffering. Research into how *people with ALS* construe suffering is required (Foley et al., 2014c).

8.10 Contexts of certainty and uncertainty in Irish healthcare services for people with ALS

Participants' uncertainty about healthcare services is a key finding of the study. As described in Chapter Six, the majority of participants had had little need to engage with healthcare services prior to the onset of ALS and many admitted that the Irish healthcare system was a challenging system to negotiate. Participants' uncertainty about healthcare services centred on uncertainty about what healthcare services offer to people with ALS, uncertainty about how the healthcare system functions as a whole, and uncertainty about the future of the Irish healthcare system.

Han et al. (2011) argue that alleviating uncertainty among healthcare users is an important aspect of healthcare provision. Researchers in other fields of care (e.g., cancer, multiple sclerosis) found that effective communication and provision of information enabled service users to cope (Nanton et al., 2009; Thorne et al., 2004) but that fragmentation in service delivery and in knowledge exchange between service providers and terminally ill service users exacerbated service users' concerns about services (McIlfatrick, 2006). Prior to the onset of ALS, few participants were cognisant of how their healthcare services functioned. Indeed, few participants in this study felt they were knowledgeable about the structures that comprise the Health Service Executive (HSE). Those who reported having searched for information felt that HSE public-interface sources (e.g., HSE website including HSE descriptors of how services are rendered) were of little benefit to them. Overall, the majority of participants felt "disconnected" from the HSE and they struggled to negotiate their way around the HSE.

Babrow (2001) argues that effective communication between service providers and healthcare users is essential to alleviate uncertainty among healthcare users. In this study, participants were uncertain about the HSE but they engaged with service providers in the clinical encounter 'to become more certain' about healthcare services in order to adapt to loss. Participants felt that service providers who had communicated effectively with them had alleviated their uncertainty and that service providers who did not communicate openly with them, failed to alleviate their uncertainty. As described, the researcher drew from an analytical strategy that enabled her to consider both micro- and macro-contexts (Corbin & Strauss, 2008) that shape how people with ALS engage with services. In this study, 'becoming more certain' about care or 'feeling uncertain' about care, centred more

on the type of *interaction* between participants and service providers (i.e. meso-level) than on the high level of ambiguity most participants experienced in the wider context (i.e. macro-level) of the Irish healthcare system. In many cases, healthcare professionals had a positive impact on the lives of participants. The findings reveal that healthcare professionals are also active agents in ALS care.

8.11 Inequity in healthcare for people with ALS: The Irish perspective

As described in Chapter Six, the Irish healthcare system is complex (Ryan et al., 2009; Smith, 2010). It comprises a hybrid system of public and private care and has complex user charge structures that allow private healthcare to take place within public hospital facilities. A medical card entitles service users to public healthcare services free of charge, and is means-tested. Primary care services are delivered by public sector healthcare professionals (excluding general practitioner [GP] services who work in a private capacity) and service users who access GP services without a medical card are charged. Access to other primary care services without a medical card is, in many cases, restricted (Nolan & Smith, 2012). Indeed, this study found that participants encountered difficulty accessing community-based allied healthcare and nursing services, without a medical card.

Access to healthcare services is a human right (Papadimos, 2007). The basic human right to healthcare means there is a collective and moral obligation on societies to ensure that people have access to healthcare; that healthcare services are owed to people who have that right; and as a human right, the right to healthcare is ascribed to all (Denier, 2005). Equity in healthcare includes *access* to healthcare, and because access to healthcare is a human right, ethicists argue that access to healthcare should be universal (Bradley, 2010). Central to egalitarian ideologies is that consumption of healthcare should not be based on ability to pay. In liberal systems, in contrast, healthcare is distributed in the private market based on ability to pay and individuals have exclusive rights to consume what they pay for.

In the Republic of Ireland, the 'public versus private' healthcare debate is intrinsically linked to discourse surrounding access to healthcare services (Burke, 2009). The mix of public and private provision and financing across different services has created a system where egalitarian *and* libertarian principles operate for different people in different services (Smith & Normand, 2011). As described in Chapter Six, participants with private health insurance reported few delays in accessing services from private-based

neurologists (including the diagnostic tests that followed) and inpatient care in private hospitals. Participants encountered restrictions and delays in accessing a broad range of primary care services without a medical card (or prior to obtaining a medical card) regardless of 'private' or 'public' status. Specialised multidisciplinary care was restricted, primarily, to the public sector. The Irish healthcare system discriminated against *both* 'public' and 'private' service users depending on what types of services they needed and how these services were financed. People with ALS in the Republic of Ireland encounter difficulty obtaining services in both public and private healthcare sectors.

8.12 Recommendations for health services research in ALS

As described, the researcher set out to identify key parameters of healthcare experiences among people with ALS. This study captured variation in ALS service users' experiences of healthcare services in an Irish context but at substantive level, the findings also enable comparisons between different systems and further development of theory in relevant substantive areas.

Research on experiences of healthcare services among people with ALS drawing on life-course theory might shed more light on how people with ALS make decisions about care and why specific interventions (including the *timing* of these interventions) mean more to people with ALS than other interventions. This study is the first to report directly on how life-course trajectories of people with ALS impact on the decisions they make about their care. Further research into the dynamics of family relations in ALS care with particular attention to the role of parenthood at different life stages, would shed further light on the complexity of healthcare provision in ALS care.

Participants' perceptions of service providers were formed predominantly in the clinical encounter. Despite variation in service allocation among participants, the researcher discovered that "personal" and meaningful interactions with service providers helped the majority of participants cope with ALS. There is a dearth of literature in the ALS field focused directly on key psycho-social processes that underpin positive (or negative) experiences in the clinical encounter, and that which exists has focused predominantly on disclosure. Guidelines on the clinical management of ALS (Andersen et al., 2012) provide direction on how to 'communicate the diagnosis' and how to communicate with service users who develop communication difficulties, but little attention is paid to what

conditions overall might enable service providers to communicate empathically with people with ALS in what is a "human" encounter. Research into the idiosyncrasies of the clinical encounter in ALS care during *and* beyond the diagnostic phase of care, would be of benefit to clinicians who aim to support people with ALS in the clinical encounter.

Perceptions of palliative care among people with ALS and among the community of ALS healthcare professionals, warrant further investigation. The researcher discovered that participants, for the most part, construed palliative care as care rendered during end-of-life care only. Participants indicated that care should focus on symptom relief but none suggested that their care had been palliative from the point of diagnosis. People's (with ALS) understandings of palliative care shape the clinical encounter but underlying tendencies that trigger how and when people with ALS engage with specialist palliative care services remain unclear. Research on the meaning of palliative care among service users and service providers in ALS care would illuminate how, when, and why people with ALS engage with palliative care services.

Loss and control emerged as *the* central concepts from this study. Living with ALS means living with loss and never regaining what is already lost. In this light, how can service providers measure the impact of healthcare services for people with ALS if unremitting loss is central to the experience of ALS? The notion that people with ALS do not regain what they have lost is new and likely to challenge healthcare professionals who believe they have the capacity to sustain what is important to people with ALS. People with ALS resign themselves to loss and they exert control in healthcare services in order to adapt to loss. A measure focused on how having a say over healthcare services enables people with ALS to cope with ALS was warranted. As reported in Chapter Three, a scale has been developed (Appendix F) from the data and it is being used in a study aimed at mapping out optimal care for people with ALS. This scale measures the control domain in ALS care. The measurement of this key domain will enable service providers to test relationships between what is most important to people with ALS in healthcare and other outcomes of care.

Research on *how* healthcare services are delivered to people with ALS in Ireland is necessary. This study identified key variables that shape how people with ALS engage with services but researchers have not yet mapped out how key structures within Irish

healthcare services impact on outcomes in service delivery for people with ALS. This study found that people with ALS construct their own trajectories as they live with ALS but engaging with healthcare services also shapes their experience of ALS. Harwood and Clark (2012) argue that healthcare systems do not determine how people make decisions about health but structures provide conditions that facilitate and constrain people engaging with services. As described in Chapter Seven, an ALS health services research programme has been funded by the Health Research Board of Ireland (Academic Unit of Neurology, 2013). This research programme will describe the care pathway in ALS based on a multifaceted matrix of clinical, cognitive, existential, social, and economic factors. The intended outcome of the research package will be a framework for the delivery of healthcare services to people with ALS.

8.13 Concluding remarks

In Chapter Two, the researcher postulated that the terminal nature of ALS might in some way be context to how people with ALS engage with healthcare services. As described in Chapter Three, a key tenet of critical realism or complex realism (Clark et al., 2008) is that it seeks to explain outcomes in the natural world where underlying relations and tendencies (e.g., patterns of disease progression) can shape events and outcomes (e.g., health service delivery) in the real world. However, the researcher as a critical realist is also attuned to the personal meanings people with ALS construct (e.g., awareness of dying) as they live with ALS. This study has shown that loss is central to the experience of people with ALS and that it is the "human" perspective that shapes how people with ALS engage with healthcare services.

How people with ALS engage with healthcare services *is* complex but the researcher has demonstrated that it is possible to capture complexity of healthcare experiences among people with ALS. Attention to the personal experiences of people with ALS and contexts (micro and macro) in which people with ALS adapt to ALS and engage with healthcare services to adapt to ALS, was central to capturing this complexity. Critical realists argue that qualitative research methods are well suited to understanding complexity in the 'real' world because critical realists seek to understand complexity at a cumulative *and* individual level (Clark, 2009). It was important to capture the individual perspectives of people with ALS about their healthcare services because a greater understanding of the

service user perspective enables (or indeed, places expectations on) service providers to respond to what is important to people with ALS.

This study reveals that people's (with ALS) perceptions of living with ALS and of the healthcare services they engage with to cope with ALS, shape the clinical encounter. However, practice parameters in ALS care (Andersen et al., 2012; Miller et al., 2009a, b) are based on the consensus of experts in the clinical field and on evidence which, for the most part, does not include the service user perspective on care. This thesis captures the perspectives of people with ALS who access healthcare services and the study is the first to map out key parameters of healthcare experiences 'grounded' in the lives of people with ALS. People with ALS value healthcare services but family, parenthood, and life stage are primary contexts to how people with ALS in Ireland engage with healthcare services.

This thesis has described, explained, and conceptualised healthcare experiences among people with ALS from the viewpoint of people with ALS. This study is the first to explain in substantive terms, how and why people with ALS engage with healthcare services. This research is important because it has identified real-life concerns among people with ALS as they face mortality and engage with healthcare services. In Chapter One, the researcher argued that the primary goal of healthcare services in ALS is to achieve meaningful outcomes for people with ALS. Responding to what is most important for people with ALS is challenging for service providers because so much of what shapes how people with ALS engage with services exists outside of healthcare services. Nevertheless, people with ALS rely on healthcare services to cope with loss and healthcare professionals have the potential to bring about meaningful outcomes in the lives of people with ALS. The challenge ahead for researchers and healthcare professionals in ALS care is to measure the benefit of healthcare for people with ALS by incorporating into such measurements the domains of care that matter to people with ALS.

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Appendix A

Reference	*n*[1] *(N)*	Location / sampling	Method	Aims of study	Main findings
Achille & Ogloff 2003	n=44	multiple locations; United States, Canada & United Kingdom	Quantitative cross-sectional; questionnaire and multiple instrument measures	To investigate attitudes towards assisted suicide among ALS patients. To describe the characteristics of ALS patients who consider assisted suicide.	The majority of participants felt that assisted suicide was morally acceptable but only 7% of participants indicated they would use it if available. Participants' contemplation of assisted suicide was associated with feelings of hopelessness and depression.
Albert et al. 1999	n=121	ALS tertiary centre NY, USA	Quantitative longitudinal (prospective); clinical evaluation, multiple questionnaire and instrument measures	To determine if ALS patients' preferences for care relate to later treatment choices.	Patients made choices about care and followed up on choices consistent with their preferences. Preferences for care also changed over time. Patients who had a greater attachment to life found life-sustaining treatments to be acceptable.
Albert et al. 2009 [Rabkin et al. 2006]	n=71	ALS tertiary centre NY, USA	Quantitative longitudinal (prospective); clinical evaluation, multiple questionnaire and instrument measures	To analyse the use of medical and supportive care by ALS patients prior to death or tracheostomy.	Patients had a desire to live on and use services up to the point of death or tracheostomy. Choosing long-term ventilation was determined by patients' own perceptions of their health status, feelings of optimism and hope, positive appraisal of life and satisfaction with life. Only 50% of patients remained hopeful and optimistic after an average of 33 months on ventilation.
Baxter et al. 2013	n=20 (N=37)	ALS tertiary cente Sheffield, UK	Qualitative; semi-structured interviews	To explore the experiences of non-invasive ventilation among ALS patients and their carers following physicians' recommendation to use non-invasive ventilation.	Positive effects of non-invasive ventilation included improved breathing and energy maintenance. Patients reported a range of barriers to using non-invasive ventilation including sleep disturbance and discomfort using technical appliances.
Beisecker et al. 1988	n= 41	ALS / Muscular Dystrophy tertiary centre Kansas, USA	Qualitative; semi-structured interviews	To identify ALS patients' perspectives on care providers.	Patients reported expectations of multidisciplinary care in the form of information, emotional support and assistive devices. Patients expected healthcare providers to convey hope and to communicate honestly and respectfully with them.

