A qualitative study into the communication surrounding the initiation and withdrawal of non-invasive ventilation (NIV) in people with Motor Neurone Disease

Charlotte Chapman

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A qualitative study into the communication surrounding the initiation and withdrawal of non-invasive ventilation (NIV) in people with Motor Neurone Disease

This thesis is presented for the degree of

**Doctor of Philosophy**

**Charlotte Chapman**

Edith Cowan University
School of Nursing and Midwifery
2019
Declaration

I certify that this thesis does not, to the best of my knowledge and belief:

i. Incorporate without acknowledgment any material previously submitted for a degree or diploma in any institution of higher education

ii. Contain any material previously published or written by another person except where due reference is made in the text of this thesis

or

iii. Contain any defamatory material
Acknowledgements

This study was undertaken with the intention of improving my understanding, communication and ability to nurse and care for people with motor neurone disease (MND). During my nursing career, I have encountered several people with MND and each time I have felt humbled by their bravery in facing the inevitability of such a merciless disease. This thesis is dedicated to all those who have been personally affected by MND, their families and caregivers.

Contributing to my knowledge for this study was the courageous journey and ultimate death from MND of my brother’s good friend, “K”, in England. My brother put me in touch with his friend’s bereaved partner who generously communicated his experiences and thoughts via email. Whilst he was never interviewed as part of this study, his poignant emails have contributed to my knowledge of MND. My sincere thanks to K and her partner who provided rare and insightful thoughts adding to my learning and understanding of this relentless, cruel disease.

The initial study intentions were broad and wildly enthusiastic on my behalf and only intervention from my supervisors managed to curtail this thesis to a manageable entity. During the past four years I have understood the feelings of elation when a chapter has been returned from all three supervisors with praise and the feeling of despair when the equivalent of red writing and three differing thoughts have appeared in the markup area of my hard work! However, the fortitude of my supervisors to read this thesis many times requires something very special, and I am eternally grateful for their ongoing support and for concealing any boredom they may have endured. Therefore, I would like to thank Professor Anne Wilkinson, Professor Moira Sim and Dr Sara Bayes for the support and time they have given to me, and for their patience with my endless questions and dyslexic idiosyncrasies.
My greatest thanks go to all the participants who agreed to be interviewed at such length and who gave incredibly candid and at times, emotional replies to my questions. My appreciation goes to the one brave participant who was so determined to be a part of this study and whose experiences gave such insight. May you rest in peace, my friend; it was my privilege to have been able to spend some time and learn from you.

There are many people who through no fault of their own, became entwined in my study. My children Will and Nick who with great tolerance managed to share the study when home from university, risking their mother’s wrath if any journals or research papers were moved. The transcription writer Marilyn whom I happened to meet when we walked our dogs together one evening and who subsequently typed all twenty-six interviews verbatim. The kind understanding from my friends whose offers of food were gratefully received and who, despite my antisocial obsession with my studies, are still my friends. My sincere thanks go to my friend and colleague Robyn, who was studying for her own PhD whilst encouraging me to keep going despite the setbacks. Lastly my dear mother who provided me with the skills in life to “try” and who sadly died during my study.

I would like to acknowledge and thank the organisations who enabled me to spend time, interview willing participants and ‘snowball’ my research to others with interest in MND: Silver Chain Group, WA; St John of God Hospitals Bunbury and Murdoch, WA; Wisdom Hospice UK; Neurological Council of WA, and willing clinician participants in Sydney, NSW. Finally, I would like to thank Edith Cowan University for the opportunity to study and the ECU student support centre, SOAR, for their support and direction.
Abstract

Introduction

Motor neurone disease (MND) is a degenerative disease that adversely affects the nervous system and muscular control. Eventually respiratory muscles weaken, causing breathing, communication and swallowing difficulties, and ultimately, respiratory failure and death. Improved quality of life and potentially a short extension of life can be provided with non-invasive ventilation (NIV), which is offered to people with MND when symptoms of respiratory distress become evident.

It is recommended that end-of-life communication, encompassing the benefits and burdens of symptom-relieving interventions (NIV and percutaneous gastrostomy tube to assist with nutrition), NIV withdrawal (proposed when continued use is considered futile) and other respiratory distress-relieving interventions (e.g., opioids), is initiated either before respiratory symptoms emerge or at that time. Little is known, however, about whether, when and how this communication occurs.

Methods

This qualitative research sought to address this gap in knowledge by determining the content and timing of end-of-life options clinicians communicate to people with MND and their families. Nineteen clinician participants and six families, all experienced with MND, were interviewed. Clinicians’ accounts of their communication were compared to bereaved families’ recollections of communication by clinicians. A framework incorporating patient-centred care principles and evidence-based medicine was used to link clinicians’ communication to the most recent MND guideline recommendations. The data collected were coded and categorised manually for each participant transcript and again using NVivo 10 software. Trustworthiness was established through independent coding of randomly selected participant interviews by one of the candidate’s supervisors. Symbolic interactionism and interpretive description
provided the theoretical lens and methodology, respectively, through which the data were interpreted.

**Findings**

Despite the existence of comprehensive evidence-based guidelines reflecting international consensus, this research found some clinicians were unaware of the recommended timing and content of end-of-life communication, some chose to depart from or adapt the recommendations and some found the recommendations too difficult to implement. Clinicians stated they had insufficient time to communicate and reiterate the benefits and burdens of care options to ensure understanding. The recommendation to refer people with MND to palliative care soon after diagnosis was reported to happen infrequently. Clinicians stated that they felt discussion recommending early palliative care referral was confrontational for people with MND and their families; family participants confirmed this view.

**Recommendations**

Despite their ethos of patient-centred care, clinicians were concerned that early end-of-life communication would take away any hope the person with MND and their family may have. Consequently, many of the family participants appeared unprepared for the consequences of NIV, not having understood the potential of NIV withdrawal, with several turning to the Internet for answers. Compounding confusion was the disjointed nature of the care provided to people with MND often involving several health care providers.

This research recommends the establishment of a progressive interactive timeline, incorporating a case manager and reducing the number of external care providers particularly in the terminal phase of the disease.
List of Presentations

Oral Presentations

1) September 2014: Palliative Care Nurses Australia Conference, Sydney, NSW. *Communication surrounding the initiation and withdrawal of NIV in people with MND*: A 20-minute verbal description of the concept and planning of the study

2) September 2016: Palliative Care Nurses Australia Conference, Canberra, ACT. *Communication surrounding the initiation and withdrawal of NIV in people with MND*: A 10-minute speed talk with questions about the study and early results.

Poster Presentations

1) July 26-29th 2017: Asia Pacific Hospice Conference, Singapore. *Communication surrounding the initiation and withdrawal of NIV in people with MND*: a poster with study aims, research questions, method, results and recommendations. Three question and answer sessions relating to the poster were incorporated during breaks in the four-day conference program.
List of Abbreviations

AHD: Advance health directive

AIHW: Australian Institute of Health and Welfare

ALS: Amyotrophic lateral sclerosis

BMAA: Beta-N-methylamino-l-alanine

COPD: Chronic obstructed pulmonary disease

EAP: Employee Assistance Programs

ECU: Edith Cowan University

ECUHREC: Edith Cowan University Human Research Ethics Committee

EFNS: European Federation of Neurological Societies

FTD: Frontotemporal dementia

GP: General practitioner

ID: Interpretive description

MDT: Multidisciplinary team

MND: Motor neurone disease

MNDA: Motor Neurone Disease Association (U.K.)

MNDAust: Motor Neurone Disease Australia
MND NSW: Motor Neurone Disease Association of New South Wales

MNDAWA: Motor Neurone Disease Association of Western Australia

NHMRC: National Health and Medical Research Council

NICE: National Institute for Health and Care Excellence

NIV: Non-invasive ventilation

OT: Occupational therapist

PEG: Percutaneous endoscopic gastrostomy

PIG: Per-oral image guided gastrostomy

RIG: Radiologically inserted gastrostomy

SI: Symbolic interactionism

SRQR: Standards for Reporting Qualitative Research

WA HEALTH: Western Australia Department of Health

WHO: World Health Organisation
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Glossary of Terms

A “good death”: A dignified death in which symptoms are well controlled, the patient’s choice of end-of-life care is known and followed and emotional support for the patient and family/significant other and clarity of communication and explanation are provided (Steinhauser et al., 2000).

Advance care planning: A care plan put together by an individual, often in discussion with family and doctors. The plan directs a person’s medical care for a time when poor health and verbal communication may be adversely affected.

Advance care plan: A request for end-of-life care, whilst not a legal document (less formal than an advance health directive), clinicians are required to follow if known to exist.

Advance health directive: A legal document that ideally clearly stipulates a person’s end-of-life care preferences and acceptable interventions.

Caregiver: Those employed through organisations to provide professional personal care and assistance to patients with MND and their families. This may also include a member of the family who is employed by the person diagnosed with MND and their significant other, or it could be a person volunteering their time to attend to the patient and assist the family. In a hospital situation, a caregiver would be a personal care assistant, or assistant to the nurses.

Clinician: Health professional involved with people with MND at the stage of respiratory decline and/or involved with the terminal stage of the disease process when the withdrawal of the NIV is considered. The type of clinician may differ depending on the region where the patient with MND resides. In more rural areas, initial discussions and subsequent palliation may occur with general practitioners, whereas in larger urban locations, respiratory physicians, neurologists, palliative care specialists, palliative care nurses, neurology nurses and allied health professionals may all be involved as part of a multidisciplinary team.

Content of NIV communication: The information provided to the person with MND and their family regarding NIV use: benefits, burdens, limitations, process of initiation and withdrawal, the relatively temporary symptom control due to the
aggressive nature of the disease at the stage of respiratory distress, and medication use as alternative and/or in addition to NIV. Inclusion of NIV withdrawal and end-of-life choices is recommended (National Institute for Health and Care Excellence, 2016).

**Diagnosis:** The identification of a health issue by examining the signs and symptoms (The Oxford English Dictionary, 2013).

**Dyspnoea:** Difficulty in breathing or breathlessness (Mosby, 2002).

**Effective communication:** When the person with MND and family have received explicit communication from a healthcare professional regarding NIV, PEG and end-of-life choices, and the patient and family have understood the benefits, burdens and limitations of NIV, the temporary nature of its effect, and know what to expect in the process of initiation and withdrawal of NIV (National Institute for Health and Care Excellence, 2016).

**Epidemiology:** The study of the distribution, incidence and prevalence of health-related events or diseases and the application of this information to the recognition and improvement of health outcomes (Mosby, 2012)

**Hospice care:** All care required for a dying person, often delivered within a specific hospice unit during the terminal phase of life (Connolly, Galvin, & Hardiman, 2015). Hospice care may also be delivered at a dying person’s home, aged care facility or within a palliative care unit.

**Hyperreflexia:** Over extended and increased reflexes (Mosby, 2002).

**Inspiration:** The action of the diaphragm contracting and pulling down which draws air into the lungs (Mosby, 2002).

**Motor neurone disease (MND):** A progressive, degenerative disease affecting motor neurones (Oliver, Borasio, & Walsh, 2006). MND is used to describe the disease entity in Australia, the United Kingdom and other Commonwealth countries. Amyotrophic lateral sclerosis (ALS) is the most common form of MND. ALS (Lerum, Solbrække, Holmøy, & Frich, 2015) is used interchangeably with MND in the global literature but is a favoured description of MND in the United States, where MND is also referred to as Lou Gehrig’s disease after the famous U.S. baseball player (Oliver et al., 2006).
**Multidisciplinary team/care:** A team of multidisciplinary health professionals (MDT) generally including physiotherapists, social workers, dieticians, specialist palliative care doctors and nurses, occupational therapists and, often for people with MND, respiratory specialists and neurologists. Multidisciplinary teams may vary with availability of health professionals, and patients may only be able to access a few of the specialists relevant to their disease. Ideally, MDT professionals work as a team within an organisation and can meet to discuss preferred treatment and management of a patient.

**Non-invasive ventilation:** Used in people with MND for symptoms of respiratory distress, often commenced to alleviate nocturnal respiratory symptoms with increasing dependency as the disease progresses. Generally, bilevel positive airway pressure (BiPap) is offered. This is a method of non-invasive ventilation that delivers inspiratory and expiratory pressures at a variable rate via a face or nasal mask to help a patient breath. Regular respiratory functional testing, including the sniff nasal inspiratory pressure (SNIP) test, is used as a screening tool for the deterioration of the respiratory muscles and this may be used by clinicians to offer or increase the use of NIV (NICE, 2010). Eventually the respiratory muscles are unable to respond to NIV and its use becomes futile. NIV is withdrawn at this stage and death occurs.

**Occupational therapist:** A health professional who assists and enables people with their daily life activities. This may include provision of equipment such as walking frames, patient specific chairs, ramps and handrails.

**Opioids:** A broad term used to describe both natural (opiates e.g., morphine) and synthetic medications (opioids e.g., oxycodone) which act on the brain’s opioid receptors to relieve pain (Doyle, Hanks, Cherny, & Calman, 2005).

**Palliative care:** The World Health Organisation (WHO) describes palliative care as: “... an approach that improves quality of life of patients and their families facing the problem associated with life-threatening illness, through the prevention of suffering by early identification and impeccable assessment and treatment of pain and other problems, physical, psychological and spiritual” (World Health Organization, 1998).
**Palliative care management for people with MND:** A specific system of care structured around the priorities and needs of the patient and their significant others, incorporating a multi-disciplinary approach where possible, and where the benefits, burdens and limitations of all options have been considered (Motor Neurone Disease Australia, 2012).

**Paresis:** A partial paralysis caused by nerve damage which may affect any muscles of the body (Mosby, 2002).

**Patient:** A person receiving treatment for a particular ailment or disease, or a client using a healthcare service (Mosby, 2002).

**Percutaneous endoscopic gastrostomy tube:** A permanent feeding tube inserted into the stomach in an attempt to maintain nutrition and hydration when swallowing food becomes difficult. In Australia, two types of gastrostomy tubes are commonly used, differing only in their method of insertion: percutaneous endoscopic gastronomy (PEG), inserted via the stomach wall and radiologically inserted gastronomy (RIG).

**Process of communication about NIV:** How, when and where discussions about NIV take place and who is involved in the communication, as this is defined above.

**Prognosis:** The Oxford Dictionary defines prognosis as “An opinion, based on medical experience, of the likely course of a medical condition” and an opinion which could outline the outcome of the disease (The Oxford English Dictionary, 2013). An explanation of the course and outcome of a disease, in this case MND, is not necessarily a prediction of time.

**Respiratory failure:** Inadequate gas exchange in the lungs due to the nerves which activate muscles (in particular the diaphragm) involved with intake and expulsion of air in the lungs becoming too weak to function adequately. This may result in generalised fatigue, headaches and, ultimately, death.

**Significant others:** Family and friends who are significantly and emotionally involved with the person with MND.

**Successful communication about NIV:** The patient and family have had the content of communication regarding NIV explained effectively and have understood the benefits, burdens and limitations of NIV, the temporary nature of
its effect and know what to expect in the process of initiation and withdrawal of NIV.

**Terminal care** and **end-of-life care**: The dignified management of a patient who has reached the stage of disease progression where prognosis is one of imminent decline and death (Doyle et al., 2005).

**The literature**: For this thesis, this includes information written in English, peer reviewed prior to publication from January 2000 to July 2016 and published either as an electronic or hardcopy journal article or guidelines in a reputable journal or website.
A qualitative study into the communication surrounding the initiation and withdrawal of non-invasive ventilation (NIV) in people with motor neurone disease

Chapter 1

Introduction

Motor neurone disease (MND), of which amyotrophic lateral sclerosis (ALS) is the most usual form, is a degenerative disease affecting the nervous system which in turn adversely affects muscular control (Borasio, Voltz, & Miller, 2001; Gent, 2012; Oliver et al., 2006). The disease is most commonly diagnosed after the age of 40 years and is usually slowly progressive over 3-5 years, with an average life expectancy of approximately three years. The average length of survival from MND symptom onset is approximately 20-48 months although there is considerable variability within the disease. Most studies report a survival rate of 10 years or more in approximately 5-10% of people with MND (Chiò et al., 2013; Motor Neurone Disease Australia (MNDAust), 2014).

1.1. Epidemiology

The most recent published estimate of the global incidence of MND is 1.5 – 2 per 100,000 per year (Doyle et al., 2005), with an approximate lifetime risk of developing the disease of 1 in 800 (Phukan & Hardiman, 2009). A more recent systematic review of European literature suggests an incidence rate of 2.08 per 100,000 (Chiò et al., 2013). In a population-based study by Alonso, Logroscino, Jick and Hernán (2009), which utilised data from the General Practice Research Database (UK), the incidence was found to be higher in men than women with a lifetime risk of 2.7 per 1000 in men and 2.1 per 1000 in women, and the peak age at diagnosis to be 75-79 years (Alonso, Logroscino, Jick, & Hernán, 2009).

In 2013, 787 persons died from MND in Australia compared to 592 in 2001 (AIHW, 2015). According to current MNDAust statistics, two people are diagnosed with MND and two people die from MND daily (MNDAust, 2013).
Approximately 1,900 people have MND throughout Australia, and the MNDA estimates a further 14 people per person with MND live with the consequences of caring for a family member with MND (MNDAust, 2013). Therefore, approximately 26,600 people live with or have lives affected by a person living with MND at any time. In Western Australia, the Motor Neurone Disease Association of Western Australia (MNDAWA) reported that it assists approximately 145 people with MND and their families at any time, including approximately six months grief support (Department of Health, 2008 [WA Health]).

Despite ongoing research, the cause of the disease is unknown, and it remains incurable. However, autosomal dominant genetic defects that appear in over half of the 5-10% of familial cases diagnosed with mutations in the SOD1, TDP-43, FUS and C90RF72 genes have been closely linked to the disease (Gent, 2012; Oliver et al., 2006; Oliver & Faull, 2013). In addition to the suggestion that autoimmunity and viral infections are involved in the causation of MND, more recently a bacterial toxin, beta-N-methylamino-l-alanine (BMAA), has been found to create a faulty protein within cells, causing protein clumping and ultimately cell death (Dunlop, Cox, Banack, & Rodgers, 2013; Oliver et al., 2006). Dunlop et al. (2013) suggest that motor neurones appear to be susceptible to BMAA, and that the effect may be linked to neurodegenerative diseases such as MND.

1.2. Clinical Presentation and Disease Trajectory

The clinical presentation in MND is muscle weakness, which may involve all muscles (general paralysis), or partial paralysis corresponding to a single muscle group, with possible hyperreflexia (over-responsive reflexes) and neurone degeneration affecting both the upper and lower neurological pathways (Gent, 2012; Huether & McCance, 2014). The progressive neurological degeneration causes muscles to weaken, leading to communication and swallowing difficulties, potential aspiration and choking, respiratory distress and
ultimately, respiratory failure and death (ProGas Study Group, 2015; Skelton, 2005). The main cause of death from MND is respiratory failure (inadequate gaseous exchange in the lungs) caused by respiratory muscle weakness and often complicated by respiratory infection (Gruis & Lechtzin, 2012; Oliver et al., 2006). These infections may be caused or complicated by dysphagia (the inability to swallow effectively) resulting in involuntary aspiration of oral secretions, food or hydrating fluids.

Given the devastating implications of the disease certainty of diagnosis is sought. The current lack of a single diagnostic test means there may be a protracted period of symptoms, often up to a year, with exhaustive testing to eliminate alternative diseases prior to confirming a diagnosis (Andersen et al., 2012; Oliver et al., 2006).

1.3. Symptom Management

Unlike cancer there is no cure for MND, so interventions to improve symptom management are recommended to ensure optimum levels of comfort prior to the stage when the deteriorating patient can no longer benefit from such interventions (National Institute for Health and Care Excellence [NICE], 2016; NICE, 2010; Palliative Care Expert Group, 2010). These symptom-modifying treatments include non-invasive ventilation (NIV) for respiration assistance, gastrostomy tube placements (for nutrition and hydration when the ability to swallow is adversely affected) and medications to reduce pain, respiratory distress, secretions and anxiety.

1.3.1. NIV

The onset of declining respiratory function, reflected in symptoms such as headaches on waking or dyspnoea (difficulty in breathing), is a distressing and at times frightening experience for a person, and can severely diminish their quality of life (Rafiq, Proctor, McDermott, & Shaw, 2012). At this stage, NIV
to support a person’s failing respiratory effort is generally offered by respiratory physicians.

Respiratory failure cannot be prevented or reversed; however, improved quality of life and potentially a short extension of life can be provided by palliating the symptoms with NIV (Bourke et al., 2006; Rafiq et al., 2012). NIV delivers positive airway pressure to assist breathing and is administered via either a face mask covering the nose and mouth or by a nasal appliance. In MND, the positive pressure is usually delivered with a changeable inspiration and expiration pressure, known as a bi-level positive airways pressure (Rafiq et al., 2012). NIV is normally offered to people with MND via a device called a Bi-level Positive Pressure Airway Device often referred to by the registered trade name BiPap (Phillips Respironics, Inc., Murrysville, PA). These machines are programmed to assist inspiration (helping the diaphragm to contract and pull downwards) by delivering a precise airway pressure and can be set for either spontaneous or timed inspiration, depending on the strength and ability of a person with MND to breathe (Gruis & Lechtzin, 2012). NIV is thought to facilitate some resting of the respiratory muscles and improve comfort and quality of life by easing symptoms related to respiratory function, thereby potentially improving survival (Faull, Rowe-Haynes, & Oliver, 2014; Rafiq et al., 2012).

A Cochrane systematic review (Radunovic, Annane, Rafiq, & Mustfa, 2013) provides evidence that NIV prolongs the life expectancy of people with MND who do not have severe bulbar (lower cranial nerve involvement) involvement. The reason for improved survival in people with MND using NIV is not fully understood. However, it has been suggested that by resting and reducing the workload of the respiratory muscles, oxygen intake and absorption are improved; thus, NIV may contribute to a small increase in life expectancy (Rafiq et al., 2012). There is widespread agreement that NIV increases potential life expectancy due to improvement of symptoms associated with respiratory insufficiency, as stated in both the NICE (this organisation has formally be known as the National Institute for Health and Clinical Excellence
but will be referred to by its current name in this thesis) clinical guidelines (2010) and the more recent NICE MND Assessment and Management Guidelines (NICE, 2016), but not by improving the underlying disease progression (Doyle et al., 2005; Hardiman, 2011). Assuming no bulbar involvement, for which the research indicates no survival improvement with NIV, a median survival improvement of 205 days has been suggested (Bourke et al., 2012). Bourke et al. (2012) note that the timing of commencement of NIV may influence the length of survival, but the optimal timing of NIV initiation remains uncertain. As Bourke et al. show, early introduction of NIV may also be linked to poor adherence to NIV.

Approximately 20% of people with MND in Western Australia, who are physically suitable for and able to use NIV, will choose to use it in the latter stage of their disease (Baxter et al., 2013). Research has shown that almost all people diagnosed with MND will develop respiratory problems. Regardless of whether NIV is used or not, respiratory complications are the most common cause of death and one of the most accurate prognostic indicators (Miller et al., 2009; Rafiq et al., 2012). A small proportion of people with MND (five per cent) present with respiratory failure on diagnosis, whilst 30% present with bulbar dysfunction. NIV may be contraindicated in this instance due to disturbed swallow and gag reflexes resulting in increased secretions and risk of aspiration (Hardiman, Van Den Berg, & Kiernan, 2011).

The progressive weakness of the muscles of the neck region involved with swallowing and upper respiratory airway function (bulbar muscles), can adversely affect the symptomatic advantages of NIV due to an increased inability to clear secretions and risk of aspiration (D. Oliver et al., 2006). Whilst the majority of people with MND will experience respiratory symptoms and be offered NIV management, many do not accept nor tolerate NIV (Hardiman, 2011). The tightly fitting mask has the potential to cause areas of pressure discomfort often resulting in sores. Hardiman (2011) reports that some people
find the mask claustrophobic and may trial the nasal appliance or decline further use.

A proportion of people who tolerate NIV in hospital do not continue to use it at home due to a decline in the manual dexterity necessary to adjust the mask for comfort, increased secretions which may cause aspiration difficulties or a person’s general deteriorating functional level (Hardiman, 2011). An increase in symptoms and declining dexterity may be countered by increased caregiver support. However, this extra care may not necessarily be available, nor is it always part of a person’s care preferences as it increases the responsibilities and burdens of the caregiver.

Declining respiratory function becomes increasingly burdensome for people with MND and their caregivers and is often used as an indicator of nearing the terminal phase of the disease (Eng, 2006; Hardiman, 2011). Importantly, the time at which NIV commencement is indicated is generally acknowledged to be an opportunity to discuss patient preferences in end-of-life options including the use of medications, and the benefits and burdens of NIV including withdrawal (MNDAust, 2014; NICE 2010, 2016).

Some people living with MND may elect not to commence NIV at the time of respiratory distress, deciding instead to have well medicated symptom control and perhaps a shorter life (NICE, 2016). Comfort-giving and symptom-relieving palliating medications may be chosen because the person with MND does not wish to prolong their inevitable suffering, or perhaps because the person does not want to become an increasing burden on their family (Ando et al., 2014; Faull, Phelps, Regen, Rowe-Haynes, et al., 2014). Increased dependency on NIV is inevitable with disease progression as the respiratory muscles continue to weaken (Faull, Phelps, Regen, Oliver, et al., 2014). NIV is withdrawn when the respiratory muscles fail completely and prolonging its use becomes futile, or at the patient’s clear request. Death then typically occurs within minutes or hours of ceasing NIV (NICE, 2016). The withdrawal of NIV can
be an extremely traumatic and ethically challenging experience for both families and attending clinicians (Baxter et al., 2013; Phelps, Regen, Oliver, McDermott, & Faull, 2015). Families and some clinicians may have the perception that withdrawing NIV is a form of euthanasia, despite the futility of using NIV further and the terminal stage of the disease (Baxter et al., 2013; Faull, Rowe-Haynes, et al., 2014).

1.3.2. Swallowing and nutrition

People with MND may experience swallowing difficulties resulting in poor nutrition and weight loss in addition to respiratory weakness. The insertion of a percutaneous endoscopic gastrostomy (PEG) tube prior to the commencement of NIV is recommended, as later PEG insertion may increase respiratory complications (NICE, 2016). Further risks of late gastrostomy placement include potential insertion difficulties, which are associated with a higher incidence of mortality, and difficulty maintaining nutrition and hydration. However, as insertion of a PEG offers little or no life extension, and may cause possible discomfort, some people with MND reject the idea (Andersen et al., 2012; ProGas Study Group, 2015). Discussion of gastrostomy tube insertion and its implications early in the course of MND is recommended particularly if swallowing is affected (NICE, 2016). Despite the use of interventions, the disease progresses relentlessly toward the terminal stage.

1.3.3 Choices during disease progression

Figure 1.1 represents the course of MND in relation to the options of NIV initiation or refusal and the timing of suggested PEG insertion and outlines the progression of the disease to palliation. The flow chart is based on statistics obtained from the Motor Neurone Disease Association of Western Australia (MNDAWA, 2008) and does not necessarily reflect the percentages in other parts of Australia or the world. The intervention points for the communication recommendations (Figure 1.1) are based on recommendations from the NICE MND Assessment and Management Guidelines (‘the NICE Guidelines’) (NICE, 2016), which are discussed further in Chapter 2. The incidence of PEG tube
insertion is added to present a more complete picture of the progression of MND as the respiratory muscles start to fail, and as an indication of recommended communication points. Within each stage of the disease progressive an outline of the recommended communication, according to the NICE (2016) Assessment and Management Guidelines, is included. The red arrows indicate advancement to the terminal stage and end-of-life.
Figure 1.1. Disease Progression and Non-invasive Ventilation Flow Chart

(Flow chart created by the candidate with information from the Motor Neurone Disease Association of Western Australia, 2015)
1.4. Communication Experiences of People Living with MND

Research into the experiences of people with MND and their families frequently suggests that clinician communication is generally inadequate, particularly when delivering the diagnosis (Aoun, Connors, Priddis, Breen, & Colyer, 2012; Belkora, 2003; Faull, Rowe-Haynes, et al., 2014). For people with MND, deteriorating health and increasing symptoms mean that treatment decisions involving potential life-sustaining technologies are often faced early in the person’s clinical care. Thus, accurate information about the benefits and burdens as well as the life extension limitations of such treatments needs to be communicated early and revisited as the disease progresses (NICE, 2016).

A potential complication lies in how MND is perceived and characterised by clinicians. The variants of MND and the sometimes unpredictable timing of progression of the disease can mean that the point at which a person is perceived as having a chronic and terminal disease becomes difficult to predict. Whilst MND cannot be understood to be other than a terminal disease, it does have an indefinite illness course with a small prospect of technology extending life for some people, but not halting the disease progression. The point at which chronic illness becomes defined as terminal may confuse the timing of support required by the person with MND and their caregivers (Lerum et al., 2015). A further challenge for clinicians is the consideration of when to involve palliative care services. These may be restricted by the capacity of local amenities to support the necessary technology, the preparedness of caregivers for palliative care team involvement and the quality of life the patient desires. Given the relentless progression of MND and the common misunderstanding that “palliative” means “no hope”, there is uncertainty among clinicians as to when to introduce the support of the palliative care team to the patient and family. The World Health Organisation (WHO) states that early palliative care reduces unnecessary hospital admissions and that health professionals’ lack of palliative care training and knowledge poses a real barrier to admission to a specialist palliative care service (WHO, 2015b).
Anecdotally, multiple health professionals and several different service organisations may be involved with the person with MND from diagnosis, which has the potential to cause confusion and lead to conflicting messages. The often unpredictable nature of the disease and the changeable requirements of people with MND requires easily accessible support to maintain the person at home, less disruption for patient and family, and to prevent lengthy and costly admissions to acute hospitals. The most recent WA HEALTH data available on this topic is from 2008, when an average length of stay at a tertiary hospital for a person with MND was between two and four weeks at an approximate cost of between AUD10,000-AUD25,000 (Department of Health, 2008). Consequently, the report recommended early involvement of palliative care specialists and other members of a multidisciplinary team (MDT) in order to maintain people with MND within their home environment and to closely monitor their changing requirements those of their caregivers (Department of Health, 2008).

1.5. Death from MND

Patient preferences in relation to choices for end-of-life care require sensitive, explicit discussion and clear documentation by clinicians to reflect and achieve patient wishes (Smith & O'Neill, 2008). Hogden, Greenfield, Nugus and Kiernan (2013) found that the caregiver is as occupied and affected by the complex decisions surrounding patient care as the patient, and acknowledge that communication between patient, caregiver and clinical team is vital to achieve optimum patient care (patient-centred care). While there are guidelines regarding general end-of-life care containing recommendations specific to people with MND, these guidelines are mostly general in nature, broadly concerned with “what should be done”, and do not specifically define, describe, or detail the implementation steps for recommended best practice (National Health and Medical Research Council [NHMRC], 2012).
As death occurs soon after the removal of NIV and opiates are used to ease the distress associated with extreme difficulty breathing, families and significant others may feel confused and concerned if they have not understood the information provided to them or expected the events leading up to the end of life. Therefore, clear, unambiguous communication addressing the link between initiation and withdrawal of NIV and the end of life is critical for people with MND and their families. In order to make a fully informed decision about NIV, discussion and a shared understanding of all salient considerations prior to the onset of respiratory distress and loss of verbal communication is vital. This communication must include the limitations of NIV on disease prognosis.

Death can cause distress and fear for the patient and their family. Ideally, therefore, caregivers facilitate an early understanding for the patient and their family that, whilst death is inevitable, a “good death” is possible. Over the past 40-50 years end-of-life research has led to an agreement on what constitutes good end-of-life care and, through a series of exploratory studies, a consensus between clinicians, patients and caregivers has emerged on the definition of a good death (Steinhauser et al., 2000). Early studies show a remarkable degree of agreement concerning what components of clinical, psychological, financial, and social support are deemed important to patients and families at the end of life. Complexities arise around differences in ethnicity, gender, religion, spirituality and culture (Meffert, Becker, Körner, & Stößel, 2015). In 1969 Kubler-Ross published a book “On Death and Dying”, which identified patient choices and personal decisions relating to terminal care including place of end-of-life care and treatment. Kubler-Ross suggested that the dying and aged tend to be isolated, as they are reminders of the inevitability of life, but she argued more resources should be made available to ensure a more caring end of life at home which might ensure a good death (Kubler-Ross, 1969).

In 2000, Steinhauser et al. conducted a study into what constitutes a good death by collecting descriptions of areas of importance from patients and families experiencing end-of-life care. Six factors were identified: symptom
management, clear communication for decision-making, preparation for death, attention to family and close friends, reciprocity of care and concern for the wellbeing of others and recognition of the patient as an individual rather than a disease entity (Steinhauser et al., 2000; Steinhauser, Voils, Bosworth, & Tulsky, 2015). In the later study by Steinhauser, Voils, Bosworth and Tulsky (2015) a more comprehensive form of end-of-life care, including understanding and supporting the choices of care made by the patient and family, is identified as contributing to a good death.

Understanding a person’s end-of-life choices is particularly important when their ability to clearly verbalise or communicate their wishes has been adversely affected. The clinician/patient interaction is especially relevant in MND when clear communication is often extremely difficult due to the person’s disease progression. Consequently, to achieve a good death, clinicians need to engage in clear, honest and comprehensive communication in order to develop a shared understanding of the goals of care for the patient and their family. Therefore, clinicians must not only be compassionate but clinically informed and, ideally, experienced if they are to conduct such conversations effectively (McConigley et al., 2012; NICE, 2016). Determining when a person with MND and their family are ready to accept and receive such honest communication, however, adds further complexity.

1.6. Hospice Care and Specialised Palliative Care

Palliative care management for people with MND is focused on the symptoms, support and needs of the patient and their family. Ideally, this management would integrate an MDT approach where the benefits, burdens and limitations of all options would be discussed between all health professionals, the patient and their family (MNDAust, 2012). The integration of a centralised, rather than a delegated community-based system, MDT incorporating palliative care has been found to reduce hospital admissions and have a positive effect on the survival of people with MND (Rooney et al., 2015).
Specialist palliative care has become more accessible throughout Australia over the last 30 years and should be central to discussions concerning end-of-life communication. The following section describes the development and significance of hospice and specialised palliative care in Australia. The role of specialist palliative care in the delivery of treatment options and the importance of unambiguous communication is discussed. This historical overview of hospice and palliative care provides contextual background to the current study.

The history and significance of the concept of “hospice” care dates back to the 12th century, when religious orders offered a place of rest to those severely ill or dying (Palliative Care South Australia, 2015). The idea of a medical specialty focused on caring for the dying was developed after the Second World War by Dame Cicely Saunders, who established St Christopher’s Hospice for the terminally ill in London. She presented her idea in a lecture in 1963 at Yale University in the United States and her thinking laid the foundations for end-of-life care as an area of expertise, a medical speciality and the development of modern hospices. Dame Saunders recognised the many facets, depths and causes of pain and the need for psychosocial and psychological support for the patient and their family. In 1975, the term “palliative care” was introduced in Canada by the physician, surgeon and academic Balfour Mount, who was concerned the term hospice evoked places of impoverishment and hardship (Palliative Care South Australia, 2015).

In 1990, each state and territory in Australia formed the Australian Hospice and Palliative Care Association which was renamed Palliative Care Australia in 1998 (Palliative Care South Australia, 2015). Palliative Care Australia has provided recommendations for the standardisation of palliative care within Australia as well as assisting with end-of-life research and the development of quality standards. Historically, palliative and hospice care focused on cancer patients and most research focused on efficacy, service provision and cost (Palliative Care South Australia, 2015). More recently
Palliative care has come to be considered relevant to and appropriate for all people with a life-limiting, progressive disease (e.g., heart and lung disease, other neuro-degenerative diseases) (National Palliative Care Strategy, 2010).

Hospice care and palliative care are terms often used interchangeably. However, palliative care may be considered a specific system of patient care ideally introduced when a life-limiting disease is diagnosed, whereas hospice care could reasonably describe palliating care at the terminal stage of a disease (Connolly et al., 2015). Both community- and hospital-based specialist palliative care focus on the quality of a person’s end-of-life, their physical and emotional suffering and the appropriateness of ongoing treatments (Palliative Care Australia, 2005; Palliative Care Expert Group, 2010).

Hospice and palliative care in Australia are provided in various settings: acute, specialist, aged care facilities, medical/generalist and community. Funding for these services comes from a complex mix of state and federal monies plus donations and bequests (Community Affairs References Committee & Siewert, 2012). The cost of providing a community palliative care service is less than that provided in an acute care setting and is often the preferred place of treatment and death (Hodgson, 2010). Hospice and palliative care, whether as a community service or in an aged care setting, has been shown to decrease health care costs by reducing admissions, inappropriate diagnoses, re-admissions and length of hospital stay (Allen, O’Connor, Chapman, & Francis, 2008; Hodgson, 2010). Community-based palliative care improves care and symptom control in a preferred place of care and adds to provider clinician satisfaction (Hodgson, 2010; Palliative Care Australia, 2005).