[1] '*n*' denotes ALS participants. '*N*' denotes total sample [i.e. includes carers and/or service providers and/or other diagnostic groups].

Appendix A

Belsh & Schiffman 1996	n=64	USA (81%) and other countries / *ALS Digest*	Survey (via email); questionnaire	To investigate the frequency of misdiagnosis in ALS based on patient response. To identify demographic and epidemiologic factors that distinguish ALS patients who are initially misdiagnosed.	27% of patients reported at least one misdiagnosis. The mean length of time to receive a diagnosis was longer for patients who received an initial misdiagnosis compared to patients who were not misdiagnosed. Misdiagnosis may have resulted in refusal to participate in clinical drug trials.
Bennett et al. 2009	n=27	ALS tertiary centre Preston, UK	Audit; questionnaire	To analyse the benefits associated with specialist nurse lead hospice-based ALS clinics.	Patients expressed satisfaction with hospice-based clinics and valued the intimate environment of the hospice setting.
Bolmsjo 2001	n=8	Neurology clinic Lund, Sweden	Qualitative; semi-structured interviews	To explore existential issues for ALS patients in palliative care.	Existential issues are of great importance for ALS patients in palliative care. Central to care is the need to be respected.
Brown 2003	n=6 (N=21)	3 counties southern England (Motor Neurone Disease Association UK register) UK	Qualitative; semi-structured interviews	To explore and compare lay and professional values in ALS care.	The professional view is detached from the experience of living with ALS. Patients' needs are broad and person focused.
Brown et al. 2005	n=11 (N=37)	3 counties southern England (Motor Neurone Disease Association register) UK	Qualitative; structured interviews[2] Questionnaire based survey[3]	To investigate whether services meet the needs of ALS patients and their carers. To explore expectations of service delivery for ALS patients, carers and service providers and compare these with services provided.	Patients expressed dissatisfaction with timeliness of services. Patients identified positive qualities in care professionals. Patients reported a lack of knowledge among service providers and deficiencies in multidisciplinary care including: specialised equipment; availability of care co-ordinators; and respite care.
Budych et al. 2012	n=20 N=107	Specialist centres (rare diseases) across Germany. Associated voluntary organisations	Qualitative; structured interviews	To describe the experiences of patient-physician interaction among people with rare diseases (including ALS).	Patients perceived physicians as lacking in expertise and they expressed dissatisfaction with how physicians interacted with them.

[2] Interviews with *ALS* participants only
[3] Not used with *ALS* participants

Appendix A

Caligari et al. 2013	n=35	Italian ALS website http://www.slaitalia.it Five major eye-tracking communication-device (ETCDs) trading companies in Italy	Quantitative; multiple questionnaire and instrument measures	To evaluate the effect of ETCDs on patient disability and quality of life (QoL). To evaluate the effect of ECTDs on patient satisfaction as an assistive-technology user.	The use of ETCDs was associated with an increase in QoL among participants and with an increase in participants' satisfaction as assistive-technology users
Callagher et al. 2009	n=23	ALS tertiary centre Preston, UK	Audit: questionnaire	To evaluate the impact of a fast-tracking system for diagnosing ALS against traditional pathways.	Patients expressed satisfaction with privacy, time, and sensitivity when provided with a diagnosis.
Cazzolli & Oppenheimer 1996	n=75	Pennsylvannia, Ohio (local ALSA chapter registers) ALS support group, West Virginia USA	Qualitative; structured interviews	To compare the use of tracheostomy ventilation and nasal-positive-pressure ventilation among ALS patients.	All patients who chose nasal-IPPV expressed satisfaction with choosing nasal-IPPV. The majority of patients using long-term mechanical ventilation received tracheostomy without advance directives in place. The majority of patients would not have chosen long-term mechanical had they known the burden of care associated with long-term ventilation.
Chio et al. 2008	n=60 (N=120)	ALS tertiary centre Turin, Italy	Survey; questionnaire	To evaluate preferences for communication and information seeking behaviour in ALS patients and their carers.	Patients reported satisfaction with communication of the diagnosis. Patients had a desire to obtain information and search for information both within and outside healthcare services.
Foley et al. 2007	n=5	ALS tertiary centre Dublin, Ireland	Qualitative; semi-structured interviews	To explore the concept of QoL for ALS patients and effect of healthcare services on perceived QoL.	Patients reported healthcare services to have a positive effect on QoL.
Ganzini et al. 1988	n=100	ALS tertiary centre Oregon, USA	Quantitative cross-sectional; multiple questionnaire and instrument measures	To determine attitudes about assisted suicide among ALS patients.	Feelings of hopelessness predicted patient preference for assisted suicide. Patients who preferred assisted suicide had greater distress, were less religious and had a lower quality of life than those who did not wish to hasten death.
Gruis et al. 2011	n=63	ALS tertiary centre Michigan, USA	Survey (via telephone); questionnaire	To identify patients' satisfaction with low-level assistive technology.	Patients reported high levels of satisfaction with assistive devices.

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Hardiman et al. 2003	n=94 (N=282)	Ireland (Irish Motor Neurone Disease Association register)	National survey (via telephone); questionnaire	To identify access to healthcare services for ALS patients in Ireland.	Patients reported low to moderate levels of access to community-based services. Private health insurance was of no advantage to obtaining services or specialised assistive devices and patients accessed services from the voluntary sector to supplement their care. Patients were unaware of the specific roles of healthcare professionals.
Hirano et al. 2006	n=184	Japan (Japenese ALS Association register)	Mixed method; Semi-structured interviews (n=27), multiple questionnaire and instrument measures (n=157)	To examine needs and experiences of ALS patients on long-term ventilation.	Patients reported fear about using mechanical ventilation and about the potential for ventilation to increase burden of care on their families. Psychosocial support with ventilation fostered hope.
Hirano & Yamazaki 2010	n=50	Japan (Japanese ALS Association register)	Mixed method; Semi-structured interviews, multiple questionnaire and instrument measures	To examine decision-making for ALS patients in mechanical ventilation.	Patients' decision to undergo mechanical ventilation was associated with family support. Patients who did not receive sufficient information about mechanical ventilation reported conflicting feelings about ventilation and/or regret about having chosen ventilation. Patients who received education had similar levels of hope pre- and post ventilation. Those who did not receive education had low levels of hope.
Hocking et al. 2006	n=7	Auckland, New Zealand	Qualitative; semi-structured interviews	To explore ALS patients' experience of living with ALS and of their care.	Patients reported feelings of losing control when confronted by numerous healthcare professionals. Maintaining relationships with service providers required time, effort and trust.
Hogden et al. 2012	n=14	Two ALS tertiary centres New South Wales, Australia	Qualitative; semi-structured interviews (face-to-face, telephone and email)	To explore ALS patient decision-making in multidisciplinary care.	Participants felt that their decision-making in care was supported via collaborative relationships between them and their healthcare professionals. Participants valued support provided by the multidisciplinary team but their focus on living in the present contrasted with healthcare professionals' effort to plan for future care.
Hossler et al. 2011	n=17	ALS tertiary centre Pennsylvania, USA	Pre- and post-intervention: Validated and non-validated measures	To explore the feasibility of an interactive computer-based decision aid to assist ALS patients in advance care planning.	Participants' overall satisfaction with the decision aid was high. Participants judged the amount of information provided by the intervention appropriate.

Appendix A

			(satisfaction with advance care planning, self-determination, decisional conflict, and satisfaction with decisions made)		The intervention encouraged participants to discuss advance care planning with their family and physicians.
Hugel et al. 2006	n=13	Neuroscience tertiary centre Liverpool, UK	Qualitative; semi-structured interviews	To explore the experiences surrounding the diagnosis of ALS for ALS patients.	Despite satisfaction with the delivery of the diagnosis, patients experienced delays in receiving their diagnosis. Patients reported poor coordination between services following diagnosis.
Hughes et al. 2005	n=9 (N=29)	ALS tertiary centre London, UK	Qualitative; semi-structured interviews	To explore the lived experience of ALS patients and their carers including their experiences of healthcare services. To identify how delivery of services could change to meet user need	Patients reported uncertainty about services and poorly timed multidisciplinary care. Patients reported concerns about professionals' detached approach and their lack of knowledge about ALS. Patients reported delays in receiving a diagnosis which resulted in problems accessing entitlements to care.
Kristjanson et al. 2005, 2006	n=119 (N=876)	Australia (Victoria, Queensland & Western Australia) – voluntary association registers	Survey; questionnaire with multiple instrument measures	To identify supportive and palliative care needs among progressive neurological patients (including ALS) and their carers. To determine the extent to which services meet the needs of patients and their carers.	Patients who received tailored services were the most satisfied. ALS patients were more receptive to care compared to other neurological groups. ALS patients were the least satisfied with services compared to other neurological groups. ALS patients emphasised more needs across a range of services (information, specialised equipment and financial assistance) than other neurological groups.
Krivickas et al. 1997	n=98	ALS tertiary centre, Cleveland, USA and Eastern Ohio ALSA chapter	Survey; questionnaire, structured interview (via telephone)	To assess the utilisation and availability of homecare services for ALS patients	Over 50% of patients surveyed did not receive homecare services. Patients who received home care perceived services to be inadequate and poorly timed.
Lemoignan & Ells 2010	n=9	ALS tertiary centre Montreal, Canada	Qualitative; semi-structured interviews	To explore the decision-making process in assisted ventilation for ALS patients.	Patients' decision-making was determined by their perceptions about outcomes (symptom relief versus life sustaining), severity of illness, social and financial supports, personal values (relationships, autonomy and quality of life) and by fears about adapting to ventilation.

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Lopes de Almeida et al. 2010	n=[4]	Neurological rehabilitative centre Lisbon, Portugal (servicing catchment ALS clinics)	Preliminary trial with clinical evaluation	To evaluate the feasibility of a telemedicine assistive device for home-ventilated ALS patients.	Patients reported satisfaction with the telemedicine assistive device.
McCabe et al. 2008	n=15 (N=138)	Australia (Victoria, Queensland & Western Australia) – voluntary association registers	Qualitative; semi-structured interviews	To investigate the types of support networks and services used by patients with progressive neurological disorders (including ALS) and their carers.	ALS patients reported a strong desire for basic care services and for a broad range of professional expertise.
McCluskey et al. 2004	n=144 (N=257)	Pennsylvania (local ALSA chapter register) USA	Survey; questionnaire	To evaluate how ALS patients and their carers rate their experience of receiving the diagnosis of ALS.	The majority of participants rated physician performance as average to poor. Patients had greater satisfaction when more time was given to disclosing the diagnosis.
McKim et al. 2012	n=26 (N=52)	Respiratory rehabilitation centre Ottawa, Canada	Survey (prospective); questionnaire	To evaluate a single-session hands-on education programme on mechanical ventilation for ALS patients. To determine whether formal education about ventilation predicts real-life choices about ventilation.	Participants' choices surrounding ventilation following formal education on ventilation predicted real-life choices. Formal education on ventilation had no effect on participants' self-perceived wellbeing. Participants demonstrated a significant improvement in knowledge about ventilation as a result of formal education.
Moss et al. 1993	n=19 (N=71)	Specialist ALS centres and non specialist services Illinois, USA	Quantitative longitudinal (prospective); structured interviews	To identify outcomes, costs, and attitudes towards assisted ventilation among ALS patients, their carers and providers.	The majority of patients were glad they had chosen assisted ventilation Patient s valued extended life provided by ventilation and they were positive about ventilation despite its physical limitations. Assisted ventilation placed burden on families.
Moss et al. 1996	n=50	Specialist ALS centres and non specialist services (Illinois, Ohio, Pennsylvania, West Virginia) Ventilator care programme (California), USA	Quantitative cross-sectional; structured interviews	To examine advance care planning and care outcomes for ALS patients.	Patients valued additional time afforded by ventilation but recognised limitations of ventilation on sustaining quality of life. The majority of patients completed advance directives and nearly all patients wanted them. Most patients wanted to stop ventilation under certain circumstances.

[4] Not reported

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Munroe et al. 2007	n=42	Neuromuscular clinic Boston, USA	Quantitative longitudinal (retrospective); chart review	To determine when end-of-life issues are discussed with ALS patients.	Decisions about end-of-life care were delayed for patients. Patients' decision-making was independent of disease severity.
Murphy 2004	n=15 (N=28)	Scotland (Scottish Motor Neurone Disease Association Register)	Qualitative; semi-structured interviews[5], video recordings	To explore ALS patients' and their partners' perceptions of AAC.	Patients reported dissatisfaction with poorly timed provision of AAC and with inadequate training to use AAC. Patients found AAC restrictive because it lessened intimacy in communication with their partner.
Narayanaswami et al. 2000	n=8 (N=19)	Tennessee, USA (Long-term ventilation domiciliary care)	Survey; questionnaire and multiple	To assess neuromuscular disease patients' acceptance of (and quality of life on) long-term ventilation.	Patients with ALS were more ambiguous towards assisted ventilation than patients with Duchenne Muscular Dystrophy. Patients reported concern about the costs associated with assisted ventilation.
Ng et al. 2011	n=44 (N=37)	ALS tertiary centre Victoria, Australia	Survey (prospective); multiple questionnaires and instrument measures	To identify healthcare needs for people with ALS and their carers in the community setting. To identify potential gaps in service provision.	Patients identified the need for more psychological and physical support. 43% of ALS participants reported gaps in rehabilitation services. 25% of ALS participants relied solely on family members for homecare.
Nijeweme-d'Hollosy et al. 2006	n=4	ALS rehabilitation clinic Enschede, The Netherlands	Questionnaire; response categories	To investigate ALS patients' use of and attitudes towards telemedicine.	Patients expressed satisfaction with the use of telemedicine but felt that face-to-face contact with physicians was still required to discuss psychosocial and emotional issues.
Nolan et al. 2008	n=16 (N=32)	Specialised teaching hospital Baltimore, USA	Qualitative (descriptive); structured interviews with 'control preference' measure Survey decision-making questionnaire and qualitative interviews[6]	To compare ALS patients' preferences for involving family in the decision-making process at end-of-life care and the actual involvement by family at time of death.	The majority of patients issued an advance directive. Patients who opted to make decisions independently were more likely to have family report that decisions were made in this style. Patients who preferred shared decision-making with family were more likely to have family report that decisions were made more independent of a shared decision-making style.

[5] Interviews with *ALS* participants only
[6] Not conducted with *ALS* participants

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O'Brien et al. 2011a,b; O'Brien et al. 2012; [Whitehead et al. 2012]	n=24 (N=52)	ALS tertiary centre Preston, UK	Qualitative; semi-structured interviews	To explore ALS patients' and carers' experiences of care between symptom onset and diagnosis. To identify factors related to uptake of social care services for ALS patients. To explore experiences of death, dying and bereavement among people with ALS and their caregivers.	Some patients reported a lack of sensitivity among physicians when communicating the diagnosis. They reported delays in diagnosis due to slow detection of early symptoms by general practitioners. Patients' desire to maintain control and normality and their uncertainty around service provision delayed their use of social services. They reported poor continuity of homecare services and most participants felt that social services were unfamiliar with ALS. However, they expressed satisfaction with multidisciplinary care at specialised ALS centres Patients felt their needs were not adequately met in the final stages of the disease and they reported anxiety and distress during this period. Carer burden was excessive and exacerbated patients' distress and desire for hastening death.
Peters et al. 2013	n=890 (N=5209)	Nationwide England, UK MND Association	Cross-sectional survey; structured questionnaires	To investigate experiences of healthcare services and access to care among people with long-term neurological conditions in England (including ALS).	Participants reported problems in the planning and integration of their care including delays with the diagnosis and poorly integrated hospital care. ALS patients reported fewer problems in their care compared to Multiple Sclerosis (MS) and Parkinson's Disease (PD) patients.
Silverstein et al. 1991	n=38	ALS tertiary centre Chicago, USA	Quantitative longitudinal (prospective); structured questionnaires	To identify if ALS patients seek information about end-of-life treatments. To identify if decisions made by ALS patients about their care remains stable overtime.	Patients' desire for information and participation in decision-making remained stable. Preferences for end-of-life treatment changed overtime. Preferences for care were independent of functional status and demographic variables.
Stutzki et al. 2013	n=66 (N=128)	Tertiary ALS clinic St. Gallen, Switzerland Tertiary ALS clinic Munich, Germany	Quantitative (prospective); Questionnaire on end-of-life care. QoL questionnaire, depression scale & ALS functional rating scale [Closed question (yes/no) completed by carers on supporting the patient to hasten death]	To determine the prevalence and stability of ALS patients' attitudes to hasten death.	50% of participants indicated that they could envisage requesting assisted suicide or euthanasia and 14% of participants expressed a wish to hasten their death at baseline participation. Only 55% and 27% of participants were in favour of gastrostomy feeding and invasive ventilation, respectively but 75% of participants were in favour of non-invasive ventilation. Their preferences remained stable over 13 months. Participants' wish to hasten death was predicted by depression, anxiety, loneliness, perceptions of being a burden on others and a self-perceived low QoL. Two-thirds of patients had communicated their wish to hasten death with their family.

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Sulmasy et al. 2007	n=32 (N=147)	Two specialised teaching hospitals Baltimore, USA New York, USA	Quantitative longitudunal (prospective); control preference measure, QoL questionnaire & general health questionniare	To determine how terminally ill patients (including ALS) opt to have carers and physicians participate in their decision making and how this changes overtime.	Patients mostly opted for shared decision-making but leaned more independently from carers. Decision control did not change significantly in the proceeding 6 months. Patients opted for physicians to make best-interest decisions at time of diagnosis but overtime, some patients opted for physicians to make decisions based on their judgements.
Sundling et al. 2009	n=7 (N=15)	University Hospital Huddinge, Sweden	Qualitative; semi-structured interviews	To explore ALS patients' and carers' experiences of non-invasive home ventilation.	Patients viewed home ventilation positively but had contradictory emotions about being dependent on a ventilator. Patients' perceptions of the benefits of ventilation were strongly influenced by home support.
Trail et al. 2001	n=42	ALS tertiary centre Houston, USA	Survey; questionniare	To determine ALS patients' preferences for wheelchair mobility.	Patients reported that wheelchair mobility had a positive impact on their wellbeing and facilitated greater independence in everyday life.
van Teijlingen et al. 2001	n=153	Scotland (Scottish Motor Neurone Disease Association Register)	National survey; structured interview questionnaire	To examine service use and needs of ALS patients and their carers in Scotland.	Patients reported long waiting periods for multidisciplinary care. The majority of patients reported unmet needs in services and they accessed support from the voluntary sector to alleviate burden of care.
Vitacca et al. 2010a, b	n=40 (N=80)	ALS tertiary centre Brescia, Italy	Multiple outcome study (satisfaction assessed by structured telephone interview)	To examine the benefit of telemedicine care for a cough-assist programme for ALS patients and their carers.	75% of patients reported high levels of satisfaction with the programme. Use of the programme reduced the frequency of hospitalisation and cost of care for patients.
Ward et al. 2010	n=32	ALS / Muscular Dystrophy tertiary centre North Carolina, USA	Survey; questionnaire	To determine characteristics of and satisfaction with powered wheelchair mobility for ALS patients.	Patients reported high levels of powered wheelchair utilisation and they expressed high levels of satisfaction with powered mobility. Patients valued the capacity of powered wheelchair mobility to accommodate to their physical disability.
Wicks & Frost 2008	n=247 (N=334)	PatientsLikeMe.com internet website	Survey (via email); questionnaire	To indentify what ALS patients and their carers know about ALS. To identify if ALS patients and their carers thought they received sufficient information about ALS.	Patients reported that they received information about physical symptoms as opposed to cognitive symptoms. Patients indicated a preference for more information about all symptoms of ALS.

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Young et al. 1994	n=13	ALS tertiary centrre Vancouver, Canada	Mixed method: Qualitative; semi-structured interviews Quantitative; questionnaires and instrument measure	To explore ALS patients' perspectives on mechanical ventilation.	Patients' decisions to avail of mechanical ventilation were determined by their views on: the potential of ventilation to improve quality of life; ability to control discontinuation of ventilation; and likelihood of ventilation to sustain life. Decisions against ventilation were influenced by patients' perceptions about potential adverse effects of ventilation on quality of life and by the burden of care associated with mechanical ventilation. Few patients sought comprehensive medical opinion on ventilation. Measure of disability was not associated with decision making.

Appendix B

Participation Information Leaflet

Study

Understanding and Use of Health Care Services by People with Amyotrophic Lateral Sclerosis / Motor Neurone Disease.

Principal Investigator: Geraldine Foley

Title: Health Research Board (HRB) Research Fellow

Telephone Number: 086 8049125

Email: foleyg3@tcd.ie

Voluntary Participation:

You have been invited to participate in this study. Before you decide whether or not you wish to take part, you should read the information provided below and if you wish, discuss it with family and friends. Take time to ask questions. Do not feel under any obligation to participate. If you choose not to participate, this will not have any negative impact on the quality of care you receive. If you choose to participate, you may change your mind at any time (before the start of the study or even during your participation) without having to justify your decision. This of course will be fully respected. You should clearly understand the procedures involved in this study so that you can make a decision that you feel is right for you.

Introduction:

The aim of this study is to explain how people with motor neurone disease understand their health care services and how they use health care services. It is hoped that findings of this study will give healthcare providers a better understanding of how people with motor neurone disease make sense of health care services and so help

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Appendix B

service providers meet the care needs of people with motor neurone disease. The Principal Investigator is also undertaking this study for the purpose of obtaining a research doctoral degree with Trinity College Dublin. If you agree to participate in this study following nomination by myself or by a colleague with a specialty in motor neurone disease, I would like to talk to you about how you experience your healthcare services.

Procedures:

It is planned that I will meet with you at home or where you live to record what you think about health care services and how you make sense of the services you receive and/or have received. It is planned that our meeting may last one to two hours. You may of course take breaks if you choose to do so as you may feel tired or simply wish for breaks to gather your thoughts. The interview may cause you to feel tired and you may stop the interview at any time. The interview will be recorded by a digital audio recorder. The interview will then be transcribed word for word. The recorded interview will be kept securely with me and will be destroyed following completion of the study. Neither the digitally recorded interview nor the typed up interview will be made available to anybody outside the researchers in this study without your explicit (written) consent. Your typed up interview will also be kept securely with me for the duration of the study and be labelled with a pseudonym (false name). The transcript will subsequently be stored in a secure location on completion of the study. Your transcript, in storage, will have no link to your name and it will be labelled with a pseudonym (false name). This is in keeping with the data protection legislation. You will be provided with a copy of the typed up interview. I am aware that the information you may provide may at times result in mixed emotional responses about

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living with motor neurone disease and about your experience of care. The information that you may share with me will be kept confidential and will only be disclosed to the investigators in this study. Your thoughts and feelings will be fully respected at all times. I may on a needs basis contact you after the interview should I need to clarify things with you. You may of course contact me at any time during the study. Your General Practitioner will be contacted by me informing him/her about your participation in this study.

Benefits:

It is envisaged that recording and analysing your thoughts about your experiences of health care will result in an increased knowledge of how persons with motor neurone disease understand their health care services and subsequently how they use health care services.

Exclusion from participation:

You may only participate in this study if you have a diagnosis of motor neurone disease.

Confidentiality:

Your identity will remain confidential at all times. Your name will not be published and will not be disclosed to anyone outside the study. This study includes the following researchers; Geraldine Foley (principal investigator), Trinity College Dublin, Dr. Virpi Timonen (co-investigator), Trinity College Dublin and Prof. Orla Hardiman (co-investigator), Beaumont Hospital & Trinity College Dublin.

Compensation:

This study is covered by standard institutional insurance. Nothing in this information leaflet affects your rights.

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Appendix B
Permission:

The Beaumont Hospital Ethics (Medical Research) Committee and the Research Ethical Approval Committee at the School of Social Work and Social Policy, Trinity College Dublin have approved this study.

Stopping the study:

You understand that the investigators may withdraw you from the study if it is deemed not to be in your best interest. This will of course be discussed with you beforehand. You may contact me at any time that is convenient to you.

Principal Investigator: Geraldine Foley

Contact details: Tel: 086 8049125

Email: foleyg3@tcd.ie

Co-investigator: Prof. Orla Hardiman

Contact details: Tel: 01 8092174

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Appendix C

Informed Consent Form

Study:

Understanding and Use of Health Care Services by people with Amyotrophic Lateral Sclerosis / Motor Neurone Disease.