The main components of palliative care services include symptom control, the management of disease and spiritual and psychological support for patients and their families in order to allow a good death and choice in where to die (Palliative Care Australia, 2005). Implicit within this description is the support and care of a patient’s family and significant others.
Early palliative care team involvement assists the development of trusted relationships between clinicians, patients and families by establishing good communication, understanding patient preferences and optimising care. Ideally early trusted relationships facilitate discussion and documentation of end-of-life treatment choices, known in the literature as advance health directives (AHD) (WA Health, 2011). Generally, there has been some attempt at a cure prior to the offer of such care but MND does not come with hope of cure, and it is, therefore, argued that palliation should commence at diagnosis (Aoun et al., 2012; Connolly et al., 2015; Motor Neurone Disease Association, 2012; Motor Neurone Disease Australia, 2010). In order to monitor disease progression and optimise the quality of life for people with MND and their family members, a care plan incorporating palliative care from the time of diagnosis is recommended to improve communication and support (Connolly et al., 2015; MNDA, 2012; MNDAustr, 2014). However, barriers to this approach exist, for both clinicians and people with MND.

Consideration if the broader approach to patient (and family) care within the healthcare system is relevant because it impacts service users’ and carers’ (and clinicians’) readiness to accept or propose palliative care. One popular approach to the timing and content of end-of-life communication is through the framework of a patient-centred model of care This is discussed in the following section.

1.7. Patient-Centered Care

A patient-centred model of care is incorporated into current medical teaching, but there is some ambiguity around how it is defined (Bertakis & Azari, 2011). Patient-centred care is generally understood to encapsulate a holistic approach within which the patient is an integral decision maker in care choices. The term was first introduced in the United Kingdom. It emphasised the importance of an individual’s involvement in their own medical care and
required explicit clarification for the patient of their disease prognosis and ongoing care (Balint, 1969). In 1995 patient-centered care was explored as a model of care more from the doctor's perspective (Schofield, 1995). Patient-centred care incorporates the idea of enhancing the doctor-patient relationship through a sharing of health issues whilst understanding the individual within their social and familial context. Incorporated within this model are health promotion and prevention, plus consideration of the doctor’s time and resource restrictions.

A systematic review undertaken more recently identified a failure to define patient-centred care within articles which either incorporated the term in the title or within the abstract (Scholl, Zill, Härter, & Dirmaier, 2014). From the articles that did offer a definition, Scholl et al. (2014) were able to categorise 15 dimensions to patient-centred care. Clinical communication was identified by Scholl et al. as an important element of patient-centred care. However, characteristics of the clinician, coordination and continuity of care and the psychosocial perspective are all aspects of the patient-centred care model (Scholl et al., 2014).

In Australia, the definition of patient-centred care includes understanding the patient within their social/familial context as well as being responsive to the preferences and principles of the individual. Incorporating informative communication, emotional and physical support and respect is the definitive holistic approach to patient-centered care in Australia (Australian Commission on Safety and Quality in Health Care, 2011). One of the recommendations from the Australian Commission into the delivery of care, is that research funding and its distribution acknowledge the importance of patient-centered care (Australian Commission on Safety and Quality in Health Care, 2011).

Similarly, the definition of patient-centred care in the United Kingdom encourages consulting with a patient to ascertain their preferences and incorporating shared control and management of their health issues (Farrell,
In the United States, an analysis into the determinants of patient centered-care found the more educated and healthier patients cared for by their family physician were more likely to have participated in patient-centered care (Bertakis & Azari, 2011). Bertakis and Azari (2011) suggest that patients with more serious illnesses are more encumbered by their medical needs and have less time to partake in patient-centered communication. Berkaki and Azari, however, found no correlation between increased patient satisfaction and patient-centred care.

Patient-centred care stresses clarity in communication between clinicians and those they are treating. The communication, whilst being informative, should uphold the dignity of the person affected by the health issue. The WHO promotes people-centred health by educating and supporting patients and their families to make informed decisions based on their health requirements, expectations and, importantly, preserving dignity (WHO, 2015a). However, this ideal health care model can only be delivered with correct training of care providers and a setting conducive to sensitive and dignified communication (Pringle, Johnston, & Buchanan, 2015).

The following section explains the development of evidence-based guidelines and identifies the inclusion of patient-centred care within these guidelines. The interpretation of how recommended evidence-based, patient-centred communication is delivered and understood is central to the research reported in this thesis.

1.8. Evidence-Based Guidelines

Following the initial work by Steinhauser et al. (2000) in defining a good death, researchers began to investigate what the best clinical practices were to achieve elements identified as important to patients and families at the end of life. Clinical guidelines have appeared, based primarily on clinical and expert
opinion, first for cancer patients and more recently for non-cancer and chronic illness end-of-life care (Steinhauser et al., 2000; Thomas & Free, 2006).

Evidence-based clinical guidelines provide up-to-date information and direct best clinical practice and recommendations for care in a particular field. Within the Australian clinical setting, NHMRC guidelines provide direction for clinical care and inform clinical decisions on a variety of disease areas (NHMRC, 2016). The NHMRC guidelines, as well as guidelines in other developed countries, are based on a thorough review of research evidence and current clinical consensus. Multidisciplinary panels incorporate experts within their field, educational institutions, consumer groups and professional bodies all of whom contribute to inform best practice.

In the United Kingdom the NICE, an independent public organisation, provides evidence-based recommendations to improve and guide health and social outcomes. The development of the guidelines is based on accepted international standards of research and assessment, undertaken by clinical experts, NICE committee members and stakeholders (NICE, 2014). The American College of Physicians’ development of clinical guidelines includes systematic reviews of the research evidence plus committee support in much the same way as the NICE and NHMRC guidelines (Qaseem, Snow, Owens, & Shekelle, 2010). The NHMRC and NICE clinical guidelines are commonly updated every 5-10 years (NHMRC, 2016; NICE, 2014). In the United States the clinical practice guidelines are withdrawn or considered invalid if not updated within five years after publication (Qaseem et al., 2010).

However, there are shortcomings with evidence-based guidelines, not least their implementation into practice (Greenhalgh, Howick, & Maskrey, 2014). As an example, Greenhalgh, Howick and Maskrey (2014) state that despite the lack of evidence for arthroscopy in all but those who medically need it, arthroscopies continue at an extremely high rate in a much broader population. A further disadvantage is that the sheer volume of evidence may have become
too great and time consuming to be absorbed by already busy clinicians. In addition, clinicians often fear that clinical expertise founded on the accumulation of years of medical practise could be overshadowed by the fear of litigation if specific evidence-based pathways are not followed (Glasziou, Moynihan, Richards, & Godlee, 2013). Nonetheless, clinical guidelines provide a way to consider the issues in providing effective care.

In MND this includes communicating symptom control options such as NIV initiation and possible removal and end-of-life options (Andersen et al., 2012; NICE, 2016). The recommendations for timing, content and delivery of communication within the most recent MND guidelines (NICE, 2016) incorporate patient-centred care in their ethos. The combination of patient-centred care, evidence-based care and a good death is represented in the most recent MND recommendations and guidelines and provide the framework for data analysis for the current study.

1.9. Rationale for the Research

Over the past 10 years throughout Australia there has been a rise in the number of deaths from MND, although the cause of the increase is not known (Motor Neurone Disease Australia, 2013). Global statistical data on MND/ALS is not uniformly collected and the statistics within this thesis are sourced primarily from Australian and United Kingdom publications.

In MND, early discussion of end-of-life symptom relieving choices has been found to be particularly pertinent, as from diagnosis the person with MND and their family need to understand there is no cure and that the progression of disease can be rapid and unpredictable. Respiratory failure is the most debilitating and critical symptom in MND and is the most common cause of death. Non-invasive ventilation (NIV) is offered for respiratory symptom relief in people with MND but has its limitations. Therefore, communicating symptom
control alternatives and their limitations is recommended to occur soon after diagnosis and certainly by the onset of respiratory symptoms.

Current palliative care guidelines recommend that MDT including clinical specialists, physiotherapists, dietitians and social workers are involved with people with a life limiting illness to ensure that all aspects of patient care are covered (Bede et al., 2011; Rooney et al., 2015). A systematic review by Oishi and Murtagh (2014), however, highlighted uncertainty about responsibility for coordinated end-of-life care in non-cancer patients. Oishi and Murtagh found that some of the clinician roles and responsibilities were unclear both to the patients and caregivers and in some cases to the clinicians themselves (Oishi & Murtagh, 2014). Arguably, this uncertainty may affect the nature of the timing and content of any discussions relating to symptom control options such as PEG and NIV.

Established patient-centred, evidence-based medicine and palliative care clinical guidelines have all recommended that communication for people with MND surrounding the initiation of NIV should occur early and include discussions relating to gastrostomy tube insertion into the stomach (NICE, 2016). It is important to understand that the impact of these interventions on people with MND and their caregivers can be significant, and the quality of life gain may be small (ProGas Study Group, 2015). It is suggested by others that these discussions should clearly inform people with MND and their families/caregivers of their options, so they can make informed choices and share these with all involved. This will in turn minimise the influence of the healthcare professionals’ personal attitude toward initiation and potential later discontinuation of treatment (Motor Neurone Disease Australia, 2014; Ruffell et al., 2013). Unambiguous discussions about preferences relating to nutrition and hydration via gastrostomy insertion are recommended to occur well in advance of respiratory symptoms, but there is uncertainty in relation to the actual content and timing of these discussions within the literature (Connolly et al., 2015; Oliver, Campbell, Sykes, Tallon, & Edwards, 2011). Further, it is not clear
whether intervention to maintain hydration and nutrition is discussed as a life-prolonging measure despite the minimal evidence (Connolly et al., 2015), nor whether the benefits and burdens of PEG insertion are included in the content of effective communication. It is acknowledged that predicting the exact course of an individual’s disease can be difficult; the call to avoid ambiguity, therefore, refers only to the content of communication relating to symptom control options, AHD and end-of-life choices delivered by health professionals.

Despite the existence of specific MND guidelines recommending early and explicit communication about end-of-life care choices, there is a perception among health care professionals that for some people with MND and their families, symptom control and end-of-life options are neither well communicated, understood, nor documented (Baxter et al., 2013; Faull, Phelps, Regen, Oliver, et al., 2014). Unclear communication has the potential to lead to confusion and emotional uncertainty for families and their health care professionals, and to result in insufficient information to make and document informed care choices. Of concern, and explored within this research, is the perceived difficulty in communicating and comprehending vital information to people with MND and their families, which acts as a barrier to informed symptom control and end-of-life choices. The research reported in this thesis investigated overall accountability for communicating information to people with MND and their families to uncover how this may have affected the clarity of their understanding. It is these issues of communication surrounding respiratory failure and symptom control (especially NIV) in people with MND that are the focus of this study.

More specifically, the objective of this research is to determine whether patient-centred care, evidence-based medicine and guideline recommendations relating to symptom control in respiratory failure and end-of-life choices were being adequately practised by clinicians working with people and families affected by MND. Of particular interest is the timing and content of
communication, how effectively and comprehensively clinicians felt they delivered the required information and consequently how they perceived people with MND and their families understood the intended meaning of these communications. In addition, the study examines whether clinicians involved with people with MND were concerned that, as clinicians, they may not fully understand their patients’ wishes for palliation and end-of-life choices.

The first part of this chapter has explained the background to the current study. Next, the focus of the research will be presented, followed by an overview of the subsequent chapters.

1.10. The Research Focus

The focus of the research presented in this thesis is clinical communication about respiratory failure and symptom control at the end of life in people with MND. Specifically, the research questions generated, and data collection conducted for this thesis were designed to explore what information clinicians communicate regarding NIV and end-of-life issues and what the experience and understanding of that information was for the MND caregiver after the death of the patient. The research sought to ascertain whether MND communication content explaining treatment options conforms to the available recommended guidelines (National Institute for Care and Health Excellence, 2016; National Institute for Health and Clinical Excellence, 2010). The aims of the research reported in this thesis are described below:

1. To describe clinician communication surrounding NIV including end-of-life choices.
2. To understand how clinicians check the level of understanding among patients and family members of the benefits, burdens and potential of NIV withdrawal prior to NIV commencement.
3. To understand the impact of clinician communication about the benefits, burdens and limitations of NIV on family members’ choices in relation to NIV introduction.
4. To understand what timing, content and style of communication most effectively helps the families of people with MND to make decisions about NIV initiation and withdrawal.

5. To compare the content, process and outcomes of NIV communication as described by clinicians and families of people with MND within this study against the currently available recommendations relating to commencement and withdrawal of NIV and related end-of-life care options.

The following questions were answered where possible by the literature review and formed the foundation for the questions asked of the participants.

1.0.1. Research questions

1. In considering the available literature and guidelines, what are the most salient components of best practice relating to the communication about NIV in MND prior to NIV initiation and withdrawal?

2. What is the evidence that the best practice communication content (benefits and burdens of NIV, continued disease progression, explanation and offer of a PEG, end-of-life discussions and understanding the potential for NIV withdrawal prior to the commencement of NIV) is incorporated into the actual communication by clinicians to people with MND and families at the time of offering NIV?

3. What do MND families/significant others recall about the communication (benefits and burdens, continued disease progression, PEG insertion, potential NIV withdrawal and end-of-life discussions) provided by clinicians at the time of respiratory failure and the offer of NIV?
4. To what degree do MND families/significant others feel that the NIV communication provided by the clinicians at the time of the offer of NIV prepared them for and helped them to understand the implications of potential NIV withdrawal resulting in the death of their family member?

5. What if any unmet needs for information, support, palliative care, or other services do the clinicians and the families/significant others (or patients themselves) with experience of MND identify?

6. Following the literature review and clinician and family/significant other interviews regarding NIV communication, what recommendations can be identified that would improve end-of-life communication and process for people with MND, families and clinicians?

Communication content surrounding the initiation of NIV and encompassing the potential of NIV withdrawal is sparsely covered in the literature and is an area where further research has been recommended (NICE, 2010). The most recent NICE MND Assessment and Management Guidelines (NICE, 2016) recommend including discussions relating to “advantages and disadvantages” of NIV when discussing its initiation. The guidelines recommend that those diagnosed with MND have adequate information relating to what NIV can offer, that it can be stopped at any time and that the discussions occur sensitively and at an appropriate time (NICE, 2016).

This research examines the content of communication between clinicians and people diagnosed with MND and their families which, if based on the evidence-based clinical guidelines, should incorporate symptom control and end-of-life issues. The study identifies two distinct participant groups: those that provide the communication (the clinicians) and those that receive the communication (the people with MND and their families). The research questioned the clinicians involved as to when they imparted the relevant information and whether they felt they had done so with enough clarity to allow
the person with MND and their family to make informed choices. In particular, clinicians’ views about understandings held by people with MND and their family members around the possibility of NIV withdrawal followed by death when NIV was deemed to be of no further benefit were explored. Whether and how the implications of commencing NIV were understood by the bereaved families of a family member with MND were also investigated. In addition, the research explored whether the families/significant others involved with the person with MND felt able to ask all the questions they wanted to ask, whether they felt adequately informed and whether all the participants could offer suggestions to improve communication between clinicians and people with MND and their families.

This study provides a much-needed contribution to the literature on the experiences of caregivers and families who have had a family member die from MND and had experience with, or refusal of, NIV as a palliation measure. Family and clinician experiences with the withdrawal of NIV are included. The study offers valuable insights into clinicians’ experiences of communicating with each other within an MDT, and with people with MND and their families. The thesis explores perceived barriers to clear communication both between clinicians as well as between clinicians and people with MND and their families. The variation of care available to rural and metropolitan people living with MND is also highlighted and discussed.

1.11. The Structure of the Thesis

Much of the existing literature on MND and end-of-life contains terms that may be unfamiliar to those outside the clinical/practice domain concerned with supporting and caring for those with a life-limiting condition. Definitions are, therefore, provided within the glossary of terms (page XIX) to help the reader navigate the remainder of this thesis.
The thesis comprises six chapters. In Chapter 1 the context and broad rationale for the study have been indicated, and the research focus, aims and questions have been presented.

Chapter 2 contains a comprehensive review of relevant literature on the topic of clinical communication relative to MND. This review identifies the precise gap in knowledge that the research questions in this thesis have been designed to address. A fully replicable literature search and selection strategy is also detailed.

The study methodology and methods are the subject of Chapter 3. The compatibility of interpretive description (methodology) and symbolic interactionism (theoretical framework) are introduced. First, the purpose, aims and significance of this investigation are restated. Next, a discussion of possible approaches to this study and justification for the one chosen is provided. The steps followed to conduct the research are then presented. In this chapter, the involvement of a stakeholder advisory group formed to confirm and refine the research questions and participant interview questions is reported, as are the ethical considerations and measures used for trustworthiness.

Chapter 4 reports the findings of the study, which include clinicians’ reasons for not always following recommendations, and barriers to communication between clinicians and families/caregivers of people with MND from the perspective of both cohorts. Illustrative clinician and family participant quotations from interview transcripts are included where relevant. Misunderstandings around definitions of some words used in communicating, as reported by many of the participants, are discussed.

Chapter 5 includes the discussion of the study findings. The theoretical perspective that guided this study is also considered. This chapter also compares existing knowledge about MND communication to the new
information this study reveals, and incorporates any relevant literature published since the completion of the literature review (Chapter 2 and Appendix A).

In conclusion, Chapter 6 brings the research and the thesis to a close with recommendations for further research and suggestions for clinical practice. Limitations identified within this study are also discussed. Chapter 6 concludes with a reflective account of the candidate’s experience of conducting this study (within the Epilogue). Lessons learned are identified for use in future research endeavours.
Chapter 2

Literature Review

2.1. Section 1: Chapter Overview

In this chapter, the argument for the current study is further developed. Chapter 1 provided the context for this research and a broad overview of the current study. This chapter is presented in five sections, beginning with an overview of the context of end-of-life communication in cases of MND and identifying the need for a focused review of the literature on communication between healthcare professionals and those with MND and their family members in Section 1. Section 2 provides a summary of the steps taken in a systematic (black) literature search, retrieval and selection process, conducted to answer the question “What is known about communication between caregivers and families about NIV in MND prior to NIV initiation and withdrawal?” Section 3 reports and discusses the results of this process in the form of narrative critical review. The report of an environmental scan conducted in which clinical guidelines pertinent to the phenomenon of interest were retrieved and summarised is provided in Section 4. This is followed by a statement about what is and is not known and discussed in relation to the introduction, use and withdrawal of NIV in MND. The need for a study to address current knowledge gap is then summarised, and the research questions are listed in Section 5.

2.1.1. Epistemological and ontological position

To understand the nature and effect of communication in relation to end-of-life treatment choices delivered by clinicians to people with MND and their families, both the study reported in this thesis and the review of literature are positioned within a relativist ontology and a subjective epistemological position. These two decisions reflect an understanding that delivery of vocal communication depends upon the clarity and specificity of the language of the
individual delivering it, and that how the “receiving” individual understands the delivered communication depends on a range of influencing individual factors (Raskin, 2008).

2.1.2. The context of end-of-life communication in MND

The importance of clear communication in which the benefits, burdens and limitations of NIV are explained to people with MND and their families has been recommended by many in the field of MND care (Baxter et al., 2013; NICE, 2010, 2016). Explicit communication enables people with MND to make informed treatment choices. Studies show the use of NIV in people with MND with good bulbar function can prolong a person’s life by 7-18 months (Bourke et al., 2006; Jenkins, Hiollinger, & McDermott, 2014; Radunovic et al., 2013). Nevertheless, with or without the use of NIV, complications can arise for people with MND such as an inability to clear oropharyngeal secretions (respiratory secretions which can become copious or viscous in MND) leading to chest infections, or increased difficulty breathing, which in turn commonly lead to death by respiratory failure (Rafiq et al., 2012). Additional considerations, such as clarifying and documenting the patients’ and families’ wishes for end-of-life treatments, such as incorporating AHDs into advance care planning, are recommended for clear direction to clinicians. Patient comfort, caregiver support and the value the individual places on a potentially longer life in relation to the possible trade-off of poorer quality of life are recommended as essential components of conversations. This communication must be undertaken when proposing potentially life extending technologies such as NIV (Lerum, Solbrække, & Frich, 2016; NICE, 2016). Thus, the inclusion of end-of-life issues and documented patient wishes in communication surrounding NIV is vital for ensuring a clear and shared understanding between all concerned.

Recent end-of-life and clinical palliative care literature suggests that NIV is being increasingly employed as a palliating intervention for dyspnoea, not only in MND, but also in other progressive respiratory diseases such as lung cancer and emphysema (Gifford, 2014). NIV can also be used as a palliating
measure in illnesses with a long trajectory such as chronic obstructive pulmonary disease (COPD) (Gale, Jawad, Dave, & Turner, 2015). Although NIV has been recommended for palliating respiratory symptom relief in MND, it is unclear whether clinicians who specialise in all progressive respiratory conditions are communicating clear and precise information about its initiation and potential withdrawal to patients and their caregivers. Understanding what this communication involves, at what stage the discussions occur, how well the necessary information is being delivered and comprehended by people with MND and their families is, therefore, becoming increasingly relevant to other diseases (Gifford, 2014; Quill & Quill, 2014). The benefits of relieving respiratory failure associated with MND and the burdens of wearing an NIV mask (associated feelings of claustrophobia and discomfort) appear similar in people with MND and those with chronic respiratory disease. People with MND and those with COPD experience increased dependence on both NIV and caregiver support (Ando et al., 2014; Gale et al., 2015).

2.1.3. The timing of palliative care involvement and triggers for NIV and end-of-life discussions.

The WHO recommends a palliative approach at the end of life, to ensure quality of care for all life-limiting illnesses (Sawatzky et al., 2016). The WHO (1998) describes palliative care as:

…an approach that improves quality of life of patients and their families facing the problem associated with life-threatening illness, through the prevention of suffering by early identification and impeccable assessment and treatment of pain and other problems, physical, psychological and spiritual.

The WHO definition does not specify an individual’s anticipated life duration for referral to a specialist palliative care organisation. In MND, not knowing the best time for referral of those living with the disease to specialist palliative care can make the transfer difficult (Dharmadasa, Matamala, & Kiernan, 2016).
Whilst MND is generally associated with a life expectancy of approximately 2-5 years, some people live with the disease for more than 10 years, which potentially changes the prognostic category to chronic (rather than terminal) disease (Lerum et al., 2015). The MND recommendations referenced within this study discuss referral to specialist palliative care at diagnosis. However, consideration of a palliative care approach which incorporates a GP and organised community care as opposed to early incorporation of a specialist palliative care service may be appropriate. This may be particularly relevant when considering life-limiting chronic diseases, which in the instance of MND may describe the course of some variants of the disease (Burgess, Braunack Mayer, Crawford, & Beilby, 2013). Burgess, Braunack, Crawford and Beilby (2013) suggest a referral to specialist palliative care is based on patient needs as well as their symptoms rather than diagnosis alone.

The Standards for Providing Palliative Care in Australia (Palliative Care Australia, 2005), which are generally accepted in principle throughout the world, include acknowledging the unique holistic requirements of the patient and their family. Ongoing assessment and communication are particularly important. According to these standards, palliative care should be available to all but is difficult for those with a life-limiting disease irrespective of diagnosis, based on clinical need. The availability of coordinated assisted care to lessen the caregiver burden, provide comfort and meet individual requirements for end-of-life care should be considered (Palliative Care Australia, 2005). In MND there is no potential of a cure but a prolonged physical and mental decline. The timing of referral to palliative care for people with MND has been addressed in the general palliative care literature and current recommendations, although there is confusion as to the optimal timing (Dharmadasa et al., 2016). Whether specialist palliative care is introduced from diagnosis or when MND symptoms worsen and symptom management technology (such as NIV) is needed is dependent on when the patient and family are judged ready to receive such information. Leaving referral too late can lead to hospital admissions and crisis
situations, should clear information not have been forthcoming (Connolly et al., 2015).

Opinions about when specialist palliative and end-of-life care are introduced abound with little consensus, due in part due to the wide variation in the trajectory of diseases (e.g., cancer versus non-cancer). However, most experts agree that palliative care should be started soon after diagnosis and thus, end-of-life communication needs to start early (NICE, 2016; Oliver et al., 2006). A study in the United Kingdom investigated whether the Preferred Priorities for Care document developed as a communication tool by the End of Life Care Strategy (Lancashire and South Cumbria Cancer Network, 2007) had any impact on the actual end-of-life care offered to people with MND. This plan was developed as a patient-held advance care planning tool principally to assist patients to make their preferred place of death clear. However, this document was not found to improve patient end-of-life care and many health care professionals seemed unaware of its existence (Preston, Fineberg, Callagher, & Mitchell, 2011). Another study examined clinical support tools. The Lothian National Health Service in the United Kingdom developed what it called the ‘Supportive and Palliative Care Indicator Tool’, which suggests general and clinical indicators to identify those in need of immediate palliative care (Boyd & Murray, 2010). This indicator tool does not recommend commencement of palliative care when the neurological diagnosis is given. It suggests that palliative care services should become involved when symptom deterioration such as breathlessness, swallowing difficulties or communication issues occur.

Other researchers have investigated the timing of discussions between clinicians and patients. In an article that suggests early open and honest communication of end-of-life issues in ALS, Connolly, Galvin and Hardiman (2015) examined published scientific literature to identify end-of-life issues pertaining to the management of MND. They suggested that early symptom control communication may enable patients and families time for informed discussion and to plan and prepare, and that these discussions may prevent
unwanted symptom control interventions such as NIV, gastrostomy tubes and the potential of invasive ventilation. Other studies tend to support these conclusions (Dharmadasa et al., 2016). Thus, honest communication surrounding end-of-life options for care is recommended from initiation of symptom control interventions (NIV/PEG), particularly if such discussions have not previously occurred, and especially as MND has an unpredictable disease course.

The introduction of palliative care soon after (or at) diagnosis is discussed by Connolly et al., (2015) and is encouraged for alleviating symptoms, improving communication, providing support to patients and families and enhancing quality of life in a terminal disease. Some health professionals may lack experience and confidence to manage the complex and sensitive discussions associated with ongoing care in a relatively rare disease such as MND, and some may perceive a sense of hopelessness should end-of-life care be discussed too soon (MNDA, 2012). As the disease progresses and symptoms of respiratory distress develop, the person with MND should be referred by their primary health care provider to a respiratory physician or neurologist for detailed specialist management and discussions regarding appropriate forms of psychological support, ventilation options, palliating alternatives and guidance by an MDT (NICE, 2016).

An MNDAust submission made to the Australian Senate Committee inquiry into palliative care in Australia advised that access to MND specific information for patients and their families is required to maintain a palliative care approach and to support clear communication about end-of-life issues (MNDAust, 2012). The submission also recommended referral to palliative care services as soon as possible after diagnosis to optimise quality of life and professional support to inform and assist with advance care planning. MNDAust recommends that a palliative care approach to MND is initiated on diagnosis, with discussions incorporating end-of-life decisions commencing when the person with MND is ready (MNDAust, 2010). Although the MNDA agrees that
the timing of such discussions is uncertain and ought to be individualised, triggers for commencing such discussions are suggested, with one indicator/prompt being dyspnoea (difficulty with breathing) and offering NIV (NICE, 2010).

A survey amongst the members of the European ALS Study Group on standards of palliative care found discrepancies in care offered amongst palliative care providers, such as the timing of the introduction of palliative care and MDTs, and the availability and maintenance of NIV (Borasio, Shaw, Ludolph, Sales Luis, & Silani, 2001). Discussions relating to terminal care were reported to occur at the request of the patient or when the patient was close to death, highlighting the possibility that the patient and family may have worried over symptoms such as choking in respiratory failure for the duration of the illness. The authors recommended development of a set of common standards for the management of ALS (Borasio, Shaw, et al., 2001).

The European Federation of Neurological Societies (EFNS) have recently revised guidelines for the care of people with ALS and made recommendations for further research such as studies into the quality of life relating to NIV and PEG use (Andersen et al., 2012). Recommendations for care within the guidelines include input from a palliative care team early after diagnosis, initiation of end-of-life discussions when an individual is ready and at the time of provision of potentially life-sustaining interventions. Respiratory distress symptoms ought to prompt end-of-life discussions and trigger the AHDs. More recently, a paper presented at the 24th Meeting of the EFNS (European Federation of Neurological Societies and European Association of Palliative Care, 2015) reiterated the need for communication regarding end-of-life issues prior to any impairment of communication abilities. A review of the current international palliative care guidelines indicates that clear communication between medical specialties and people with MND is required (NICE, 2016). The recommendations include the need for international palliative
care MND/ALS guidelines incorporating respiratory failure options (Bede et al., 2011; NICE, 2016).

Finally, the NICE clinical guidelines (2010) for the use of non-invasive ventilation in the management of MND recommend that respiratory physicians should be part of the MDT and this is reiterated in the NICE (2016) MND Assessment and Management Guideline. Eng (2006) suggests a general acknowledgement amongst clinicians specialising in MND that involvement of a palliative care service at the stage of respiratory decline, if not before, is important. Hardiman (2011) and Borasio, Voltz and Miller (2001) agree a palliative care approach should commence from the onset of symptoms and initial diagnosis of MND, and recommend discussions involving the patient, significant others and the medical team to optimise appropriate ongoing care, understanding and support. Borasio et al. (2001) also suggest discussion and information for families and people with MND about the terminal stage should commence, at the latest, with the onset of symptoms of dyspnoea.

Whilst there is agreement in the literature for early palliative care involvement and the potential usefulness for AHDs (Borasio, Voltz, et al., 2001; Hardiman, Bede, Oliver, & Obrannagain, 2011), the optimal time for involvement of the specialist palliative care team from the perspective of the patient or family remains unclear. The concept of improved communication surrounding the implementation of NIV and ongoing disease progression is acknowledged as an area requiring further research (NICE, 2010).

In summary, people with MND can present with different palliative care issues compared to cancer patients. Clear communication surrounding the introduction of palliative care and symptom control delivered at a pace directed by the person with MND and their family is an important element in quality end-of-life care and is explored within this thesis.
2.1.4. Patient and family perspectives on end-of-life communication

The literature published over the past 16 years has been reviewed for this chapter. It identifies clear communication as a priority of end-of-life patients and their families. Understanding the disease process is essential for them to make appropriate and timely symptom control and end-of-life decisions. People with MND and their caregivers can be assisted to make clear decisions relating to their ongoing care if they know the facts of their disease and if the support is available to ensure their wishes are upheld. In a retrospective study of 42 people with MND, Anderson, Kuru, Munroe, and Sirdofsky (2007) agree that end-of-life discussions should commence from the time of the person’s MND diagnosis and continue over the duration of the disease especially as patient preferences often change over time. Anderson et al. found that people with MND are generally agreeable to participate in end-of-life discussions particularly relating to invasive measures soon after diagnosis, but also found that this does not necessarily occur. Despite guidelines for clinicians recommending honest discussions relating to interventions for symptom control prior to the need for those interventions (Motor Neurone Disease Australia, 2014; National Institute for Health and Clinical Excellence, 2010c), one study has identified that these discussions may be experienced as “pressure” by some people with MND (Greenaway et al., 2015).

In a systematic review to find the most important aspects of end-of-life care identified by patients and their families in an inpatient setting within Australian hospitals, Virdun, Luckett, Davidson and Phillips (2015) identified clear communication and the ability to contribute to decision-making as priorities in end-of-life care. From the patient’s perspective, multiple studies have suggested that most would welcome more honest and up-front discussions concerning prognosis, benefits and burdens of various treatment options and their effect on patient and family quality of life (Astrow et al., 2008; Virdun, Luckett, Davidson, & Phillips, 2015). Several studies show that people with MND would like the opportunity to discuss their treatment options and would like
as much information as is available, believing that doctors should initiate such conversations (Anderson, Kuru, Munroe, & Sirdofsky, 2007; Blackhall, 2012).

A comparative study of decision-making in patients with advanced cancer (n= 60 patients) and MND/ALS (n= 32 patients), found communicating end-of-life issues was significantly less well discussed in the advanced cancer patients than in MND/ALS patients (Astrow et al., 2008). Astrow et al. (2008) suggests this may be due to the variable prognosis in cancer, with periods of remission and the potential of a cure. Astrow et al. propose that as people with MND have fewer treatment options and a more predictable prognostic pathway than cancer patients, the discussions relating to the end of life occur earlier during the disease. Astrow et al. (2008) reports a lack of evidence on whether a patient’s diagnosis influences end-of-life discussions. Reflecting the 1999 American Academy of Neurology standards which recommend discussions well in advance of the terminal stage, Astrow et al. (2008) encourage further research into the relationship between diagnosis and the impact of end-of-life communication. This is supported by others in the field of MND research (Miller et al., 2009; Miller et al., 1999).

Communication regarding ongoing support and potential complications are acknowledged as vital for all concerned, and advance care planning should be continually revisited along the disease continuum (Anderson et al., 2007; Oliver & Faull, 2013). Hardiman (2011) and Doyle et al. (2005) stress the importance of keeping people with MND informed about their disease progression and reassuring them that appropriate care and medications will be available to ensure a comfortable death. When NIV is suggested by the clinician either for a trial or for ongoing use, it is recommended that the benefits and limitations of NIV are fully discussed with the patient and significant others (NICE, 2016).

2.1.5. Content of end-of-life communication in MND

A U.K. telephone audit of specialist palliative care doctors caring for people with MND in the United Kingdom (Oliver et al., 2011) found the majority
of palliative care specialists interviewed were only involved at the terminal stage of the patient’s care, by which stage, discussions relating to interventions which may have an influence on patient symptoms and quality of life should have occurred. However, this audit found wide variation in the understanding of specialist roles and low levels of referrals to palliative care specialists by other doctors, resulting in some patients not being offered interventions to improve their quality of life (Oliver et al., 2011). Nearly 16 years ago a report by the Quality Standards Subcommittee of the American Academy of Neurology (Miller et al., 1999) recommended communicating patient information on respiratory care in MND/ALS to ensure appropriate decision-making. The committee recommended that such communication occur well in advance of declining respiratory function to enable patient understanding and avoid inappropriate emergency care (Miller et al., 2009; Miller et al., 1999). Using a combination of focus groups and interviews with clinicians having some experience with palliative care, McConigley et al. (2014) explored the complex communication requirements provided to people with MND and their families. Acknowledging the often disjointed care for people with MND in Australia and the unpredictable speed at which MND can progress, the results from this study showed the need for improved clinician communication, and education for clinicians providing care (McConigley et al., 2012; McConigley et al., 2014).

The EFNS recommends that the diagnosis and prognosis of MND/ALS could be communicated to patients and families, and that this as a skill a clinician needs to develop. The EFNS recommends allowing adequate time for a clinician who knows the patient and who has ascertained the patient’s level of knowledge to provide information. The recommendations also suggest providing written information, or a support organisation website relating to MND, with the choices of symptom control and reassurance of ongoing support. The EFNS highlights the importance of effective communication between patients, caregivers and all members of the MDTs, yet patients and families are not always satisfied with the communication they receive (Andersen et al., 2012).
An American study completed in 2004 found age and level of education had a significant influence on whether a patient wished to actively participate in their healthcare decisions; those who were older and more highly educated expressed a greater desire to participate (Levinson, Kao, Kuby, & Thisted, 2005). Clinician perceptions of patient preferred end-of-life care preferences and the extent of honest communication can be unrepresentative, as indicated by recent research from Taiwan (Huang et al., 2015). Huang et al. (2015) collected data from 314 terminally ill patients (diseases unspecified) and 177 physicians involved with their end-of-life care via a questionnaire. Their questions pertained to honesty with patients about prognosis and treatment preferences. Whereas most of the study patients preferred to be well informed (94.3%), only 80% of the physicians believed that their patients wanted this. The research indicated a discrepancy between 90% of the patients preferring supportive care only at the terminal stage, compared to 15.8% of the physicians who felt this was their patient’s preference, and 33% who thought active treatment was their patient’s preferred option.