Principal Investigator:

Geraldine Foley, HRB Research Fellow, School of Social Work & Social Policy, Trinity College Dublin

Co-investigators:

Virpi Timonen, Associate Professor, School of Social Work & Social Policy, Trinity College Dublin

Orla Hardiman, Consultant Neurologist, Beaumont Hospital Dublin & Professor of Neurology, Trinity College Dublin

Declaration:

I have read this Information Leaflet (Version 3 dated 25/04/11) or I have had this Information Leaflet (Version 3 dated 25/04/11) read to me and I understand the contents. I have had the opportunity to ask questions and all my questions have been answered to my satisfaction. I freely and voluntarily agree to be part of this research study, though without prejudice to my legal and ethical rights. I understand that I may withdraw from this study at anytime, and without this decision affecting my future treatment or quality of care. I understand that my identity will remain confidential at all times and that my interview will not be made available to anybody outside the study without my explicit consent. I understand that, on completion of the study, my

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typed up interview (transcript) will be stored in a secure location and it will have no link to my name. I understand that I will be given a copy of the transcript of my recorded interview. I have been given a copy of the study Information Leaflet and this Consent form.

Participant's name: _____
(Name in block capitals)

Contact details: _____

Participant's signature: _____

Date: _____

Statement of Investigator's responsibility:

I have explained the nature and purpose of this research study, the procedures to be undertaken and any risks that may be involved. I have offered to answer any questions and fully answered such questions. I believe that the participant understands my explanation and has freely given informed consent.

Investigator: _____
(Name in block capitals)

Investigator's Signature: _____ Date: _____

Qualifications: _____

Contact details: Tel: 086 8049125

Email: foleyg3@tcd.ie

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Appendix D

Geraldine Foley – Participant #31 130712

Duration: 93mins 38sec

G: **Right, it's recording there.**

P#31: Can I just say to you before we start?

G: **Yeah sure.**

P#31: How I see this MND, how I see the disease myself. I read a very interesting book some years ago by Stephanie Meyer and she was talking about, it was an alien species came down to earth and what they did was that it was like, it was like sort of a butterfly, a gossamer, sort of like little maybe little octopus, little octopus and what happened then, you inhaled it and it took over your brain, took over your body. Well, I often think that MND is like that. I feel like I've been taken over by a sort of an alien presence and it's, it's in my brain and I think it's like an alien that has nuclear weapons, that it can do all sorts of things to me and I'm combating it with bows and arrows because as far as I'm concerned the only weapons that I have are either both Rilutek and a high protein, high fat diet. Nothing else, now allied to that I have the support, the services. So, so I'm, this alien has taken me over who has all these great, all this great arsenal of weapons that can use in my brain against me. For instance, I'm getting paralysed down one side, it can press a button and I'll get paralysed on the other side. I've dropped, one of my feet is dropped and I'm having difficulty walking and this, this MND, this alien in my system now can do all of these things to me and it came to earth in 1843 as somebody said and since then little, very little progress as far as I can see with it. Because all I have is the Rilutek which, and you consider in the time all the cures for cancers and AIDS and all of that that happened in all that time. And yet, this MND, there's nothing like that in my view has happened that I can see. I know there's research going on, but that's where we are. So I find myself in a constant battle, day after day because this, this MND alien that has taken over my body and that I have not too much defence against. Normally in, in a war you win some battles, I'm losing all the battles and I know for a fact I'm going to lose the war. The war, the war is lost already because MND is going to take me anyhow, but I'm not winning any battles because I feel that I don't have, the arms that I'm provided with in no way can in any way defeat or even hold back this alien that is taking me over. Does that make any sense to you?

G: **Absolutely, yeah, it's a really interesting analogy. So is it a sense then @@Martin## that you feel that you are constantly losing?**

P#31: Oh absolutely. Last Christmas, in last February we were 50 years married. I was able to go up to the pub, walk into the pub, sit and have a few drinks with my friends, wife and my friends, celebrate, that was last February. And here I am now and I'm at the point now where I'm really faced with a wheelchair.

Appendix D

G: In February of this year was it, or February?

P#31: Yes, yeah, now I was diagnosed in the end of October coming into November last year. I was diagnosed and at that time all I had was very slight numbness in the side, in the sides of my legs and because all, from the time I was 10 years old I was an athlete and progressed through all sorts of running into marathons and ultra-marathons and then I became a cyclist, ultra cycling and all that I know my body very well. I read my body very well and when I am, when I was getting this numbness I got all sorts of tests done. I went to, for a vascular, a series of vascular thing or whatever, but deep in the back of my mind there was some, something really not right here so eventually ended up with a neurologist called @@consultant neurologist## down in @@Dublin acute regional hospital##.

G: How did you end up with him? What was the path to him?

P#31: Well because I, I went down, I was attending a cardiologist and I got a stent done in @@Dublin acute regional hospital## and I was attending a cardiologist, @@consultant cardiologist## and I was talking to him and I said to him, I said I'm a little bit worried about this, he had me on, he had me on a Lipostat and I said to him, I'm a little bit worried, I'm getting some, I'm getting some problems with my muscles or I said I don't know whether it's a numbness or what it is in my legs. I said would it be related to the medication I'm on. And he says I don't think so he said, and I said well I'm, I'm a little bit worried about it because it's on going now I said and I've had, I said I've had a vascular done, I've been to an acupuncturist. I said I've been to a physiotherapist and they're all a little bit, not quite knowing what's going on.

G: Were you suspecting something serious yourself, at that time?

P#31: In the back of my mind I was saying I don't like this, you know. So, and at one stage somewhere the, the January before my wife had been in hospital in, in the @@private hospital## for three months and then she was in a wheelchair and she came out in a wheelchair for a few, for a couple of months and at one period when she was in hospital I was up in the shops and I was walking from the shop over to go up and my legs went completely dead on me and I, I sat on, on the front of a window in a shop, in one of the shops and I rang my daughter to get me. But I came home here and I went to bed and next morning it was gone, but that episode repeated itself two or three times and that's what brought me down first of all to the acupuncturist, physiotherapist for a vascular scan but when I went to see @@consultant cardiologist## and I said to him about this numbness in my legs I asked him was it in relation to the medication I was on and he said I don't think so he said. But he said I'll tell you what he said, in this building he said there's a neurologist and he said if you like I'll get my secretary to make an appointment for you because he says it's obviously bugging you a bit. So I said okay, so I went in to see the, I went to see the neurologist about two weeks afterwards.

G: Was this in a private capacity?

P#31: It was.

G: @@consultant neurologist## is it?

P#31: Yes, yeah. So I went to him, so he, he looked at me, examined my legs, said where's the numbness, I told him. He said is it a weakening or a numbness and I said it's a numbness as far as I'm concerned. And he said to me what are your worst fears and I said three things. I said, I said Parkinson's, multiple sclerosis, I said or motor neurone. And he says okay, he said I accept your worry he said. I don't know what's wrong with you by the way he said, but he said I'm going to give you a prescription he said for Rilutek, so he said from today you start taking it and he said I'm giving you that as a precaution he said because one of the tests I'm going to send you for is MND. Now he said I'm ruling out multiple sclerosis straight off, not that, he said I'll have you tested for Parkinson's and I'll test you for MND. So within probably a week he had me down getting a nerve and muscle test and within another week he had me back and he said I've really bad news for you he said, he said I think the only diagnosis I can give you at this stage is MND.

G: You said you had three conditions in your head, Parkinson's, MS or MND. Was there any particular reason why you suspected MND? Why did you include MND in that? Had you heard of MND before or?

P#31: Yeah, I had a friend with Parkinson's. I had a friend, his brother multiple sclerosis, I didn't know. I didn't know, the only thing I knew about MND was that many years ago I read up a film star who had a horrific death from MND and I remember reading about him. He played James Bond in Casino Royale, the very first one, he was, had a little moustache, he was, I can't think of his name at this very moment. He played an older James Bond in it but he had a dreadful death, he, because people apparently around him didn't understand the condition too much. But I don't know why, maybe because I was going to see a neurologist.

G: What do you think, how do you think having MND might compare to having other conditions like the ones you mentioned or other conditions where you can be physically disabled? Because MS and Parkinson's can give you physical disability as well.

P#31: I think the medication for MS and the fact that you can get remission makes it a different ball game. I know with Parkinson's you can also have, the medication can be very effective. They are the two basic differences that I see between my condition and those other two conditions.

G: I just want to make sure I'm correct because if I'm not tell me, that because there are some conditions like MS and Parkinson's there are available treatments that can really have some positive effects and sometimes you're right, change the course of the condition for a while, like MS you mentioned. And with MND there isn't. Are you saying there's more hope in those conditions or?

P#31: Well let's say I see less hope.

G: Less hope.

P#31: Yeah, in MND, I mean I'm trying to remember who said it to me, about being progressive, incurable and terminal. Now that, that is, that is to me, that's the purest way of expressing this disease.

G: I think it was Colm Murray who said that, wasn't it?

P#31: Maybe.

G: He said it in a TV documentary and he used those words actually to describe it.

P#31: Yeah and I think that is the purest way of describing it because as I see it, it is progressive and more quickly progressive than I would have thought myself.

G: Are you shocked by how fast it is?

P#31: Oh absolutely, because all of a sudden all the things, now I was lucky that I'd done some things since this. I was able to go to a solicitor and get power of attorney and all of that done in relation to our financial affairs with my daughters. I should have got the wet room done earlier. There's a number of things I should have done earlier. I, the car you see outside is an automatic Golf. I bought that in October, just before I was diagnosed, I can't drive it now, you know. And like, I did drive it up to about, about six weeks ago but in that time I progressed to not being able, because I can't lift, that's as far as that arm will come up now so I can't turn the wheel you know.

G: You mentioned the shower room. You got your official diagnosis earlier this year and we're now in July and you're thinking now, god should you have started this work back then versus more recently.

P#31: Yeah.

G: But just going back to that time, it was probably a massive shock was it? The whole thing was a shock?

P#31: No, believe it or not I didn't. No, it wasn't. I don't know why it wasn't a huge shock. Now, maybe one of the reasons is because any ambition that I wanted to fulfil in my life I more or less had done that. You know, like my life would have been kind of, physically active life in relation to all my running and cycling and all of that and, and for to keep your own mind working I played bridge three times a week. So I did all, there was nothing left literally that I felt...

G: You hadn't done.

P#31: That I hadn't done.

G: And on the shower room, looking back to February.

P#31: Yeah.

G: Did you think to yourself okay, I need to start doing this or is it because it's just that the condition has been so much faster than you thought that prompted you to do this? I

suppose my question is did you make this change with the shower room because MND is progressing in your head fast for you versus at the beginning you thought I probably don't need to do this now, you know, we'll wait and see how things go?

P#31: That's exactly how it was, that is, I couldn't express it better myself. I was kind of hesitating and saying you know, do I have to spend twelve grand getting the job done and you know, and like is it absolutely necessary or should I consider a stair lift instead, all of that and then, and then I kind of felt myself, I got a few builders in and they all take their time about sending you back a quote and that. And then I could feel myself progressively getting, whereas on this side I was, I had been, that arm and leg were weak and I was depending on this side and all of a sudden I was getting problems with this hand and this leg. I said I better get moving on this and, and that's when I made the decision, I'll get it done straight off. And even in the time it has taken, in the two weeks I have felt myself going downhill in the two weeks.

G: And you mentioned there also you're 74.

P#31: Yeah.

G: You seem to be quite satisfied with what you've achieved in life, at this point, quite active and so on.

P#31: Yeah.

G: Do you think that makes a difference in terms of how you might cope with MND or how you might accept it, not accept it versus perhaps somebody who hasn't done as much as you've done, who is younger, you know, hasn't had the life experience?

P#31: Oh I've no doubt at all, it must be, it would be, I retired when I was 55 and if I had been diagnosed at 55 would have been a totally different ball game. It would have killed me, it would have destroyed me because like as you see @@Emily## is 42, her sister's 44, they have their families. I have no kind of responsibilities towards them. If I had young children I'd, this would be the most devastating thing that would have ever happened.

G: Is that because if you were if you were younger with MND you've more of a possibility of having children that are dependent on you?

P#31: Yeah, and the fact that I may not be there for them either.

G: Yes, I see what you mean. So you do really see a difference between that?

P#31: Oh I do see a huge difference there. And as well as that, if I had of, in my mind said I'm, I will not die until I climb Kilimanjaro you know, I haven't got any regrets like that. Like I used to run, I used to run marathons and I got sick and tired of running. That's the gods honest truth and I used to run with a group of guys and on Sunday morning we used to run a thirty mile run, every second Sunday morning. So a marathon wasn't a big, twenty-six miles was a big deal then, you know, it really wasn't, you know. So we were discussing this one day and I said to one of my pals I said I really am kind of sick and tired of running marathons I

said, and he said why don't we get it out of our systems and we won't run any more marathons. So in a three month period we ran six, every second week we ran them. We found a marathon and ran it and after we ran these six marathons we both said that's it, you know. And I never ran a marathon after that, that was the end of it. So I was happy enough with that.