Similarly, a German study found the majority of terminally ill patients wanted and expected clear communication from their health providers about their treatment despite the authors acknowledging that death and dying are not widely discussed in German society (Meffert et al., 2015). Blackhall (2012), in a review of palliative care and ALS/MND literature, suggests, as many palliative care experts do, that usual care and specialist physicians who have not had much exposure to palliative care education or training may avoid end-of-life discussions with those with MND. Blackhall (2012) suggests this may be because they fear it could lead to a sense of hopelessness, and often wait until the patient initiates the subject. This hesitation to discuss end-of-life issues by clinicians and their belief that such discussions should be initiated by the patient is singled out as a particular problem in end-of-life care medicine by Lemoignan and Ells (2010), who recommended further research into this phenomenon.
2.1.6. Recommended communication and frontotemporal dementia

The most recent guidelines relating to MND assessment and management, published in February 2016, recommend information about prognosis and management of symptoms is provided at diagnosis and delivered by a highly trained and experienced specialist such as a neurologist (NICE, 2016). Health professionals are urged to ask patients how much information they wish to have explained at diagnosis and whether they wish their families to be involved. In relation to PEG insertion, which is recommended prior to the introduction of NIV, it is suggested that advice and support is offered either at the time of diagnosis or when any concerns about swallowing or weight loss emerge. The NICE (2016) recommendations also suggest that prior to any decision on PEG insertion, assessment should be made of the patient’s ability to give consent and whether frontotemporal dementia (FTD) has been diagnosed.

Oliver et al. (2011) have argued that the impact of cognitive changes on communication, decision-making, and understanding should be recognised by health professionals to avoid inappropriately implementing symptom control systems such as NIV and gastrostomy tubes. Frontotemporal dementia (behavioural or language disorder) has more recently been recognised as an overlapping disease component of MND, with clinical implications for patient management (Phukan, 2007; Phukan et al., 2012). Phukan et al. (2012) suggest 14% of people diagnosed with MND/ALS may present with dementia at diagnosis, with more than 40% of patients experiencing cognitive changes but without evidence of dementia. Cognitive changes, which can be subtle, have potential implications for the capability of a patient to either understand or have the ability to communicate adequately with clinicians, particularly in relation to treatment options (Phukan et al., 2012). The process and ability to assess for the presence of cognitive changes in people with MND was considered by the candidate to be a substantial area of research so was not included in this study.
How FTD impacts on families was touched upon by the clinician participants and considered by some of the family participants in this study. This is discussed further in Chapter 4.

2.2. Caregiver and Patient End-of-Life Concerns

2.2.1. NIV communication from initiation to withdrawal incorporating advance health directives and multidisciplinary teams.

There is little in the general MND literature covering what specific information should be discussed surrounding initiation, non-initiation or withdrawal of NIV, although several studies suggest detailed communication should occur (Faull, Rowe-Haynes, et al., 2014; Lemoignan & Ells, 2010; NICE, 2010; Oliver & Faull, 2013; Preston et al., 2011).

The NICE (2016) Assessment and Management Guidelines for MND recommend that the person with MND, their family, the respiratory service/physician and those incorporated into the MDT, should draw up a detailed plan which includes regular opportunities to discuss withdrawing NIV (NICE, 2016). Anderson et al. (2007) suggest that signs of respiratory distress should prompt discussion and documentation of patient preferred treatment alternatives including options for the terminal phase.

The American Academy of Neurology (2012) recommend that all people diagnosed with ALS/MND should have documented discussions relating to NIV options with their clinician annually, and any reasons for not having such discussions should also be clearly documented (American Academy of Neurology, 2012). Information relating to NIV commencement remains unchanged from the NICE (2010) guidelines. It is suggested that discussions about NIV be “appropriate to the stage of a person’s illness” and delivered in a sensitive manner (NICE 2016: Recommendation1.14.2, page 23). The latest version of these guidelines recommends the patient should be reassured that NIV can be stopped at any time and given many opportunities to discuss the
option of withdrawal. It is not made clear, though, who should initiate these discussions or the content of the discussions (NICE, 2016).

Preston, Fineberg, Callagher and Mitchell (2011) interviewed 11 bereaved family members/MND caregivers and found that despite patient preferences of care being documented and read by family members, there was an apparent lack of awareness among health professionals of the care priorities for people with MND. Early effective communication was acknowledged to enable advance care planning, offering peace of mind to people with MND and their family members. However, the content of the communication between clinicians and people with MND to enable treatment choice was not reported (Preston et al., 2011).

Oliver and Faull (2013) recommend ongoing discussion with the patient and family from the commencement of NIV, offering support and covering all aspects of care including emergency care and advance health care planning. Oliver and Faull note the lack of published literature regarding the deterioration of patients using NIV and associated communication. For example, the understanding of deteriorating symptoms and medications required to relieve them, the possible medication side effects of increased sedation, and the implications of NIV withdrawal are rarely addressed. This lack of evidence inevitably makes conversation about these issues with patients and significant others difficult (Oliver & Faull, 2013). However, despite considerable agreement on the need for discussions to include withdrawal of NIV as part of usual care provision and some suggested trigger points in the patient’s disease trajectory for honest end-of-life discussions, in practice, it remains unclear as to who has or should have these discussions, their content, the impact these discussions have on the patient, significant others and clinicians at the terminal stage of the disease, or when patients and families feel these discussions should occur.

2.2.2. Triggers for communication

Based on a consensus of expert clinical opinion, triggers for initiating clinician and patient discussions relating to MND end-of-life discussions are
recommended in the NICE (2010) NIV guidelines and re-iterated by Mitsumoto and Rabkin (2007), Rafiq et al. (2012) and NICE (2016) MND Assessment and Management Guidelines. These triggers include, for example, pain requiring high dosages of analgesia, dyspnoea or symptoms of hypoventilation when NIV would be offered, or when the patient or family requests information relating to end-of-life care (Mitsumoto & Rabkin, 2007; NICE, 2010; Rafiq et al., 2012).

Another discussion in the MND literature is whether NIV initiation/respiratory failure would be an appropriate time for explicit end-of-life discussions, however, the input of the person living with MND or the views of their families has been minimal (Baxter et al., 2013; Eng, 2006; National Institute for Health and Clinical Excellence, 2010, 2016).

2.2.3. Communicating the benefits and burdens of NIV

Experiences with NIV for people with MND appear to be as varied as the communication that surrounds the intervention. Lemoignan and Ells (2010) interviewed nine people with MND, all of whom were suffering adverse respiratory symptoms, regarding their decisions to use NIV. Those interviewed felt they had a poor understanding of how respiratory failure, NIV technology, and their own disease progression could affect their quality of life. Fear relating to the possibility of choking, or fear of being a burden to their family members were found to be factors in a patient’s decision-making process relating to NIV in the study by Lemoignan and Ells (2010). Their study concluded that the consideration of when to discuss NIV and provide information should be guided by the person with MND and significant others, suggesting further research regarding decisions surrounding NIV is necessary.

In another study, nine people with MND with respiratory distress were offered NIV but declined for reasons of loss of self-control and a perception that NIV was not of value to them (Ando et al., 2014). Ando et al. (2014) concluded that the prolonging of life was less important to these patients than their “sense of self” and the preservation of control and dignity (p. 341). Some participants in the study had negative experiences using NIV with discomfort and anxiety.
relating to the mask and forced air, and some had poor experiences with health services and technological support. Some of the participants in the study by Ando et al. seemed to doubt their breathlessness was really a part of the disease or felt that they could control the symptom without the burden of NIV. Greenaway et al. (2015), in their study involving 21 people with MND diagnosed for longer than six months, suggested that timely communication to ensure adequate information is received to enable an informed choice relating to NIV and gastrostomy feeding tubes is an individualised, changing process often influenced by relationships with healthcare providers and previous medical experiences.

Recognising the difficulty healthcare professionals have when trying to assess the amount and depth of information to provide to a person when offering NIV, Greenaway et al. (2015) imply that concise information which includes the benefits and burdens may alleviate some patient and caregiver fears and assist the decision-making process. However, the study also recognises that clinicians move toward interventions may be perceived by people with MND as being pressured into an option. Greenway et al. suggest that individualised and patient-centered symptom control alternatives may be preferred to strict adherence to guidelines as the patient decision-making process for interventions can be influenced by complex relationships with family and clinicians. Whilst the participants in the study identified an apparent “lack of accuracy of information” (p.1008) to enable a decision for or against NIV/PEG from the clinicians, the actual content of the communication verbalised by the clinicians surrounding the offering of interventions was not reported.

Communication about treatment options at all points in the disease trajectory is influenced by a myriad of complex factors, including the distinctive individual characteristics of the participants in the conversation, the content of the information provided, what was heard and understood by those present, and the discussion and individual decision-making mechanisms resulting from the information. Such complexity requires that discussions regarding the benefits,
burdens and limitations of NIV be specific and provide clarity for the patient and their significant others. There is a great deal of ambiguity in the literature regarding how and when clinicians should discuss significant end-of-life decision-making issues with patients and their significant others. In addition, the literature suggests that the benefits and burdens of various end-of-life treatment options including the use of palliating comfort and symptom-relieving treatment alternatives should be discussed and patient decisions relating to end-of-life care understood and documented (Faull, Phelps, Regen, Rowe-Haynes, et al., 2014; National Institute for Health and Clinical Excellence, 2010, 2016; Oliver & Aoun, 2013).

Because of the progressive neurological deterioration and subsequent decrease in independence of the person with MND, the reliance on caregivers increases. This dependence includes the adjustment of the NIV mask, attention to potential pressure ulcers particularly where the mask fits over the nose and being able to remove the mask when necessary. Caregiver burden and stress have been highlighted in recent MND literature, with a focus on a need for improved contact with palliative care services for both patient and caregiver, and an acknowledgement of complex decisions regarding symptom control and quality of life (Aoun et al., 2013; Hogden, Greenfield, Nugus, & Kiernan, 2013; Phukan & Hardiman, 2009). In an extensive review of the literature over 10 years (January 2000-April 2011), Aoun et al. (2013) identified respiratory distress as a difficult symptom for caregivers to cope with, particularly in the terminal stage of MND. Their review also emphasised the importance of clear communication for patients and caregivers by health care providers, and the need for improved access to palliative care (Aoun et al., 2013). However, the potential of NIV, its benefits, burdens, limitations and eventual withdrawal, or how discussion affected caregivers, were not clarified in the review by Aoun and colleagues.

Reasons for not initiating or discontinuing NIV may include the desire to not prolong patient suffering or discomfort relating to its use, which may become
more pronounced as the patient becomes increasingly immobile and
incapacitated. The stage at which the respiratory muscles start to weaken and
palliation by medications and/or initiation of NIV is chosen, may be considered
to mark the commencement of the terminal stage of life (Eng, 2006). This
highlights the need to discuss all potential outcomes of NIV including the end of
life and the eventual need for end-of-life care, at the commencement of NIV.
This will ensure the clinicians are confident that caregivers/family understand
what happens when NIV is removed or if the MND patient dies of co-morbidities
whilst remaining on NIV (some people die of co-morbidities such as pneumonia
prior to withdrawal of NIV). Thus, there is a gap between recommended
discussions relating to MND and NIV use in guidelines and the reality of what
some family members and people with MND hear, remember or understand.

2.2.4. The impact of poor health communication on patients and
families

Uncertainty related to the diagnosis or prognosis of MND combined with
poor communication and understanding of technical treatment details and
information can cause severe stress and strain on the patients and their
families/significant others (Belkora, 2003; Jarvis, 2014). This uncertainty and
confusion may lead those diagnosed with the disease to attempt to collect as
much information as possible and seek out several doctors and other resources
to find answers and inform decisions. This information-seeking has the potential
to result in competing and potentially conflicting care providers with their distinct
views of the case, perhaps adding to confusion (Belkora, 2003; Borasio, Voltz,
et al., 2001). In addition, Belkora (2003) discusses the possibility for patients
and families to be overwhelmed when a diagnosis of a life-limiting disease is
communicated, and who may, therefore, leave decision-making to those with
clinical expertise. Decision-making is both an emotional and cognitive process,
and clinicians could adopt a decision dialogue process to assist people with
their choice of management and care (Belkora, 2003; Jarvis, 2014). Further
research into the content of communication between patient,
caregiver/significant other and clinical team may provide insight.
The MND literature does not appear to incorporate or reflect the viewpoint of the person with MND, nor their significant others, regarding when and how communication explaining NIV initiation and potential withdrawal is imparted. Foley, Timonen, and Hardiman (2014) identified a lack of research surrounding patient understanding of health services and studied how people with MND engage with healthcare professionals. In their study of 34 people with MND, each person was interviewed once, and each had involvement with medical and support care. However, only eight of these participants had accepted either NIV or PEG. The study found that interviewees believed NIV and PEG were interventions to sustain life and not palliating interventions to alleviate suffering (Foley, Timonen, & Hardiman, 2014). This poses a question as to whether people with MND and clinicians may have varying views on “suffering”; people with MND tend to perceive NIV as assisting symptoms of respiratory distress and potentially extending their suffering, but clinicians perceive NIV as relieving their suffering by assisting their symptoms.

In a study by Lerum et al. (2016), 25 MND caregivers were interviewed to explore their experiences of caring for a person with MND. The aim of the study was to understand the difficulties and priorities of care. Lerum et al. found caregivers’ attitudes to seeking clarity of MND information was mixed. Some participants found a sense of control through clarity of information, whilst others found it spoilt the present moment which they treasured. Others used knowledge to challenge the diagnosis (Lerum et al., 2016).

Martin et al. (2014) followed 78 people with MND and 50 MND caregivers regarding circumstances which influenced and enabled decision-making relating to options of symptom control. Martin et al. explored the complexity around whether to accept interventions such as NIV and gastrostomy in people with ALS/MND. It appeared that decisions were related to variables such as the health status of the patient; those who were more unwell were more likely to decide to use an intervention. People with MND and their caregivers with more years of education and a higher IQ, according to Martin et al., were more
capable of seeking information and more likely to decide to use an intervention. The study also found employment status, disease management in the early stages of illness and caregivers with a well-balanced and strong psychological approach to the care of their family member influenced some of the decisions made by the person with MND. It is recommended, therefore, that healthcare professionals assess the coping ability of the caregivers and consider this along with other decision-making factors, when advising patients about treatment choices (Martin et al., 2014). However, the research by Martin et al. did not include research into specific communication used to deliver the treatment choices nor who should deliver the information.

In summary, the amount of information both the person with MND and the family wish to hear from clinicians, and when they wish to hear it, is unclear from the literature. Avoidance of difficult discussions by clinicians relating to disease deterioration and progression and particularly the withdrawal of NIV complicates this issue and is discussed by Faull, Rowe-Haynes and Oliver (2014). Their research suggests that the prognostic conversations are overwhelming and are laden with grief and loss, and so are avoided by patients, significant others and clinicians alike (Faull, Rowe-Haynes, et al., 2014).

2.3. Unresolved Issues Relating to End-of-life Communication

2.3.1. Controversies and clinician/patient difficulties with NIV withdrawal and end-of-life discussions

There is minimal evidence in the literature indicating whether or how people with MND evaluate their quality of life or gauge their quality of death. A lack of communication that encourages effective discussion between patients, families and health care professionals surrounding the terminal stage and death of a person with MND is a significant finding of a secondary data analysis combined study conducted in Australia and England (Ray, Brown, & Street, 2014). Despite the use of advance care planning, Ray et al. (2014) highlight the difficulties in initiating conversations about ongoing care requirements and state
that the development of strategies for clear communication between patient, clinicians and families is vital to ensure patient preferences of care.

A small study of eight MND caregivers identified possible conflicts of interest between patients and caregivers when prioritising work, family and the needs of the person with MND (Hogden et al., 2013). The complexity of changing care requirements for people with MND, communicating with and involving the caregiver plus gauging caregiver level of involvement can present barriers to optimal care and challenges for clinicians (Hogden et al., 2013). Furthermore, a correlation has been found between respiratory function in people with MND and their caregivers' level of distress, affecting the person with MND (Pagnini et al., 2012). Pagnini et al. (2012) suggest the perceived level of social support and caregiver psychological well-being may influence the respiratory symptoms of a patient with MND. Conversely, Pagnini et al. (2012) also suggest that well managed caregiver depression and anxiety may positively affect a patient’s respiratory symptoms.

Whilst there is research which extends the knowledge and understanding of clinicians’ difficulties with the withdrawal and ongoing use of NIV as well as literature acknowledging the emotional burden of caregivers, there is little work on the actual communication surrounding the initiation of NIV, non-initiation/refusal or withdrawal of NIV, and the patient and caregiver understanding of the benefits, burdens and limitations of commencing on NIV. The gap in understanding the end-of-life expectations of the person with MND in relation to their treatment alternatives, and the recommendation that patients should be provided with all their palliative care options, is a finding in an investigation into depression and quality of life in people with MND/ALS (Kübler, Winter, Ludolph, Hautzinger, & Birbaumer, 2005).
2.3.2. Ethical issues

A complex ethical situation arises when the person with MND severely deteriorates and NIV is no longer able to provide a level of support that is effective and positively contributes to the person’s quality of life (Faull, Rowe-Haynes, et al., 2014; Phelps et al., 2015). Faull, Rowe-Haynes et al. (2014) and Phelps, Regan, Oliver, McDermott and Faull (2015) discuss the emotionally challenging aspects and practical implications of the experience of doctors who work in palliative care relating to the withdrawal of NIV in people with MND. They recommend detailed guidelines and conclude that the complex issues affecting the doctors may be difficult to solve. The loss of the ability to communicate with people with MND as their disease progresses is suggested in the literature as a trigger for NIV withdrawal by doctors, and while the wishes of the patient are considered essential, they are not always known (Faull, Phelps, Regen, Oliver, et al., 2014; Faull, Rowe-Haynes, et al., 2014). There is minimal literature relating to patient and family understandings of the limitations of NIV and its potential withdrawal, although fears of choking and symptom control have been discussed (Neudert, Oliver, Wasner, & Borasio, 2001).

As a consequence of disease progression, the patient may end up in what has been labelled as locked in syndrome. This is more common with tracheostomy and invasive positive ventilation but acknowledged to occur with NIV when used constantly, or where the inability to move occurs and clear verbal communication becomes impossible but the brain remains intact and active (Back, 2001; Borasio, Voltz, et al., 2001; Oliver et al., 2006). An ethical dilemma occurs at this point as to the appropriateness of prolonging life with the continued use of NIV or the possibility of an emergency procedure of tracheostomy to maintain respiration with positive ventilation, if the consequences of these choices have not been clearly explained to the patient and their significant others (Faull, Phelps, Regen, Oliver, et al., 2014).

Discussions to ensure full comprehension of the consequences of various treatment choices including a patient’s choice to decline potentially life-
prolonging interventions such as NIV, and the patient’s and family’s decisions for treatment or refusal, should be documented in their medical record early in the disease process (Back, 2001; Oliver et al., 2006; Rafiq et al., 2012). Clear communication from clinicians to those with MND relating to end-of-life issues, which may not only influence their treatment options but help prepare them for a potentially unpredictable disease course, have been found in several studies to be welcomed by most people with MND (Anderson et al., 2007). In a study of 42 people with MND, Anderson et al. (2007) found that end-of-life discussions relating to invasive mechanical ventilation via tracheostomy treatment options were poorly covered by clinicians despite the patient wanting open and honest conversation. Whilst NIV is not as medically invasive as mechanical ventilation, it has burdens, so it is reasonable to suppose that honest and explicit communication is important to clarify the patient’s preferences for treatment.

2.3.3. Patient autonomy: NIV withdrawal

On the first page of the Motor Neurone Disease Australia Patients’ Rights document (MNDAust, 2008) the right of each patient to choose “to accept, refuse or discontinue treatment within the legal framework of each state or territory” is emphasised. In a study by Baxter et al. (2013), healthcare professionals associated with people with MND using NIV in the terminal stage found caregivers/families experienced some confusion about when to withdraw NIV and the impact. Health professionals interviewed within the study expressed concern that families/significant others had a perception of killing the patient on withdrawal of NIV. Misunderstanding relating to management of people with MND and NIV was found in this study to result in some patients receiving emergency resuscitation attempts at the end of life. Whilst indicating that NIV has a generally positive effect on the end of life for people with MND using it, Baxter et al. highlighted the importance of end-of-life directives and making the wishes of the patient known.

Some research literature supports people with MND, who wish to discuss end-of-life issues, receiving information to enable them to make informed
choices (Faull, Phelps, Regen, Rowe-Haynes, et al., 2014). The literature identifies potentially stressful and controversial issues that may arise with the cessation and withdrawal of NIV (Baxter et al., 2013; Faull, Phelps, Regen, Rowe-Haynes, et al., 2014; Faull, Rowe-Haynes, et al., 2014; NICE, 2010). For example, Baxter et al. (2013) identify the impact of the withdrawal of NIV on the clinician’s psychological state, regardless of how symptoms are managed and whether death is instantaneous. Faull et al. (2014) identify the communication and emotional difficulties clinicians encounter with NIV withdrawal, particularly NIV withdrawal at the request of the patient. In a recent retrospective study from England, the ethical and legal issues arising from patient requested NIV withdrawal were explored (Phelps et al., 2015). The study highlighted both the challenge for clinicians to provide honest and clear communication, and the difficulties for clinicians in having to deal with the emotional responses from both patients and families confused by the NIV withdrawal decision process.

Patient and/or caregiver confusion and anguish has the potential to affect end-of-life patient care should healthcare providers be uncomfortable with the concept of NIV withdrawal (Phelps et al., 2015). If seen as an assisted death by the clinician, or if they are unable to communicate the expected outcome clearly, end-of-life care may be adversely affected (Phelps et al., 2015; Ruffell et al., 2013). The literature suggests the opportunity to discuss end-of-life decisions and potential issues is taken at the initiation of NIV and should be reiterated throughout its use (Baxter et al., 2013; NICE, 2010, 2016).

Within the NICE (2016) MND Assessment and Management Guidelines is a new recommendation to ask people with MND how much information they wish to receive about the disease and whether they wish this information to be shared with their families/caregivers (National Institute for Health and Care Excellence, 2016). Ensuring and acknowledging patient care and comfort is a priority as is understanding that clinicians require high level communication skills to establish the trust required for patients to make informed decisions.
Further research which may assist this situation was a recommendation of the NICE MND Assessment and Management Guidelines (2016).

By highlighting the need to support all those involved with NIV withdrawal and to eliminate emotional challenges for both clinicians and families, Faull, Rowe-Haynes et al. (2014) recommend detailed discussions which include the timing, how withdrawal happens, and symptom management thereafter. The responsibility of who is to be involved (family, clinicians) with the NIV withdrawal should be included within these discussions and patient, family and clinicians should have a documented plan of end-of-life care. The suggestion by Faull, Rowe-Haynes et al. that an ethical statement incorporating the clinical aspects and outcome is included within these detailed discussions, and incorporated within MND guidelines for NIV withdrawal, is proposed to help alleviate the ethical and potentially legal dilemma associated with NIV withdrawal for clinician and family. The recent NICE MND Assessment and Management Guidelines (2016) recommend that the healthcare professional commencing NIV can ensure there is legal and emotional support available for other health professionals who may be involved with the withdrawal of NIV. The guidelines also recommend that prior to the decision to withdraw NIV, the patient and family should have support and clear legal and ethical information from clinicians experienced with NIV withdrawal. However, whilst this may assist the clinicians ethically, it may not assist the family members, or the clinicians emotionally with the consequences of NIV withdrawal (Faull, Rowe Haynes et al., 2014).

This overview has identified some gaps in the literature on the topic of communication surrounding the initiation and withdrawal of NIV in MND and forms the background to the research reported in this thesis. In the next section, the methodology used to undertake a sharply focused review of clinicians’ NIV-related communication practice is detailed. The review was conducted with reference to the process outlined in the Australian Journal of Nursing’s “Systematic Reviews, Step by Step” series of articles (Aromataris & Pearson,
The question asked of the literature was formulated using the Population (P), phenomenon of Interest (I), Context (Co), or “PICo” design (Forrest & Miller, 2001), and provided the basis for a systematic search and selection of quality scientific literature that justifies the current study. The report of the review follows the PRISMA checklist (Moher, Tetzlaff, Liberati, & Altman, 2010), modified for reviews of qualitative literature (see Appendix S).

2.4. Section 2: The Research Question and Systematic Literature Search Strategy

2.4.1. Step 1: Developing the question to be answered by the literature review

As stated previously, the question for this review of the literature was developed using the adapted PICo design in which the population, interest and context of scientific studies sought are denoted (Forrest & Miller, 2001; Moher et al., 2010). The question developed for this review was: “What is known about communication (I) between caregivers and families (P) about NIV in MND prior to NIV initiation and withdrawal (Co)?”.

2.4.2. Step 2: Type of studies included

Information for this focused review was collected from qualitative studies in which data were collected via questionnaires, face-to-face interviews, focus groups and telephone interviews.
2.4.3. Step 3: Developing the logic grid

The logic grid classifies the search terms or synonyms used for the literature search. The search terms were identified from and related to the PICo question.

Table 2. 1 Logic grid and search words

<table>
<thead>
<tr>
<th>Population</th>
<th>Phenomenon of Interest</th>
<th>Context</th>
</tr>
</thead>
<tbody>
<tr>
<td>caregiver*</td>
<td>communication</td>
<td>NIV</td>
</tr>
<tr>
<td>clinician*</td>
<td>discussion</td>
<td>non-invasive ventilation</td>
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<tr>
<td>nurse*</td>
<td>decision</td>
<td>initiation</td>
</tr>
<tr>
<td>doctor*</td>
<td>choice</td>
<td>withdrawal</td>
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<tr>
<td>physician*</td>
<td>mnd</td>
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<tr>
<td>famil*</td>
<td>als</td>
<td></td>
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<tr>
<td>relative*</td>
<td>motor neurone disease</td>
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<td>partner*</td>
<td>amyotrophic lateral</td>
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<td>end of life</td>
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<td></td>
<td>palliative</td>
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</table>

2.4.4. Step 4: Defining the search strategy

Boolean operators (AND,OR) were applied between keywords and categories. Within Medline and CINAHL MeSH terms were used in the context of alternate words with similar meaning.

SEARCH 1: caregiver* OR clinician* OR nurse* OR doctor* OR physician* AND famil* OR relative* OR partner* OR loved one* AND communication* OR discussion* OR decision* OR choice* AND mnd OR als OR motor neurone disease OR amyotrophic lateral sclerosis AND end of life OR palliative AND NIV OR non-invasive ventilation AND initiation AND/OR withdrawal
SEARCH 2: caregiver* OR clinician* OR nurse* OR doctor* OR physician* AND famil* OR relative* OR partner* OR loved one* AND communication* OR discussion* OR decision* OR choice* AND mnd OR als OR motor neurone disease OR amyotrophic lateral sclerosis AND end of life OR palliative AND NIV OR non-invasive ventilation

SEARCH 3: communication* OR discussion* OR decision* OR choice* AND mnd OR als OR motor neurone disease OR amyotrophic lateral sclerosis AND end of life OR palliative AND NIV OR non-invasive ventilation

SEARCH 4: mnd OR als OR motor neurone disease OR amyotrophic lateral sclerosis AND end of life OR palliative AND NIV OR non-invasive ventilation.

SEARCH 5: mnd OR als OR motor neurone disease OR amyotrophic lateral sclerosis AND NIV OR non-invasive ventilation

2.4.5. Step 5: Parameters applied to the search

The following limits were applied to the literature search:

- Adult age of participants
- Years of publication: 1990-2016
- Peer reviewed journal articles
- English language

The search inclusion and exclusion criteria are defined more explicitly in Table 2.2. The criteria by which the included full articles were evaluated is shown within Table 2.3.
### Table 2. 2 Inclusion and exclusion criteria for the initial literature search

<table>
<thead>
<tr>
<th>Inclusion Criteria</th>
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<tbody>
<tr>
<td>Peer reviewed journal articles</td>
<td>Non-reputable or non-peer reviewed articles</td>
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<tr>
<td>Articles written in English</td>
<td>Articles not written in English</td>
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<tr>
<td>Literature with concise titles and abstracts relevant to the research</td>
<td>Literature with ambiguous or vague titles and/or abstracts</td>
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<tr>
<td>Method considered rigorous and well defined with sufficient participant numbers</td>
<td>Method poorly described with few participants</td>
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<tr>
<td>Discussion and conclusion well defined and argued</td>
<td>Discussion and conclusion considered to be written to fit the initial objective/hypothesis and not the results as found</td>
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<tr>
<td>Articles able to generally clarify research thesis topic; found via search keywords</td>
<td>Articles considered too general after reading abstract and articles where necessary</td>
</tr>
<tr>
<td>Specific to literature review chapter: Articles able to help identify and clarify research questions</td>
<td>Articles with some relevance to MND but not sufficiently specific to clarify research questions</td>
</tr>
<tr>
<td>Where ‘communication’ related to communication between clinicians, people and families relating to MND disease management and end-of-life care</td>
<td>Where communication related to items to assist people with MND to communicate such as whiteboards, electronic devices and the research surrounding such innovations</td>
</tr>
<tr>
<td>Literature which related directly to the search question</td>
<td>Literature which was considered minimally relevant to the search questions</td>
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</table>

#### 2.4.6. Step 6: Deciding the databases to search

MEDLINE, CINAHL, ProQuest Research Library, and the Cochrane Library of systematic reviews were searched for relevant published research journal articles. Initially article titles were reviewed for relevance, and those that did not resonate with the review question were discarded. The abstracts of the remaining articles were read again and those that did not reflect the review question were rejected. Finally, the full content of the remaining articles was
reviewed, and only those that would likely enable the review question to be answered were retained for quality appraisal (n=18). A hand search of the reference lists of the articles did not identify any new abstracts for review. The final number of articles retained for quality appraisal that met the inclusion criteria was n=8.

2.4.7. Step 7: Example of full electronic search strategy

The following is an example of the five-phase search protocol used for the systematic review. The Medline database search is used to illustrate and identify several peer reviewed research journal articles. The results of each search are shown in bold at the end of each search string:

SEARCH 1: caregiver* OR clinician* OR nurse* OR doctor* OR physician* AND famil* OR relative* OR partner* OR loved one* AND communication* OR discussion* OR decision* OR choice* AND mnd OR als OR motor neurone disease OR amyotrophic lateral sclerosis AND end of life OR palliative AND NIV OR non-invasive ventilation AND initiation AND/OR withdrawal: (0)

SEARCH 2: caregiver* OR clinician* OR nurse* OR doctor* OR physician* AND famil* OR relative* OR partner* OR loved one* AND communication* OR discussion* OR decision* OR choice* AND mnd OR als OR motor neurone disease OR amyotrophic lateral sclerosis AND end of life OR palliative AND NIV OR non-invasive ventilation (Total= 6 – 1 excluded as duplicate from CINAHL, 4 excluded as irrelevant by title; 1 kept for abstract/full article review)

SEARCH 3: communication* OR discussion* OR decision* OR choice* AND mnd OR als OR motor neurone disease OR amyotrophic lateral sclerosis AND end of life OR palliative AND NIV OR non-invasive ventilation (Total= 6 – 5 excluded as duplicates from CINAHL; excluded 25 either repetitive within the search or by title = 1 included for abstract/article review:)

SEARCH 4: mnd OR als OR motor neurone disease OR amyotrophic lateral sclerosis AND end of life OR palliative AND NIV OR non-invasive ventilation.
(Total 52-36 excluded from systematic review but 6 retained for further review guidelines and policy statements and retained for environmental review; 16 retained for abstract review)

SEARCH 5: mnd OR als OR motor neurone disease OR amyotrophic lateral sclerosis AND NIV OR non-invasive ventilation (Total=232-duplicate or rejected on title 224; 6 retained for abstract review, 2 retained for scoping review and as part of the environmental scan).

2.4.8. Step 8: Study selection

The research literature retained for quality appraisal as a result of following the steps reported above could be categorised as qualitative, and as such was assessed using the Standards for Reporting Qualitative Research (SRQR) (O’Brien, el al, 2014). The COREQ checklist (Tong, Sainsbury &Craig, 2007) and criteria for reporting qualitative research has been completed (Appendix T).

The candidate and one supervisor assessed the literature proposed for inclusion in this review: first by title relevance and then by abstract significance to the search question and, by extension, the candidate’s research. Lastly the full articles were read by both the candidate and the supervisor and then appraised for quality using the SRQR checklist. The quality appraisal process resulted in three of the 18 retained articles being discarded.