G: **I'm just reflecting back on other interviews @@Martin##, is it the case that the certainty of having MND, experiencing physical change that you are aware of yourself, prompts you to actively plan your care, engage with care, plan future care? Do you think that's the case?**

P#31: Yeah, I think that, when I got MND first, I joined the MNDA, the IMNDA, I joined the association, they sent me out a great folder, a lot of information in the folder, maybe too much somebody said to me, but I didn't find it was too much by the way, I, you know, I was happy enough to get the information. I was happy with that. I was happy with the fact that I saw @@the consultant neurologist at the national ALS clinic## and her team and I saw her once and then when I went back the second time I didn't see her. I saw @@Beaumont clinic researcher5## and he's, he mostly spoke about research and stuff like that, and I didn't really get a chance to discuss my condition. He said to me oh you're stabilised. I didn't think I was at the time, and then he made an appointment for me not to see the Professor for three months. And when I came, and I found three months was too long. I have very little input with the local service at that stage.

G: **This is after the diagnosis, so initially you weren't linked in with the local services?**

P#31: No. Now I was seeing a physiotherapist and my wife was involved with the primary care team up in @@nearby town##, the team that I'm involved in now. But I wasn't like, the district nurse would come in and say to me how are you today and I'm pretty okay, because you don't, you look pretty okay, you know. So but I felt, I felt I was kind of floating on my own and this thing is progressing and I didn't know what the, like I rang @@consultant neurologist## one day, the neurologist and I said to him, I just rang and said, I said I'm a little bit worried about my condition and the very first thing he said to me, are you alright, can you still walk. And like at that stage I could and I thought he was being very extreme, you know I said I can still walk, and he said oh I thought you were in trouble he said. I said well I feel I'm in trouble.

G: **So it's not just about walking or not walking?**

P#31: Yeah, I was just, it was just that I was starting to get a feeling of like I couldn't lift, this arm was coming to the point I couldn't lift it and I couldn't lift the kettle, I still can, whereas I could with this one, but now I can't lift the kettle. And my breathing is getting slower, not as good, but in that, in that three month period I felt a complete disconnect. I hadn't got, the support services were doing nothing for me really. I felt I was too far away from

Beaumont. I didn't know what would happen if I, say for instance if I became unwell, did I go to Beaumont, do I go to @@local catchment hospital##, you know.

G: **So what did you do to change that because it seems that you are linked in with services? How did that change?**

P#31: Well what happened then was that on the three month, when the three months came around and I went down. I said to them I feel a complete disconnect and so I feel that I, I said I know there are supports out there, I said to @@the consultant neurologist at national ALS clinic##, look I said, I know that you pride yourself on the service that you personally and your team give to MND people. But I said I have to say, to be perfectly honest, I'm not seeing it. I said last time, I didn't see you, I was here three months ago, I didn't see you I said, I didn't have a chance to really to discuss my condition, and I said I'm kind of apprehensive about a lot of things and I said I felt three months was too long to be out there floating, as I'd see it, floating and I said I've had very little contact with the local services. So she said three months was too long, for it to be left where you are at this stage, too long she said, you should have had a six week appointment and she said what I'll do I'll organise a conference, @@ALS specialist nurse## will organise a conference with the people up in the primary care team in @@nearby town##.

G: **Did that happen?**

P#31: It did, so @@ALS specialist nurse## and, and @@IMNDA specialist nurse## from the Motor Neurone Association arrived in @@nearby town – primary care team## and they met the occupational therapist, the physiotherapist and the, the local nurses up there. And they discussed, they discussed me and my condition and then they brought @@Emily## and myself in, or @@Ann## my other daughter and myself in and they discussed with me what they had been saying about me.

G: **How did you find that?**

P#31: I found, I found that was pretty okay.

G: **Looking back now do you think that only happened because you shared your feelings at the clinic about your concerns? I mean if you had gone to the clinic and not said anything, do you think things would have changed or, or was it the fact that you were being missed do you think?**

P#31: Yeah I think it would have been, there would have been a disconnection whereas I think that the fact that @@ALS specialist nurse##, you may have experienced this and I find it all the time, even my own doctor, my own doctor is a practicing GP for 20 years. I'm his first MND patient and the very first day I met him look he says, I have to say to you, I have to be very honest now, I know nothing about your condition. He said I'll have to read up about it, I'm depending on you to tell me. Well my impression of the @@local primary care team## is that they had seen very little of my condition either, although the physiotherapist had.

But whereas when I went to physiotherapist first I could do a whole range of exercises that she organised for me, but very quickly I couldn't do the exercises and at this stage my, my connection with her is minimal because she can't really do much for me. Because this has progressed so quickly, you know.

G: So you've had mixed experiences really so far, more recently good experiences maybe?

P#31: Yes.

G: Because you seem to me quite eager to engage with services, get the help that's out there to help you live with MND. I'm just looking around. You have a number of aids.

P#31: Well I have, funnily enough some of it is my wife's, my wife's, that's my wife's, yeah so I use that *(P#31 referring to rollator)*. The little wheelchair you see there, we bought on the internet because it's a very, it's a lightweight, the girls found it hard dragging a big wheelchair so that's handy for shopping. The scooter you see there just arrived today. Now these are all, these are all as a consequence of, of, of the motor neurone nurse and the IMNDA nurse meeting the primary care team in @@nearby town## because these things weren't happening.

G: Just as a matter of interest @@Martin##, the scooter that arrived today, has that been funded by the IMNDA or was it funded by the HSE?

P#31: No, by the HSE.

G: So you've a medical card?

P#31: I've a medical card. Now, also, I have, once I get into bed I can't get out of bed, because once I'm laying any way flat, I find it difficult first of all to turn over, but I can't get out of the bed and where initially the occupational therapist gave me, it's a round thing to sit on the side of your bed, and you lever yourself out.

G: Bed lever.

P#31: Yeah, but it didn't work. So but since, since the meeting with the team, @@ALS specialist nurse## says to me why don't you organise a hospital bed for me that, an electronic bed, so that's in the pipeline. In the meantime I still can't get out of bed but my wife is a very frail woman, with her help and holding on to this thing I can get out of bed. Now also since the meeting, I have a daily help comes in to help, to wash and dress in the mornings, but at this stage I'd be up before she does, she arrives in because of all the building going on and to let them in the door and all that. So that has happened as well.

G: So a lot has happened recently.

P#31: A lot has happened since the meeting with the primary care team.

G: At the moment so are you generally satisfied with services or do you still think that services could do more for you?

P#31: No, at the moment I am, I am reassured with the services. Once I'm in, I felt, I felt although I was, I felt I wasn't kind of, I wasn't part of the whole. I felt I was like kind of floating on my

own whereas now, I am connected and it's a team effort. Now I'm able to sit down and discuss it with them.

G: **Do you think teams work better?**

P#31: Oh absolutely, yeah.

G: **One thing you mentioned there at the beginning of the interview, from your analogy that you feel absolutely no control over the condition.**

P#31: Absolutely no control.

G: **Do you feel you're in control of healthcare services? Or is it all the same thing?**

P#31: That's a good question. How would you, how, could you put that a different way?

G: **Yeah, so it seems to me that you definitely feel that you've no control over the progression of the condition. The condition is a progressive condition as you know. There is nothing you can do to change that course, there's nothing you can do to alter the change to your body.**

P#31: Yeah and the medication doesn't either.

G: **Yeah, so you feel you've no control over that. But do you feel in control of how your healthcare services are delivered to you?**

P#31: Well yes, I think that, like I know for instance that, that @@ALS specialist nurse## over in Beaumont said don't ever hesitate to ring me, don't feel that you can't phone. Now the, the nurse from the motor neurone association said exactly the same, twice I rang, I rang @@IMNDA specialist nurse##, I rang @@IMNDA specialist nurse## once and I rang and I got the girl that was standing in for her to help and they both, they came over to the house to see me and I thought it was, that was pretty okay because whatever concern I had I was able to sit down and discuss it with them. And I do think myself that, that since the meeting the occupational therapist has certainly pulled her weight for me and the district nurses are very good and have organised the meals on wheels because my wife's not very, I used to do all the cooking anyhow. So they have organised that for me as well and I know as well is that if I ring them they'll come down and I'm very happy with all that now, yeah.

G: **So you are feeling in control of the services at the moment. Not initially, but more so now that things are working better for you. You also mentioned there even though MND is progressing for you, that you're happy that you've had the opportunity to sort out power of attorney?**

P#31: Yes.

G: **That's something that came into your head to do?**

P#31: No, it was, it was suggested in the motor neurone book, you know, because, because my wife has, is in the early stages of vascular dementia. So I discussed it with my doctor and I said to him, I said with this condition I have, there may come a time when I won't be able to speak. I said I know that @@Pam## is getting, at the moment now where she's lucid

now, if it came on. She went down to the @@catchment day hospital## down in @@local catchment hospital## deal with, with elderly people and the doctor down there advised me as well, she said have you got all your legal affairs in order and all that because you have motor neurone, she has vascular dementia and I said no, but I will. And I said but what will I do and she said well get in touch with a solicitor, talk to your doctor, get in touch with your solicitor and speak to your solicitor about it and then involve your family. And that's exactly what I did.

G: **Looking back at that now, do you think you primarily did that because of a fear of losing your speech or would you have done it anyway, with MND?**

P#31: I would have done it anyway with MND, yeah.

G: **Anything else that comes to mind at all about your experiences of services so far? Anything at all, just eager to get your perspective.**

P#31: Well the other thing was that when I fell here, now I also wear, you know this thing, we have you know the button that you press.

G: **Pendulum alarm.**

P#31: I have that, that was St. Vincent DePaul provides that, the local St. Vincent De Paul. When I fell I pressed the button here and he, he came on and my wife was able to tell him I needed an ambulance and, and lucky enough I had my mobile phone in my pocket and I was on the floor there so I rang my other daughter. I knew she wasn't working all day. So I was able to ring her and she came around and one of the questions I asked down in Beaumont, one of the questions I'd asked at the conference, I said if I became ill for any reason, do I go to Beaumont or do I go to, to @@local catchment hospital##. I'm in the @@local catchment hospital## catchment area and they said, @@ALS specialist nurse## said to me well look, if you're unwell for any reason and somebody puts you into a car, they can bring you over to us here in Beaumont. If you ring the emergency services they will bring you to @@local catchment hospital##. And I said, and all my records were over here in relation to MND and she said yes, and I said but if I arrive in @@local catchment hospital##, I said and I said I know that not all medical staff are familiar with MND, I said what happens then. And she said, well, what they would normally do, if you can tell them you have MND they will, you could either tell them to contact us or they will do, the consultant over there will do it himself. So I got a chance to see this in action because when I fell, when the ambulance men arrived he said to me can you get up off the floor I said no, I've MND. And he said okay, he said, he said we'll, he obviously knew, he said we'll do all this now, we'll look after you, just you stay there. So they bandaged me up on the floor and that and then they got me up to a chair. When he brought me down to @@local catchment hospital##, as he brought me in the door he was saying to them this man has MND, he said he can't be left in the corridor or whatever, and almost immediately I was put in on a trolley. So and then, I

wasn't left too long on the trolley and they put me into, because I was, I've very little flesh on me now and all my muscles along my back are gone. I'm very bony and even lying with the trolley after about an hour I was starting to ache and it does take time because they're very busy. So, what they did then obviously because I have MND, first of all they transferred me on to a more comfortable bed and put me over beside the nurses' station, which was pretty okay. And the, the consultant told me he had been in touch with, with Beaumont, he said I know you've motor neurone he said, the fall, was the fall related to motor neurone and I said ninety percent, I said because I just thought, so he said okay he said, he said so he said I'll attend to your injuries he said and he said if you've any other needs he said tell the nurse and I'm on, I'm on duty. Now, the only thing I found in, in @@local catchment hospital## was that although the trolley or the bed I was on has arms on it, because of my weakness I couldn't lift myself up and I was kind of static for about eight or ten hours and my back was aching. Now because the cutbacks down there is, at one time the nurse had aides and the nurse's aide would lift you, you know, but those girls all work on their own now. Tough old gig down there. So for me to kind of shift position or to be, try and sit up a bit trying to take the pressure off my back I have to get the nurse on duty who was very busy and I kind of hated doing that, you know. So when the morning came, when the consultant came back he said to me I'm going to keep you for another day now he said because he said you're, I have to stitch all your injuries he said because you're bleeding so much because I was on Plavix, which is an anticoagulant. And he said, and I said I'm sorry I really have to go home, I said I couldn't lie here now for the rest of the day and tonight and he

G: **There was no bed, no?**

P#31: No. But he recognised that I was having problems so what he did was, he got me out of the bed sitting on a chair and he said would you be able to sit on the chair and I said no, said I want to go home. So he said okay he said, I'll have you home as quick as I can and he did. My daughter came out and collected me and brought me home. But that was my experience of @@local catchment hospital##. It wasn't a bad experience, you know, they looked after me really well.

G: **You said yourself they seem to be under pressure.**

P#31: Yeah, but it was the fact that like I had been, when I really got that numbness I was talking about in my legs I couldn't walk, at one stage I arrived down in @@local catchment hospital## and stayed overnight down in the A&E as well but I could turn in the bed, I could shift around, I could sit at the side of the bed. When you have motor neurone and you are, you can't move and you're lying on the flat of your back all night, that gets difficult.

G: **Is it a case then @@Martin## that when you feel more certain about services or certain about how things might turn out for you, it nearly makes you feel, how will I say ... less**

stressed about the whole thing? You know you can go into hospital and be cared for relatively well and you're confident that these people will do the best they can for you, that that somehow makes it more bearable?

P#31: It does, yeah.

G: So certainty of the services and confidence in people to do what they say they'll do for you.

P#31: It does make it more bearable, yeah.

G: Would you say now you have in general, confidence in the healthcare system or in healthcare professionals to care for you?

P#31: At this moment in time, I have, yeah, I have. Now to be honest with you, in relation to MND I have to be very realistic about this. The healthcare professionals do very little for me in relation to the disease because they can't. They've done as much as they can, they have me on the Rilutek and they have me on high protein and high fat diet, after that they just react to whatever problems I run into and it may be more bearable, make the disease more bearable. I mean I see @@the consultant neurologist at national ALS clinic## and her team as like a group of shepherds who, who are shepherding me towards you know, towards my, towards my death really, you know. And what they're doing is they're, they will in so far as they can make it more bearable for me.

(Interruption – P#31 talking to builder)

P#31: Now I was saying to you, like I see, I see like @@the consultant neurologist at national ALS clinic## role and her team as, as I said like, like my shepherds to move me as gently and as bearably possible towards where you know, when I die. And I know they'll do that very, with compassion and efficiency and indeed the local services would be, will be the same but that's about it really.

G: So in that sense @@Martin## you see services for people with MND as primarily symptomatic, supportive. Do you think that's because it's such a rapidly progressive condition?

P#31: Where I'm concerned, yes. But now, from, from reading and listening to people I know it doesn't necessarily be as rapid as it's happening for me. I know that it can be slower. I saw a man in a wheelchair recently on the BBC documentary where, they did, they rebuilt his house for him. He'd MND and he was, he was, he had, he was three years in a wheelchair, had it three years. I don't see myself in that category, to me its come too quickly.

G: It's interesting because I've spoken to, you are the 31st participant I've spoken to and you know, people have different views, people have different experiences. You said yourself I

don't see myself living that long, compared to the man on the TV who was living with it for three years and doing well. Do you think about time frame then?

P#31: I, in my own mind, when I spoke to @@consultant neurologist## about the condition, when he spoke about, he said maybe the condition is coming on quicker than you thought it was, you weren't aware the condition was coming on, he said it could have been last June maybe, so if that's so, I'm at this stage a year and a bit into the condition and my own view is that my timeframe would probably be less than two years.

G: From now?

P#31: No, no, from, from June of last year and it's progressing so quickly now. I mean I know now that because my diaphragm and my abdominals, the muscles under my ribs are all weakening now, that I, I, my breathing is not as good as it was because my diaphragm is weak so I can't stand up for any length of time. I know myself when all of these things fail that I'm going with them, they're not exclusive you know, they don't work apart from me. So it's going so quickly now that I am, that's my time frame.

G: And have you readjusted that time frame as you said, you said earlier in the interview I think even the last few weeks since you've been getting this work done to the house that you've noticed even more of a change in the last two weeks. As you continue to see more change or experience it, do you readjust that time frame?

P#31: I do actually, I do but I'm not saying oh, if I make it to Christmas I'll be okay, but yes I do, I do I'd say this is going more rapidly than I thought.

G: So you're not thinking specifically about months or dates?

P#31: Oh no, no.

G: You think about it in a general sense, I see what you mean. Just on that, are you okay talking about this?

P#31: Hmm.

G: You said you feel that @@the consultant neurologist at national ALS clinic## role and her team is that they shepherd you towards the inevitable. Do you think that's the best healthcare professionals can do for you, in the context that MND is a terminal condition?

P#31: Well unless they come up with something new, you know. I read there's research going on but like, I'd find it difficult to find a different role for the, for all the team in view of the fact that it is terminal and that it is progressive and that it is incurable. Like, when I think about it I say it's not like if you had AIDS and there's a whole raft of medication you can take and I mean Freddie Mercury would still be alive.

G: You're right, he would be, yeah.

P#31: Had the medication been available at that time, but it wasn't. So, so you know.

G: Is it because MND at the moment is a terminal condition, quite progressive for most people, as you said there are variations, that it dictates the care approach for people

with MND and healthcare professionals' responsibility is to fit in with that approach - that it's the nature of the condition influences what healthcare professionals can do for you?

P#31: Yeah.

G: Okay.

P#31: That's about right.

G: You've made some, you have done the power of attorney thing and you would do that anyway whether or not you encounter problems with your speech. Do you think about any other things around, for around that time? You know some people for example with MND are asked to make certain decisions around end-of-life care, do you think about that at all?

P#31: Yeah, I, there's a, I know that down in, in @@local catchment hospital## there's a new unit has been opened down there, a new hospital actually and the, there's a palliative care team down there. Now, the last time I spoke to the motor neurone association nurse, I said to her that I would like to alert the palliative care team to my condition and, because they just opened a new hospice down in @@local catchment hospital## and she said to me do you not think it's a bit premature to involve the palliative care people. And I said well I'm not so much talking about involving them in my condition at this stage, but to acquaint them with the fact that I may be, may have to involve them at a later stage. Now, at that stage it wasn't coming on as rapidly as it is now, but now I'm reassessing my situation in relation to, to asking my GP to alert the palliative care people perhaps to come in and have a chat with me some day. Does that make any sense?

G: Absolutely. It comes across to me that you are reassessing all the time depending on what's happening, you know, you're judging the progression yourself because you're experiencing that progression and so you're thinking, hmm, okay maybe it's time to do X, Y and Z. So I suppose in a sense you're planning for the future.

P#31: Yeah, well that'd be correct, yeah.

G: Have you had any discussions, specific discussions @@Martin##, around life-sustaining treatments? You know, when, if and when you have respiratory problems you know.

P#31: @@the consultant neurologist at national ALS clinic## when I spoke to her last time she said to me, on my last visit six weeks ago said to me when I'm speaking to you, you are slightly breathless, and she says so I'm going to send you for a lung test and I said, and what's the, what's the reason for that and she said because if you're not getting enough oxygen in your system you'll be very flat and I'm very flat, very little energy and she said, and she said if, if your lung tests shows that your oxygen levels are going down, we will provide a machine that you take for six hours a night, you breathe through this machine. And I said it sounds like a medieval torture, you know and she said but if it helps you to get

around, you know. But as it happened, the, the lung test was pretty okay. And then when I was down there yesterday she sent me for a different lung test, which is pretty okay as well. So thankfully that hasn't happened. In relation to the feeding, I can still, I can still eat, I still chew my food and swallow and I'm not getting this feeling of being blocked up at the back although I have read about the feeding and I spoke to a man down in the clinic one day who was on that, on that, that abdominal feed. So no, at this stage I'm, at this stage I'm not at that stage.

G: **Do you have any particular feelings about that, because do you see them as life-sustaining treatments in themselves? Life sustaining - I mean sustaining life time wise. Do you have any feelings about them in the context of a condition like motor neurone disease?**

P#31: Well, let me put it to you this way. If for some reason I went into a coma tomorrow, I would not want to be resuscitated. I don't want to come back. I don't want to come back to face all this all over again, I'd be happy enough. I wouldn't say this in front of my family by the way.

G: **Yeah, sure.**

P#31: Although I may say it to @@Emily## who, who's going to be my carer. She works in a bank and she's, she's taking two years leave of absence, but she won't need that much, but that's, that's what they are allowing her, two years leave of absence so she'll be officially my carer. She lives just about less than a mile away. If, if I for any reason went into a coma I would not want to be resuscitated, I'd be happy enough not to come back.

G: **And continue to face this?**

P#31: Yeah, yeah.

G: **So for example if for some other reason tomorrow, god forbid you had a heart attack, independent of MND that was to put you in...**

P#31: Because I have a heart condition, I have two stents.

Q **Oh do you, oh right.**

P#31: I should have told you that.

G: **So yeah, that's a good example. Let's say you suffered a heart attack tomorrow which threatened your life and put you into a coma as you said. You would be happy to go then, versus come back and face this?**

P#31: Yes, no I'm on, the only medication I take any day, the only thing, is two Rilutek a day, I take nothing else, I take nothing, no heart medication at all.

G: **Should you encounter some more respiratory problems, maybe this machine for night time would be useful for you, it's called NIPPI – would you be open to that then? I mean I know it's not the situation right now but do you think about that?**

P#31: I would have, I would have great difficulty with any kind of artificial aid to keep me alive.

G: **You're thinking you might have some problems with that?**

P#31: Yeah

(Interruption – P#31 talks to builder who enters room. P#31 and G move into a different room to continue interview – Builder needs to work within kitchen)

G: **Okay, I'm just going to bring this out here I'll leave it running so I don't have to turn it off. Okay ... mind the step there now Michael. Will I go over here?**

(Interruption – moving into different room)

G: **Okay?**

P#31: It's a bit warm here, is it, will I open this?

G: **I'm okay, unless you want to now.**

P#31: No I'm okay.

G: **Right, I'll just leave that there.**

P#31: Get yourself comfortable there, Geraldine.

G: **You've plenty of rooms in the house anyway.**

P#31: Yeah I spend all my time out here, it's lovely.

G: **It's a lovely house you have. So the NIPPI, you might, you think you might have problems going for that?**

P#31: I think, I think to be honest if I were to, I'm not a great sleeper anyhow, if I were to spend, if I had to spend six hours a night just to give me a little of extra energy in the day and time I wouldn't do it, I really wouldn't do it.

G: **And what if it was the case, for example I know that @@the consultant neurologist at national ALS clinic## advocates that aid if people from a medical point of view require that. One might say that @@the consultant neurologist at national ALS clinic## is quite an expert in that area and so if she encouraged you, you would be swayed?**

P#31: Well I had this discussion with her about the high fat diet because all of my life, my body's programmed for a low fat high carbohydrate...

(Phone Rings)

P#31: All my life I have been programmed for to keep very, very thin, keep my heart rate down so my feeling is I'm, excuse me...

(Interruption – Phone Call)

P#31: That's UPC, I have Sky television and I'm changing it to UPC because I've no broadband in the house and I have to get it.

G: Yeah I had that done last year and just to get everything on the one bill.

P#31: That's it exactly, yeah, because I'm paying Eircom, I'm paying Sky and...

G: I know, I just did it for convenience sake, you know. It's fine, you know, they just have to come out and do the cable thing.

P#31: I don't think they're quite as good as Sky but anyhow. Now I was saying to you that I, like I started running when I was 10 and I finished running when I was 50 because I broke a kneecap and then I took up cycling and in all that time I got to know my body really well because if you go run marathons on a regular basis you really have to know yourself, you have to know how to, you have to know about your nutrition, you have to know what kind of training to do, how to get your body ready. I mean I got my heart rate down to 46 to do long distance and then I got my weight under 10 stone, which is about perfect for marathon running, you know, big heart, small body. So when I spoke to @@the consultant neurologist at national ALS clinic##.

G: About the diet?

P#31: She said to me about, she said you, she said all your, your preconceptions of what was best for you now you have to forget all that, so I'm going to put you on chips and burgers and I said I can't do that. I said I, I said I just can't do it because I said, I said apart from my body coping my mind is also programmed to reject all those things, I will not be able to do that. Now I did in a limited fashion, I you know, I started eating full fat butter and I had chips, but I never got to the point where I could actually do what she wanted me to do.

G: Was that because you think your whole mind frame before MND came into your life, it's just really difficult to completely leave that behind?

P#31: Yeah

G: Enter a new mind frame. I see. Even though you understood her reason for advising you to do that?

P#31: And she didn't quite understand.

G: Did you sense that?

P#31: She didn't quite understand why I would resist change to this. She said but you should be delighted because now you can go out and eat your ice creams and eat your chocolate and I said, sorry I just could not do that, you know.

G: So you're unsure. There's your daughter back.

P#31: There's @@Emily##.

G: Busy house! You're unsure so about whether or not, if needed, you would use the NIPPI, which is the breathing machine at night time. It's possible that if you didn't undergo this

particular treatment, if clinically advised, you would have less time. I mean it is considered a life-sustaining treatment.

P#31: Yes, yes.

G: So even though it could be life sustaining in a sense of time, you still wouldn't necessarily jump at it just for that sake?