The quality appraisal results are summarised in Table 2.2. The quality of the papers is described more fully later in this chapter and in Table 2.3. In summary, the journal articles identified in red in Tables 2.3 and 2.4 were disqualified from the quality review as they did not comply with the research criteria.
Table 2. 3 Alignment of the 21 standards for reporting Qualitative research (SRQR)

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<th>Study</th>
<th>S1 and S2</th>
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### Table 2. 4 Research journal articles retained or discarded for review at quality appraisal stage

<table>
<thead>
<tr>
<th>Concise Title</th>
<th>Specific research purpose</th>
<th>Methods</th>
<th>Results/finding</th>
<th>Discussion/limitations</th>
<th>Funding/Conflicts</th>
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<tbody>
<tr>
<td>1) End-of-life decision making in 42 patients with ALS (Anderson et al.2007)</td>
<td>Purpose stated: Timing of discussion of EOL issues: (qualitative study)</td>
<td>Retrospective chart review of people with ALS from diagnosis and then each 3-4 month. Ethics approval</td>
<td>Data collected from June 1999-September 2004. 40/42 patients had EOL discussions at first visit</td>
<td>Retrospective analysis. Limitations: All discussions by one clinician therefore personal bias may have been present</td>
<td>No conflicts of interest</td>
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<td>2) Withdrawal of ventilation at a patient’s requests in MND: a retrospective exploration of the ethical and legal aspects that have arisen for doctors in the UK (Phelps et al.2014)</td>
<td>Purpose clearly stated: To explore and identify legal and ethical issues of ventilator withdrawal: (qualitative study)</td>
<td>Retrospective thematic analysis. 24 clinicians interviewed; 5 by phone,19 face to face. Email recruitment to doctors and via MND clinical study groups. Stated ethics approval</td>
<td>Data taped and transcribed/coded. Saturation after 15 interviews. Ethical challenges for HCP when withdrawal of NIV requested by a patient.</td>
<td>Previous studies identified and integrated. Suggestions for scope of application discussed. No limitations identified.</td>
<td>One of the authors on NICE 2015 NIV/MND guidelines. Funding identified MNDA grant and LOROS (hospice UK)</td>
</tr>
<tr>
<td>3) Physiological as well as illness factors influencing acceptance of NIV and gastrostomy in ALS (Martin et al.2014)</td>
<td>Purpose stated: Identify factors relating to NIV and gastrostomy acceptance in people with ALS: (qualitative study)</td>
<td>A prospective study; 78 people with ALS and 50 caregivers. A descriptive analysis of baseline information. Ethics approval. Written consent from participants</td>
<td>Main finding, those with fewer years of education and lower IQ may be more passive in their decisions regarding gastrostomy and NIV, rather than only influenced by symptoms and illness variables.</td>
<td>Study identified potential limitations: interpretation of findings; no follow up data on those refusing NIV. NICE 2010 guidelines published mid study and may have influenced results. Previous studies identified and incorporated into study.</td>
<td>No conflicts of interest. The research was supported by multiple associations/organisations</td>
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<tr>
<td>Concise Title</td>
<td>Specific research purpose</td>
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<tr>
<td>4) Decision making about gastrostomy and NIV in ALS (Martin et al. 2016) Qualitative Health Research 26(10), 1366-1381.</td>
<td>Purpose clearly stated: Participants were subset of larger study. Interviewing participants occurred at point when they accepted or declined gastrostomy/NIV (qualitative study)</td>
<td>Thematic analysis used to investigate symptom relieving options in ALS. Ethics approval obtained. Written consent from 20 HCP (varied) participants who were selected representatives of people with ALS.</td>
<td>Transcribed interviews with Nvivo 9 used to code and organize data. Findings suggest themes relating to decisions, and previous work integrated into results.</td>
<td>Limitation of study: potential bias in the patient’s selection of the HCP who represented them at interview. Few interviews conducted with patient’s who refused NIV mainly because they died before they could nominate a HCP to represent them</td>
<td>Declared funding conflict by authors: grants and salaried support from trusts and organisations. Views expressed by authors not necessarily Dept of Health UK</td>
</tr>
<tr>
<td>5) Decision-making for gastrostomy and ventilatory support for people with MND across UK hospices (Oliver et al.2011). Journal of Palliative Care 27(3), 198-201.</td>
<td>Purpose clearly stated: Involvement, attitudes and knowledge of specialist palliative care consultants with NIV and gastrostomy discussions. Are the guidelines known and/or applied?</td>
<td>Studies undertaken to use two different methods:62 records analysed regarding symptom control in MND (previous study) then telephone audit (structured questionnaire) of palliative care consultants who had cared for the patients in previous study: referred for ethics approval but deemed unnecessary as considered an audit.</td>
<td>Previous work integrated into findings. Study shows clear differences in timing of discussions of NIV/PEG by palliative care consultants and timing of involvement of consultants. The findings support other research previously published that clinicians are not always aware of guidelines.</td>
<td>No limitations identified by the authors.</td>
<td>No conflicts of interest identified by the authors</td>
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<td>Concise Title</td>
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<td>6) ALS and assisted ventilation: How patients decide</td>
<td>Purpose of study clearly stated: to better understand the experience of decision-making in people with ALS</td>
<td>Qualitative phenomenology methodology: 10 semi-structured interviews with people with ALS and their caregivers. Ethics approval granted and consent from each participant.</td>
<td>Interviews with patients or caregivers using open-ended questions, were taped and transcribed. Six main themes identified and were verified by the participants: meaning of intervention, the importance of context, fears, values, information and adaptation and acceptance</td>
<td>Timing of ventilation discussions and relevant information/evidence/QOL to be given at time to suit individual and caregiver. Limitations include that the interviewer knew the participants already. Some aspects of the interviewer’s clinical role may have influenced the findings. Small sample size.</td>
<td>No declared funding or conflict of interest</td>
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<td>patients decide (Lemoignan, J., Els, C. 2008)</td>
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<td><em>Palliative and Supportive Care</em></td>
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<td>7) Why don’t they accept NIV? Insight into the interpersonal perspectives of patients with MND (Ando et al.2014)</td>
<td>Purpose of study stated: To understand why people with MND decline or stop using NIV</td>
<td>Qualitative research: From a cohort of 35 people with MND who were offered NIV, 9 participated in study. Semi-structured interviews and phenomenological analysis. Participants given information sheet and consent. Ethics approval granted</td>
<td>Themes identified (4); preservation of self, negative perceptions of NIV, negative perceptions of health care services and not needing NIV.</td>
<td>Authors identify that their interpretation of data may not be the only interpretation. Limitations identified: restricted verbal communication in some patients, almost all the data collected from patients who had decline or withdrawn from NIV at an early stage.</td>
<td>No declared conflicts of interest identified</td>
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<td><em>British Journal of Health Psychology</em></td>
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<td><em>20</em>(2), 341-359.</td>
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<td>9) The initiation of NIV for patients with MND: patient and carer perspectives of obstacles and outcomes (Baxter et al. 2012)</td>
<td>Purpose of study stated: patient and carer perspectives of obstacles with NIV in MND</td>
<td>Interviews with 20 patients and 17 caregivers in the first month following introduction of NIV. A qualitative study undertaken until saturation of data.</td>
<td>Study identified themes for acceptance of NIV and the potential barriers which may be anticipated by clinicians to assist patients make decisions.</td>
<td>Limitation identified by authors: sample was weighed towards older males and patients with limb-onset disease and tolerance may be a consideration.</td>
<td>Authors report no conflict of interest. Research funded by research programs; views expressed are authors not Dept of Health.</td>
</tr>
<tr>
<td>10) The use of NIV at the end of life in patients with MND: a qualitative exploration of family carer and health professionals experiences (Baxter et al. 2013) Palliative Medicine 27(6), 516-523.</td>
<td>Purpose of study clearly identified: to describe carer and health professional experience of end-of-life care in people with MND using NIV</td>
<td>A qualitative longitudinal study incorporating in depth face to face interviews with patients and carers. Data collected between 5/2010 and 4/2012, until saturation of data. Interviews audio-recorded and transcribed verbatim.</td>
<td>Study reports on 10 patients followed through to their terminal stage. Total of 24 participant interviews: HCP nominated by carers, plus carers relating to the 10 NIV users, were analysed. Study confirmed importance of end-of-life discussions.</td>
<td>Study consistent with other studies advising early end-of-life discussion/ initiated at commencement of NIV Limitation: a relatively small study of carers and HCP in one small area of UK. Reported information from successful NIV users rather than those who discontinued or decided not to use.</td>
<td>No conflicts of interest identified. Funding under grant: ref: PB-PG-1207-15122.</td>
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<td>11) The impact on the family carer of MND and intervention with NIV (Baxter et al. 2013) Journal of Palliative Medicine 16(12), 1602-1609</td>
<td>Purpose of the study clearly identified: to explore carer burden of person with MND using NIV and how if NIV impacted on family/carer</td>
<td>A mixed methods approach using questionnaires and qualitative interviews. Quantitative data analyzed using SPSS 19.0. Qualitative data analyzed using thematic analysis. Ethics approval granted. All participants had information sheets and signed consent.</td>
<td>Carer Strain Index used to assess level of carer strain. 20 patients recruited; 15 carers completed. Little evidence found of increased carer burden with NIV as disease progressed.</td>
<td>Authors report and advise caution when interpreting very small sample for a quantitative study. Previous studies referred to. This study supports previous research in advising support for carers should start early although inundation of services soon after diagnosis was also reported.</td>
<td>No conflicts of interest identified. Funding provided under Research for Patient Benefit, UK.</td>
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<td>12) Healthcare professionals' views on the provision of gastrostomy and NIV to ALS patients in England, Wales and Northern Ireland (Ruffell, N et al. 2013) Journal of Palliative Care 29(4), 225-231</td>
<td>Purpose of the research clearly stated: study aimed to quantify HCP views on the provision of gastrostomy and ventilatory support for people with MND.</td>
<td>An online survey of 177 (1298 clinicians invited to participate) HCP's in the UK following email invitation to participate. Questionnaire piloted by the clinic that helped to develop it and wording changed as necessary. Ethics approval granted.</td>
<td>Demographic data collected and tabulated. Statistical data imported into SPSS 20.0 clearly described. Differing views between medical and allied health professional regarding the timing of initial discussions around NIV and gastrostomy. Implications of NIV withdrawal included</td>
<td>Limitations as identified by the authors: Because the collection method was anonymous, unable to follow up on non-responders. Low response rate could be due to lack of experience with MND. Practical consideration given to time constraints of clinicians; results may have been influenced by recent changes to NICE NIV guidelines. Too many limitations to be included in quality appraisal</td>
<td>Authors acknowledge funding from MNDA UK and other funding grants. No conflict of interest identified.</td>
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<td>13) Accepting or declining NIV or gastronomy in ALS: patients' perspectives (Greenaway et al. 2015) Journal of Neurology 264(4), 1002-1013.</td>
<td>Purpose of study identified: to identify factors associated with decision making in patients accepting or declining NIV and gastrostomy in ALS</td>
<td>32 participants recruited via ALS register as part of larger prospective study. Semi-structured interviews and analyzed by inductive thematic approach. Ethics approval granted</td>
<td>Three main themes identified: Personal experiences related to patient centric factors; external influence of HCP and family; the concept of time and the effect on decision making</td>
<td>Limitations as identified by authors: study findings may generalize from a clinic population. Patients with communication difficulties were represented by their care givers so views may not have been exact.</td>
<td>Authors state no conflict of interest</td>
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<td>14) Staying just one step ahead: providing care for patients with MND. (McConigley et al. 2013) BMJ Supportive &amp; Palliative Care 4(1), 38-42.</td>
<td>Unclear title: Purpose of study clear in abstract; to determine the experience of and need for, HCP education to enable care for people with MND particularly at end stage</td>
<td>Descriptive qualitative design as part of a larger study. 250 HCP attending MND conference (2008) sent invitation and interviews and focus groups conducted; audiotaped interviews were transcribed verbatim and data managed using Nvivo 8. Data collected until saturation achieved. Ethics approval identified.</td>
<td>Key theme identified: plan and prepare for disease changes. HCP had to be able to predict changes in an unpredictable disease. Acknowledgement that HCP care for patients can be fragmented and that communication vital to enable informed care choices.</td>
<td>Limitations identified: Small number of participants within focus groups held within a conference with competing sessions.</td>
<td>No competing interests. Research funded by NHMRC (425565)</td>
</tr>
<tr>
<td>Concise Title</td>
<td>Specific research purpose</td>
<td>Methods</td>
<td>Results/finding</td>
<td>Discussion/limitations</td>
<td>Funding/Conflicts</td>
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<td>15) Identifying who will benefit from NIV in ALS/MND in a clinical cohort (Berlowitz et al. 2016) <em>Journal of Neurology, Neurosurgery and Psychiatry</em> 87(3), 280-286</td>
<td>Purpose of the study included within background section in abstract.</td>
<td>Retrospective analysis of a prospectively collected cohort of people with ALS/MND. A quantitative study. Ethics approval identified.</td>
<td>NIV was found to increase life by 13 months compared to other symptom control methods.</td>
<td>Study did not capture information on prolonged survival on QOL in people with MND on NIV.</td>
<td>Research funded by two grants; authors state funders had no role in study design, analysis or results.</td>
</tr>
<tr>
<td>16) Standards of palliative care for patients with ALS: results of a European study (Borasio et al. 2001) <em>Amyotrophic Lateral Sclerosis and Other Motor Neuron Disorders</em> 2, 159-164.</td>
<td>Study purpose identified as an investigation of care for people with ALS between and within countries.</td>
<td>A questionnaire was mailed to 110 members of European ALS group. 73 were completed and returned, from 18 countries. Questions focused diagnosis, treatment, PEG, respiratory support, community services and terminal care. Ethics approval not reported.</td>
<td>This study acknowledged NIV can increase survival but was not widely available in all areas/countries. Study suggested many patients with tracheostomies had them fitted in an emergency: validates need for early and improved communication.</td>
<td>The results found a high level of care for people with ALS however was focused on information from specialist centers. Paucity of published controlled studies was identified (pre-2001). Areas not covered by study include pain and psychological issues. Questionnaire only validated by experts.</td>
<td>No conflict of interest identified.</td>
</tr>
<tr>
<td>Concise Title</td>
<td>Specific research purpose</td>
<td>Methods</td>
<td>Results/finding</td>
<td>Discussion/limitations</td>
<td>Funding/Conflicts</td>
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<td>18) Issues for palliative medicine doctors surrounding the withdrawal of non-invasive ventilation at the request of a patient with motor neurone disease: a scoping study. Faull, Haynes, Oliver D (2014) BMJ Supportive &amp; Palliative Care 4(1), 43-49</td>
<td>Purpose of study clearly described: to identify challenges that palliative doctors encounter in relation to NIV withdrawal in people (at patient’s request) with MND.</td>
<td>Primary research: An electronic questionnaire sent via email to palliative care specialists (993) within UK and Ireland after pilot trialled with registrars in palliative medicine. Mixed methods: thematic analysis of free text. Ethics approval sought but not required as study ‘service evaluation’</td>
<td>134 (4 discarded as not from doctors) respondents of which 76 had experience with NIV withdrawal in MND. Practical, ethical and emotional challenges acknowledged but advance care planning may help. Better understanding required to inform future guidance for doctors.</td>
<td>Limitations not clearly identified by authors although mention made that survey very superficial exploration of this difficult area. As only palliative care doctors surveyed, others involved with care of people with MND (nurses, allied health, respiratory specialists) could add further insight. Those that responded may be those with most challenging experiences in study area</td>
<td>One of the authors of this study was a member of the NICE (2010) guideline committee on NIV. Research funded from LOROS and the Wisdom Hospice</td>
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</table>
2.5. Section 3: Search Results

Initial database (SEARCH 1-5: n=968) and manual journal reference list searches (n=16) identified a total of 984 possible relevant articles (n=984). However, articles were excluded using the selection criteria, leaving a total of 18 articles retained from the initial search review for full article review, after which a further two were discarded. The number of articles retained for inclusion in the final review was eight. The literature review illustration below (Figure 2.1), adapted from the PRISMA Flow Diagram (Moher et al., 2010) summarises the method used for this literature search and its results.

Articles were excluded at various stages due to the following reasons: the irrelevance of their titles to the key word search and the search question, non-English language, duplication of title or article, lack of specificity to MND and ALS and/or only mentioned in the context of many diseases with non-specific aspects of palliative care, single case studies or an ill-defined methodology. Some articles had been superseded by the same author with updated research and were excluded. Many articles were editorials, and only the most relevant according to the selection criteria were selected for the environmental literature review. The candidate acknowledges there may have been some relevant journal articles missed due to the quantity found within the initial search and since some articles were disregarded if their title appeared too vague.
Records identified through database searching (n = 968)

Additional records identified through other sources (reference lists) (n = 16)

Records after duplicates removed (n = 84)

Records screened by abstract (n = 52)

Records excluded (n = 34)

Full-text articles assessed for eligibility (n = 18)

Full-text articles excluded as not sufficiently relevant (n = 10)

Studies included in qualitative synthesis (n = 8)

Studies included in qualitative synthesis (meta-analysis) (n = 0)

Figure 2. 1 Literature Search and Retrieval Process (Moher et al., 2010)
2.6. Overview of Retained Articles

The literature generated through the systematic searches and selection process outlined above identified knowledge gaps within the topics relevant to this thesis. Several of the journal articles (n=15) included within the systematic review reflected more than one theme. The themes identified within the eight research articles retained for this review include:

1. The communication and moral challenges doctors face when discussing the possibility of NIV withdrawal with people with MND and their families
2. The role of healthcare professionals and influence of families on people with MND making symptom management (NIV) decisions
3. The importance of well-timed, effective and individualised communication by health care professionals to people with MND and their families.

**Theme 1:** The communication and moral challenges doctors face when discussing the possibility of NIV withdrawal with people with MND and their families.

The withdrawal of NIV is not well documented within the literature. Within the systematic search, only one article was found which explored the potential dilemmas doctors face when asked to withdraw NIV from a person with MND. In the United Kingdom, a retrospective qualitative analysis of 24 doctors with experience of ventilatory withdrawal from people with MND were interviewed (Phelps et al., 2015). The doctors experience of withdrawal of NIV (n=16) undertaken at a patient’s request was explored. Whilst this research was predominantly exploring the complexity of legal and ethical issues arising for the doctors requested to withdraw NIV, individual perspectives and emotional issues relating to the families concerned were not explored. However, Phelps et
al. (2015) acknowledged that the influence of such complex emotional issues for doctors might have an impact on families of and people with MND.

Theme 2: The role of healthcare professionals and influence of families on people with MND making symptom management (NIV) decisions.

The decision by people with MND as to whether to use symptom relieving measures such as NIV and gastrostomy may be influenced by a healthcare professional’s own perspectives (Martin et al., 2016). In the study by Martin et al. (2016) healthcare professionals were nominated by people with MND to represent their decision to use (or not) NIV and gastrostomy. Whilst quality of life was related to the practicality of the intervention, the pressures involved with making multiple decisions because of clinical need was expressed. The healthcare professionals found the timing of discussions relating to NIV and PEG difficult, with some stating early discussions were vital. All acknowledged that such discussions depended on the emotional and social factors relating to the individual with MND. The study by Martin et al. (2016) is limited by the small number of NIV related interviews, and by possible bias in the selection of the healthcare professionals who took part.

Decision-making for NIV and gastrostomy in MND and how these decisions may be influenced by healthcare professionals, is further explored by Oliver, Campbell, Sykes, Tallon and Edwards (2011). Their study focused on the involvement of 22 palliative care consultants who took part in a telephone audit. Only a small group of consultants (n=5) was involved with early discussions with people with MND and involved an MDT approach to discussion. The other 17 consultants were either involved at the terminal stage of the disease or reported that the consultant was rarely involved with a patient’s decision-making. The
consultants with a special interest in MND or within a specialist MND unit were more likely to involve an MDT and acknowledge the need to develop a rapport with the person with MND. The study by Oliver et al. (2011) suggested that the guidelines for these symptom control measures may not be considered (or known) by healthcare professionals involved with the care of people with MND.

**Theme 3:** The importance of well-timed, effective and individualised communication by health care professionals to people with MND and their families.

In two articles by Baxter et al. (2012, 2013), family and carer experiences of the initiation and use of NIV in people with MND are explored. In the first qualitative study, 20 people with MND using NIV and 17 carers were interviewed within one month of the commencement of NIV. The aim of the study was to investigate personal attitudes and initial concerns around using NIV. Some of the perceived barriers to NIV, as voiced by people with MND in the study, included the sensation of air being blown over the face, fear of the machine itself, sleep difficulties due to machine noise and dryness of mouth, lips and nose. Most people with MND within the study stated the need for perseverance with NIV. The study concluded the importance of pre-empting potential difficulties of NIV, the need for accessible advice plus the importance of discussing NIV in detail with people affected with MND and their families. However, this study did not discuss the content of the communication delivered by the health care professionals to the study participants prior to the initiation of NIV.

In the second qualitative study by Baxter et al. (2013) the difficult issues associated with potential NIV withdrawal in people with MND were explored. Interviews were conducted with nine bereaved family carers and 15 healthcare professionals, in relation to 10 people with MND using NIV at the end of their
lives. The often unexpectedly rapid deterioration of the person with MND led in two instances to resuscitation attempts by emergency departments. This reiterates the need for clear direction in end-of-life care planning. Rapid deterioration within the terminal phase was described as a surprise by some of the health professionals within the study by Baxter et al. (2013) who reiterated the NICE (2016) guideline recommending that communication of end-of-life issues should be ongoing once NIV is initiated.

Identifying factors which influence decisions relating to accepting or declining NIV and gastrostomy was studied through face to face interviews with 21 people with MND by Greenaway et al. (2015). Within a thematic analysis three themes were identified; patient-centred issues such as fear, control and need; the role of and information provided by health care professionals; and the concept of time. Greenaway et al. found that some health care professionals directed their patients towards some interventions stressing the consequences of late decisions. In some instances, this was perceived as being pressured into a decision by the person with MND and their family. A lack of knowledge about MND and the disease process among health professionals was also found which caused some participants to feel under supported. Acknowledging the complex communication dilemma for health professionals was highlighted by Greenaway et al. Offering clarity relating to quality of life and the intervention itself, before and after commencement, may assist people with MND make their decisions.

The need for health professionals who have the appropriate experience and education to assist and care for people with MND is explored in a study of 31 health professionals involved with MND and palliative care (McConigley et al., 2014). Communication within the care team as well as a rapport between health care professional and the person and family coping with MND was highlighted by McConigley et al. (2014) and other studies (Faull, Rowe-Haynes, et al., 2014; Oliver et al., 2011). The introduction of palliative care to the person with MND and their families was admitted as being met with resistance at times,
particularly by those families unwilling to acknowledge the terminal stage of the disease. Skilled and accurate communication was found to be essential to navigate the complexity, unpredictability and possible speed of change of the disease.

Members of a professional body specifically involved with palliative care and with experience of people with MND were asked to complete an electronic questionnaire relating to NIV withdrawal (Faull, Rowe-Haynes, et al., 2014). The research corresponded with the study by Phelps et al. (2015) in highlighting the emotional and ethical challenges doctors face when asked to withdraw NIV. Of more importance to the candidate’s study, is the research by Faull et al. (2014) which found communication regarding the benefits and challenges of NIV should be (but not always are) incorporated earlier in the disease rather than when deemed necessary. Discussions relating to AHDs may assist patient care relating to NIV withdrawal but can be overwhelming and, therefore, avoided by families and health professionals alike.

2.6.1. Summary of retained articles

It is not clear whether or how frequently non-initiation of NIV is discussed, when referral to a multidisciplinary palliative care team is offered, whether this option is practically available (for example in rural Australia), and what level of comprehension of the prognosis and options the patient and their family have. Furthermore, when do clinicians routinely discuss the emotional, psychological and physical impacts of NIV? Is the possibility of withdrawal of NIV and subsequent imminent death discussed as the present guidelines recommend? Whether the amount of extra life that NIV therapy typically offers is discussed, but if the relentless disease progression, despite the NIV, is explained are there also questions as yet unanswered? Finally, whether clinicians routinely introduce the idea of not initiating NIV to avoid prolonged suffering and offer alternatives such as medication for symptom control is unknown.
2.7 Section 4: Environmental and Targeted Scans

More generally relevant literature sourced from the environmental scan was included in the background to the research literature. Google scholar produced 9,990 results for the initial search which was reduced (n=292) after eliminating irrelevant titles, repeat articles within the same search, and duplicates. The remaining abstracts considered broadly relevant were read (n=50). Any peer reviewed articles considered generally relevant which could enlighten the research question and identified gaps within the literature were included within the background of the literature review (n= 32). These articles are listed and described in Appendix A.

2.7.1. Environmental scan

As NIV is relevant to MND-related respiratory failure as well as several other chronic diseases, an environmental scan of contextual literature about the issue in relation to both MND and a variety of other healthcare conditions was conducted. This helped to determine what care is recommended in relation to end-of-life communication concerning NIV in people with a life-limiting health conditions including MND. The scan resulted in a review of the contextualising grey literature that proved essential to the candidates overall understanding of the topic of interest and further confirmed the need for the current study (Martin-Misener et al., 2012).

2.7.2. Targeted scan

The targeted search was aimed at data bases where published recommendations for care could be found. The databases accessed included NICE, NHMRC, WHO and the Motor Neurone Association databases for the United Kingdom and Australia. Of particular interest for this research was the unanswered question of how the present MND NIV Assessment and Management Guidelines, previously the Use of Non-invasive Ventilation in the Management of Motor Neurone Disease (NICE, 2010), in relation to the benefits and burdens of NIV and end-of-life communication are being reflected in practice (NICE, 2016). The NICE Assessment and Management Guidelines
(2016) were published toward the end of the research process for this study. The targeted search identified several MND recommendations including the European consensus on palliative care in neurological disease (European Federation of Neurological Societies and European Association of Palliative Care, 2015) and a guide to ALS patient care for primary care physicians (Amyotrophic Lateral Sclerosis Society of Canada, 2010). However, the NICE guidelines (initially version 2010 and later 2016) were found to be the most comprehensive and agreed with other MND management guidelines found (Amyotrophic Lateral Sclerosis Society of Canada, 2010; Andersen et al., 2012).

The following focus areas were identified in the documents retrieved:

1. General palliative care/MND specific palliative care research, including patient-centered care, evidence-based medicine, clinical guidelines and recommendations regarding best practice in end-of-life care and a good death, and ethical issues/communication surrounding NIV and end-of-life
2. The timing of palliative care service involvement in the illness trajectory of the person with MND and triggers for NIV and end-of-life communication for people with MND and their families
3. Content of communication in NIV and end-of-life discussions for people with MND and their families
4. Factors influencing decisions, for example, MND related dementia and the barriers to essential communication
5. Caregiver and patient concerns at the end of life and during MND and the identification of various issues in the delivery of end-of-life care that are still unresolved in the literature and clinical practice
6. Clinician concerns with communicating NIV benefits and burdens and end-of-life concerns
7. The impact of insufficient NIV/end-of-life communication in people with MND.
As each of these focus areas is considered, what remains unknown about each is also explored. This informs the development of a research question to address that gap. As these focus areas are inextricably linked, they are discussed together within the summary of evidence.

2.7.3. The development of clinical guidelines and recommendations regarding best practice in end-of-life care

Clinical guidelines incorporate expert clinical opinion and research on “how patients actually die” to determine optimum standards for palliative care. For example, the WHO Policy Statement (WHO, 2015b), the NICE quality standards for end of life care (NICE, 2011), the Gold Standards Framework for palliative care (Thomas & Free, 2006) and guidelines from Palliative Care Australia (The National End of Life Framework Forum Planning Group, 2010) all agree that the following elements are vital to good end-of-life care: communication, co-ordination of care, symptom control, continuity of care, caregiver support and adequate practical physical and psychological care in the terminal phase. Although there is consensus across a wide variety of clinical specialties and disease categories on what kinds of care should be provided to those facing the end of life, implementing these guidelines for patients across the continuum of care has proven difficult.

2.7.4. MND specific palliative care guidelines

The few end-of-life recommendations identified specifically for people with MND tend to mirror those guidelines identified for general end-of-life patients. However, the literature points to some special circumstances that people with MND and their significant others face, specifically around the initiation, non-initiation, or withdrawal of NIV (National Institute for Health and Clinical Excellence, 2010, 2016). For example, the earlier The Use of Non-Invasive Ventilation in The Management of Motor Neurone Disease Guidelines (NICE, 2010) provided evidence-based recommendations regarding treatment and care of people with MND.
The guidelines recommended that the choices of symptom-relieving measures which may be beneficial to patients, including NIV, be honestly communicated to people with MND at a time appropriate to the individual’s capacity to emotionally comprehend the information. The later NICE (2016) Management and Assessment Guidelines, which supersede the NICE (2010) MND NIV guidelines, recommend being “sensitive about the timing of discussion” (p.14; section1.7.1) particularly relating to any cognitive or communication issues, without providing further detail to assist clinicians in interpreting and applying these recommendations. In addition, these recommendations suggest that when considering the process of initiation, non-initiation or withdrawal of NIV, patients and significant others should be reassured that all comfort measures and medication will be provided to prevent distress from symptoms. While this aspect of care is widely discussed in the literature (Borasio, Voltz, et al., 2001; NICE, 2010, 2016; Oliver et al., 2006), insufficient detail about the content of such discussions to guide clinicians was identified.

People with MND have been found to feel more demoralised and hopeless than cancer patients, with increased dependency on social support and prolonged and ever worsening, physical incapacity (Clarke, McLeod, Smith, Trauer, & Kissane, 2005).

Tables 2.5 and 2.6 summarise and compare general palliative care guidelines with motor neurone specific recommendations found within a targeted literature search. The two tables highlight the recommendation in all the guidelines relating to palliative care, that clear communication is one of the most important components of good end-of-life care. This enabling communication style allows patients to make informed treatment choices relating to their end-of-life care. The most pertinent take-away message from this comparison is the uncertainty about when palliative care should commence in MND, whereas it is understood to be relevant for cancer patients as their condition becomes unresponsive to curative treatment. There is notable
similarity between the recommendations in general palliative care and MND relating to optimum symptom control and the availability of end-of-life information. In MND, communication surrounding NIV and end-of-life care is recommended to occur prior to the initiation of NIV and to be documented within an AHD (NICE, 2016). Within the general palliative care guidelines individualised care plans are recommended, although AHDs are encouraged by the clinicians to clarify the patient's end-of-life care wishes.

Table 2. 5 MND Specific Palliative Care Recommendations

<table>
<thead>
<tr>
<th>MND Palliative Guidelines</th>
<th>Palliative Care Recommendations</th>
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<tr>
<td>European Federation of Neurological Societies (EFNS) guidelines on the clinical management of amyotrophic lateral sclerosis (MALS) - revised report of an EFNS task force (2014).</td>
<td>Percutaneous endoscopic gastrostomy tubes should be placed before respiratory insufficiency develops. Non-invasive positive-pressure ventilation also improves survival and quality of life. Maintaining the patient’s ability to communicate is essential. During the entire course of the disease, every effort should be made to maintain patient autonomy. Advance directives for end-of-life care should be discussed early with the patient and caregivers, respecting the patient’s social and cultural background (Andersen et al., 2012).</td>
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### Table 2.6. General Palliative Care Recommendations and Policy Statements

<table>
<thead>
<tr>
<th>General Palliative Guidelines/Policies</th>
<th>Palliative Care Recommendations</th>
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<tr>
<td>World Health Organisation (World Health Organization, 1998) Definition of Palliative Care</td>
<td>Provides relief from pain and other distressing symptoms. Affirms life and regards dying as a normal process. Intends neither to hasten nor postpone death. Integrates the psychological and spiritual aspects of patient care. Offers a support system to help patients live as actively as possible until death. Offers a support system to help the family cope during the patient's illness and in their own bereavement; Uses a team approach to address the needs of patients and their families, including bereavement counselling, if indicated. Will enhance quality of life and may also positively influence the course of illness. Is applicable early in the course of illness, in conjunction with other therapies that are intended to prolong life, such as chemotherapy or radiation therapy, and includes those investigations needed to better understand and manage distressing clinical complications (World Health Organization, 1998).</td>
</tr>
<tr>
<td>Therapeutic Guidelines; Palliative Care (Palliative Care Expert Group, 2010)</td>
<td>Provides relief from distressing symptoms. Neither hastens or postpones death. Includes all aspects of culturally sensitive support and care for patient, families and caregivers. Avoids futile interventions. Acknowledges death as a normal process. Uses a multidisciplinary team to maximise requirements of patients/families/caregivers.</td>
</tr>
<tr>
<td>National Institute for Care and Health Excellence (2011) Quality standard for end of life care for adults</td>
<td>Local availability of end-of-life information. Symptom relief; information relating to care options. Sensitive and clear communication. Advice freely available to patients and caregivers including support groups.</td>
</tr>
<tr>
<td>Department of Health (UK) (2010) End of life strategy; quality markers</td>
<td>Action plan for delivery of high-quality care. Ensure people approaching end-of-life offered care plan. Ensure effective identification of those nearing end-of-life. Ensure patients preferences and choices are documented and expressed when they wish. Ensure that the needs of caregivers are addressed. Ensure 24 hr coordinated care for patients.</td>
</tr>
<tr>
<td>Palliative Care Australia (4th Ed) (2005) Standards for providing quality palliative care for all Australians</td>
<td>Care planning based on individual and on respect for patient, caregiver and family. Sensitive communication skills used by providers to ensure holistic needs of the family. Ongoing assessment and care planning. Co-ordinate care with good communication. End-of-life issues discussed honestly with patient and family. Primary caregiver given information and support. Access to palliative care.</td>
</tr>
<tr>
<td>Thomas, K., &amp; Free, A. (2006). Guidelines in Practice, 9(6). <a href="http://www.goldstandardsframework.org.uk">www.goldstandardsframework.org.uk</a></td>
<td>To improve the quality of care for people near the end of life, in line with their preferences; assess patient physical and psychological needs. To develop the generalist workforce, leading to better collaboration and coordination of care; anticipate patient requirements. To improve cost effectiveness by decreasing hospitalisation. Clear communication between patients, families and clinicians.</td>
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2.8. Section 5: Summary of Evidence

The lack of clarity in the current literature regarding the content and process of communication between clinicians, people with MND and their significant others, as well as the level of comprehension of this information by the patient and the family at the time of discussing NIV, forms the basis of the research reported in this thesis. Most guidelines and recommendations are based upon expert specialist clinical opinion rather than scientific research. Further research regarding the withdrawal of NIV in people with MND and specific practice guidelines may need to be developed to ensure optimum patient care and understanding, and this remains the case as little has emerged since this recommendation was made by Eng (2006) and endorsed by the Motor Neurone Disease NIV Guidelines (NICE, 2010). Subsequent endorsement in the Motor Neurone Disease Assessment and Management Guidelines (NICE, 2016) is based on much of the same expert clinical opinion.

The procedure and communication surrounding NIV withdrawal is included as a recommendation for further research in the NICE (2010) clinical guidelines into the use of NIV. The importance of clear communication delivered in a sensitive manner, is emphasised in the NICE (2016) Motor Neurone Disease Management and Assessment Guidelines, with recommendations on delivering concise information when the person with MND is ready to accept it. Moreover, the Mechanical Ventilation for Amyotrophic Lateral Sclerosis/Motor Neurone Disease Cochrane review (Radunovic et al., 2013) suggests that further research is required in order to understand the personal factors surrounding the use of NIV.

The influences on clinicians in initiating, conducting, ensuring patient and family comprehension of MND respiratory symptom control and focusing on NIV are extremely diverse. They include the factors that influence when to have the conversation as well as what the patient and/or family is ready to hear about in terms of the benefits, burdens, limitations and potential impacts of disease progression and symptom control. The identification of which clinician is
responsible for ensuring that the patient and family are told the appropriate information relating to end-of-life care, symptom control and quality of life issues and their level of understanding of this information also appears uncertain at this point. While it is clear that there are accepted trigger points in illness trajectories at which honest discussion should occur - the proposal to initiate NIV is one - it is also evident that these discussions are often delayed as health professionals perceive a lack of readiness to engage in such discussions on the part of the patient or family and they may be unwilling to acknowledge that when the patient’s condition deteriorates these discussions are necessary (Mitsumoto & Rabkin, 2007).

In addition, whilst MDTs are seen as important for effective symptom control and support for the person with MND, the timing of the introduction of multidisciplinary palliative care also remains uncertain and unpredictable (Bede et al., 2011). Early palliative care is generally accepted as preferred practice (Andersen et al., 2012; MNDAust, 2014); however, there are barriers to this occurring, including ineffective communication within the MDT and between the MDT, MND patient and their family. Some identified communication barriers to an early palliative care referral include health professionals’ reluctance to confer a lack of hope of recovery on the person with MND and lack of appreciation of the potential for rapidly changing patient needs, support and symptom management (Boersma, Miyasaki, Kutner, & Kluger, 2014).

The availability of a group of specialists to form an MDT within the same health facility, inclusive of a palliative care specialist, who have the capacity to meet and communicate as an interconnected team including the person with MND and their family may prove difficult. This MDT is more likely to occur in larger hospitals and cities. The absence of this in rural areas may prove to be a barrier to aspects of necessary care and overall responsibility for the person with MND and their family members. Palliative care MDTs, the variability in the quality of the palliative care available and the inconsistency of palliative care provision in urban versus rural areas may all create barriers to the move to
palliation for people with MND. These have been explored within the parameters of this research and are discussed later in the thesis.

The potential for timely, effective and explicit discussions relating to AHDs, symptom control and end-of-life preferences are vital prior to the time when speech becomes adversely affected and while clear communication remains possible. Whilst these discussions are generally considered to be the domain of the palliative care specialist, aspects relating to end-of-life care for people with MND are considered appropriate for discussion shortly after diagnosis so that the patient and their family can make choices prior to any deterioration of symptoms. Particularly in rural areas, the person with MND may not have local access to a palliative care specialist or MDT or may not be referred by their GP. These difficult discussions become the responsibility of the GP or are delayed until the patient’s disease progresses sufficiently that palliative care become involved. There is a possibility for assumed responsibility for the end-of-life discussions, and this aspect is also explored within this research and identified within the discussion chapter. Palliative care, according to international guidelines, should be offered to all patients when identified with an incurable, life-limiting disease in order to alleviate adverse symptoms and promote quality of remaining life (NICE, 2011; WHO, 2015b). It follows that people with MND and other neurological diseases, as well as those with chronic diseases such as renal or heart failure, should not only have equal access to palliative care but their care should reflect evidence-based treatment pathways and recommendations, much as cancer patients’ care is informed by evidence-based guidelines (National Institute for Care and Health Excellence, 2011; Palliative Care Australia, 2005).

However, as MND is a rare neurological disease, many clinicians are relatively inexperienced with the symptom control issues, the delivery of difficult prognostic information and with optimal timing for referral to palliative care specialist teams. Palliative care guidelines relative to those with cancer include recommendations that value and encourage partnerships within health care
organisations and the formation of a framework according to which care is delivered wherever the cancer patient wishes to die (National Institute of Clinical Excellence, 2004; Palliative Care Expert Group, 2010). Recommended end-of-life communication for cancer, chronic disease and neurological patients all include well-coordinated patient-centered care and symptom control, psychosocial support for patient and families and the provision of generally detailed information about disease trajectory (NICE, 2013; Palliative Care Australia, 2005).

The 2016 NICE guidelines recommend early referral of people with MND to palliative care and an MDT (Anderson et al., 2007). The guidelines encourage early discussion with the person with MND and their family about end-of-life issues and the complications surrounding the initiation and withdrawal of NIV. The description of points to include within conversations, previously described as honest communication (National Institute for Health and Clinical Excellence, 2010), is now more prescriptive; it is now considered necessary to ensure the person with MND understands what NIV is and what it can achieve. While the recommendations have become more precise, the in-practice content of clinician communication between people with MND and their families about NIV and end-of-life choices is still not known. It is this gap that the research reported in this thesis seeks to address.

2.8.1. Research questions resulting from the literature review

The research questions were formulated from the gaps evident in the black and grey literature reviewed and are summarised as follows (Table 2.7). The first question has been answered by the literature reviewed in this chapter.
Table 2. 7 The Research Questions

1. In considering the available literature and guidelines, what are the most salient components of best practice relating to the communication about NIV in MND prior to NIV initiation and withdrawal?

2. What is the evidence that the best practice communication content (benefits and burdens of NIV, continued disease progression, explanation and offer of a PEG, end-of-life discussions and understanding the potential for NIV withdrawal prior to the commencement of NIV) is incorporated into the actual communication by clinicians to people with MND and their families at the time of offering NIV?

3. What do MND families/significant others recall about the communication (benefits and burdens, continued disease progression, PEG insertion, potential NIV withdrawal and end-of-life discussions) provided by clinicians at the time of respiratory failure and the offer of NIV?

4. To what degree do MND families/significant others feel that the NIV communication provided by the clinicians at the time of the offer of NIV prepared them for and helped them to understand the implications of potential NIV withdrawal resulting in the death of their family member?

5. What if any unmet needs for information, support, palliative care, or other services do the clinicians and the families/significant others (or patients themselves) with experience of MND identify?