P#31: That would be, what would be my last resort, I wouldn't, no, that'd be the last thought I'd have, I just wouldn't now, the breathing, the breathing thing is probably something that I would, I would certainly not do, just to give myself some extra energy during the day and a little time. Now another reason is because I sleep in the same room as my wife, we sleep in separate beds, but our beds are beside each other because she's a brittle diabetic and she often has hyperglycaemic reactions during the night and the only way I'm conscious of it is if I'm in proximity to her. If I had a machine there that was making any kind of a noise, she would not be able to sleep and I wouldn't be able to sleep anyhow. So for all of those reasons.

G: Its many factors really, isn't it?

P#31: Yeah, yeah. The feeding tube is different, I wouldn't, I wouldn't like to be on a feeding tube to prolong, to give me a little bit of extra, to prolong my life. At the same time I wouldn't want @@the consultant neurologist at national ALS clinic## or others think he's starving himself to death, by not taking the tube. Because I don't know what, I don't know what alternative, if they recommend the feeding tube, I don't know what the alternative is if you can't swallow or if you can't digest food, I don't know.

G: I mean if a person is at a point where they really can't swallow, you're not at that point, you know. You may never be at that point but some people just can't swallow and they lose weight rapidly, really rapidly now, talking months.

P#31: Yes and by the way, neither would I want to feel that I'm lying in bed waiting to die and in a way committing a sort of a tacit suicide, I wouldn't want that either. You know, the fact I'm not going to eat, you know, and in that condition, in that situation I may consider if I were in the situation I may consider the feeding tube at that stage, but I wouldn't, I would never take it to prolong my life.

G: Even though by taking it, it might?

P#31: Yeah

G: But the choice, the reason why you might opt for a feeding tube isn't life sustaining. I see what you mean. So you could potentially consider a feeding tube, not because you want to sustain your life, but because by the alternative, you would be starving yourself and for you, that might be some form of assisted suicide?

P#31: Yeah

G: Even though technically, by definition it's not?

P#31: I'd have to argue that out in my own mind because I have the, I'd have to be very sure in my own mind. I'm not a religious person as such, although I am you know, I am a moral person. I wouldn't want to put myself in a situation where I thought myself that I were committing suicide. But having said that, I would recognise that if, if the natural way to nutrition is to swallow food and you can't do that, well then the moral argument about whether I should use a feeding tube or not has a different dimension.

G: **So are you suggesting then @@Martin## that there is a moral dilemma in relation to maybe an expectation or responsibility to continue to live with MND, within what treatments are available and to use them because it allows you to live? Or are you saying then by declining certain interventions like that you are giving up?**

P#31: No, neither of those reasons, I don't see, I don't see what I'm thinking in what you're saying. I would have to give it more thought really, you know.

G: **Yeah, it's complex.**

P#31: Yeah it is a bit complex, yeah.

G: **Anything else that comes to mind at all that you can think of or? One thing some people have mentioned to me is when they think about MND and it's progression, some people have fear about, fear of suffering, physical suffering with MND. Do you have any fears around that?**

P#31: I've a high tolerance to pain. I'm terrified of being of my life being in everybody else's hands except my own.

G: **Control then, is it?**

P#31: Yeah, the thought of like sitting in a wheelchair paralysed and depending on other people to do for me, that to me that's, that's a total nightmare, that is my worst nightmare, I would hope before I got to that condition that I would, I would die. It's not in my own hands, but I, I hate the thought and I always said in my whole life, I always thought I was keeping myself healthy by my running and cycling and good diet and the great fear I had in my whole life was that I'd end up in the medical professions' hands and here I am, that's exactly where I am. I hate that.

G: **Do you think that's primarily a control issue? It's the potential of not being in control that frightens you versus the situation, it's not necessarily progressing with MND, it's the question of as you progress with MND, you give some more control over to services to look after you. It's just inevitable that the less you physically can do, somebody else does for you and the balance changes nearly, you know?**

P#31: The balance is changed even now, because when I'm using that walker, for, if @@Pam's## not here I can't even make a cup of tea for myself now, because the, all the things you have to do, to get a cup, I can't lift the kettle first of all, if I could to pour the water over here, to get the milk over there, bring the milk back and do all that, but then I have to get the cup

to the table. And when I, it's very difficult, you know and already I'm losing control over all of that. I can't dress myself now, you know.

G: So you are aware of that. Just thinking back on some stuff I read about people, people have looked at this in MND, people's coping strategies. Is it that you're feeling less in control do you think?

P#31: Yeah, absolutely.

G: And even though you feel less in control, are you aware of any strategies you might use to try and get that control back or try and be in control?

P#31: Well I still try and do whatever I can myself, but I'm, I'm beginning to realise that you know, I'm not going to be able to control all these things and I really don't like the fact that I will depend so totally on people to do things for me. And nobody does things the way you do things yourself anyhow, you know.

G: Are you a perfectionist?

P#31: No, no, but like say for instance let's say god I'd love a cup of tea now, I'd get up and make it. If I said to @@Pam## will you make me a cup of tea I could be waiting 15 minutes, you know, she's doing other things anyhow and you're sitting there saying.

G: So is it the simple things of life nearly, really, for you, you know we all take for granted?

P#31: Yeah, and all going by the board.

G: So in that context then, do you think there's a huge amount of loss in MND? People experience loss?

P#31: Loss of control?

G: Loss of anything, loss can mean different things for different people.

P#31: You lose everything, absolutely everything, yeah, yeah. Now we used to, I, this maybe, may not be due to the physical condition of MND, but it most certainly is related psychologically. I used to love in the evening to sit down and have a glass of wine, bit of cheese, no real desire now. No, no feeling that I'd like to as I say, would you like a glass of wine, I say ah, not really you know and yet that's only happened in the last maybe, last month. I've no feeling of wanting to do stuff like that. Now we used to go to the movies every Sunday morning, we'd get the, take the car down the town and we'd go into the movies down in @@local town##, but even though the girls would put me in the wheelchair now and bring me down, I'd have no interest in going to a movie. And very little interest in even going out you know, that's all related to the MND. And yet I would feel myself, if I was speaking to somebody else who had the condition and if they said to me, say come on, I'll bring you up for a pint and if they said to me ah I don't want to, I don't think so, ah come on what's wrong, come on.

G: So is it that you have to experience it to really understand it?

P#31: Yeah.

Appendix D

G: Do you think healthcare professionals can, healthcare services, healthcare professionals can ever understand it then if they don't have it? I mean we can be helpful and so forth, but if you don't?

P#31: I think there's more happening than people can see or realise. Personally I think it's, I think it's my, my own, my own body is withdrawing from my old life into this new situation and I'm, I'm discarding all the things that, that I used to do, used to enjoy. I'm not doing it deliberately, up here it's happening, my body is saying you know, pull back from that. And that's what's happening.

G: I can't think of another question, unless you've got one, it's been really interesting. Just one more thing actually, one thing that crops up from previous interviews is people talk a lot about what they view as caring, people have different interpretations of what caring is. What do you think caring means?

P#31: What caring is?

G: Caring, yeah. When I speak to people they have different interpretations of what they view that as. What do you view caring as?

P#31: Many answers to that, I suppose really caring for me is the, to be available when I need somebody that they're available. To do whatever you know, I actually need. You often, people often come in and will tell you what you need, not necessarily me telling them what I need, I think caring is that. If I say to somebody I need you to do this, that they would do what I asked them to do, well in so far as possible rather than somebody saying to me we'll do this for you, whether you want it or not, you know.

G: So respecting your view point, not just respecting it, but following through with it?

P#31: Yeah, yeah.

G: Do you think that you value services more, value healthcare professionals more when you think they're caring?

P#31: Oh absolutely, yeah.

G: And if you feel like that, does it make it easier to engage with services?

P#31: Yeah.

G: Right, so it's not all about just coming in and giving you information saying X, Y and Z.

P#31: No, and it's, and what I find as well is that I like, I like somebody that comes in and has maybe a kind of a sympathetic approach, rather than a business-like approach. Okay business-like is okay if you're running your business, but if you're, if you're dealing with somebody who feels vulnerable with this kind of illness, it's nice if someone can come and would have like, would sit down and say simple things like you know, how are you today and not say to you, this really kills me now, when I meet some of my friends and like maybe put on a suit or maybe put on a polo shirt and a pair of jeans and meet and they say Jesus you look good enough to run a marathon, you know. That kills me.

G: **You mean this sort of, what do you call it, patronising, is that the right word?**

P#31: Maybe, probably not meant to be patronising.

G: **Unintentional patronising?**

P#31: Unintentional patronising or somebody saying to me god you look better than I do, you know, please don't say that. I say to myself, okay I do look okay on the outside, well not today, but before I got this *(P#31 referring to facial bruising from fall)*, I mean I looked exactly I did maybe five years ago, you know. So, but and I do, I kind of understand, understand when people say to you know, because you're sitting down somewhere in the pub or in a restaurant or something, having a drink or something and they say god you look okay to me or you know. And you feel like saying well I'm, you know, in here I'm not okay. But I don't do that but I kind of simmer a bit. I say to myself I wish you didn't say that. And funny enough I stop meeting people after that, that I felt that I didn't want people to be saying to me oh you're looking great today, you know.

G: **Because one thing you just mentioned there @@Martin## was you prefer people who listen versus people who come in with a business-like approach. You mentioned earlier on in the interview that you started off in the private sector, you know, you have private insurance, even though you have the medical card now you went to see the neurologist first in a private capacity because you normally would have done that, I understand. Do you think there is a difference, I mean number one do you think public healthcare services should, can ever be a business? I'm just trying to figure out, you know some people talk about public healthcare as, it's not a business, you can't generate profits, private healthcare has to generate profits to survive and the state funds public healthcare. Do you have any thoughts on that, in terms of how services are delivered?**

P#31: Well I know, my wife was in the @@private hospital## for three months and as a private patient, and I thought they were hugely business like. But there wasn't an awful lot of sympathy, all the way down, from the very top down to the very bottom, right down to the consultants, the doctors on the wards, the nurses, all the way down, even the physiotherapist, she was unable, she was in there for three months, she walked into the hospital and came out in a wheelchair. I was able to get a physiotherapist myself who got her walking within three weeks of coming out of the, whereas the professionals who were dealing with her, they were so business-like in their attitude towards her that she was not able to, to start walking again. And I felt, that was the private sector. I find in my dealings with the public sector there's a whole lot more sympathy going on, there's a whole lot more, I see in relation to my wife and myself that there is a different attitude, it's more of a, a person to person rather than I'm doing a job, you know.

G: **It's interesting, some people say that, some people don't but yeah, I wonder sometimes is that got to do with the fact as you, as I said private healthcare is a business, it has to be a business, otherwise it doesn't exist, yeah. It's a different model, isn't it?**

P#31: Now there are exceptions of course.

G: **Of course, absolutely, there's always exceptions. Anything else at all, I mean before we wrap up or if there's anything else that's really important to you that you haven't talked about yet or we haven't even touched on?**

P#31: Only that I find that, I found, and I have found from the vary start with MND that it's kind of smoke and mirrors, like everybody, no one is prepared to say to you, like for instance where are you in the disease.

G: **Have you asked those questions? Have you said to @@the consultant neurologist at national ALS clinic## where do you think I am right now, in terms of prognosis, timeframe? Where do you honestly think I am?**

P#31: Oh well I do.

G: **What has she said?**

P#31: They all, everyone says, well everybody's different, you know, and, and I said well how do I gauge where I am if everybody's different, well you gauge yourself against yourself. Well how do you gauge against yourself, well how are you, how were you this time last month and how are you this month and how, have you, have you progressed further into the disease, in that period of time. And I say, well, yeah but you know that still doesn't tell me, and at this moment in time that's where I am, you know, we're all different.

G: **Let's say if somebody was prepared to say to you right, would you, do you want to hear it?**

P#31: Well I think they're all afraid to say it, in case you can't, in case you can't or won't take it on board and I do understand that because I was down, having acupuncture with @@acupuncturist## his name is, down in @@nearby suburb##, he's an acupuncturist and doctor and he's been a friend of, of mine for many years and he also has an interest in a number of nursing homes and because he knows, because he knows about @@Pam's## oncoming dementia and the fact that she's, she's quite unwell herself a lot of the time. He said to me I wouldn't have this conversation with many people he said, but I'm going to have it with you he said because I think you, you are able to have this conversation he said. He said have you considered between now and end of life what you're going to do, because he said one of the alternatives he said if I were you, I'd consider, first he said one of your daughters now is going to be a carer he said not only for your wife as well and he said she would need a carer on her very quickly, you know. And he said you will probably need a carer on your own, so he said it's the most onerous job possible he said, being a carer and he said whatever about her working but he said, he said I know you had

problems the other night. If your wife has a, a diabetic reaction during the night, he said you will have to ring her he said and she's not living in your house. Very difficult, he said, he said and in view of the fact where you are and it's progressed so quickly he said, he said and he said I'm not trying to drum up business. I don't need to but he said would you not consider a nursing home. He said where you will be, he said where you will be cared for, he said you might very quickly get to the point where to get out of bed you might need a hoist to get you out of bed. A hoist, a wheelchair all the difficulties about the toilets, he said all of that he said, that can be easily taken care of in a nursing home and he said a good nursing home, not talking about you know, he said a good nursing home all that'd be taken care of, he said and it would leave your daughter to be able to look after your wife. Which in its own time is going to be a full time job anyway. And he said would you ever consider all of that and I said, he said I'm not saying that to you lightly and I wouldn't say it to many people, but he said whatever time you have left, that he said that you'd be comfortable, you know your wife is well looked after and that you'll be looked after. And he said, and because the nursing home's that I'd recommend to you are within fifteen minutes of where you live, he said it'd mean that your family have access to you. You know and I thought about that and I couldn't, I, I couldn't make a decision on it.