6. Following the literature review and clinician and family/significant other interviews regarding NIV communication, what recommendations can be identified that would improve end-of-life communication and process for people with MND, families and clinicians?
2.9. Theoretical Framework: Domains of Influences on the Phenomenon of Interest

The impact of MND on family members has been described in the literature as potentially devastating. The lack of any possible cure for MND restricts the communication to the patient and family to issues of adequate symptom control, comfort and caregiver support. The communication surrounding NIV initiation and its potential removal resulting in death, deciding if and when to remove NIV and whether or not to have a PEG inserted prior to weakening of the swallowing reflex and respiratory muscles, are all highly emotional issues. In the context of a disease whose course can be so diverse and individualistic, trying to remove all ambiguity from these complex discussions is difficult. There are many interacting factors that influence the effectiveness of the communication surrounding NIV, and these are summarised in the Domains of Influence matrix provided in Figure 2.2.

It is necessary to consider the complexity of the communication recommended to occur at various trigger points by the clinicians, the link between commencing NIV and its potential withdrawal and death, and the way in which communication may be interpreted by those who hear it. The Domains of Influence framework derived from the existing literature and developed by the candidate, is used to present and contextualise the discussion of findings of this study in Chapters 4 and 5.
Figure 2. 2 Domains of influences.

References: (1) (Anderson et al., 2007); (2) (Andersen et al., 2012); (3) (Foley et al., 2014); (4) (Martin et al., 2014); (5) (McConigley et al., 2014); (6) (National Institute for Health and Care Excellence, 2016); (7) (Phelps et al., 2015)
2.10. Chapter Summary

As respiratory failure is the most common cause of death in people with MND, alleviating respiratory distress either with technology or medications is important. The clarity of communication which surrounds initiation of NIV and the end-of-life care choices of the person with MND is, therefore, vital. Extensive review of the literature to fully understand the implications of what the guidelines recommend is imparted to people with MND and their families has been undertaken and the gaps in knowledge about this phenomenon identified. A tabulated description of the most relevant peer-reviewed journal articles is provided in Appendix A.

Whilst there is extensive literature and numerous guidelines concerning general end-of-life care, and specific recommendations guiding the use and recommended communication surrounding NIV in people with MND, there is a paucity of research examining whether and how the communication recommendations specific to NIV in MND are applied in practice. The existing literature does not elucidate the actual content of discussions between clinicians, people with MND and their significant others regarding the initiation, non-initiation or withdrawal of NIV; the benefits, burdens, and limitations of NIV; alternatives to NIV; nor their impact on the quality of life of the patient and family. Moreover, it is not apparent from the literature whether these discussions are reiterated at any time during the disease progression and with whom they occur, nor how much the patient and family comprehend the realities regarding NIV. The lack of specific research into content and detail of communication surrounding the experiences of people with MND, their families and their clinicians points to the need for further research (NICE, 2010; Oliver & Faull, 2013).

This chapter has provided the foundations of this thesis and has presented the five research questions to be answered, relating to the communication surrounding initiation and withdrawal of NIV in people with MND. Investigation of the literature to find how well caregivers and families of people with MND understand the benefits, burdens and limitations of NIV prior
to commencement; how this impacts their end-of-life decisions and subsequent experience of their family member’s death; and how clinicians communicated has exposed knowledge gaps and answered the first research question. Literature concerning PEG tubes and, more broadly, on hydration and nutrition has also been included in this literature review. The inclusion of communication surrounding PEG tube insertion was considered relevant as PEG tubes are generally considered for symptom control at approximately the same time as NIV, have related clinical benefits and limitations and are recommended to be incorporated into communication surrounding end-of-life issues. In the following chapter, the research methodology and methods used in this study are defined, an explanation as to why the approach taken was chosen is provided, and the study processes and considerations are detailed.
A qualitative study into the communication surrounding the initiation and withdrawal of non-invasive ventilation (NIV) in people with motor neurone disease

Chapter 3

Method and Methodology

3.1. Chapter Overview

In Chapter 2 the need for this study, its purpose and aims and the specific research questions to be asked of the data, which were informed by the gap in available evidence, were presented. Although the published literature touches on many aspects of MND, NIV and end-of-life care, little evidence was found that enabled comparison of the present guideline recommendations relating to the timing and content of a clinician’s delivery of NIV communication with actual patient and caregiver understanding and recollections of communication. The complexities of communicating respiratory failure symptom relief and end-of-life choices for people with MND are, therefore, the focus of the research questions asked in the current study.

Whilst clarity of communication in any context is difficult to verify and measure, for this research the most recent published guidelines and recommendations for communicating respiratory failure symptom relief and end-of-life choices to people with MND were used as an indicator of what these conversations should feature. Therefore, this study investigated what information clinicians report they communicate, when and how they assess patient and family understanding of the information presented to them, how its presentation compares to guideline recommendations, and what was understood by the recipients of this communication, that is, family members of people with MND. This information, along with insights from an expert advisory panel, or stakeholder advisory group (SAG), described below, helped inform the approach to data collection in the current research.

To answer the questions and aims posed for this study, clinicians working with people with MND and members of families of those with MND
were asked in-depth questions about communication relative to NIV and related end-of-life care issues. Their responses were then analysed qualitatively and made sense of in the context of the literature on this topic. It is these research processes and outcomes that are reported in this chapter and the two that follow.

This chapter comprises two sections. The first section is concerned with the philosophical, methodological and theoretical approach underpinning the current study, and the second section reports the methods and processes followed in the conduct of this research.

Section 1 (Methodology) begins with an overview of the four predominant research paradigms within which contemporary health researchers work, and the ontological and epistemological tenets of each. The broad approach to the conduct of research by scientists working within each paradigm is then described. Those employed for the current study are then identified as naturalistic-interpretivism and the qualitative approach, and the rationale for their selection is provided.

Several specific methodologies that reflect the qualitative approach are then visited, and the one selected for the current study – interpretive descriptive (ID) methodology - is presented and defended. Examples of theoretical lenses through which health researchers make sense and meaning of the data they collect are then provided. This section concludes with a statement about the theoretical framework that was chosen to underpin the current study – symbolic interactionism (SI) - and the reasons for its selection.

In Section 2 (Methods) the fully replicable steps taken in the current study are detailed. This section includes information about sampling and recruitment processes, data collection, analysis and sense-making processes and measures taken to ensure the study is ethical and trustworthy.
3.1.1. Research questions and aims

As a reminder to the reader, the research questions and aims pursued in this study were as follows (the first question has already been answered in Chapter 2):

1. In considering the available literature and guidelines, what are the most salient components of best practice relating to the communication about NIV in MND prior to NIV initiation and withdrawal?

2. What is the evidence that the best practice communication content (benefits and burdens of NIV, continued disease progression, explanation and offer of a PEG, end-of-life discussions and understanding the potential for NIV withdrawal prior to the commencement of NIV) is incorporated into the actual communication by clinicians to people with MND and families at the time of offering NIV?

3. What do MND families/significant others recall about the communication (benefits and burdens, continued disease progression, PEG insertion, potential NIV withdrawal and end-of-life discussions) provided by clinicians at the time of respiratory failure and the offer of NIV?

4. To what degree do MND families/significant others feel that the NIV communication provided by the clinicians at the time of the offer of NIV prepared them for and helped them to understand the implications of potential NIV withdrawal resulting in the death of their family member?

5. What if any unmet needs for information, support, palliative care, or other services do the clinicians and the families/significant others (or patients themselves) with experience of MND identify?

6. Following the literature review and clinician and family/significant other interviews regarding NIV communication, what recommendations can be identified that would improve end-of-life
communication and process for people with MND, families and clinicians?

The study aims to be answered by the research questions are as follows:

1. To describe clinician communication surrounding NIV including end-of-life choices
2. To understand how clinicians check the level of patient and family members’ understanding of NIV benefits and burdens and the potential of NIV withdrawal prior to NIV commencement
3. To understand the impact of clinician communication about the benefits, burdens and limitations of NIV on family members’ choices in relation to NIV introduction
4. To understand what timing, content and style of communication most effectively helps the families of people with MND to make decisions about NIV initiation and withdrawal
5. To compare the content, process and outcomes of NIV communication as described by clinicians and families of people with MND within this study against the currently available recommendations relating to commencement and withdrawal of NIV and related end-of-life options for care.

3.2. Stakeholder Advisory Group (SAG)

Once the research questions were derived from the identified gaps in the literature, the questions for the semi-structured interviews were developed. These were informed by a small SAG of leading MND clinicians convened for this purpose. The SAG provided valuable insights into the experience and care of those with MND and of end-of-life clinical practices in a range of contexts and settings. This yielded information that was extremely useful when considering the possible reach of the study findings and recommendations both within and outside Australia.
Approval to convene the SAG as a process step in the current study was obtained from both the Wisdom Hospice in Rochester, Kent, United Kingdom (Appendix R) and Edith Cowan University Human Research Ethics Committee (Human Research Ethics Committee, Approval number 12099: Appendix B). Two aspects of clinician expertise were explored in the SAG conversations. The first aspect concerned explanations about NIV and how it was offered to and initiated for people with MND. The second aspect was about care, symptom control and ongoing communication relating to the end-of-life wishes of the person with MND and their family. Based on the information gleaned in these conversations, the nature and order of the clinician participant questions was refined to ensure relevance to the research aims and to reflect participants’ likely thought processes; this included the omission of two questions originally proposed that were found to be repetitive.

3.3. Significance of the Study

The literature suggests that for some families, end-of-life treatment options and the availability of palliating medication alternatives are not well communicated or comprehended, leading to an end-of-life decision-making crisis and potentially creating emotional indecision and confusion for families, significant others and the clinicians involved (MNDAust, 2012; NICE, 2010; NICE, 2016). The overall objective of this thesis is to determine to what extent the communication between health professionals and people diagnosed with Motor Neurone Disease (MND) incorporates symptom control and end-of-life choices. The significance of such a study is to ascertain whether recommended patient-centred best care communication is occurring and whether those involved with delivering and receiving the recommended communication consider this to be the best timing and most useful communication regarding end-of-life choices for people with MND. Importantly the communication experiences of those affected by a diagnosis of MND have been explored and compared to the clinician experiences. This study is significant as it identifies
areas for communication improvement and consequently the end-of-life care for those living with MND.

The findings of this study are presented in Chapter 4 (Results)

3.4. Section 1: Methodology

3.4.1. Positivism versus interpretivism

Health research is most commonly situated within one of two scientific paradigms depending on the philosophical stance of the researcher and on the research problem. Those who subscribe to the positivist worldview assert that there is one truth, and that research should measure and statistically “make sense of” data about phenomena of interest to reach that truth. Researchers working in this paradigm almost exclusively use quantitative methods to deduce “what is going on”, having first developed a hypothesis about the topic of focus. In contrast, the naturalistic-interpretive worldview takes the position that truth consists of multiple realities that arise through individuals’ constructs of it as a result of their experiences and interactions, and that the aim of research conducted within this paradigm is to explore and understand personal experiences and the meaning of social or individual situations to reach those truths (Burns & Grove, 2007; Streubert & Carpenter, 2011). Researchers who take a naturalistic-interpretive stance most usually subscribe to inductive qualitative methodologies in which no prior assumption is made about “what is” in relation to the topic of interest, but rather the understanding of the phenomenon of interest emerges from the data itself.

An inductive, learning-by-understanding technique was used for this study to analyse ideas and experiences as they emerged from clinician and bereaved caregiver answers. To ascertain the most effective method to capture the required information, several approaches were considered. As MND is a relatively rare disease some difficulty was anticipated in collecting enough data to enable research questions to be answered through quantitative means.
Therefore, a qualitative method was considered the most appropriate approach; although quantitative participant data may be limited the depth of qualitative interviews would likely be informative and personal (Wertz, 2011).

The study reflects the lived experience of the participants and relies on information collected from in-depth semi-structured interviews with clinicians and bereaved families of people with MND. Additional information, generously given by an individual diagnosed with MND who asked to be part of the research, offers valuable insight into the MND experience. This supplementary information provided a deeper understanding of the topic not anticipated in the original design of the research protocol. The research process for this study has four main steps with further steps within in each of these. The theoretical rationale (naturalistic-interpretive / qualitative), initially described within the Chapter 2, is expanded on further in this chapter.

Given the likelihood of an insufficient quantitative sample size and having considered the gap in knowledge and the research questions posed to address that gap, it was determined that a quantitative approach would be inappropriate for exploring the complexities of the communication that takes place around the emotionally-laden topic of end-of-life care options. Rather, the phenomenon under investigation demanded a naturalistic-interpretive / qualitative approach to enable the candidate to fully explore participants’ experiences and views. However, some quantifiable demographic data was collected to help demonstrate the representativeness of the sample, and this is presented within Chapter 4 (Tables 4.1, 4.2 and 4.3).
3.5. Overview of Predominant Theoretical Perspectives for Health Research and Their Epistemological Tenets

In selecting the most appropriate approach for this study, the following criteria were considered: of most importance, the methodology should enable the inquiry to achieve the most useful outcomes; it should also identify the techniques of the research; and the theoretical or conceptual perspectives should provide rational context and enlighten the methodology and epistemology, or the theory of knowledge. Several methodological options are available to the naturalistic-interpretive researcher (Crotty, 1998). Among others these include phenomenology, grounded theory, ethnomethodology (Crotty, 1998), and ID (Thorne, Reimer Kirkham, & MacDonald-Emes, 1997). The key features of the four qualitative perspectives considered for this study and their benefits and disadvantages in relation to it are outlined in Table 3.1.
Table 3.1. Four possible qualitative research methodologies for a study investigating end-of-life care decision-making in relation to MND

<table>
<thead>
<tr>
<th>Perspectives</th>
<th>Advantage</th>
<th>Disadvantages</th>
<th>Selected/Rejected</th>
</tr>
</thead>
<tbody>
<tr>
<td>Phenomenological Methodology</td>
<td>Based on the lived experiences as expressed by participants.</td>
<td>1) Includes imagination, emotion, thought, sensory perception but not specifically communication. 2) May be unaware of an experience if experienced by the semi-conscious.</td>
<td>Rejected: Personal experiences considered too imprecise for this research.</td>
</tr>
<tr>
<td>Grounded Theory Methodology</td>
<td>How social norms are investigated which when developed generally build a theory from specific gathered data.</td>
<td>1) Data gathered with intention of building a theory which was not necessarily the intention of this research. 2) Questions tend to be unstructured.</td>
<td>Rejected: Specific questions were required to be asked of participants and intent was not to seek a theory.</td>
</tr>
<tr>
<td>Ethnomethodology Methodology</td>
<td>Behaviour of a group may be changed by their social constructs.</td>
<td>1) Very specifically studying a group who perhaps may not even think about their behaviour as it is influenced by their social norms.</td>
<td>Rejected: Too specific to social groups and cultures.</td>
</tr>
<tr>
<td>Interpretive Description Methodology</td>
<td>Knowledge gained through the research can result in immediate change in practice. As a methodology, works in conjunction with SI.</td>
<td>1) Involves critical analysis and understanding of current clinical recommendations and guidelines (theoretical knowledge). 2) Results must be practical to enable change.</td>
<td>Selected: Provides information by understanding and therefore informing, by seeking personal experiences.</td>
</tr>
</tbody>
</table>

3.5.1. Phenomenology

Phenomenology is a philosophical method which studies the lived experience of individuals and how they interpret their experiences, originating from the work of Edmund Husserl (1889-1938) and variously developed creating a broad method for qualitative research (Streubert & Carpenter, 2011). Much of Husserl’s work was incorporated into lectures and research manuscripts rather than books. His ideas in developing phenomenology have been further incorporated into more recent literature, for example by the philosopher Martin Heidegger (Fieser & Dowden, n.d.). Phenomenology
describes an experience of an individual’s life which, when used as a method of research, gathers those experiences to describe and understand a particular occurrence (Streubert & Carpenter, 2011). The theory is based on the concept that events and objects in life create an individual’s sense of reality as they occur and as perceived by the individual (Edgar & Sedgwick, 2008).

Phenomenology emphasises an individual’s singular view of the world and does not attempt to reach a consensus view about phenomena of interest (Streubert & Carpenter, 2011). It was rejected for this reason. One of the key imperatives of this study was to discover information that could be used by all stakeholders in end-of-life care decision-making; it was, therefore, important to use an approach that would enable a consensus about what is going on to be reached. Phenomenology may be useful as a methodological option if MND was being explored to see whether the person with the disease thought that they may defined by the label of MND when perhaps they perceived the disease as only a small part of who they were. In this situation, phenomenology could be used to explore the individual’s feelings and mechanisms for coping with MND.

3.5.2. Grounded theory

Grounded theory is an approach developed in the 1960s by Anselm Strauss and Barney Glaser with a view to developing a theory by collecting data from interviews, observation and evidence relevant to the study. Grounded theory involves purposely selecting a group with similar lived experiences which may provide the researcher with contextual understanding of an event or circumstance (Schreiber & Stern, 2001). Historically, grounded theory developed from two schools of thought, one emerging from Chicago University and the other from Iowa University. As a student of Chicago University, Blumer (1969) described and developed social interaction theory, the philosophical underpinning of grounded theory. The concept of an individual taking meaning from oneself as well as through interactions with others, became the foundation of SI (De Chesnay & Banner, 2015). Using grounded theory as a research framework, the research questions are open-ended rather than specific,
allowing for the research to broaden and change direction depending on the data gathered (Maltby, Williams, McGarry, & Day, 2014). For this research, which requires specific information, a grounded theory methodological framework was considered too broad and ill-defined.

3.5.3. Ethnomethodology

Ethnomethodology, which was founded by Harold Garkinkel in the 1960s, is a methodological perspective derived from a collective of people or a social group who have unique or specific practices or ways of life (Maltby et al., 2014). In the context of MND this method could be used if the study focused on one particular group, and to study the group as a whole would enlighten all others who may be diagnosed with MND. Using ethnomethodology would not be suitable for this research, however, as I wanted to illuminate the phenomenon of interest from a range of sources.

3.5.4. Interpretive description

To understand the phenomena of interest, namely, the end-of-life/symptom control communication between clinicians and people with MND and their families, an inductive analytic approach was selected. Interpretive description was developed principally as a process for obtaining knowledge within the context of clinical events to elicit change (Thorne et al., 1997). Originally designed as an alternate research method to understand the subjective individual experience for clinical nursing research, ID has been developed over the past 20 years. Interpretive description starts with an analysis of current clinical knowledge and understanding. By purposeful sampling and capturing subjective insights by interviewing participants, themes develop which can inform clinical norms.

Interpretive description incorporates aspects from grounded theory and ethnomethodology in its approach to purposive data collection; however, it rejects the sometimes restrictive development of a theory (Thorne, Reimer Kirkham, & O’Flynn-Magee, 2004). How people act, interact and communicate
in their lives is the foundation of this research wherein what the individual considers to be true is uppermost; ID throws light on a collection of expressed truths to inform a practical change. For these reasons, ID was selected as the most appropriate methodology for the current study.

3.5.5. Theoretical perspective

The social interaction of interest in this study is the communication between the clinician and the person with MND and their family, and the interpretation of meaning ascribed to this communication as remembered by participants. By understanding and appreciating the meaning of events and participants’ interpretations of the communication engaged in, this study attempts to identify the factors that influenced the choices of symptom control and end-of-life care made by the person with MND. Symbolic interactionism was selected as the most appropriate theoretical perspective for this study. By listening and questioning people with experience of MND, understanding and meaning were created through an interpretive process.

Communication can be confusing and misinterpreted if not clearly delivered or left open to supposition. The understanding of words and the nuances of interactive verbal and non-verbal delivery can lead to communication problems and this in turn can change the meaning an individual may give to a situation. The theory of SI involves the interaction of the meaning individuals attribute to a situation or person, the language used to comprehend, and the thought which allows interpretation of meaning and language.

The methodology selected for this study, ID, and SI are highly compatible for the conduct of qualitative research studies that could effectively inform nursing practice. In fact, the underlying assumptions of ID are considered symbolic interactionist, and SI is considered an important part of ID’s theoretical heritage (Oliver, 2012).
3.6. Understanding the History of Symbolic Interactionism

The origin of SI is attributed to George Herbert Mead (1863-1931) despite the further development of the perspective by Mead’s student, Herbert Blumer. Symbolic interactionism focuses on the meaning and interpretation ascribed to communication and social and interpersonal interaction (Oliver, 2012). The basic principle of SI is that how an individual perceives the world and acts is based on their individual interpretation of the meaning of the influences on their actions (Blumer, 1969).

Since the inception of SI, there have been several variations emanating from different universities, including from philosophical leaders such as John Dewey and Charles Cooley from Chicago University. The University of Iowa developed their own variant of SI largely founded on the thoughts of Manford Kuhn and Carl Couch (De Chesnay & Banner, 2015). Kuhn differed from the Chicago school of thought, believing that there were some basic meanings for an individual which remained constant despite external influences (De Chesnay & Banner, 2015).

Mead was influenced by the work of Charles Darwin and the theory of evolution, sharing his belief that human behaviour is a constant adaptation to the environment (Mead & Morris, 1934). Mead considered that social interaction influenced the way in which humans learnt or were influenced to react (Mead & Morris, 1934). Within this social construct, Mead differentiated between mind as an essentially social process and the brain as a human organ.

Cooley was a sociologist who developed the concept of the “looking glass self”: how an individual perceives themselves is based on how a person believes others sees them (Cooley, 1998). The individual appears to others in a way that reflects how that individual believes they are seen, which in turn involves a perceived judgement based on appearance, and an imagining of how others feel about the individual. According to Cooley, this concept can lead to
behavioural change based upon the perception of others toward an individual (Cooley, 1998).

John Dewey, also from Chicago University, suggested that the way an individual responds to a situation relates to learned responses within a social order (Dewey, 1922). Dewey maintained that personal awareness of a social situation by attributing certain characteristics is a *habit* (trait) of social classification resulting in a personal judgement (Dewey, 1922). Dewey described mind as intrinsically involved with the environment in its ability to adapt behaviour. In particular, language and communication were identified by Dewey as responses to environmental factors (Dewey, 1922). The perception of instinct as a cause of social behaviour was rejected by Dewey who thought spontaneity affected old habits and that consequently thought deliberation could change a person’s behaviour (Dewey, 1922).

The development of SI at Chicago University throughout the 20th century continued to be based on the concept of reality as constructed by social interaction (Blumer, 1969). Following the teachings of Blumer, Straus commenced development of a research method based on the concepts of SI. Straus transferred to the University of California where he and Barney Glaser developed grounded theory in the 1960s (De Chesnay & Banner, 2015). Grounded theory is the structured collection and analysis of data which has the potential to lead to the development of a theory (Ezzy, 2013). Grounded theory investigates social normalities which when the data has been gathered and analysed enables a theory to be developed.

The intellectual theories, particularly emanating from the Chicago school of SI, have informed the choice of theoretical perspective used for this study. The exploration of communication between those involved with MND, the interpretation of this communication as heard and recalled, and the meaning assumed by the individuals all fit within the scope of SI. The meanings participants constructed, particularly from verbal communication, and to a lesser
extent, non-verbal communication, is presented throughout this thesis. This study has drawn on the SI concept of meanings influenced by situations and communication, and how individuals’ interpretations influence their life choices.

3.6.1. Limitations and examples of SI and ID in research

When SI was considered for this study, any reported limitations or criticisms of the approach were explored to ensure its appropriate use. The approach has been criticised as insufficient to address the influences of social structure, historical events, culture and power (Dennis & Martin, 2005; Meltzer, 1975). Symbolic interactionism has also been accused of not considering the influence of human emotions or the influences of organisations, both factors which have the potential to change human life (Meltzer, 1975). However, Dennis and Martin (2005) counter-argue that SI does explain the phenomena of power, citing studies in the areas of education and deviance where the consequences of power are enforced within social processes. Roles and behaviours can be influenced by circumstances and an individual can be defined by behaving in a way that is consistent with that circumstantial situation (Cast, 2003). Cast (2003) suggests that an individual may be able to influence the behaviours of people by accepting the influences of others. Cast gives an example of intravenous cannulation (IVC) and how children may be influenced by parents and health care providers who describe the impending situation. This has the potential of influencing the situational behaviour the child has ascribed to the meaning of the procedure. Cast used SI as a theoretical perspective to study and understand the communication between health care provider and parents throughout IVC procedure in children. On balance, SI was confirmed to be appropriate for the current study.

Interpretive description was similarly considered and was confirmed to be suitable on the basis of its application in other nursing research. In a study exploring the moral experience of health professionals in humanitarian work, the strengths and challenges of using ID as a qualitative method are discussed (Hunt, 2009). Using ID in his doctoral research, Hunt (2009) found coherent
structure to enable his research development and others have effectively employed this methodology in the exploration of nursing topics (Kimber, Georgiades, Jack, Couturier, & Wahoush, 2015; Nkulu Kalengayi, Hurtig, Ahlm, & Ahlberg, 2012; Thorne, Con, McGuinness, McPherson, & Harris, 2004; Williams & Haverkamp, 2015).

The broader interpretation of a situation and how it influences action was addressed by Blumer in his discussion of the importance of organisation on human society (Blumer, 1969). Blumer appeared to acknowledge aspects of a broad environmental influence and how aspects of it may influence human behaviour. An example in this research is the influence of the commonly held understanding of palliative care being a place of death and how this environmental aspect influences human behaviour. The compatibility between ID (framework) and SI (theoretical perspective) is used in the candidate’s research to understand and make meaning of the subjective participant insights and to potentiate change (Oliver, 2012). The following section incorporates and discusses SI within the research design, and how SI is used to understand the communication which influences end-of-life choices made by or for people with MND.

3.6.2. Theoretical perspective of SI within the research design

Initially using a perspective based on SI as a study of social interaction, concepts and behaviour, a construction for explaining and understanding each person’s decision-making process, and subsequent course of action relating to the initiation and withdrawal of NIV in people with MND was developed (Blumer, 1969; Stryker, 2002). For example, studying individual participant interpretation of communication surrounding NIV initiation, its benefits, burdens and limitations, and how this communication related to the experience of relief from respiratory failure and subsequent death in a person with MND, has provided participant shared agreement or non-agreement of what constitutes useful communication. Studying the content of communication and understanding among the participants at the time when the person with MND was offered and
either accepted or refused NIV, had NIV withdrawn or died of co-morbidities, has informed several aspects of this research. These include whether communication between participants resulted in similar memories and understandings of any discussions relating to commencement and withdrawal of NIV, and how their individual interpretation influenced their decisions.

Building upon the perspective of SI, and based on the literature review, a more prescriptive framework was developed to understand the complexities and ambiguities of NIV end-of-life communication. This enabled further refinement of the research questions, design, and analysis plan for this study. During data collection, the researcher found that while the theoretical perspective of SI provided a working guide, the interacting variables created the potential for verbal communication imprecision and misinterpretation. For example, a number of factors interacted to complicate communication: the complexity of the language used and the education level of participants, the capacity of the patient and family to question the clinicians, finding and interpreting information from the Internet by some and participants’ financial and social situations. The additional and often undisclosed or undiagnosed cognitive changes which can present in MND added further challenges for the person with MND, their families and clinicians in ensuring that what was said and heard was understood. Therefore, the theoretical perspective is grounded in an understanding of the unique experiences and individual interpretation of each participant.
3.7. Section 1: Summary

The first section of this chapter focused on the importance of selecting a methodology and theoretical framework. Various theoretical stances and methodological approaches have been examined and their advantages, disadvantages and relevance for this research identified. A subjective epistemology and SI were selected for this study as the theoretical perspectives and ID was selected as the framework for informing change. The development of a framework representing the influences on the phenomenon of interest was presented as the basis for the research questions. The research questions, in turn, were designed to enable the accumulation of participant perspectives which would inform change, broaden how communication may be delivered and influence the care choices of people with MND and their families. The following section of this chapter outlines how this qualitative study was designed.

3.8. Section 2: Methods

The research process is diagrammatically shown in Figure 3.1 which identifies the order of steps taken in the study once the first draft of the research questions had been developed.
Literature review; identify the knowledge gap and develop research questions

Apply Research Ethics approval

Slight amendment to clinician questions; no change to family participant questions

Clinic participants recruited via invitation through place of work

Reliability: Recorded interviews transcribed verbatim by professional transcriber

Clinic participants: collect data using semi-structured questions in the setting of their choice (generally their office at place of work)

Validity: transcripts returned to participants for veracity of content

Clinic participants: Data analyses using Nvivo and researcher coding of themes

Analysis

Family participants recruited via invitation through the MNDA (Australia) newsletter

Relate themes to original research questions and short summary of results to clinic participants

Family participants: collect data using semi-structured questions in the setting of their choice (generally their home)

Ensure reliability, validity, and transferability of research data: Discussion with limitations of research identified. Conclusion: recommendations and suggestions for further research

Family participants: Data analyses using Nvivo and researcher coding of themes

Relate themes to original research questions and short summary of results to family participants

Clinic participants recruited via invitation through place of work

Stakeholder Advisory Group

Clinic participants: Data analyses using Nvivo and researcher coding of themes

Family participants: Data analyses using Nvivo and researcher coding of themes

Figure 3. 1. The Research Process
3.8.1. Setting

The research was initially confined to interviews within Western Australia. However, due to insufficient participant numbers, the research was broadened to include New South Wales. In respect of the sensitive nature of the research, the clinician interviews all took place within the clinician participants’ workplace in a suitably private room, or via a telephone/Skype recorded interview. All family participants could determine a suitable venue of their choice and were encouraged to bring a support person along if they felt this was appropriate given the nature of the conversations. Only one of the participants had a support person present, and as this was another family member who had been involved with the care of the person with MND, they requested to be interviewed jointly. The family and caregiver interviews all took place in their respective homes, as chosen by each participant.

3.8.2. Qualitative data collection and analysis

A generic approach to qualitative data collection was employed for this study. Qualitative data collection is used for various lines of inquiry which allows the respondent the freedom to provide more than just a quantitative answer. For example, conducting open-ended and/or semi open-ended interviews allows the respondent to give detailed information in answering questions. This can add meaning or context to experiences and a deeper understanding of the way aspects of life work. These are important to inform and instruct process change that becomes transparent and accessible to the interviewer. The process of inquiry for qualitative research may be from interviews, observation or material collection such as questionnaires or analysis of official records. For this study, data were collected from semi-structured interviews with two types of participant. Data were analysed to ensure rigor, reliability and minimisation of researcher bias to enable others to replicate the research (Munn et al., 2014).

3.8.3. Semi-structured interviews

The interviews conducted for this study were intended to answer the research questions shown in Table 3.2. It was anticipated that by using a
personal semi-structured interview method, the researcher would be able to obtain in-depth responses. Semi-structured interviews have a flexible structure which, despite pre-organised questions, allow for detailed and personal answers and accounts (Streubert & Carpenter, 2011). Semi-structured interviewing requires that the researcher builds a trusting relationship with the participant, so the participant feels listened to and able to describe their experiences with the assurance of complete confidentiality.

The ability to observe the participants’ reactions and emotions when describing their recollections enabled the researcher to ensure the willingness and appropriateness of the participant continuing with the research questions. The observation of the participants and any degree of distress dictated whether the interview should pause or terminate, had the participant appeared emotionally distressed. Individuals were invited to rest or cease the interview and seek support through their place of work, or through the MNDAWA or MND NSW emotional support staff who kindly agreed to support this research. The researcher explained to the participants at the commencement of the interview that they may elect not to answer any of the questions they found distressing. It was also explained that they could choose to continue with the interview and seek support from MNDAWA or MND NSW or they could choose to continue with the research without seeking further support.

3.8.4. Development of the interview schedule

When considering the aims of this research, and after extensive exploration of the literature, the research questions (Table 3.2) were formulated and the most searching and pertinent participant questions constructed with the assistance of a SAG (Appendices D and E). The decision to interview participants was chosen in preference to a format of questionnaires. The candidate felt a more personal approach may elicit more insightful information, particularly as the subject matter was deeply personal and may cause some emotional distress (Streubert & Carpenter, 2011). The candidate was concerned that questionnaires may restrict the participant responses and whilst
a larger number of participants could have responded, the information may not have been as broad and informative. The candidate also felt that questionnaires may be given less in-depth thought by the participants and any clarification of questions or tangential thoughts would be less easy to obtain (Streubert & Carpenter, 2011).

3.9. Sample Selection: Participants

3.9.1. Clinician participant identification and recruitment

Purposeful sampling was used to identify and obtain the most pertinent group of participants. This method of sampling ensures that the participants targeted are representative of the information required for the research (Patton, 2015). In this case, clinicians involved with the effects of adverse respiratory symptoms in people with MND, clinicians who are involved with the initiation of NIV, those involved with end-of-life care for people with MND, and bereaved family/caregivers involved with people with MND were identified as best positioned to provide information about the topic of interest. By targeting and selecting participants with MND experience and with extensive knowledge of the research topic, optimal insight throughout the study was ensured. Purposive sampling and snowball sampling (information discussed and communicated to others not previously in the study, enabling relevant and interested others to participate) were used as a means of directing the research to the most relevant clinicians.

3.9.2. Clinician participants: inclusion criteria

Two participant groups were identified within the clinician participant cohort: those who were responsible for the conversation regarding initiation and use of NIV (respiratory physicians, or neurologists) and those who interacted with people living with MND in the terminal phase of care and were involved with withdrawal of NIV. Community neurological and/or palliative care nurses and allied health workers who provided ongoing support for the person with MND and their families were included as communicators of ongoing care. All
clinicians, whether at initiation or withdrawal of NIV in MND, were asked the same set of questions to ascertain any areas of assumed knowledge, or communication supposed to have occurred. The interview protocol included questions being asked in the same order for each participant, although because of the open-ended questions at the beginning of each interview, some parts of the questions were answered at that time and were not asked again. Protocol also demanded the consent of the participant for the interview to be recorded (Appendix F).

3.9.3. Clinician participants: recruitment of interview subjects

Clinician participants were identified through their association with MND in palliative care services, hospitals, specialist centres (neurologists, respiratory physicians and specialist nurses) and invited by email, letter or face-to-face to partake in the research. Any interested clinical participant was emailed or sent the Clinician Respondent Information Sheet (Appendix G) explaining the project and an Informed Consent Form containing further information regarding participants’ involvement (Appendix F). All participants were provided with a written letter explaining the purpose of the research, how the research was to be undertaken and how long each interview was anticipated to take. Prior to the interview commencing, participants were also provided with a written consent form which they were asked to read and sign. Confidentiality was assured through the process of de-identification of each interviewee and allocation of a code (for example P1). Only the researcher had access to the link between the identity of the participant and their code. Some of the participant quotes have been used in the thesis to capture sentences or statements to lend meaning to themes that have resulted from the research.

3.9.4. Family participants: recruitment of interview subjects

Bereaved families and caregivers involved with a person with MND were targeted through advertising in MNDAust newsletters (Appendix G) and word of mouth among the interviewees. A web-based invitation to participate was posted on the MNDA WA website and incorporated into their monthly newsletter
(Appendix H). The invitation was advertised twice and included details of the research project and the expected timeframe, methods of responding to the researcher if interested in participating and an assurance that participants’ privacy would be protected. As participant numbers were low in Western Australia, an application was made to the ECU Human Research Ethics Committee to extend the research via both MND NSW and interested doctors to disseminate details of the research to bereaved caregivers. Once an interested caregiver participant contacted the candidate, a letter/email with a Family Respondent Information Sheet explaining the project (Appendix J) and Informed Consent Form (Appendix F) containing further information regarding participants’ involvement was sent to the participant. Entry into the study only occurred after verbal and written informed consent had been confirmed. The initial research design did not incorporate people with MND as it was thought that the candidate’s questions relating to end-of-life decisions may cause distress. However, a person with MND who was using NIV who wished to explain and discuss his experiences contacted the candidate and asked to be included. An amended ethics application and subsequent approval from the Edith Cowan University Human Research Ethics Committee (ECUHREC) was obtained (Appendix C). Therefore, one interview was conducted with a person with MND. These data were used to illuminate and contextualise the two primary data sets.

A convenient appointment date and time was subsequently organised by the candidate in response to each potential participant’s initial responding email, with further communication with the participant via their preferred method of email or telephone. It was anticipated this invitation would be discussed between family members and significant others with experience with MND and NIV who might have wished to participate and share their communication experience producing a snowball effect. This approach produced one further family member participant. Within this cohort, there were six respondents and one person with MND wishing to assist the research.
3.9.5. Clinician participants profile: initiation of NIV

The first cohort of interview participants were respiratory clinicians, respiratory nurses and social workers responsible for the conversations regarding use of NIV and palliative care options, specifically at the time of commencing NIV when respiratory muscles of the person with MND weaken causing breathing difficulties (e.g., when NIV is offered to alleviate distressing symptoms). A gastroenterologist was incorporated into the study and asked the same set of questions as all the clinicians regarding communication surrounding gastrostomy tube placement and how this communication related to NIV commencement. Clinicians from the first group were asked what specific information they communicated to people with MND about the benefits, limitations and potential withdrawal of NIV; PEG insertion; when they discussed the involvement of palliative care and end-of-life choices; and how they made sure that the information they had given the person with MND and their families, had been understood. Eight respondents were interviewed in this cohort.

3.9.6. Clinician participants profile: withdrawal of NIV

Clinicians from the second group - palliative care specialists, allied health (generally as part of an MDT) and nurses involved with palliative care or neurological nursing - were asked to recall episodes of care for people with MND where NIV was withdrawn during the terminal phase of the disease. Of specific interest in relation to this group were their observations and feelings surrounding the withdrawal of ventilation support in people with MND and how this affected the caregivers and families. They were asked to reflect on their perception of the preparedness of families and significant others in relation to understanding the outcome of withdrawing NIV, even if not directly involved with the withdrawal. The clinician participants were also asked to reflect on their feelings the preparation and communication from an MDT had on their capacity to provide holistic support to the families and significant others at the time of withdrawal. Any difficulty in people with MND accessing an MDT or palliative care specialist was noted. Within this cohort of participants there were 11
respondents interviewed. Two of the first cohort, those responsible for the initiation of NIV, were also in a team of clinicians responsible for the removal of NIV; however, only one of the palliative care clinicians had referred a person with MND for NIV prior to the terminal phase.

3.9.7. Family/caregiver participants profile

Families and significant others, who have lived with a person with a diagnosis of MND, were asked to recall their experience(s) of the conversations which may (or may not) have occurred prior to the commencement (or refusal) of NIV and communication relating to placement of gastrostomy tubes for their family member diagnosed with MND. They were encouraged to reflect on how these conversations may (or may not) have prepared them for the outcome associated with the choice to initiate or not to initiate NIV, the limitations of NIV as explained to them, end-of-life discussions as they occurred and withdrawal of NIV at the terminal phase of care. Six bereaved family participants were interviewed, in one instance a brother and sister interviewed together (F1 and F2).

3.10. Data Collection: Clinician and Family Participants

Data was obtained using individual semi-structured interviews from the three groups of respondents outlined above, each of whom were asked to consider two points in time along the disease trajectory: the first point was when the respiratory muscles of the person with MND weakened and breathing became difficult and NIV is offered and the second point was when NIV is withdrawn. As discussed in Chapter 1, communication relating to PEG placement and the commencement of NIV are closely linked and are recommended to occur prior to respiratory distress (NICE, 2016). Therefore, questions relating to PEG insertion and use were incorporated within the semi-structured questions.
The long, in-depth interviews were recorded with the consent of each participant and transcribed verbatim by an appointed professional transcriber (transcription confidentiality agreement: Appendix Q) who identified the participants by codes known only to the researcher. Statistical analysis and demographic data were collected and collated: clinician roles, MND years of experience, whether working within an MDT and familiarity with the most recent NICE (2016) MND Assessment and Management Guidelines. Four of the interviews (three clinicians and one family participant) were conducted via recorded audio/visual SKYPE interviews. There were no interruptions or disturbances during these interviews and they were all transcribed verbatim. Both telephone and the face-to-face individual interviews lasted for between 60-90 minutes. The interviews were terminated when the participant indicated they had nothing further to add. A thank you email was sent to each participant within 24 hours.

In considering the methodology, research questions (Table 3.2.) and participant semi-structured questions, the research method was developed to obtain the most accurate and dependable information.
<table>
<thead>
<tr>
<th>Research Question</th>
<th>Strategy and Method</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. In considering the available literature and guidelines, what are the most</td>
<td>Extensive search of published literature through databases, guidelines and recommendations, reference lists in published articles and conference material. Answered in Chapter 2.</td>
</tr>
<tr>
<td>salient components of best practice relating to the communication about NIV in</td>
<td></td>
</tr>
<tr>
<td>MND prior to NIV initiation and withdrawal?</td>
<td></td>
</tr>
<tr>
<td>2. What is the evidence that the best practice communication content (benefits and</td>
<td>Search of Australian and International MND guidelines relating to MND and symptom control particularly relating to end-of-life care.</td>
</tr>
<tr>
<td>burdens of NIV, continued disease progression, explanation and offer of a PEG,</td>
<td>Semi-structured interviews with clinicians a) to understand what, how and when recommended information is being communicated at the time of respiratory failure and how clinicians assess patient understanding of communication and b) what the clinicians involved at the end-of-life experience with patient and family comprehension of prior recommended communication</td>
</tr>
<tr>
<td>end-of-life discussions and understanding potential NIV withdrawal prior to</td>
<td></td>
</tr>
<tr>
<td>commencement of NIV) is incorporated into the reported communication by clinicians</td>
<td></td>
</tr>
<tr>
<td>by clinicians to people with MND and their families at the time of offering NIV?</td>
<td></td>
</tr>
<tr>
<td>3. What do MND families/significant others recall about the communication (benefits</td>
<td>Semi-structured interviews including ‘their story’ with families to understand the communication as heard and understood at the time of respiratory decline particularly relating to end-of-life</td>
</tr>
<tr>
<td>and burdens, continued disease progression, PEG insertion, potential NIV</td>
<td>Included within this research was a person with MND describing his experience with communication</td>
</tr>
<tr>
<td>withdrawal and end-of-life discussions) provided by clinicians at the time of</td>
<td></td>
</tr>
<tr>
<td>respiratory failure and the offer of NIV?</td>
<td></td>
</tr>
<tr>
<td>4. To what degree do MND families/significant others feel that the NIV communication</td>
<td>Semi-structured interviews with the bereaved families and caregivers at least 3 months post death of a person with MND</td>
</tr>
<tr>
<td>provided by the clinicians at the time of the offer of NIV prepared them for and</td>
<td></td>
</tr>
<tr>
<td>helped them to understand the implications of potential NIV withdrawal resulting</td>
<td></td>
</tr>
<tr>
<td>in the death of their family member?</td>
<td></td>
</tr>
<tr>
<td>5. What, if any, unmet needs for information, support, palliative care, or other</td>
<td>Suggestions made in answer to an open question asking participants of their experiences and recommendations</td>
</tr>
<tr>
<td>services do the clinicians and the families/significant others (or patients</td>
<td></td>
</tr>
<tr>
<td>themselves) with experience of MND identify?</td>
<td></td>
</tr>
<tr>
<td>6. Following the literature review and clinician and family/significant other</td>
<td>Reviewing the data and comprehensive analysing and coding of the interviews to identify and clarify any recommendations verbalised by participants. This area of the study has identified limitations of the research and areas which may be considered for further research</td>
</tr>
<tr>
<td>interviews regarding NIV communication, what recommendations can be identified</td>
<td></td>
</tr>
<tr>
<td>that would improve end-of-life communication and process for the patients,</td>
<td></td>
</tr>
<tr>
<td>families and clinicians involved with the end-of-life care for people with MND?</td>
<td></td>
</tr>
</tbody>
</table>
3.10.1. Bracketing and reflexivity

An ability to reflect and be self-aware has assisted the candidate to minimise researcher bias. By understanding the social, educational, cultural and personal influences in her own life, a generally more empathetic stance has been taken on interviewing techniques and analysing data (Patton, 2015). Personal perspectives and preconceived ideas, where they existed, were put aside, and the data collected was analysed without prejudice. The researcher made every attempt to exclude researcher bias by using the method of "bracketing" (Patton, 2015). This involves a process by which constant reflection (reflexivity) by the researcher and discussion with university supervisors, reduces and tries to eliminate any researcher preconceptions. The credibility of the candidate's research is inextricably linked to the researcher's ability to justify and analyse the evidence without bias.

3.11. Data Handling and Management

3.11.1. Data storage

The digital recordings obtained during the interview phase of this research were transcribed from the digital recorder and onto the researcher’s password protected computer which remained locked in the candidate’s home office. The recorded interview data were erased once transcribed onto the candidate’s password protected computer. As per NHMRC guidelines, all hard copy data collected for this project and records of any work associated with this research have been kept securely locked in a designated area in the student’s home or at Edith Cowan University retained on the candidate’s password protected computer. At the completion of the research transcripts and analysis data will be kept for five years (as per NHMRC (2012) requirements) on an Edith Cowan University password protected computer which is located inside a locked office that is in turn located in a swipe card protected area.
3.12. Data Analysis

Descriptive demographic information was collected for analysis, including clinicians’ specialisation and experience, whether part of an MDT and the number of NIV withdrawals and MND deaths the clinicians had been involved with. The gender and age of the person with MND as stated by the family participant, and whether the person with MND was seen by an MDT is included in information for this research. The tabulated data are presented in Table 4.1 in the discussion chapter of this thesis as an overview of participant demographics and distribution of specialised services.

Data analysis commenced following the first participant interview, with relevant and emergent themes noted. The NVivo10 software package was used to assist this process. All the interviews were de-identified, coded and transcribed verbatim. The quantifiable raw data were tabulated and are contained within the results in Chapter 4 (Tables 4.1 and 4.2).

3.12.1. Stage 1 analysis: coding

The first phase of analysis included the identification of codes, or phrases in the interview transcripts that related to the research questions. These were colour coded on the printed verbatim transcripts, examples of which are provided in Table 3.3. and contained within the appendices of this thesis (Appendix M). These phrases were then extracted and alike codes were grouped (Stage 2). Several exemplar codes are used to illustrate the findings in Chapter 4.
Table 3. Example of first level analysis

<table>
<thead>
<tr>
<th>Raw data</th>
<th>Level 1 codes</th>
</tr>
</thead>
<tbody>
<tr>
<td>1) So his wife wanted a definite sign of when she would stop the PEG fees, if he was no longer able to communicate. I haven’t seen the Advance Health Directive, but she did say there was a special sign that they would have.</td>
<td>1) Hydration and nutrition; discussions about disease progression and timing of gastrostomy</td>
</tr>
<tr>
<td>2) No. I think there’s very little work done around AHDs, because there is just not enough knowledge, experience or time.</td>
<td>2) Advance health directives</td>
</tr>
<tr>
<td>3) They really do need to think about withdrawal of NIV, which I don’t know is discussed with clients in detail, or if it is they’re not processing that information. I’m not involved in any of those discussions, so I really don’t know what has been said. But when I have asked if that has been discussed, people have said, “No.”</td>
<td>3) NIV withdrawal; is this discussed at the time of commencement of NIV prognosis and limitations of treatment included in ongoing honest discussions timing of end of life discussions.</td>
</tr>
</tbody>
</table>

Table 4. Example of second level analysis

<table>
<thead>
<tr>
<th>Codes</th>
<th>Sub-category</th>
</tr>
</thead>
<tbody>
<tr>
<td>1) NIV initiation; is end of life care discussed at this time</td>
<td>1) Family satisfaction with end-of-life care</td>
</tr>
<tr>
<td>2) NIV benefits, burdens and limitations discussed including increased dependency</td>
<td>2) Other clinicians involved</td>
</tr>
<tr>
<td>3) NIV withdrawal; is this discussed at the time of commencement of NIV</td>
<td>1) Patient and family understanding</td>
</tr>
<tr>
<td></td>
<td>2) Experience of refusal of NIV</td>
</tr>
<tr>
<td></td>
<td>1) Withdrawal experience</td>
</tr>
<tr>
<td></td>
<td>2) Tracheostomy as emergency procedure by ED</td>
</tr>
</tbody>
</table>

3.12.2. Stage 2 analysis: sub-categorising

The second phase of analysis involved grouping of alike codes into sub-categories. The points at which no new sub-categories were emerging from participant interview codes was when initial saturation of the data was assumed, and no further interviews took place once each sub-category was sufficiently “thick” (representative of the sample). Table 3.4. provides an example of the sub-categorising second stage of analysis, and additional examples are provided in Appendices K and L.

Table 3. 4. Example of second level analysis
A list of all the sub-categories and categories that emerged from this stage are provided in Table 3.5. below.

Table 3. 5. Categories and sub-categories included in data analysis

<table>
<thead>
<tr>
<th></th>
<th>1) Advance health directives</th>
</tr>
</thead>
<tbody>
<tr>
<td>2)</td>
<td>Hydration and nutrition; discussions about disease progression and timing of gastrostomy:</td>
</tr>
<tr>
<td></td>
<td>hydration</td>
</tr>
<tr>
<td></td>
<td>how is this described</td>
</tr>
<tr>
<td>3)</td>
<td>Multidisciplinary team:</td>
</tr>
<tr>
<td></td>
<td>responsibility for overall care</td>
</tr>
<tr>
<td></td>
<td>interdisciplinary communication</td>
</tr>
<tr>
<td>4)</td>
<td>NIV benefits, burdens and limitations discussed including increased dependency</td>
</tr>
<tr>
<td></td>
<td>patient and family understanding (NIV): Guideline recommendation (NICE, 2016)</td>
</tr>
<tr>
<td></td>
<td>experience of refusal of NIV</td>
</tr>
<tr>
<td>5)</td>
<td>NIV initiation; is end of life care discussed at this time: Guideline recommendation (NICE, 2016)</td>
</tr>
<tr>
<td></td>
<td>other clinicians involved</td>
</tr>
<tr>
<td></td>
<td>family satisfaction with end-of-life care</td>
</tr>
<tr>
<td>6)</td>
<td>NIV withdrawal; is this discussed at the time of commencement of NIV: Guideline recommendation (NICE, 2016)</td>
</tr>
<tr>
<td></td>
<td>prognosis and limitations of treatment included in ongoing honest discussions</td>
</tr>
<tr>
<td></td>
<td>timing of end of life discussions</td>
</tr>
<tr>
<td></td>
<td>withdrawal experience</td>
</tr>
<tr>
<td></td>
<td>tracheostomy as emergency procedure by ED</td>
</tr>
<tr>
<td>7)</td>
<td>Prognosis and limitations of treatment included in ongoing honest discussions</td>
</tr>
<tr>
<td></td>
<td>introduction to palliative care (Symbolic Interactionism: influence and understanding)</td>
</tr>
<tr>
<td></td>
<td>ethical dilemma</td>
</tr>
<tr>
<td>8)</td>
<td>Understanding and obtaining the MND diagnosis family perspective</td>
</tr>
<tr>
<td></td>
<td>symptoms of frontotemporal changes</td>
</tr>
<tr>
<td></td>
<td>age at diagnosis</td>
</tr>
<tr>
<td>9)</td>
<td>Timing of end of life discussions</td>
</tr>
<tr>
<td></td>
<td>barriers to clear communication</td>
</tr>
</tbody>
</table>
3.12.3. Stage 3 analysis: categorising
Once all sub-categories were developed and saturated (no new information found from the interviews), those that resonated with or reflected with others were collapsed into major categories. Table 3.5 provides the list of the final major categories and their component sub-categories.

3.13. Trustworthiness

3.13.1. Trustworthiness of the research candidate
The candidate conducting the interviews is an experienced registered nurse working in palliative care, where sensitive communication and interviewing of both patients and families is a vital skill. The candidate understood and had the ability to separate the two roles involved with palliative care nursing and research interviewing: to be empathetic, reflective and understanding of the participants’ experiences whilst remaining supportive and respectful in the researcher role. The researcher had read extensively on sensitive interviewing techniques and followed appropriate guidelines on interviewing (Patton, 2015). Of concern, was the potential for participants, particularly the clinician participants, to supply information thought to be useful to the researcher’s background as a palliative care nurse. Whilst this may impede complete data accuracy and transferability, the researcher also recognised that understanding clinical terminology and a familiarity of clinical issues may have enabled greater engagement with the participants. To ensure that data collection and analysis was conducted as objectively as possible and reflected the research questions, the candidate and her supervisory team had regular discussions regarding interpretation of the data. To verify the candidate’s results, independent analysis of a selection of interview transcripts was conducted by one of her supervisors. Comparing the findings of both the candidate and the supervisor found the themes to be compatible.
3.13.2. Research credibility

Guba and Lincoln (1994) suggested four criteria, which if adhered to, should confirm the rigor of research: credibility, dependability, confirmability and transferability. The trustworthiness, or credibility, of the research was aided by clarity of the questions asked of the participants, which were designed to encourage the participants to describe the conversations surrounding the initiation, use and ultimate removal of NIV in patients with MND (Streubert & Carpenter, 2011). The professionally transcribed interviews (Appendix Q) were returned to the participants to ensure validation and approval for use in the data analysis and to establish trustworthiness and credibility of the researched information (Polit, Beck, & Hungler, 2001; Pope & Mays, 2006; Streubert & Carpenter, 2011). Analysis of a sample of the interviews was undertaken by one of the candidate’s supervisors post analysis by the researcher, in order to verify credibility of the results. Data and supporting documentation allow for an audit trail ensuring dependability of the research. The audit trail includes interview transcripts, analysis of data, notes relating to the process and any difficulties encountered with the method, personal notes and all copies of drafts relating to the final work presented (Guba & Lincoln, 1994; Polit et al., 2001; Pope & Mays, 2006). Confirmability was conferred on the study by an independent reviewer’s verification of the data analysis decisions and research audit trail. Further confirmability will be assured by other researchers substantiating the findings as a result of their own research in the future. Finally, the transferability of the outcomes of the research is proven through the resonance of the findings with clinicians from a range of practice contexts through several member-checking opportunities. These included sharing the finding that communication between healthcare providers and people with MND can be poor and potentially affect an individual’s decision for end-of-life care. The finding in this research that families find multiple healthcare providers to be confusing and to affect their communication and decisions has been discussed by the candidate in her work setting with agreement from palliative care patients and families alike.
3.14. Ethical Considerations

3.14.1. Ethics approvals and considerations

The Human Research Ethics Committee at Edith Cowan University (ECUHREC) (Number 12099: Appendix B) approved commencement of participant interviews in June 2015. Subsequently, ethics approvals were sought from Silver Chain Hospice Nursing (Appendix N), St John of God Hospital Bunbury and Murdoch, WA (Appendix O) and the Neurological Council of WA (Appendix P) and granted in mid-2015. Ethics approval from ECUHREC was further obtained to include data from people with MND who volunteered to participate after two people diagnosed with MND responded to the advertisement in the MNDAWA newsletter targeted at bereaved family members. The participants wished to tell their stories to assist others with MND, and ethics approval was subsequently granted to allow their experiences to be heard (Appendix C). Only one of the people with MND was ultimately interviewed. The second potential MND participant was considered too unwell and emotionally fragile to participate without the possibility of causing the participant further adversity. As a means of increasing participant involvement, subsequent ethics approval was sought and granted from ECU to include clinicians from New South Wales via snowball effect, word of mouth and email information between clinicians. All amendments to the original ethics application to ECU are included in Appendix C.

3.14.2. Ethical considerations: bereaved caregivers

Recalling conversations surrounding the initiation and withdrawal of NIV may prompt mixed memories for the caregivers, with feelings of sadness, anger and loss. Consideration was given to the potential for feelings of profound sadness and grief during the interviews with the bereaved families/significant others. The candidate was particularly aware of any possible emotional ill effects the one person with MND who chose to be interviewed for this study may have suffered. The style of interview differed from the other participants. The interviewee told his story and was periodically prompted rather than using
the schedule of semi-structured questions. The interview was conducted at a pace which suited the participant and finished when indicated by the participant.

The candidate ensured that the person with MND and bereaved family participants were all clients of the MNDAWA and had access to pastoral and psychological support as part of the service this organisation provides. While funding for this service provision is limited until six months after the death of the person diagnosed with MND, the Association agreed to provide this service to any bereaved caregiver participants from this research that may be outside the technically funded bereavement period. All bereaved caregiver participants and the one person with MND were provided with written information about the support offered by MNDAWA and support from the more generic support counselling service offered by Lifeline at the conclusion of the interview. This was to provide support to those who may have felt inclined to ask for emotional assistance well after the interview.

The ethical considerations for researching patient-focused palliative care include its acceptability for patients and families at the end of life, since participation in research may create further intrusion and distress at such a difficult time. For people with MND, this ethical predicament may be compounded by cognitive changes and difficulty with communication particularly towards the end of life. For the bereaved families, timing of the interviews was considered and a precedent in published research was sought. In a study which specifically investigated interviewing bereaved families of MND and cancer patients, the timing of interviews after the death of their family member was found to be an individual and crucial factor (Bentley & O’Connor, 2015). Bentley and O’Connor (2015) found that offering choice in timing of the research interviews post death of a family member enabled appropriate arrangement to be made for each individual family and led to their willingness to participate (Bentley & O’Connor, 2015). Therefore, when considering timing and the need to be respectful of the grieving process, the bereaved participant interviews for
this thesis were conducted when the bereaved families contacted the researcher, but all more than three months post death of their family member.

3.14.3. Ethical considerations: clinician participants

Clinician participants involved with the decline of the respiratory muscles and the initiation of NIV (respiratory clinicians and allied health), and those providing end-of-life care to people with MND (palliative care specialists, allied health practitioners and nurses) were interviewed regarding the content and timing of the communication surrounding the initiation of NIV in people with MND and any experience of NIV withdrawal. Whilst this difficult conversation may include end-of-life and withdrawal of NIV issues, the research questions specifically related to the clinicians normal practice protocol. Therefore, a low level of discomfort was anticipated and found in this group. The group of clinicians specifically responsible for end-of-life and palliative care was interviewed regarding their experiences relating to MND and the withdrawal of NIV, including the subsequent death of the patient and the preparedness of the family/significant other for the outcome. The interviewing of this group of clinicians was anticipated to elicit varied feelings associated with sadness, frustration and loss, some of which were captured within the interviews but none of which caused distress to the participants. All clinician participants were reminded of their Employer Assistance Programs (free counselling programs offered by health services) through which participants are offered support and an opportunity to debrief. All participants were offered written information on Lifeline, a counselling support service which they were encouraged to access at their convenience.

3.14.4. Consent

Permission was sought from participants to undertake recorded interviews and discussions, with consideration to a participant’s state of health and mind throughout the research. It was emphasised at the commencement of the research that participants can decide to withdraw from the project at any time they chose, and that participants’ wellbeing took precedence over the
research. The participants were supplied with information regarding the structure and purpose of the research (Appendix J), information on free counsellor support and examples of the potential questions that may be included at the recorded interviews (where requested) enabling the participants to make an informed decision as to whether to participate in the research. The candidate ensured all participants had signed consent prior to the commencement of the interview (Appendix F).

Whilst it may be supposed that some unanticipated information may be forthcoming within the interviews, the participants were reassured they were under no obligation to proceed with the interview and may stop the discussion at any time. In this situation, the researcher would re-confirm consent from the participant as an ongoing process to ensure the willingness of the participant to participate, particularly if considered to be in a vulnerable condition or situation (Oliver & Faull, 2013). As the research was obtained from families and significant others some time post death of a family member with MND, the information obtained for the research was reliant on the memories and experiences of the family members, caregivers and clinical team prepared to participate in the study.

3.15. Section 2 Summary

In Section 2 of this chapter, the method used to recruit the participants for this research has been explained. The researcher aimed for sufficient participant representation through an indirect approach to ensure that those participants who became involved did so completely voluntarily. Therefore, an advertising and/or word of mouth approach was directed electronically via websites, newsletters or general information email toward the desired participant groups. As MND is a rare disease the sample population is limited, however, the small number of bereaved caregiver participants volunteering for this research is felt to be usefully representative. Another possible limitation to the study sampling (further identified and discussed in Chapter 6) may be those
without electronic information access or those not supported through the Neurological Nurses (Neurological Council of WA) who snowballed the invitation to participate in the research through their network of nurses.

The research method of an extensive literature review and in-depth qualitative interviews has started to address the gap in understanding the communication which is recommended to occur at the initiation of NIV. This has been achieved by comparing the findings of this research with the present MND assessment and treatment recommendations concerning when and what should be included in clinician communication. The clinician interview information has been compared with the understandings and recollections of bereaved families regarding the information given to them relating to their family member when symptom control measures were discussed, implemented, refused or withdrawn.

3.16. Chapter Summary

Clear communication is required for people with MND and their families so they can make informed end-of-life decisions. This chapter has outlined the method by which this research has undertaken credible and reproducible inquiry into the communication surrounding NIV by interviewing selected participant groups involved with MND.

The complexity of the communication recommended (NICE, 2016) to occur at various trigger points along the MND trajectory by the clinicians, the link between commencing NIV and its potential withdrawal and death, and the way in which such communication may be interpreted by those who hear it are all necessary to consider. The emotional and ethical considerations necessary to conduct research into the end-of-life options, particularly when considering removal of NIV, have also been discussed. The theoretical perspective of SI and the Domains of Influence framework derived from the existing literature on
the topic of interest are used to present and contextualise the findings of this thesis in Chapter 4 and discussed more fully in Chapter 5.
Chapter 4

Findings

4.1. Chapter Overview

In this chapter, the results from 26 participant interviews is presented in relationship to the research questions and the NICE MND Assessment and Management Guidelines (NICE, 2016). Seven major themes emerged from analysis of the data. These are identified in Section 4.4 of this chapter, expanded upon throughout the chapter and discussed in Chapter 5.

Quotations from participants are included where relevant to illustrate the results, and the context of the quotes is explained within the narrative. The substantial amount of information gathered from the in-depth participant interviews not only sheds light upon the research questions but provides insight into a range of associated issues. At times information not directly relating to the research questions is included where it helps to clarify and broaden the understanding of end-of-life and NIV communication in MND.

The communication difficulties and factors encountered in relation to the research questions are discussed under specific headings derived from the themes identified. The significance of the influences on the participant data analysis and outcomes identified are further considered in the Chapter 5. This chapter commences by presenting the background to participants and their involvement.
4.2. Background to the Family Participants

The seven family participants (n=7) included a brother and sister interviewed together (F1, F2), in accordance with their preference. At times the brother and sister had different recollections on specific issues and different viewpoints, making the interview particularly useful in providing depth and perspective. Four other family participants (F3, F4, F5, F6) who had experienced the death of a family member with MND were interviewed. The seventh participant (P1) was diagnosed with MND and actively sought participation. While people with MND were not the target of this study, P1 thought his experiences should be heard to assist others diagnosed with the disease, and ethics approval was subsequently granted (Appendix C). As P1 was the only person interviewed with MND, his comments have been included with the bereaved family participants. All the family participants were from Western Australia (n=7). They were asked a range of questions (Appendix E) with attention directed toward their understanding of symptom control, end-of-life choices and their involvement with palliative care.

Table 4.1 describes family participants in terms of their level of health knowledge, use of the Internet for health information, their rural/remote classification, the primary clinical responsibility for overall care and whether their care was provided as part of a structured MDT. Health literacy was not the focus of this study and, therefore, was not formally assessed. However, the participants themselves acknowledged use of the Internet where they had access to it, and if they were conversant with health and health systems. Emphasis was placed on creating a good rapport with participants to help discuss and understand their experiences.
Table 4. 1. Family Participant Demographics

<table>
<thead>
<tr>
<th>Family Participants</th>
<th>Health knowledge and/or health information from internet</th>
<th>Regional, rural or remote</th>
<th>Primary clinical responsibility for overall care</th>
<th>Part of structured MDT</th>
</tr>
</thead>
<tbody>
<tr>
<td>F 1/2</td>
<td>No</td>
<td>Regional/rural</td>
<td>Unclear: GP and neurologist</td>
<td>No</td>
</tr>
<tr>
<td>F 3</td>
<td>Yes</td>
<td>Regional</td>
<td>Neurologist in association with GP</td>
<td>Yes</td>
</tr>
<tr>
<td>F 4</td>
<td>No</td>
<td>Rural</td>
<td>GP</td>
<td>No</td>
</tr>
<tr>
<td>F 5</td>
<td>Yes</td>
<td>Remote</td>
<td>GP</td>
<td>No</td>
</tr>
<tr>
<td>F 6</td>
<td>Yes</td>
<td>Rural</td>
<td>GP</td>
<td>No</td>
</tr>
<tr>
<td>P 1</td>
<td>No</td>
<td>Regional</td>
<td>Neurologist</td>
<td>Intermittently</td>
</tr>
</tbody>
</table>

Note: General Practitioner (GP), Multidisciplinary team (MDT)

4.3. Background to the Clinical Participants

The clinicians involved in this study were from two cohorts of specialties: those involved with respiratory decline and symptom intervention (NIV/PEG) and those involved with palliation and end-of-life care. The two clinician participant groups were further subdivided into doctors, nurses and allied health providers. The medical participants included four palliative care specialists, one rehabilitation specialist, three respiratory specialists and a gastroenterologist. The nurse participants included four MND/neurological specialist nurses, three palliative care nurses and a respiratory clinical nurse specialist. The two allied health participants were a social worker and a respiratory physiotherapist, both of whom worked with people with MND within an organised MND specific MDT. At the time of interview, the participants worked in Western Australia (n=8), New South Wales (n=9) and the United Kingdom (n=2). Table 4.2 shows a summary of participants, their de-identifying codes and professions. Also included are the
clinicians' years of experience working with people with MND and the percentage of their work involving people with MND. Table 4.1 also shows whether the clinicians worked in regional, rural or remote areas and whether they were involved with NIV withdrawal and end-of-life discussions.

There are similarities and differences between clinical specialties regarding views about when and who should have the discussions about the initiation and withdrawal of NIV and end-of-life choices. Both clinician cohorts in this study had experience with the withdrawal of NIV.
Table 4. 2. Clinician Participant Demographics

<table>
<thead>
<tr>
<th>Clinician</th>
<th>Specialty</th>
<th>Years (y) of MND experience and workload</th>
<th>Involved with NIV withdrawal discussions</th>
<th>Regional, rural or remote</th>
</tr>
</thead>
<tbody>
<tr>
<td>C1</td>
<td>Palliative Care Consultant</td>
<td>10+ y Varies</td>
<td>Yes</td>
<td>Regional and rural</td>
</tr>
<tr>
<td>C2</td>
<td>Palliative Care Specialist</td>
<td>10 y Varies</td>
<td>Yes</td>
<td>Regional</td>
</tr>
<tr>
<td>C3</td>
<td>Palliative Care Specialist</td>
<td>17 y Varies</td>
<td>Yes</td>
<td>Regional</td>
</tr>
<tr>
<td>C4</td>
<td>Respiratory Specialist</td>
<td>12 y Varies</td>
<td>Sometimes</td>
<td>Regional and rural</td>
</tr>
<tr>
<td>C5</td>
<td>Respiratory Specialist</td>
<td>6 y 75%</td>
<td>Yes</td>
<td>Regional and rural</td>
</tr>
<tr>
<td>C6</td>
<td>Neurological Nurse</td>
<td>4.5 y Intermittent</td>
<td>No</td>
<td>Regional and rural</td>
</tr>
<tr>
<td>C7</td>
<td>Neurological Nurse</td>
<td>8 y Intermittent</td>
<td>Indirectly; when patient requires information</td>
<td>Regional and rural</td>
</tr>
<tr>
<td>C8</td>
<td>Palliative Care Nurse</td>
<td>16 y Intermittent</td>
<td>Yes</td>
<td>Regional</td>
</tr>
<tr>
<td>C9</td>
<td>Palliative Care Nurse</td>
<td>14 y Intermittent</td>
<td>Yes</td>
<td>Regional</td>
</tr>
<tr>
<td>C10</td>
<td>Palliative Care Nurse</td>
<td>5 y Intermittent</td>
<td>Yes</td>
<td>Regional and rural</td>
</tr>
<tr>
<td>C11</td>
<td>Respiratory Specialist</td>
<td>10 y</td>
<td>Yes</td>
<td>Regional and rural</td>
</tr>
<tr>
<td>C12</td>
<td>Physiotherapist</td>
<td>5% of annual work</td>
<td>Yes</td>
<td>Regional</td>
</tr>
<tr>
<td>C13</td>
<td>Social Worker</td>
<td>approx. 30 MND pts ongoing</td>
<td>Sometimes</td>
<td>Regional</td>
</tr>
<tr>
<td>C14</td>
<td>MND Clinical Nurse Specialist</td>
<td>14 y Ongoing</td>
<td>Yes</td>
<td>Regional</td>
</tr>
<tr>
<td>C15</td>
<td>Gastroenterologist</td>
<td>Sees 2-4 people with MND monthly</td>
<td>No</td>
<td>Regional</td>
</tr>
<tr>
<td>C16</td>
<td>MND Clinical Nurse Consultant</td>
<td>15 y Ongoing</td>
<td>Yes</td>
<td>Regional</td>
</tr>
<tr>
<td>C17</td>
<td>Respiratory Clinical Nurse</td>
<td>22 y Varies</td>
<td>Yes</td>
<td>Regional</td>
</tr>
<tr>
<td>C18</td>
<td>Rehabilitation MND Specialist</td>
<td>16 y approx. 30 MND/year</td>
<td>No</td>
<td>Regional</td>
</tr>
<tr>
<td>C19</td>
<td>Palliative Care Consultant</td>
<td>11 y approx. 30 MND/year</td>
<td>Yes</td>
<td>Regional</td>
</tr>
</tbody>
</table>
4.4. Themes Identified from the Findings

Seven major themes emerged from analysis of the categories identified from participant interviews (Chapter 3). Together these were found to characterise communication between clinicians and people with MND and their family members/caregivers and are summarised in Table 4.3 below.
Table 4. 3. Themes and Key Findings

<table>
<thead>
<tr>
<th>Major Themes Identified</th>
<th>Key Findings</th>
</tr>
</thead>
</table>
| 1) Experience of clinicians with end-of-life communication and NIV withdrawal (4.5 below) | 1) Reluctance to communicate end-of-life choices by the clinicians involved with symptom control prior to palliative care involvement.  
2) Clinicians with longer experience in their field may be less likely to be fully cognizant with the most recent recommendations and guidelines. |
| 2) Timing and content of communication about the benefits and burdens of NIV, PEGs and end-of-life choices (4.6 below) | 1) Many clinicians found the recommended timing of end-of-life communication unrealistic.  
2) Confusion with understanding the benefits burdens of PEGS and NIV by people with MND and their families.  
3) Palliative care clinicians reported patients often had end-of-life choices communication too late and not prior to their involvement. |
| 3) Time allocated for communicating and discussing end-of-life choices (4.7 below)      | 1) Respiratory clinicians state they have insufficient allocated time to communicate end-of-life whilst having to discuss how NIV works and how to use the device.  
2) Respiratory clinicians state because many of their clients are from the country and they may only meet them once, they must prioritise the mechanics of NIV above emotional issues.  
3) Time required to recognise FTD in people with MND and the implications for end-of-life choices. |
| 4) Early referral to palliative care and family understanding of palliative care (4.8 below) | 1) Clinicians reluctant to refer soon after MND diagnosed.  
2) Families overwhelmed by the concept of palliative care. |
| 5) Communication within a multi-disciplinary team (4.9 below)                           | 1) Frequent poor communication and differing opinions between members of MDT. This was found to be worse where a group of health care professionals made up a MDT from different areas and organisations.  
2) AHD poorly discussed and documented, and not re-visited when NIV or PEGS became part of a person with MNDs symptom control. |
| 6) Patient and family communication and understanding (4.10 below)                     | 1) People with MND and their families were under-prepared for the burdens of NIV, often feeling they had little choice as to whether to use it or not.  
2) Potential withdrawal prior to commencement of NIV was poorly communicated if at all. |
| 7) Communication barriers from clinician and family perspective (4.11 below and listed in Table 4.5) | 1) Clinicians report patient denial, not wanting to take away hope and lack of adequate time to have clear end-of-life communication; clinicians often wait until asked for information.  
2) Family members suggested lack of health literacy may prevent confidence to ask for information. |
4.5. Experience of Clinicians with End-of-Life Communication and NIV Withdrawal

One of the influences on the willingness and capacity to have end-of-life communication with people with MND identified from the literature review was years of clinician experience (Blackhall., 2011). All the clinician participants interviewed had many years of experience with communication surrounding end-of-life decisions (4.5-22 years), with 12 of the 19 clinicians having had 10 years or more.

Anecdotally, experience with communicating end-of-life choices to palliative care patients generally enhances a clinician’s ability to sensitively discuss options of care. Clinical experience in palliative care and respiratory medicine of the participant clinicians was, therefore, anticipated to facilitate the difficult discussions (relating to the end of life and the potential withdrawal of NIV) soon after diagnosis. In relation to MND, however, the experience of having prior involvement with withdrawal of NIV, an important aspect of MND, did not always appear to make such communication any easier for the clinicians. Despite their years of experience, the respiratory clinicians interviewed expressed the difficulty they encountered in having end-of-life discussions when offering NIV to ease symptom control; framed by three participants as giving NIV with one hand and discussing NIV withdrawal and the end of life with the other (C4, C11, and C12). As C12 said:

C12 (physiotherapist): You’re offering them something that potentially may treat their symptoms, but then at the same time saying, “Oh, by the way, you know, let’s also talk about, you know, end of life as well.”