G: **You could?**

P#31: Couldn't

G: **Oh you couldn't, right.**

P#31: I, I could see all the, I could see all the pluses for, for everybody involved, but

G: **It still didn't seem like the right decision?**

P#31: Well, I thought it may have been the right decision for me, not, not maybe, not at this stage but a little bit later but as he rightly put it, the time to make decisions when you are in control, when you can pick a room that you like, when you can tell them what you want to do, what you want them to do for you, you know. But I couldn't make that decision because I didn't feel I was that far enough down the road. But, and neither is @@Pam##, that far enough down the road.

G: **It's like you know, the decisions you make or you don't make inevitably involve your spouse.**

P#31: Yeah

G: **Yeah, it's not just you making the decisions for yourself?**

P#31: Well if, if I were living on my own I would have said yes, it would have made a lot of sense to me.

G: **So the context is that you are living with your spouse.**

P#31: Yeah, but having said that, the very fact that @@Emily## will have to look after both of us and @@Pam## is not hugely mobile either, and like we get meals on wheels delivered

now, she can't, she forgets that it's lunchtime or it's tea time and I'd have to remind her and all that and then everything she does now I'd say have you done this, have you done that, you know and I find that very stressful having to remind her all the time and almost saying I'll do it myself. And yet I can't. So like at the end of the day I think if I were in a nursing home and @@Emily## were looking after @@Pam## that might be the best solution. But I, I can't do this, if I said that to @@Pam## it'd kill her.

G: **Yes, you take her into consideration all the time, absolutely. Anything else at all, you think, we've covered a lot actually.**

P#31: Covered a lot, yeah. I hope I've been of some help to you.

G: **Absolutely, everybody's so helpful, I mean to be honest to give me your time you know. I'm travelling around the country doing this so it brings me to various places. But unless you've anything else to add that really sticks in your mind that you think is important to share?**

P#31: I think we've gone through most of it.

G: **Okay, alright will I turn this off then?**

P#31: Yes.

G: **Okay.**

P#31: Thanks a lot.

(Recording Ends)

Appendix E

Respondent no: P#23

Pseudonym: Pascal

Age 41. Spinal onset ALS. Attends BH clinic. P#23 not known to me prior to interview.

Geographical location: ****** (non-urban)

Length of interview: 1 hr 20 mins

Contact date: 16/04/2012

Today's date: 17/04/2012

Theoretical sampling √

Sampling based on:

I continue to sample for variation in experiences and remain open to new concepts including variation within categories. P#23 was theoretically sampled to at some point in the interview explore further the concept of loss in ALS. All participants so far speak of uncontrollable loss. I felt that so much of P#22 interview centered on loss. She described herself as deeply frustrated because of unremitting loss in her life. Poignant was her comment – "the future is so grim because of all the loss". I decided to sample a young participant and explore, in part, his views on the future with ALS. P#22's losses are physical and non physical but are her losses in some way dimensionalised by the extent of her physical loss? P#23 is less physically disabled than P#22. I wonder are his perceptions of the future as "grim"?

Short description of the setting, atmosphere etc:

Welcomed by P#23. P#23 now works from home (high-earning private sector professional). Some casual chat over coffee prior to recording – P#23 spoke about how he came to live in @@county## (originally from @@other county##), effort/toil in building their home (impressive home) and now less likely to move again.

1. What were the main issues or themes that struck me in this contact?

Once again, I am struck by what seems to be a struggle between accepting ALS and not yet giving into ALS. As for all other participants, P#23 is aware that ALS is terminal. In fact, P#23 touched on the terminality of ALS more so than any other participant thus far. I felt P#23 seemed almost pragmatic in his approach: spoke openly (and without visible emotion) about at least having the opportunity to settle his affairs (decide on place of death, burial vs cremation, etc). This he feels gives him "peace of mind". He is eager that his spouse will have little to sort out after he has passed. He envisages that the period after his death will be a difficult period for his spouse. P#23 disclosed that even though he feels accepting of ALS, he also feels determined to fight ALS – he continues to do the best he can to fight the progression of ALS i.e. healthier lifestyle habits, early

participation in clinical trial. He is certainly not accepting of death anytime soon. I sense that P#23 has reflected much more on his mortality than he might otherwise have done had he not developed ALS.

P#23's views on healthcare services are mixed. He is deeply frustrated by what he describes as the 'grey box' of the HSE. Similar to a number of other participants, he was not aware (prior to ALS) that a medical card is necessary in order to access primary care services in full. He is appalled that access to public health care is essentially means-tested. This he feels constitutes a violation of his human rights – access to public healthcare is a right. He has received a medical card since his diagnosis but he has thought about how many people who are over the threshold for a medical card are discriminated against simply because they have a reasonable income. P#23 feels there are many irregularities within the HSE. He admits that he encountered little difficulty in obtaining a medical card but disclosed his application to the HSE was perhaps assisted by the efforts of a local TD [politician]. P#23 is also aware that private healthcare care services have little to offer the ALS service user. He had always subscribed to VHI [private health insurance] and had rarely availed of public healthcare services in the past and/or needed to engage with public healthcare services. As for the majority of participants, P#23 is concerned about the future of healthcare services in Ireland. He spoke openly about his future care needs and he anticipates that he will require fulltime care. He feels that it is unrealistic to expect that the HSE will provide all services in the context of cutbacks and the likelihood of further cutbacks in public expenditure. In this context, he anticipates that the IMNDA will assist him and he has established links with the IMNDA and collaborated with the IMNDA in initiatives to increase awareness of ALS. Nevertheless P#23 is impressed with HCPs in the public sector. He has encountered a number of HCPs in the public sector and found them to be professional, informative, reassuring, and hard working. Similar to other participants, P#23 feels that public-based healthcare services are more than satisfactory if one can access these services. In this context, he believes that the HSE is a system failure versus a person failure. He is frustrated by the lack of information available to services users. He feels that the HSE fails to advise services users about how to access local services and he is grateful to BH clinic personnel for referring him onto primary care services.

Striking is how P#23 spoke about how he is adapting to ALS. Similar to the majority of participants, he suggests that adapting to physical loss occurs without choice. He also feels that one has no choice but to adapt to services as services are essential in order to assist him to live with ALS. Once again, I am reminded of how many participants speak of "no choice". In addition, P#23 feels that he has no choice but to plan services ahead of needing them. His preference is to request services at the point of need but suspects that he has no choice but to request services ahead of time (owing to the length of time associated with processing service requests, etc). P#23 also feels that if he

were a parent, he would more than likely have to plan services further in advance than he has done so far.

As for all participants so far, P#23 has experienced and continues to experience loss. Loss for P#23 seems very much about loss of future achievement, life ambitions, etc. However, he also speaks about loss of "mid-term" plans – the impact of ALS on his everyday life. For P#23, physical loss creates other losses and shapes the experience of LOSS – he suspects that one encounters more non-physical loss with increasing physical loss. P#23 differentiates between 'existing' and 'participating'. He suspects that come the time when he will require total care, such an 'existence' could prompt him to travel to Switzerland in order to avail of euthanasia. Similar to P#20, he admits that he is unsure whether or not he might feel differently come the time. As for my interviews with P#11 and P#20, I asked P#23 what he meant by total incapacity as it is at such a point he might choose euthanasia. P#23 feels that total incapacity equates to complete physical dependence on others. He doubts such an existence would be acceptable to him. He acknowledges that euthanasia is a complex issue but nevertheless he feels that one should have the right to choose. Interestingly, P#23 suspects that decisions pertaining to euthanasia are perhaps more difficult for people with ALS than for people with cancer (in the context that neuropathic pain is not a feature of ALS). Overall, I sense that P#23 wants to live on. However, certain conditions i.e. complete physical dependence on others might well prompt him to hasten death.

Similar to the majority of participants thus far who have encountered @@the consultant neurologist at the national ALS clinic##, P#23 is very impressed with @@the consultant neurologist at the national ALS clinic##. As for so many participants in this study, P#23 spoke about clinical encounters with @@the consultant neurologist at the ALS clinic##. When asked why he holds her in such high regard, he disclosed that he is impressed by her clinical expertise, public reputation, and research activity. I sensed that P#23 admires @@the consultant neurologist at the ALS clinic## – "matter of fact", "approachable" if not a little "brash". Yet, he also feels that @@the consultant neurologist at the ALS clinic## is genuinely caring. Caring so far incorporates many different properties & dimensions **(see codes / concepts under 'caring')**. Many participants describe @@the consultant neurologist at the ALS clinic## as "matter of fact" and "doesn't sugar coat anything". And yet, all feel she is caring and focused on the service user.

I think BH clinic, @@the consultant neurologist at the ALS clinic##, other BH clinic personnel, and the trial are significant contexts to P#23's experiences of health care services. Like some other participants, he does not associate the BH clinic with HSE services even though the BH clinic is a public clinic in a public hospital funded by the HSE! In the absence of a BH clinic, he suspects that he would be at a loss as BH clinic personnel linked him with local primary care services. He also suspects that personnel at the BH clinic are more expert in ALS care than those who work in

primary care. P#23 commenced on the clinical trial at BH. However, he had to withdraw from the trial as he experienced a rapid fall in his neutrophil count. He is disappointed that he had to withdraw from the trial. Nevertheless, he remains hopeful of a positive outcome to the trial and is reassured knowing should an effective treatment become available, he will have access to the treatment.

2. Summarise the information you failed to get from the interviewee? Why?

I felt that this interview was a 'full' interview – lengthy, open and covered a range of experiences. P#23 did not become emotional at anytime in the interview. Interestingly, he disclosed that he feels as if he is still "looking in from the outside". When prompted why this is so, he suspects that this is because he has not yet encountered significant physical disability.

3. Anything else that struck you as particularly salient, interesting, illuminating or important in this contact?

- As for many other participants interviewed thus far, P#23 inevitably compares himself to other people with ALS. He is "ecstatic" that his progression has been somewhat slower than he had expected (he suspected the "worse case scenario" at time of diagnosis). I think P#23 anticipates ongoing **change in his life** in line with the progression of ALS.
- I sense that P#23's efforts to raise awareness of ALS and assist others with ALS are not *entirely* altruistic. His disclosed that his efforts may also serve him well i.e. potential to avail of IMNDA services when required.

4. What new or remaining target questions do you have in considering the next contact?

- All participants thus far have spoken about the challenges of engaging with the HSE. The HSE is an uncertain place – continue to be alert to similar and different views.
- Are efforts to assist other people with ALS ever truly altruistic?
- Is LOSS in ALS somehow determined by the restrictions imposed on people with ALS because of physical loss?
- Do other service users move from the 'outside' to the 'inside' with increased physical disability and associated loss?
- So far it appears that people with ALS shift from fighting ALS to accepting the terminal stage when they become totally dependent on others for their care. Is this also the case for future interviewees?
- Explore further what is meant by "existing" versus "participation" – related to loss.
- So far BH clinic is a key dimension to the healthcare experiences of participants (who have availed of this service). Do some people with ALS feel differently? – I have not yet encountered a participant who feels that the BH clinic is of little benefit to them.

Appendix E

I have decided to sample a retired healthcare professional who attends BH clinic (as a tertiary-based service). Are her experiences in relation to services (negotiating the HSE, her expectations of the clinic) different? How does P#24 compare herself to other people with ALS? P#24 (middle aged / parent / grandparent) has already encountered significant disability and is at the end stage of ALS. Does she perceive things from the "inside" or from the "outside"? What does loss **[see dimensions and properties of loss]** mean to her at the end stage of ALS?

Appendix F

Scale on Control in ALS care
(Foley – Timonen – Hardiman scale)

How important is it for you to feel that you have a say over the health care services that you receive?

Very important
Important
Somewhat important
Not important

You have a say over the health care services you receive -

Never
Sometimes
Often
Always

"Having a say over health services helps to cope with MND" - Do you:

Strongly agree
Agree
Disagree
Strongly disagree