The experiences of clinicians in communicating end-of-life choices and the potential withdrawal of NIV were relatively limited. The point was also made by C2 (palliative care doctor) that as MND is such a rare disease, while the clinician may feel comfortable with withdrawing NIV and having discussions
associated with withdrawal, the nurses may be lacking in experience and confidence to do so. This possibility was confirmed by C10 (nurse), who felt the experience necessary to explain to the patient that NIV could be removed and that death could follow was beyond her experience. The nurse said she felt confident to reiterate communication relating to NIV removal only if a specialist doctor had initiated the conversation. She did, however, feel confident to talk generally about end-of-life as did the other nurse participants. The palliative care and neurological nurses interviewed agreed that they felt confident and able to discuss end-of-life issues and the possibility of NIV withdrawal if the person with MND had some prior knowledge of the possibility of NIV withdrawal (C6, C7, C8, C9, C10, C14, C16 and C17). All the nurses stated that they discussed end-of-life choices despite the difficulties, preferring an approach of gentle honesty. This is exemplified by the following quote from C8:

**C8 (palliative care nurse):** But it’s got to be about making sure that things are said, even if they’re unpalatable. And, you know, there are ways – we’ve all heard some very, you know, inappropriate ways of breaking bad news and certainly just being right out there and unemotional, but gentle, is the only way to go. People will respect that you’ve been honest with them, you know. They might hate you, but that’s okay, they need to hate someone. Someone has got to be hated.

All the clinicians were asked if they had been involved with the process of NIV withdrawal in a person with MND and all except four (C15, C18, C6, and C7) had been present at least once. Therefore, the experience of being present at NIV withdrawal, and how the experience had affected the clinicians was considered potentially relevant to future communication skills surrounding early end-of-life discussions. Although some clinicians described well controlled and positive experiences with NIV withdrawal, the experience was not always like that. Despite the best protocols and recommendations, clinicians reported difficulty with giving good information when certainty is lacking. Emotionally and clinically, being able to decide the exact protocol and timing for NIV withdrawal,
C11 (respiratory specialist): *Bloody awful, because – the when, how, pharmacotherapy, you know. What do you do? Do you necessarily withdraw NIV or do you let people die on NIV? ... I suspect it’s enormously difficult from the family’s point of view unless the conversations leading up to it have been really good and really clear.*

C14 also described the difficulties experienced with the emotional communication involved with NIV withdrawal:

C14 (MND clinical nurse specialist): *It’s a conversation that we dread, I think, to remove it, because the breathing is so essential to stay alive. And there’s no good or there’s no best timing for it. It really depends on the relationship and the time that – how much the patient trusts you to talk about it; how much the family trust you, what you’re telling them actually makes sense, not killing her – killing your mum. It’s – I think it’s more difficult than talking about stopping the PEG. I certainly feel that.*

This negative clinician experience, which might influence communicating the potential of withdrawal of NIV in other people with MND, was reiterated by C8 who, despite not being personally affected, stated that others involved had been:

C8 (palliative care nurse): *A number of staff were really confronted. A number of staff said they didn’t want to be involved, and so then we set it up for 24 hours later.*

A very specific element of clinicians’ experiences of dealing with the emotional impact of communicating NIV withdrawal was noted by a palliative care
consultant. C1 highlighted the confusion amongst families and clinicians as to whether NIV withdrawal is euthanasia:

**C1 (palliative care consultant):** *We had one example in a general hospital where we had to spend a lot of time with the whole nursing staff, and also the support afterwards – partly the family, who actually coped quite well, but also the professionals - and having a debriefing after the instant so that professionals could say how they were feeling, particularly those who perhaps had contacted defence organisations or professional organisations and been given misinformation.*

In this research, the years of participants clinical experience did not appear to make the early (recommended) discussions of NIV withdrawal any easier or more likely. To the contrary, negative experiences of NIV withdrawal and knowing the potential difficulties involved both emotionally and ethically could lead to hesitancy in communicating the information. This research was unable to ascertain the level of experience or education clinicians require to enable them to initiate difficult end-of-life communication.

### 4.6. Timing and Content of Communication about the Benefits and Burdens of NIV, PEGs and End-of-Life Choices

Clinicians in this study frequently assumed that communication about NIV withdrawal and the end of life had occurred with the person with MND and their families prior to their own involvement. The responsibility for the role of communicating difficult end-of-life choices appeared unclear to the clinicians and this task was sometimes left to the palliative care clinicians involved late in the disease process. Whilst some of the clinicians (C7, C8, C14, C16, C19, C1, and C2) were largely aware of the existence of the NICE MND Management and Assessment Guidelines published in 2016 and their content regarding MND
communication and end-of-life choices, the timing of these recommended discussions was reported to be difficult. Most of the clinician participants felt that in practice the recommended timing of end-of-life communication was not always realistic.

Clinicians who initiated NIV stated that they try to commence end-of-life discussions and the possibility of NIV withdrawal early on (C4, C11 and C12) but are often faced with hesitancy or denial from the person with MND or their family, who do not wish to discuss it:

C4 (respiratory specialist): I mean, you’re talking about end-of-life issues, which many health practitioners are uncomfortable talking about. And then I would think 20 to 40 to 50 per cent of patients will tend to say, “Well, we don’t want to talk about that. We’ll think about it when the time comes.” So even though you may have the best intentions of trying to have all this out in the open and discussed and documented, it’s not always the case that the patients wish to discuss it.

The four respiratory clinicians (C11, C12, C4 and C5) said they tried to ascertain what other specialists had already said to the person with MND, and two said they would initiate end-of-life discussions if they had not already been broached (C11, C12). One palliative care consultant considered discussing NIV withdrawal at the stage at which it was offered to be a logical and important for transparency. He maintained that this ensured informed consent for commencing and that clear information about the potential to withdraw NIV was provided at the same time (C19):

C19 (palliative care consultant): I guess the pros is a very transparent discussion, isn’t it, that you’re actually, at the time of putting the – sorry, of starting the machine, the NIV, you’re also talking about the possibility of withdrawal. I can see the merit of that. I guess on the other hand – I’m just
thinking of the emotional dimension of that, and I guess that may – I’d have to think of the sensitivity of that conversation at that time.

C19 also stated that the emotional dimension and sensitivity of that communication would have to be considered on an individual (person with MND) basis. C19 was unaware of the guideline recommendation to discuss the potential withdrawal of NIV prior to the initiation of NIV until interviewed by the candidate, despite being broadly aware of the MND guidelines.

However, the preservation of hope can underlie a family’s wish to not discuss the end of life. The following is an account of how one healthcare professional dealt with the issue:

**F3 (family):** And then when [person with MND] was, you know, eventually diagnosed, we – by that stage we’d been reading up on best practice and, you know, we knew all about the multi-disciplinary team approach, etcetera, etcetera, etcetera. And [the health care professional’s] response was, “Oh, you’re not going to need that for a long, long time. And don’t talk to MNDAWA. They’re way too negative and I’ll see you in a year.”

The nurses interviewed for this study suggested that prior to their involvement, doctors should initiate communication about the potential of NIV withdrawal and end-of-life issues. The nurses assumed, but were uncertain, that this was the case (C6, C7, C10, C14, C16 and C17). This further highlights the communication issues between groups of healthcare professionals and between healthcare organisations which is discussed in Section 4.9.

Clinicians found the timing of communication about the end of life and NIV withdrawal challenging. There was confusion about which healthcare professional was responsible for the communication and concern that patients might not be prepared for the conversations. Most of the specialist clinicians tried to ascertain how much the person with MND and their family knew about
the course of the disease and gauged their communication content on the response.

4.6.1. Communicating the benefits and burdens of NIV

The respiratory clinicians in this study indicated that communicating the benefits and burdens does not necessarily occur when initiating NIV. There are several reasons given: too much information all at once, insufficient time, fatigue of the person with MND and the priority given to necessary technical aspects of NIV (C4, C11, and C12):

**C11 (respiratory specialist):** You know, how to set up the mask, how to set up the machine, how to plug it in, what to do if it leaks, what to do if they get rain out, what to do if they get this, that and something else. And sometimes a lot of the attention can be focused on the sort of practicality of the machine, and the sort of softer, more difficult conversations often get a little bit left to the side.

**C12 (physiotherapist):** We were going to start them on non-invasive ventilation and that’s a really difficult time because trying to explain where and what NIV can and can’t do, but also the other longer-term implications. It’s a little bit hard to talk about deterioration and death and dying when it’s the first time you’ve ever seen a patient. You’re throwing a machine at them, potentially – or at least trying to get them started on it because you’ve probably got two days before they go back to the country.

Some of the burdens mentioned by the respiratory specialists and neurological and MND nurses involved with the support of country people with MND were the practicalities of obtaining an NIV machine, battery backup and emergency planning for that eventuality (C6, C7 and C11). The practicalities of NIV may become overwhelming for the families if not adequately explained and if emergency backup plans have not been formulated. The family participants F1 and F2 supported the importance of contingency planning:
F1 (family): And then if something went wrong with the machine, Dad would ring me. I’d go to their place and then I’d try and work it out and if I couldn’t I’d ring [the hospital], and often they couldn’t really answer my questions. So, the technical back-up of that was really difficult. If it didn’t work I ended up finding a chemist around here that dealt with BiPAP machines and stuff like that, so we could get new parts … Finding the filters was something I had to research, so finding replacement filters. And if something – it wasn’t really discussed if it broke down or anything like that.

C6 discussed the issue that some people with MND had a backup plan of going to a hospital with their NIV in the event of a power cut and that this was a huge worry for the families. The initial cost of the machine appeared to depend on whether the person with MND had health insurance cover, which clinic they were linked with, and where they happened to live (C6, C11). The point was made by C11 and C12 that as referral to them was often late in the disease they had to condense vital information regarding the use of NIV to fit within the brief time available before the person returned to the country.

One of the clinicians (C14) mentioned hospital admission would be necessary should withdrawal of NIV be considered, which, as reported by C6, could be very upsetting for the person who wished to die at home:

C6 (neurological nurse): The lady that couldn’t tolerate NIV, when she was going on it, was very clear that she wanted to die at home, and it was explained to her by the doctor that that would not happen if she was on NIV and had it withdrawn; it would have to be done in hospital, which was very upsetting. It didn’t eventuate, but I know that caused a lot of stress for the family.

Increased dependency on family support and caregiver strain was mentioned as a burden of commencing NIV (C5, C11 and C2) and its long-term use, as the person with MND is less able to adjust the mask or take it off:
C11 (respiratory specialist): And I upfront say that, you know, “As this disease progresses, that more and more the burden of actually putting it on and actually making it go falls more and more on the carer as people lose the dexterity in their upper limbs.” And that, you know, that’s something that they need to know about upfront.

The possible benefit of improving survival with the use of NIV was questioned by some of the clinicians who made the point that there was only one study published (at the time of the interview) (Rafiq, Proctor, McDermott, & Shaw, 2012). However, as added by one of the respiratory specialists:

C11 (respiratory specialist): it depends a bit on how savvy the patient is and how good they are at understanding the idea of a survival benefit, because I think that there probably is. But I won’t use that to push people into having NIV.

Therefore, some clinicians did not say anything about increased survival (C12, C18), however, some respiratory clinicians did discuss improved survival as a benefit of using NIV with their patients (C11, C14, C5):

C11 (respiratory specialist): If they’re saying, you know, “No, I don’t want it,” I will, you know, make some sort of allusion to the idea that there might be a survival benefit and if they want to chase it, I’ll talk about how there probably is, but, you know, for some people surviving longer with a progressive neurological disease may not be a benefit, in fact, and may be more of a burden.

All the clinician participants interviewed stated that they communicated that NIV would not stop the progression of the disease, and that the person with MND would become more dependent on it. The community nurses reiterated dependency and disease progression, and all the bereaved family participants and the person with MND agreed they were fully informed about this and understood the continuation of disease progression despite the use of NIV. However, a few of the family participants recall being told of the practical
burdens of NIV: issues of skin tears, particularly on the bridge of the nose; feelings of claustrophobia from the mask; and increased dependency on caregivers to adjust or remove the mask as muscle movement becomes more difficult (F1, F2, F5, P1). This may have been due to professional caregivers’ views of NIV, as a comment from one of the clinicians suggests:

**C18 (rehabilitation specialist):** But, you know, I’m actually in favour of it [NIV], so I don’t tend to talk too negatively about it.

As previously stated, the respiratory specialist participants in this study, whilst acknowledging the guideline recommendation of communicating the benefits and burdens of NIV and potential of NIV withdrawal prior to NIV commencement, had difficulty discussing those sensitive issues in practice:

**C17 (respiratory nurse consultant):** I mean, you can’t give the patients – if you told them every problem that might occur in the next four years they’d go, “I’m not using that.” Do you know what I mean?

Some of the clinicians involved at the time of respiratory failure and distress were honest in their admission that the “lack of hope” end-of-life discussions relating to possible withdrawal of NIV meant that these discussions were left to the palliative care service to address later (C18, C4). Some clinicians, whilst accepting that palliative care should be involved from the time of diagnosis, did not acknowledge NIV as palliative care symptom control nor explain it in such a way to the person with MND (C18, C16, C17, and C14). Virtually all the clinicians involved with initiation of NIV felt that as NIV may relieve much respiratory distress and had the potential to prolong life, it should be encouraged (C5, C4). However, C11 made the point that the potential to increase life with NIV was not always a good thing, as a longer life may equate to longer suffering. There was some uncertainty from clinicians relating to NIV use, who generally confirmed its usefulness but admitted it did not suit everyone (C19, C11, C4 and C12):
C4 (respiratory specialist): … and, you know, we talk about length of life versus of quality of life. We say that – I’ve no doubt in certain circumstances non-invasive ventilation can extend the life of some people with motor neuron disease, but that has to be balanced against quality of life.

Prolonging life and hope were considered important to the clinicians and influenced their decisions to discuss NIV withdrawal potential and end-of-life choices. However, prolonging life (and possible suffering) was not always a priority of the person with MND and their family (F1, F2, and F6). None of the bereaved family participants said that the potential of prolonging life in the context of possible suffering was alluded to or discussed. However, thinking retrospectively, some of the bereaved family participants said that their family member did suffer as a consequence of their lack of understanding that the NIV could be withdrawn, as stated by F1 and F2:

F2 (family): Just get this thing [NIV] off. I just – maybe she was thinking, “I just want to go. I’ve had enough.”

F1 (family): I think now, I would have opted for that option [NIV withdrawal] because Mum was so uncomfortable and so suffering that I would have opted to say, “Let’s, you know, ditch that, and let’s, you know” – and I think now Dad would have taken that option. But from what we knew, and what we – while we were going through it, I don’t think we would have done any different to what we did do. But do you know what I mean, hindsight’s a wonderful thing . . . I regret – and you can use that as a quote if you want – but I regret the last three nights that mum had at home. It was awful for her, for dad, for me. It was just – still burned in my memory, and it’s hard to sort of go away.’

The difficulties with NIV and the discomfort of a mask caused ongoing suffering for some people with MND, as illustrated by F1 describing her family member coping with the NIV mask towards the end of her life:
F1 (family): … putting it [NIV] on, pulling it off, and putting it on, and she was trying to stand up and sit down, and she was just all over the place. And I just said to her, “Look” – I was in tears, and I just said, “I can’t do this again. We need somebody that knows what to do for you to have you [referring to the hospice].”

Some of the respiratory clinicians stated that they provided the person with MND information regarding withdrawal at the time of offering the NIV (C5), however, this doesn’t seem to have occurred for F1 and F2’s family. All the respiratory clinicians stated they tried to gauge whether the person was ready to engage with end-of-life discussions and intended to provide withdrawal information if appropriate, time allowed and the person with MND was not too fatigued (C11, C12, and C4).

In summary, this research has found that the family participants did not understand the benefits and burdens of symptom control and any potential life-extension in relation to possible prolonged suffering. Most of the clinician participants stated that they try to discuss the benefits and burdens of symptom control measures, but that generally the benefits, rather than the burdens, of symptom control are more the focus of discussions.

4.6.2. Communication at end-of-life trigger points

Patient deterioration or implementation of symptom control measures such as NIV or PEGs are also times when end-of-life discussions are recommended to occur (NICE, 2016). Clinician participants of both cohorts (palliative care and respiratory) were asked when they commenced end-of-life communication and whether there were any trigger points used as openings for such discussions.

The respiratory clinicians varied in their responses. C5 involved the palliative care specialist at the initiation of NIV and stated that whilst some
discussion was presumed to have occurred prior to the respiratory specialist involvement, it was certainly reiterated by C5 at that time:

**C5 (respiratory specialist):** *End-of-life information is provided on the first outpatient visit. We do discuss this in great detail during the subsequent clinic visits ... through the regular reviews we do make them fully aware of the prognosis and the limitations of treatment and the likely outcome of treatment strategies.*

Respiratory clinicians C11, C12, and C4 and the social worker, C13, stated that they tried to ascertain how much the person with MND understood about their disease whenever they met them and were guided by the person as to how much information they wanted rather than waiting for recommended trigger points and proceeding regardless of the patient’s preference:

**C11 (respiratory specialist):** *Okay, you know, if you’re going to have – I kind of – as a respiratory physician it depends a little bit on who the neurologist that I’m working with is and how much they’ve done already.*

**C12 (physiotherapist):** *I guess the patient that we hate to see the most if sometimes we get outside referrals from – not from the MND clinic ****, but they’re being sent to us by another respiratory physician or elsewhere, and these patients are extremely end-stage there. It’s the first time we’ve ever seen them.*

The respiratory clinicians all stated how difficult it was to have discussions about the end of life when initiating symptom relief with NIV, suggesting that those discussions may not always occur as recommended (C12):

**C12 (physiotherapist):** *That’s a really difficult conversation because it’s – one of the things we’ve found is that non-invasive ventilation is sometimes used as –*
almost as the end point. So, people will offer it, but then not talk about the limitations of that therapy … you’re offering them something that potentially may treat their symptoms, but then at the same time saying, “Oh, by the way, you know, let’s also talk about, you know, end of life as well.”

The gastroenterologist (C15) interviewed stated that he was generally not involved with discussions relating to end-of-life:

**C15 (gastroenterologist):** If I get a sense that prognosis has yet to be discussed, or they don’t know, then I often don’t go down that path unless it’s necessary for my care, only because I think the person – the team that I guess is going to be most responsible in coordinating this person’s care should be having that discussion rather than an external specialist they may only see once.

The community palliative care nurses all said they commenced end-of-life discussions at admission to their service (C8, C9 and C10) whilst the neurological nurses (C6, C7) who were involved earlier in the disease process, said they were open to end-of-life discussions as soon as they were involved and particularly when they recognised deterioration within the person with MND. The two MND specialist nurses (C14 and C16) both said that they commenced symptom control and end-of-life discussions soon after diagnosis. C4 and C16 felt able to discuss NIV and PEGs as a positive method of controlling symptoms, and explained end-of-life choices particularly to alleviate any fear of a frightening, choking death:

**C4 (respiratory specialist):** So, yes, I do indicate that PEG tube feeding may prolong life by that mechanism but make it very clear that again that’s not a cure for motor neurone disease; ultimately it’s not going to change the outcome.

The palliative care clinicians acknowledged they were generally involved late in the disease process so tended to reiterate rather than initiate such
discussions. The palliative care clinicians made the point that end-of-life discussions when the patient was first introduced to palliative care may not be an acceptable initial conversation to have, and that, while discussions were patient-led, they did prompt the discussions as the person physically declined (C3, C2, C1, and C19). Despite the NICE (2016) guidelines and the European guidelines on the clinical management of amyotrophic lateral sclerosis (Andersen et al., 2012) recommending various trigger points for end-of-life communication to occur, one clinician did not see NIV initiation as one of those trigger points (C18). Instead, the clinician felt it was a stage a person with MND had to go through, and not a trigger for considering the end of life:

C18 (rehabilitation specialist): I don’t actually see that as end of life. I just see that as a stage in their illness that they all seem to go through. So, I’ve never actually considered non-invasive ventilatory support as a trigger for end of life [discussions].

In addition to being asked if there were any trigger points in MND that would initiate end-of-life communication, the clinicians were asked by the interviewer if end-of-life choices of care were initiated when NIV was offered:

C19 (palliative care consultant): It’s a good question. I may not necessarily bring the two together. I think at the beginning with the NIV I’m mainly focusing on the fact that, “I’m concerned about your breathing” — sorry, “concerned about your respiratory function. You’re having the NIV and I want to see how you’re feeling with that.” I may not necessarily at that time, right at the beginning, embark on a conversation about what might happen if — I guess if the future were to be that there was a discussion about withdrawing from NIV. So, I may not have that right at the beginning.

One of the nurses suggested that clinicians avoid the difficult end-of-life communication, fearing the effect the conversation may have on their patients and families:
C8 (palliative care nurse): *I think health professionals’ fear addressing those issues [end-of-life choices], and the response that they might get from the clients and the caregivers raising those issues.*

In summary, the communication guideline recommendations are generally adopted by clinicians in a manner which supports their patients and are considered the best by the individual delivering the communication, but not necessarily as the guidelines recommend (NICE, 2016). Some of the clinician participants in this research focussed on the positives surrounding symptom control rather than any prolonged suffering that may occur. The palliative care specialists stated that they often had to clearly point out, and belatedly discuss, end-of-life options which had not been triggered earlier in the disease process.

4.6.3. Communicating prognosis

Within the NICE MND Assessment and Recommendation Guidelines (NICE, 2016) it is proposed that the person with MND can expect to understand their prognosis from early diagnosis. The clinician participants were asked if they included the prognosis within early communication with the person with MND and their family. All the clinicians considered that giving the prognosis as length of survival was difficult; all said that if asked for a prognosis by the person with MND, their answer was not reflective of the course of the disease but rather an attempt to ascribe a survival time factor:

C19 (palliative care specialist): *I do say – and I couple that with the fact that, “We don’t know how long you’ll have to live; that a prognosis is difficult to know, and that we will do our very best to support you and keep you comfortable through this whole process.”*

There appears to be an unchecked assumption about what patients and a family want to know when asking for a prognosis; for example, they may have requested a description of the course of the disease rather than merely an estimate of time until death. Family member participants F1, F2, F4 and F5
described having been unclear of the specifics of MND when interviewed, or of which symptoms to look out for that may have been responsive to intervention, despite using the Internet for information.

All the clinician participants made the point that with the Internet so readily available, people with MND and their caregivers either had already researched or could have obtained information prior to their (clinician) involvement:

**C1 (palliative care consultant):** Some are [well informed]. Some are very much ahead. Some may have read a lot on the Internet. Some may have read some awful things on the internet.

**C18 (rehabilitation specialist):** And, you know, in the days of – living in the days of Google and Internet they’re not restricted to doctors’ opinions and – you know, these people have all been on the Net before they come to us. And I think they’ve formed a pretty strong opinion about where they’re heading.

Table 4.3 indicates the types of communication family participants had with clinicians prior to the offer or commencement of NIV. Specifically, the benefits and burdens, end-of-life options, PEG insertion and at what stage of the disease palliative care referral was recommended. The table also shows whether the family participants interviewed had AHDs in place and whether these had been formalised with their clinicians.

This research questioned whether the clinician participants discussed prognosis with their patients. All the clinicians stated that they did but could not give definitive timelines. Prognosis was not discussed in terms of how the disease may progress or what the person with MND could expect to experience. Throughout the course of the disease, only two families stated that they fully understood the end-of-life care options (F3, F6).
### Table 4. Family Communication with Clinician Pre-Commencement of NIV

<table>
<thead>
<tr>
<th>Family Participant</th>
<th>AHDs formalised</th>
<th>Benefits and Burdens of NIV discussed</th>
<th>Increased use of NIV discussed</th>
<th>End-of-life options discussed</th>
<th>Palliative care referral and options discussed</th>
<th>PEG insertion: timing and discussions</th>
</tr>
</thead>
<tbody>
<tr>
<td>F 1/2</td>
<td>Discussed between family members.</td>
<td>Not physical problems; some mechanical issues discussed</td>
<td>Yes, although some confusion</td>
<td>Not fully understood until palliative care involved</td>
<td>Late in disease trajectory</td>
<td>Pushed by clinicians but refused by person with MND</td>
</tr>
<tr>
<td>F 3</td>
<td>Yes, and discussed within family</td>
<td>Yes, and researched on internet</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes, pre-respiratory problems</td>
</tr>
<tr>
<td>F 4</td>
<td>No, but wishes known by husband</td>
<td>Very late and then NIV refused by person with MND</td>
<td>N/A</td>
<td>Too late; patient already in palliative care</td>
<td>Late in disease trajectory</td>
<td>Recommended by GP; patient pulled PEG out; hospital reinserted</td>
</tr>
<tr>
<td>F 5</td>
<td>No, but wishes known by family.</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>patient and wife in denial</td>
<td>Yes; prior to respiratory distress</td>
</tr>
<tr>
<td>F 6</td>
<td>Yes, with GP</td>
<td>Yes: but emphasis on machine mechanics and sorting out costs</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
<td>Initially refused but agreed RIG later when swallow affected</td>
</tr>
<tr>
<td>P 1</td>
<td>Family discussed and organised</td>
<td>No</td>
<td>Yes</td>
<td>Late in disease</td>
<td>Late in disease</td>
<td>Yes, but unsure when?</td>
</tr>
</tbody>
</table>

#### 4.6.4. Delivering the diagnosis

Research participants were not directly asked how the MND diagnosis was delivered to the family, however, several of the family participants volunteered this information. One of the main causes of distress was the way the MND diagnosis was delivered by the specialists (F1, F3, F5 and F6). F1 describes the situation where various tests had been performed on her family member and the specialist phoned F1 to arrange an appointment for the following day. F1 asked whether they now had a diagnosis and was told that
they had, but the specialist told F1 that he would not discuss it over the phone but would “deliver the news face-to-face tomorrow”. This caused much anxiety, as F1 felt that had she known the diagnosis of MND she could have prepared herself and prepared relevant questions:

**F1 (family):** Yes, I was quite frustrated when I was talking to him on the phone, because I was – I knew I was going to get bad news. I just wanted to have that so that I could research it before I spoke to Mum and Dad.

This point was confirmed when the family received the news, but it did not really register on them until some 15 minutes later. Apparently, the person with MND was only able to write on a white board at that time, and wrote:

**F1 (family):** “Is there a cure?” And he [doctor] said, “I’m afraid there’s not.” And she said, “How long do I have to live?”

Unlike the experience of F3, the unexpected diagnosis of MND for the partner of F5 was delivered by phone. F5 and the family member receiving the diagnosis of MND stated they had little idea of what MND was:

**F5 (family):** No. [person with MND] did, because he – after we got rung up on the phone, he did, you know, Dr Google.

The experience of F3 when receiving the diagnosis of her family member was equally confusing:

**F3 (family):** Oh, yeah, initial diagnosis discussions. That was horrendous. … So we were referred to a neurologist who will remain nameless. The first appointment we had with him was in early July of 2011. He told us at that meeting, “Well, either it’s bad, and I can help you, or it’s very bad and I can’t.” And our immediate reaction is panic. You know, “Please tell us more.” “Oh, no, I don’t want to worry you.”
F4 described how his family member had become progressively emotional (crying often) and developed slurred speech. Eventually, and after the affected person had seen a neurologist, the family was referred to an ear, nose and throat specialist for a speech impediment and it was he who recognised fasciculation (quivering) of the tongue and diagnosed MND.

The family participant involved with F6 described how her family member had various investigations including an MRI (Magnetic Resonance Imaging) the results of which apparently showed no abnormality. At a later date the MRI was reviewed by another specialist who found reportable changes. That the changes had not been recognised earlier was quite distressing for the person concerned and the family. F6 went on to say that the diagnosis was then confirmed many times by various means which to the family seemed “ridiculous”:

F6 (family): So, you know, there was about a consultant and three neurologists that they’d seen, and they all had a reconfirmation of diagnosis. So, it got a bit beyond a joke, for want of some other words, to be told this.

The person with MND interviewed for this study (P1) described how he was referred to a neurologist who, despite fasciculation being present in his lower legs, did not perform an MRI. Instead the neurologist believed that the fact that P1 had Type 2 diabetes explained the clinical manifestations. Ten years later the fasciculations were in P1’s upper legs so he sought a further neurological opinion. Again, he was told the issue was not MND. The partner of P1 explained to the neurologist that P1 was often “quite violent in bed” with uncontrolled movements:

P1 (person with MND): Uncontrolled movement. So, his suggestion was that we sleep in separate beds. That didn’t go down very well with either [partner] or myself. [Partner didn’t ask to go in. He [doctor] invited her in, and he was really quite rude to her [partner].
Inappropriate communication by doctors as illustrated by the dialogue above (P1) was raised again by F6. This encounter involved a doctor who had stood beside her family member’s bed and, because the number of bed days allocated in an acute setting depends on a person’s condition, proceeded to discuss aged care facilities:

F6 (family): … doctor, was very inappropriate about discussions over Mum’s bed, like it’s that not including the client as such, to say that “she would need to be out of here in a nursing home.” And that was particularly upsetting for Mum and Dad.

Given that there is no specific diagnostic test for MND, diagnosis can be arduous and protracted. However, frustration was voiced by families that MND was not considered earlier in the illness as a potential diagnosis and the person affected was not sent to see a neurologist. For example, F1 and F2 describe how their family member had post anaesthetic respiratory difficulties relating to an operation followed by heavy mucous secretions. Apparently, the person with MND went to see at least three specialists (unsure which specialties) over approximately 12 months who all stated there was nothing wrong. Finally, their family member, already at the stage of losing her voice, saw a sleep specialist who referred the person to a neurologist. As the consequences are so devastating, it is understandable that clinicians want to undertake various tests to ensure the correct diagnosis. However, this research has found that the way the diagnosis has been given to the family participants caused considerable distress and had implications for patient and family readiness to discuss end-of-life symptom control.

4.6.5. Communicating end-of-life options into NIV initiation discussions

Many of the clinicians in this study, despite their years of experience, found it extremely difficult to uphold the recommendation that discussion of the potential for withdrawal of NIV occurs at the time of the offer or prior to the
initiation of NIV (NICE, 2016). The priorities for symptom control and end-of-life care in a person with MND may not be consistent with the clinician's care aims and preferences, so clear dialogue between health professional and patient is essential. Whilst MND is a disease that progresses relentlessly, some symptom relieving therapies may offer a short extension of life; NIV is one of those therapies. However, prolonging life may not be the ultimate aim of people with MND and any prolongation may be considered by them as an extension of their suffering:

**F2 (family):** They talked about the end of life, but they [specialists] didn't talk about the fact that, you know, they [person with MND] might … be getting confused and not be able to make the decision in the end.

Therefore, early understanding of and an agreement about end-of-life care priorities and preferences is recommended to occur soon after diagnosis, and especially when there is a deterioration of symptoms (Andersen et al., 2012; Borasio, Voltz, et al., 2001; NICE, 2016).

### 4.6.6. Withdrawal of NIV communication: ethical and legal concerns

Most of the clinicians interviewed reported controlled experiences with the withdrawal of NIV, generally undertaken within the comfort and security of the family home with the full support of family members (C8, C9, C1, C2, C4, and C19). However, there were some experiences that were not so controlled and which, because of this, caused distress both to the clinicians and the families. One of the nurses detailed an experience of when she had been requested to remove NIV by a doctor without any discussion or contextual information. Although she was in a hospital situation she felt unsupported and unprepared for the outcome (C10). The nurse concerned refused to comply with the request to remove the NIV, believing that this was outside her scope of practice. The doctor making the request eventually came to sedate the patient and remove the NIV. A concern expressed by one of the palliative care
specialists was the ethical side of withdrawing NIV, and that communication had to be very clear to prevent the person’s family misunderstanding and experiencing the process as euthanasia rather than a person declining further treatment (C1, C17):

**C1 (palliative care consultant):** It's talking about end of life, which no-one wants to do. Often people don't want to hear. It’s talking about these quite complicated ethical decisions, and certainly [in the U.K.] ethical education is pretty minimal in medical, I think, and in nursing. So, people haven’t got a background.

The importance of all members of a family understanding their family member’s end-of-life choice in refusing ongoing symptom treatment was emphasised by many of the clinician participants.

**C2 (palliative care specialist):** I guess the other difficulty that I worry about is conflict within the family and if someone comes back later and says, “What were you doing?” And support for the team, you know? Clearly this is a big deal for any palliative care team to undertake at home.

**C11 (respiratory specialist):** I suspect it’s enormously difficult from the family’s point of view unless the conversations leading up to it [NIV withdrawal] have been really good and really clear.

As well as the ethical considerations, C5 highlighted the importance of understanding the legalities and policies concerned with NIV withdrawal to prevent emotional and lasting psychological issues particularly for the health professionals and family members. The fear that unclear communication may lead a bereaved family member to carry the scars of perceiving NIV withdrawal as killing was expressed by one experienced MND nurse (C14), who had in fact experienced this situation.
Ideally the explanation that NIV can be stopped at any time, and that medications can be used to alleviate any symptoms that may occur, should be part of the initial NIV communication. Questions that may continue to worry a person with MND can be answered and reiterated as the disease progresses, hopefully reducing psychological stress. One of the palliative care specialists (C13) made the point that it is difficult to know what amount of information is sufficient regarding the removal of NIV, as it depends so much on the stage of disease acceptance of the diagnosed person. Uncertainty around how or when death may occur for each individual poses an additional barrier for the clinician considering when to initiate such conversations (C13, C4):

C13 (social worker): The barrier is where they have enough information about “if I take it [NIV mask] off” and where they’re at, at what stage they’re at, “Am I going to die straight away and how is that going to be, and what symptoms am I going to have, and will I be able to cope with them,” and so forth.

The concern of providing symptom relief with NIV and later in the same conversation discussing the option of taking the NIV mask off when it was no longer useful was dreaded by some clinicians (C4, C1).

The idea of killing, or being perceived to kill, with the withdrawal of NIV was mentioned as a barrier (Table 4.4) to communicating withdrawal (C14). C11 thought that the death-defying culture in which we live was a communication barrier to discussing the end of life at the time of NIV initiation. This was reinforced by C11 who, whilst acknowledging that the clinical guidelines were a good idea in theory, felt that the real world frequently forced their hand. C12 stated that some people who were symptomatically ready to commence NIV were psychologically unprepared for the recommended communication surrounding NIV withdrawal and the end of life. Such patients would halt conversations relating to the end of life which were upsetting for them (C6, C7, C8, C10, C4, and C12). One palliative care specialist (C3)
confirmed that the potential of withdrawal of NIV and end-of-life choices are not discussed often enough at the point when NIV is started. The specialist added that this meant decisions relating to treatment choices were sometimes too late to be useful.

4.6.7. PEGs as a symptom control measure

The discussions surrounding the potential usefulness of a PEG tube and the explanation of the need for insertion prior to respiratory failure is a recommendation of current MND guidelines (Andersen et al., 2012; NICE, 2016). It is recommended that communication about this occurs soon after diagnosis and certainly when symptom control measures including NIV are first offered. However, some of the clinicians felt that communicating the benefits and burdens of PEG tubes and whether they had the potential to prolong life was inadequately explained (C2, C3, and C5):

C3 (palliative care specialist): Just touching on PEG tubes again, I think they’re often sold to people as more of a solution than they are, and I don’t think the difficulties of PEG tubes are adequately communicated: the risks of leakage, the risk of infection and the risk of aspiration. So, I think, you know, they’re kind of seen as an easy option when someone can’t swallow, and the reality of dealing with the other side of it isn’t always discussed as well.

C12 suggested that some people with MND and their families were unsure why they had been sent to a respiratory specialist prior to insertion of a PEG. They were also unsure if they wanted one and concerned they were being “pushed” in that direction by well-meaning clinicians. In a discussion about PEG insertion, one gastroenterologist stated:

C15 (gastroenterologist): [Clinicians are] always tempted, too tempted, to make value judgments about quality [of life] on behalf of other people and we really don’t have the right to.
Further, PEG use appeared to be commonly described by clinicians to patients and families in the context of potential increased survival, but the potential of increased suffering appears to be less well addressed, if at all (C8, C16). C19 and C17 did say that in some circumstances they would describe survival in the context of suffering, particularly if the person had MND with frontotemporal involvement (in which case C19 did not mention PEGs at all) or older onset of MND.

**C19 (palliative care consultant):** *The group that I don’t go down the PEG line at all is – and I don’t know whether this is just me, but I don’t do it with the frontotemporal group. I just avoid that now. Sometimes that can be an open question, “Why aren’t you speaking about it?” But I guess I’m just concerned – two things, one if the dementia – if the behavioural story becomes more difficult and the person’s now pulling at their PEG or RIG, and second is really is this the appropriate thing to be doing that may extend someone’s life?*

The benefits and burdens of PEG tubes are often inadequately discussed by clinicians leading to misunderstanding by the families. This research could not clearly ascertain if this was confusion on behalf of some of the clinicians who then passed on misinformation to the patients, or as C2 proposes:

**C2 (palliative care specialist):** *I think that the discussions around PEG tubes are done poorly, and the number of well-meaning people who think that they can prevent aspiration, whereas clearly that’s not true, whereas if you ask nearly all of my patients why their PEG tube was inserted they tell me it was to prevent aspiration pneumonia, which is clearly not true, but it’s the myth that’s out there.*

The understanding that a PEG tube will not alleviate the risk of aspiration was a point brought up by respiratory clinicians (RS1 and RS2) and the
gastroenterologist (RS5) interviewed for this study. All these clinicians said that this was frequently misunderstood by patients. Some of the families of people with MND were confused about the timing of PEG being offered, particularly as PEGs were encouraged prior to the appearance of symptoms that they might relieve (swallowing difficulties and inadequate nutrition), or symptoms which may prevent insertion of the PEG (respiratory insufficiency as an increased anaesthetic risk). According to (F5):

F5 (family): I’m pretty sure we were never told to make a decision about wanting to have a PEG, because the symptoms weren’t even slight at the beginning.

The respiratory clinician C12 made the point that some families and people with MND may not understand the potential of PEG tubes and may feel pressured by clinicians, particularly if they were asymptomatic:

C12 (physiotherapist): “Gee, everybody’s been at them at the PEG, and we’ve blundered in and said exactly the same thing, and, you know, making them feel as though, you know, maybe they’re being pushed down a pathway that they don’t want to go.” I think that’s really – you know, that’s not fair on them.

Whilst the insertion of a PEG tube is recommended prior to the respiratory muscles decreasing in efficiency and the time at which NIV is offered, communication of the benefits and burdens of both symptom control methods are recommended early in the course of the disease. The aim was generally to get a PEG inserted prior to weakening of the respiratory muscles, at which point an anaesthetic would be too dangerous:

F5 (family): Yeah, but I only learnt by reading myself, I’m pretty sure, that the PEG would have had to go in.
F1 and F2 stated that their family member with MND was offered a PEG but it was apparently too much of an anaesthetic risk by that time. Neither F1 nor F2 were sure if a PEG had been mentioned before then. None of the family participants or the person with MND interviewed recall being told of the risks of aspiration with a PEG or the possibility of infection. The dialogue below indicates that one family, despite their good health literacy, thought (incorrectly) the insertion of a PEG would improve the risk of aspiration:

**F6 (family):** She did, yeah, so in February, because of the distress caused by choking on food and fluids. And because Dad was her main carer – Mum’s main carer – he, I feel, probably more strongly advocated for it, because of the distress on him as well in managing the choking with the aspiration.

Both C3 and C15 brought up the point that patient and family understanding of medical complexities relating to NIV and PEG tubes need a degree of health literacy. For example, the understanding that a PEG tube may be offered and need to be inserted prior to any obvious swallowing difficulties in a person with MND because of the risk that respiratory decline can make the PEG insertion a significant anaesthetic risk:

**C15 (gastroenterologist):** And perhaps that’s health literacy, perhaps that’s my fault for not explaining it clearly enough. But, yeah, I mean, I guess it’s a barrier in terms of understanding the complexities of why they might need it earlier than the need to use it; that is, if their respiratory involvement, or their respiratory disease, is mild, but definitely present, but they’re swallowing beautifully at the moment, the idea of putting in a PEG now for the future – even though you don’t need to use it now – can be difficult to get across.

Family member F5 indicated that their understanding of PEG tubes was due to their own investigation and that not one health professional indicated that it would have to be fitted prior to respiratory muscle loss. Conversely, participants F6 and F3 and their family members were very aware of the course
of MND and had investigated symptom control options extensively. The health literacy of F6 and F3 enabled them to ask pertinent questions of the clinicians and make more informed decisions.

In summary, this research has found that there remains considerable misunderstanding between clinicians and families about the benefits and burdens of having a PEG in people with MND. The risk of aspiration is not improved with a PEG in situ. The understanding by families that a person with MND can develop swallowing difficulties as the disease progresses seems not to have been well communicated. This research also identified that the tardiness of referral for a PEG (described by some of the respiratory clinicians), is often too late for the person with MND.

4.7. Time Allocated for Communicating and Discussing End-of-Life Choices

Sufficient time to have detailed, emotional and difficult communication relating to end-of-life choices with a person with MND and their caregivers was considered a huge communication barrier (Table 4.4) and was mentioned by the majority of the clinicians interviewed:

C2 (palliative care specialist): Time, you know. We’re all very time poor, and these conversations take time, need to be re-visited, the patient’s cognitive ability, family issues. You know, conflict within a family. All sorts of things come into play. Like I say, when it’s easy it’s easy, and when it’s hard it’s nearly impossible.

Adequate time is needed to explain possibilities such as risk of aspiration infection if a PEG is inserted, whether to be treated by oral or intravenous antibiotics or whether to be treated at all under such circumstances (C3). C3 made the point that allowing 10 minutes for such discussions, particularly when a person with MND is already fatigued, is completely inadequate. The
expectation that the person with MND will be able to meet with the necessary clinicians in a short amount of time and be able to fully comprehend all that was said to them is another issue:

**C12 (physiotherapist):** One of the things, you know, when we see the patients, I’d like to spend a lot more time with some of them but some of them are just — by the time they’ve seen the speech therapist and the dietician that morning, and maybe spoken to [the nurse], and then they’re going to see us in the afternoon to do some lung function, they’re just exhausted. … So they’re fairly long conversations for a lot of people … the worst patients to see is when you feel like you’re being rushed and you’re trying to introduce a new therapy plus manage all the back end, knowing that you’re not going to see them again, and knowing that they may be going to an area where there’s very limited multi-disciplinary or specialist care to manage some of these more difficult issues.

Adjusting communication to a person’s cognitive ability and health literacy and ascertaining any family conflict was reported to require a great deal of time. Clinicians spoke of the necessity to re-visit the same conversations to ensure understanding but again, they find this very time consuming and not always possible for people returning to the country (C11, C1, C2, and C3):

**C11 (respiratory specialist):** There are some people that are willing to engage in this, and then there are other people where you can’t get them to engage in discussions about end-of-life care. And so, my goal is to do it early and to do it often until I feel like the patient and I, and family and I, are on the same page. But it’s sometimes, in the real practical world, more difficult than that.

**C2 (palliative care specialist):** If it’s not in your routine practice, and you look after an MND patient once every decade, then I think that’s really hard, you know? So, yeah, I think that’s a barrier, lack of experience, yeah. And I think
C11 (respiratory specialist): I can imagine that in trying to teach people how to do something that’s complicated there is a great temptation to focus on the functional at the expense of the emotional, because the emotional is difficult, and the conversations are difficult, and you have to talk about death and dying and uncertainty and difficulty and what’s going to happen when this brand new shiny machine that I’m just introducing you to doesn’t work anymore.

The family participants interviewed made the point that unless they were assertive and asked the right questions, they could miss the relevant information at the specialist appointments:

F5 (family): Because in these appointments you – particularly with a neurologist who gives you an overall reading on how the whole thing’s going, if you don’t ask questions, and you’re not an assertive sort of person, sometimes I think you could walk out missing a lot, do you know what I mean?

Family participant F6 suggested that the communication recommendations within the MND guidelines may not be relevant to all cases of MND. F6 went on to suggest clinicians involved with communicating end-of-life choices make time to listen to the person with MND and their family members:

F6 (person with MND): So, you know, some health professionals – I think periods of silence to listen to the client are needed without the health professional thinking, “I’ve got to tell them X, Y, Z, you know, because this is what the evidence-based guidelines say.”

To summarise, both the clinician and family participants made the point that significant time was required at specialist appointments to deliver the necessary information and understand the broad spectrum of emotional and
practical issues involved with MND. Having time to listen to the person with MND and their family was considered particularly important.

4.7.1. Influence of frontotemporal changes in people with MND

The need for time for communicating end-of-life choices has been identified in this research, but time and ability to recognise FTD in people with MND is a further challenge. The clinicians were asked if, in their experience, frontotemporal changes in a person with MND caused difficulties in decision-making regarding their treatment choices and end-of-life care. The clinicians generally had little experience of poor decision-making that could be directly attributed to frontotemporal cognitive changes, however, C1 did make one observation relating to FTD:

C1 (palliative care consultant): *It does feel like an iceberg really. There is an awful lot under the water of how we do the communication. We’re not very good at basic communication anyway, and then we’ve got this huge great mass that we can’t quite see yet and we’re getting inklings of.*

One of the respiratory clinicians observed that there may be increased impact on the caregivers of a person with MND if frontotemporal changes were affecting their understanding of symptom-relieving options:

C13 (social worker): *What concerns me sometimes with that [frontotemporal dementia] is that they [person with MND] don’t have a deeper understanding of the situation [as they would] if they didn’t have the FT [frontotemporal] happening, and it impacts on the carers, and that’s my concern.*

Potentially prolonging the life and suffering of a person with MND by making choices for options which may ultimately be detrimental for them was a concern of C19 a palliative care consultant (4.6.7).
4.8. Early Involvement with Palliative Care and Family Understanding

The majority of respiratory clinicians suggested that some end-of-life communication should have been attempted with people with MND from early diagnosis, prior to respiratory issues arising and their involvement in the patient’s care (C11, C17, and C4). The involvement of a palliative care team was identified as helpful in ensuring end-of-life communication occurred. C5 (respiratory physician), for example, stated that in his experience, end-of-life communication commenced at or shortly after diagnosis and that the palliative care team was involved from diagnosis:

C5 (respiratory specialist): *From the very beginning, Palliative care team can provide vital support before the end stage of the disease arrives. Their involvement should not only revolve around end of life care.*

The palliative care and respiratory clinicians interviewed for this study were asked at what stage of the disease progression they became involved, and their opinion as to whether the timing to palliative care was appropriate. The clinicians responsible for offering NIV for symptom relief were also asked when they involved or referred the person with MND to a specialist palliative care service:

C4 (respiratory specialist): *I mean, ideally all patients very early on, close to the time of diagnosis. For various reasons some patients don’t want that, and that’s okay, and it depends upon the support structure they have in place already.*

Despite guidance for early referral, there was a perception amongst some of the clinicians that, at the time data for this research was collected, some doctors involved at diagnosis or early in the MND trajectory believed that palliative care was for the terminal stage or last few weeks of life only (C18, C19):
C19 (palliative care consultant): … or has inadvertently forgotten that we [palliative care service] exist, or some other - or perhaps holds an old-fashioned view that palliative care is simply for the dying - now, of course, motor neurone disease is an incurable disease from day one, so it ought to involve us.

The palliative care specialist participants reported that people with MND were frequently referred to them when already extremely ill and nearing the terminal stage, often underprepared and lacking the ability to communicate clearly their wishes for end of life (C1, C2 and C19):

C2 (palliative care consultant): To be honest, most of my [MND] patients are referred very late, and we look after them for their end-of-life care.

This was reflected by most palliative care clinician participants who at times felt frustrated that the referral to palliative care was made later than it should be to optimise the benefits of palliative care for the person with MND and their caregivers (C8, C2, C14, C18 and C19):

C19 (palliative care consultant): It does happen, where the person is referred very late, and we think that’s disappointing because that patient has been known to have MND in our area for some time.

Some respiratory clinicians report that occasionally difficulties in communicating with the community palliative care teams regarding assistance with NIV patient issues was an obstruction to adequate patient respiratory care (C12). Frustration was apparent amongst respiratory clinicians that their involvement was too late to adequately assist people with MND with developed respiratory failure (C11, C12 and C17). The safest option for insertion of a PEG tube is prior to failure of the respiratory system. Late referral meant that this may not be possible, according to the gastroenterologist and the respiratory specialists interviewed (C15, C4, C11, and C12):
C11 (respiratory specialist): If that [PEG] hasn’t been discussed by the time the patient gets to me as a respiratory physician, I almost see it as a failure of the system.

C15 (gastroenterologist): Understanding the medical complexities of PEG tube insertion needs a fair degree of health literacy and that can sometimes be a barrier. For example, the appreciation that respiratory involvement increases your anaesthetic risk of the procedure and hence for me the need to put PEG tubes in before significant respiratory involvement and that medical need for that to happen can sometimes not be 100% understood.

Whilst all the clinician participants thought early palliative care involvement for people with MND was a good idea, a referral to palliative care did not always occur close to diagnosis (C8, C1, C2, C3, C13 and C19). The reasons for this non-referral, or non-inclusion within an MDT (where available) from diagnosis were unclear from the clinician responses, although a desire not to undermine any hope that the person with MND may have, and patient denial, were given (C4). The information gathered for this study indicates this may be due to financial constraints, organisational policies of the local palliative care service, or non-referral by neurologists and respiratory specialists. Respiratory clinicians reported that palliative care services were often stretched and understood that in their area of patient responsibility, it was not always possible to keep people for ongoing specialist palliative care for protracted periods (C11):

C11 (respiratory specialist): Now, I also know that, you know, there are a variety of workforce issues that mean that palliative care teams struggle to continue to see patients that are “on their books” for a long period of time.

In some areas resource constraints and funding rules require the limitation of access to palliative care services, for example, a pre-requisite of having a life expectancy of 3-6 months as an entry requirement:
C16 (MND nurse consultant): Because I’ve had a palliative care specialist from central **** ring me up, and he goes, “Well, you’ve made this referral to a patient. Has he got less than three months to live?”

As the course and timing of MND is so difficult to predict, life expectancy and, therefore, suitability for access to a palliative care specialist becomes virtually impossible to forecast (C9, C13, C16, and C8):

C13 (social worker): Oh. Well, that’s a – well, you see the definition of “palliative” – here at **** it’s about six months of life left approximately, right.

C16 (MND nurse consultant): But some pall. care teams are quite open to that [early intervention], but some pall. care teams have this weird definition period that they will not accept the patient until they have three months or less to live.

Recommended referral times, both to palliative care and to respiratory specialists, is discussed more fully in Chapter 5. The use of specialist palliative care too early in the diagnosis was questioned by one respiratory specialist, who said that other organisations or the GP should be able to manage the person with MND at least until symptoms became more complex (C4):

C4 (respiratory specialist): If they have good support otherwise from partner, general practitioner, allied health staff, myself, then there may not be – from my point of view and the neurology nurses, there may not be a great need for the specific palliative care services to be involved.

The clinician (C4) suggested that when more complex end-of-life treatment was required would be the time to involve palliative care specialists and nurses. Several palliative care clinician participants, whilst acknowledging early referral to the palliative care team to be important, thought that specialist palliative care involvement could remain intermittent until the person with MND became more symptomatic (C3, C16, C19, C8). This represents a departure from the recommendation in the current guidelines. The few specific MND clinics involved in this study were able to involve palliative care from diagnosis,
and then have sporadic involvement until respiratory or swallowing became problematic. However, the point was made that this presented a difficulty in knowing exactly at what stage those symptoms were occurring, and whether at times they were being missed (C1):

**C1 (palliative care consultant):** *Now, that sounds very clear in some ways but how do we know those stages are happening? And that’s why I think probably we need to be in most – a lot of the time, so we [palliative care team] can actually pick up some of those changes.*

Of the seven participants interviewed within the category of the bereaved families and people with MND, three were offered palliative care referral late in the course of the disease (F1/2, F4, and P1). Two families (F3, F6) considered the timing of their referral to palliative care appropriate (although it did not occur at the time of diagnosis). When early palliative care was mentioned to family F5, the benefits of what the service could provide were not adequately communicated to the family to alleviate their fear of what palliative care meant:

**F5 (family):** *That word “palliative” over-shone whatever they were saying to us.*

The concept of palliative care was confronting to people with MND in this research. From a symbolic interactionist perspective, the public understanding of palliative care is of death and dying and is perpetuated by clinicians who remain reluctant to discuss or refer to palliative care. Early palliative care involvement for some people with MND and their families was confusing, particularly if they were minimally symptomatic at the time of palliative care discussions. From the clinician perspective, the way in which language is used was pointed out by C19 as important:

**C19 (palliative care consultant):** *I’m particularly conscious about technical language, how difficult, intimidating and troubling technical language can be, and it just is not a good communication style. So, it’s really clarity of language.*
The confusion within families about the role of palliative care was apparent in the data. For example, participants F5 and F3 said:

**F5 (family):** *She’s a specialist. And she said, “Now, I’ve got a letter here from your GP saying that you’re in denial about all this.” And we said, “We’re definitely not in denial about what’s happening to us. We’re just in denial that we need to see a palliative care team so early.”*

**F3 (family):** *It was discussed, and because sort of our initial – you know, sort of we now know that palliative care is really the only thing that you can do with motor neurone disease. To us at that stage we were thinking “end of life” and we’re getting a bit paranoid.*

The provision of a holistic approach incorporating palliative care for people with MND and their families is still dreaded by some patients and families and adopted reluctantly by clinicians involved with diagnosis and early involvement with MND (Oliver et al., 2016). MNDA recommendations are generally accepted as a good thing by clinicians, but they are not always reflected in practice (C8, C1, C2, C3, C16 and C19). Early palliative care or, as recommended, palliative care referral from the time of diagnosis, is discussed further in Chapter 5.

A point raised by C12 as a reason for early palliative care involvement and referral from respiratory clinicians to palliative care specialists was a practical one: the cost of prescription medications. An example given was the high cost of glycopyrrolate (a medication used to assist in alleviating excessive secretions) if prescribed by a respiratory doctor compared to prescription through palliative care. This same prescribing issue exists in New South Wales and Western Australia.
In summary, there is broad agreement with the guidelines on early referral to palliative care, but this does not necessarily translate into practice. Barriers from the clinician’s perspective include the difficulty in estimating the unpredictable course of the disease, reluctance to take further hope away from the patient and family and the availability of the palliative care service to accept the patient. From the perspective of the person with MND and their family, this research identified a reluctance to be referred largely due to misunderstanding what palliative care could provide and the support that could be offered.

4.9. Communication within a Multidisciplinary Team

The information already given to people with MND and their caregivers was not always known by a subsequent healthcare professional, causing confusion between healthcare providers from various organisations involved with the same patient. One community nurse (C8) mentioned that the organisation she worked for did not give out much information on symptom control for people with MND, leaving it up to other organisations to provide this. Below, C16 describes the frustration with working within the private healthcare system and trying to access information from the public allied health system:

C16 (MND clinical nurse consultant): . . . look, I know it's public and private, or whatever it is, but there has to be some way that if, you know – and I can get that it’s probably maybe because the community allied healthcare team is busy, or whatever it is. But even having – knowing who the person has been referred to from an OT [Occupational Therapist] point of view, so I don’t have to ring, you know, three aged care teams to work out where the OT is, or even just having assessors, if I ring My Aged Care to tell me, “Okay, well, this OT has seen your patient,” so then I can follow up with that OT, I think would make it a bit easier for me so I’m not constantly chasing my tail.

The neurological and palliative care nurses working and supporting people with MND and their families within the community were described by the
family participants as an informative and wonderfully knowledgeable resource. However, the nurse participants in this study reported that they often had to initiate communication and referrals relating to symptom control including breathlessness and the potential for NIV even though they didn’t necessarily perceive that to be part of their role or professional remit. These issues had apparently not been broached by doctors:

C6 (neurological nurse): A recent example was a client who I could see was noticeably becoming breathless and hadn’t at that stage been followed up by anyone about it. So, I initiated the discussion about respiratory issues and got them information about NIV so that they could be prepared when they saw the respiratory physician for talks about NIV.

In order to ensure the person with MND and their family receive all aspects of necessary care from diagnosis to death, an MDT approach to care has been considered the most beneficial (Miller et al., 2009). A coordinated team of specialists assists with advice on symptom control and identifying the individual requirements of each person with MND and their caregivers. The communication includes discussion about the introduction of NIV and the potential for the withdrawal and consequences of withdrawal. However, this research found that communication between health professionals within an MDT was sometimes poor, particularly if the team was fragmented between organisations, or split between metropolitan and country:

C1 (palliative care consultant): I think my other fear is you can get so many teams of people involved. You’ve got specialist palliative care, GP and primary care, respiratory care, gastroenterologist who’s been involved with the PEG, the rehabilitation team, a neurologist who’s saying – you know, and the possibilities of confusion and misinformation are enormous. And the inter-team – I was going to say warfare.
Within metropolitan areas the MND MDTs were generally coordinated teams attached to a single area of healthcare. However, this was certainly not always the case:

**C19 (palliative care consultant):** *Now, the interesting thing in the city is that not everywhere has a multi-disciplinary team. So, it's a bit of a patchwork quilt, and indeed would be the whole of Australia a patchwork quilt.*

This perspective was found within urban and rural areas. Where structured and coordinated MDTs did not exist this sometimes led to multiple appointments in different facilities, and confusion as to which professional was attending to what clinical problem. Certainly, there was comment from some of the family participants that there was a lack of coordinated care, confusion with who was who and who did what, and that they felt they were repeating symptoms and issues many times to many different people (F1, F2). There was also some confusion even between family members as to who was coordinating their family member’s care and when asked if there was a specific team involved, as the following exchange illustrates:

**F1 (family):** *No … odd … they were very odd.*

**F2 (family):** *But “K” was coordinating everyone, wasn’t she?*

**F1 (family):** *No, no, she was just helping me – she’s the motor neurone group lady.*

F1 and F2 agreed that the amount of different appointments with different organisations caused their family member with MND and her husband much confusion, fatigue and wasted much precious time:

**F1 (family):** *We’d been to a few speechies [speech therapist] and dietitians, which Dad was getting confused about. So, I sort of explained to them that the dietitian was about the nutrients that she was getting; the speechy was about …*
how to swallow, and the muscles in the neck. And Dad said, “We’re saying the same things to the same people.”

**P1 (person with MND):** So, he [doctor] made some comment about, “If you haven’t heard in two- or three-weeks’ time, ring me and up and I’ll see what’s going on.” So, we didn’t hear anything from anybody else. So, for three weeks we were absolutely left in limbo, which was quite traumatic.

The speed of disease progression and the inability to find assistance with immediate requirements was a recurring theme, and concurred with the findings of McConigley, et al. (2014), who found that whilst clinicians should try to pre-empt the requirements of people with MND as their disease could progress rapidly, this was not necessarily occurring. Families and people with MND felt, for instance, that equipment needs from NIV to prosthesis requirements were too slow to materialise once a need for the particular item had been identified by health professionals (P1, F1, and F2). F5 acknowledged that their family member was provided with all the technology they may require and seemingly prior to their actual needs, which were well anticipated by their hospital and MDT providers. This is in stark contrast to P1 who felt the provision of necessary equipment was far too slow due, it was suggested, to poor communication between healthcare professionals:

**P1 (person with MND):** So by the time that we went - the private contractor had all the measurements done and the prosthesis was made - I’d gone too far to use them. So, I think that’s probably an area where, particularly with MND, we felt that things were happening and deteriorating faster than what … we could get assistance with.’

The provision of technology, services, an MDT and specific MND information and care seems to depend on where a person with MND lives or who they are able to access. Communication about and monitoring of changes in a patient’s health status and needs did not always occur within an MDT,
leading to confusion and emergency hospital admissions. The one participant (P1) with MND interviewed recalled how, following an overnight sleep study to ascertain health difficulties affecting sleep such as respiratory insufficiency, nothing was heard for several weeks and during which time P1’s breathing became more difficult. The situation was only addressed when a friend calling at P1’s home, finding P1 in respiratory distress, made a call to the MND specialist and was advised to take P1 to hospital straight away. The specialist clinicians interviewed for this study admitted that emergency presentations to hospital with respiratory distress occurred in people with MND, and at worst this led to tracheostomy insertion (C19). It was unclear from the data whether this was because of MDT confusion but certainly mixed messages from members of the MDT led to people with MND becoming confused about their care options:

**C1 (palliative care consultant):** *There was one person who was told it [MND] would – he’d known patients where it had burnt out after a few years and not progressed. So those patients – you know, it can be difficult because they may get mixed messages – different messages when they see a neurologist in a clinic who says, “Oh, you shouldn’t be thinking about a gastrostomy” when we’re already starting to talk about gastrostomy.*

Some of the specialist teams discussed and prepared their patients for emergency MND presentations to hospitals by giving cards or information with their palliative care specialist contact details. This enabled the emergency department doctors to contact them for guidance on a disease they may have had little experience with (C14, C19 and C7).

The clinician participants within this study recognised that for some people with MND, travelling between health professionals within an MDT whose members were not co-located caused practical problems, particularly as the disease became more advanced. The point was made that fragmentation of the MDT also caused difficulties communicating between team members with repercussions for continuity of appropriate care for the patient (C12).
C1 (palliative care consultant): I feel it might be better if there was more of us in the clinic together. And I know in many clinics you see the doctor, and then they see the nurse, and then they see the OT, and then they see the physio, and then they see the speech and language therapist, and then they see the psychologist, and then – you know. And I can see the positives of that, but I also feel it would be exhausting.

C11 (respiratory specialist): That’s often in the practical world really quite difficult with people who are, you know, poorly mobile, whose prognosis is very limited, and who you just want to maximise, you know, the time that they have, and you don’t want to waste their time by getting them in for lots of appointments.

The communication between organisations, particularly if there was no structured MDT incorporating palliative care or palliative care nurses with an understanding of MND, caused issues around patient “ownership” and perceived interference (C11, C12, and C1):

C4 (respiratory specialist): Look, I don’t think it’s [communication between healthcare professionals] particularly ideal at all. The communication we have is by letter, very occasionally a phone call. I don’t get a lot of – I mean, I write letters, and I think I’m communicating, but I don’t feel I get a lot of feedback, unless there’s a big problem.

C16 (MND clinical nurse consultant): Even though I might have requested certain things on referral be done, the person who picks up that referral – without even knowing the patient, and completely sometimes disregarding my clinical judgment – might just say to a patient, “Well, this person just needs a physio review, or an OT review,” forgetting all the other allied healthcare that might be needed.
Lack of co-ordinated care and communication between organisations within the same city was a concern of several clinicians, and there appeared to be a varying level of regard for the quality and approach of some services (C11, C16, and C1, C19). This is exemplified by C16:

**C16 (MND clinical nurse consultant):** It's like a hotchpotch. Like with anything, you know some palliative care teams we work really closely with; some palliative care teams don’t want to know about it … Some pall. care teams have this weird definition period that they will not accept the patient until they have three months or less to live.

Further, there were times when test result reports between health professionals appeared to be lost within a system and not available at specialist consultations (P1, C16). Families expressed concern and frustration that diagnostic reports did not ever reach their GP or specialists involved locally (F6). Delay in hearing from specialists was an issue for both health providers and families. This was raised as a concern by a person with MND who felt the delay in receiving results from a sleep test meant delay in getting the correct symptom relief measures:

**P1 (person with MND):** I didn’t get – this is one of the things that sort of upset us a little bit. We didn’t get any feedback, you know, from the overnight [sleep study] stay. And I felt afterwards that I could have been on the machine two or three months before, because of the deterioration.

Inappropriate direction to other services was another concern voiced by one of the bereaved caregivers (F4). The example they gave was of a health professional who had felt a mobility device could ease an issue that the person with MND and their caregiver were experiencing, but it wasn’t practical and caused more worry:
F4 (family): *Then he started relating to the fact that, “Oh, we got all sorts of devices. You know, we got these lifts and we got this, and we put them on there.”. But, you know, the nurses have to have two people to operate it, but, you know, you’re expected to operate it by yourself, you know?*

Other family members wondered whether it was wise of healthcare professionals to suggest additional aids or other services knowing delays to getting them were likely, as typified by F6’s comment below:

**F6 (family):** *Oh, yeah, she, you know, had all the speech, dietician. But I think at times, to be honest, people – the professionals – I think professionals should feel it’s okay to – they need to – if you can’t do anything. Or that the client has chosen they don’t want any more help as well, because it’s another person that Dad’s dealing with making appointments, receiving phone calls. So it’s almost taking the time out of the precious bit of time you’ve got left.*

This research found that the clinician participants agreed that communication between clinicians involved with the care of a person with MND could be improved and was at times confusing. The family participants made the point that time became increasingly valuable as the person with MND became more unwell. Having to repeat issues or symptoms to many people involved with the care of the person with MND took away valued time.

**4.9.1. Advance care directives or care plan to be in place prior to NIV**

Planning for the end of life and discussing and preparing an AHD is a recommendation of the current MND guidelines (Andersen et al., 2012; NICE, 2016). The AHD should detail what is to happen in the event of a concurrent infection such as aspiration pneumonia, where the person may wish to die, whether or when NIV should be removed or nutrition via PEG ceased. Being able to communicate or be understood when the ability to talk has gone, was a point illustrated by C6:
C6 (neurological nurse): *His wife wanted a definite sign of when she would stop the PEG feeds, if he was no longer able to communicate. I haven’t seen the advance health directive, but she did say there was a special sign that they would have.*

This example illustrates a problem that may exist more widely; although there was understanding between the couple about the cessation of treatment, it was unclear if this was an arrangement that had been documented or was known by healthcare professionals. C1 illustrated the challenge of ascertaining exactly what a person with MND may want, by describing a situation where he discussed when NIV was to be withdrawn with a patient if he can’t communicate. However, the issue then becomes what exactly does “communicate” mean? Clarity in documenting exactly what type of communication is acceptable to all and understood by clinicians and the person with MND (being able to talk, blink or by some special sign) in relation to ending NIV or nutrition via a PEG is vital, particularly if a concise AHD has not been documented. Some clinicians stated that to have a precise document expressing a person’s end-of-life wishes took more experience and time than they could provide (C8, C10); this further reinforces time poverty as an issue.

The clinician participants were asked at what point of the disease they discussed AHDs with people with MND and whether their families were involved with the decisions made. Clinicians appear to make the decision on when to initiate discussions about end-of-life care and document outcomes of these discussions in an AHD on an ad hoc basis:

C13 (social worker): *We go pretty much according to what the patients can cope with and what they seek, and if we feel they can deal with the information then, yes, we – we can do it even from the first visit if it comes up.*
C7 (neurological nurse): The wife was quite happy with those choices, but one of his daughters, who was a nurse, was not. So, there was a lot of conflict about this daughter not - you know, wanting all interventions: PEG feeds, etc. So, there was a lot of conflict making it distressful for the client and for the spouse.

Many of the clinicians discussed the importance of developing a rapport with their patient before discussing AHDs (C13, C14, C19), but as some clinicians only saw their country patients for one or two visits, this was not always possible (C11, C12).

The clinicians were asked whether the AHDs were reviewed when a person with MND had interventions such as a PEG or NIV. Whilst believing the AHD should probably be discussed further at such times, few actually incorporated any possible issues associated with symptom control measures into the document:

C2 (palliative care specialist): Not routinely. They should be, but I don’t think they are in clinical practice. And I think although we would assume that all of these patients do [revisit their AHDs], I think it would be very interesting to know how many actually did, because I’m not at all convinced that they do.

C18 (rehabilitation consultant): Look, we haven’t [reviewed AHDs] a lot. We should have. xx [the palliative care consultant] does them all when they are – when they do need to be done, but, no, I think that’s a hole in our service.

The clinicians were also all asked whether they found that the AHDs were sufficiently detailed in order to be useful when clarity regarding treatment was required:

C2 (palliative care specialist): Lots of people’s AHDs are very vague. They give you a – I think they give you a general direction, though, as to whether
someone wants aggressive or limited treatment. But they’re often not much more help than that.

C3 agreed with C2’s comment about the vagueness of some AHDs and gave the example of a recent document they received which had read pain relief only, but on further discussion, the person concerned actually wanted comfort measures such as medications for nausea and respiratory secretions. Apparently, the person had not been offered guidance from any healthcare professional or legal representative, so completed the document to the best of their ability. This was then signed by the GP and dated the day before the person had signed and dated it, making the document completely invalid. C11 suggested that perhaps structured AHDs were not the best option in clarifying end-of-life choices, and that to prepare a person or proxy decision-maker who had a good understanding of the likely decisions which may have to be made, may be more effective than writing down every eventuality, which may not be possible. C12 and C13 made the point that when AHDs are initially discussed, the person with MND and their family may request all treatment possible, but as time goes on, their approach to living longer may change. Conversely C16 shared the experience of caring for people with MND who initially stated they did not want any interventions, but when faced with an inability to swallow or breathe, requested PEG and NIV. C19 cited aspiration pneumonia as a good example of potential lack of consistency between what is stated in an AHD and actual patient and clinician behaviour at the pertinent time. The example he gave is one in which a patient stated that no antibiotics were to be used, but then presented to casualty with aspiration pneumonia and was given antibiotics:

**C19 (palliative care consultant):** It’s almost like pulling of the – like a scene out of a western. One person’s pulling out an advance care [directive], the other person’s giving the antibiotic [that has been declined in that advance care directive], and you’re thinking, “Okay, well, everybody’s not harmed. Okay, it’s happened. Okay, let’s now think about the future.”
A few of the clinicians either stated that they lacked sufficient experience to advise and assist with AHDs (C8, C10) or referred their patients back to the GP (C4). Several of the clinicians, like C19 above, described that in their experience, most AHDs lacked sufficient clarity to be useful (C7, C11, C15, C16, C2), although all saw the value of having them. As participant C5 says:

C5 (gastroenterologist): The guidance surrounding [the] terminal stage of care is poor and can lead to difficult decisions especially if AHD are not in place.

Despite understanding the importance of AHDs, family members F4 and F5 had not managed to encourage their family member with MND to document their end-of-life choices, and each expressed regret about this. On reflection participant F5 thought this was probably a form of denial within the family, although F4 stated it was because the person with MND simply did not want the conversation as she “did not want to die” (F4).

Three of the families interviewed (Table 4.3) did appear to either have had a formal AHD (F3) or have had an informal written agreement between the person with MND and the family members (F1 and F2) about end-of-life care. In all cases these family participants reinforced the clinicians’ perception of the AHD as usually fairly ambiguous and not always known to all relevant persons. F1, for example, stated that the person with MND had refused anything invasive and that the document was unsigned by them. This document had apparently not been shown to the doctors but was known to the community palliative care team. Despite a formal AHD, F3 did admit that the person with MND procrastinated in writing and signing the document as the person “did not want to think about it too much” (F3). F3 reported that the document contained the following:

F3 (family): If a sort of condition developed that was incapable of being sort of effectively treated then he was palliative care only.
Ambiguity and lack of clearly written AHDs was found in this research, with much of a patient’s end-of-life wishes known to the families but not necessarily to the attending clinicians. At times, this resulted in treatment that was perhaps futile and not what the person with MND had wanted.

4.9.2. Family issues and suggestions

The country and outer metro families interviewed mentioned the difficulties with travelling to appointments particularly as their family member’s health deteriorated and increased fatigue and moving becoming increasingly difficult (F1, F2, F6):

F1(family): *She was exhausted by the time she got there and she just – we couldn’t get from the car to the hospital to get a wheelchair to wheel her to the doctor’s surgery.*

One family from a rural area thought that the gap between specialist appointments was too long. Initially at least, the appointments were every three months and it felt a very long time for the family who accepted that they did not necessarily hear and fully comprehend what the specialists had said to them (F5). F5 went on to suggest that a mentoring system would be useful so that information could be obtained from people with experience of MND, particularly other caregivers. F5 observed that she would have appreciated more involvement with people who had been through the MND experience:

F5: *Yeah. I find that really difficult living in the country, because I don’t feel that a lot of local nurses or medical people have had a lot to do with it. I know they have, because we’ve had some people in our area with motor neurone. But it’s talking to carers is the biggest communication need that I have really, because they have the on-the-ground tips to give you, you know.*

Family member F3 suggested that genetic testing should become more available and included in MND communication. F3 related the experience of her
family member with MND knowing that MND can be inherited and worrying about their children and grandchildren. However, this comes with its own problems as clearly outlined by a very experienced palliative care specialist:

**C1 (palliative care consultant):** The family can’t help because they’re just terrified, and I think that’s the – the other issue, I think, is going to get worse – I mean, we’ve had it with two families now with C9orf [genetic link]. One lady had four daughters. So, while they were sitting there watching her die, two of them, in theory, would be seeing how they would die. And so, you know, the impartial family isn’t any more, because they’re even more involved than the patient, and the patient, if they have some insight, is seeing, “I’m putting this on my children who might be facing this as well.”

F6, who had much experience in negotiating the nuances of the health system and had acquired excellent health literacy, suggested:

**F6 (family):** They’ve still got to be professionally responsible, but maybe they need to start out differently by asking the question, “Do you want to eat, or do you want to have this intervention?” And it’s okay for the person to say, “No,” and the health professional has to accept that, and I suppose document … Not that I’m thinking of, because maybe we as a family do ask the right questions, and that’s a real key, is the health literacy, isn’t it, that people haven’t got.

F1 recommended greater GP awareness of MND particularly relating to referral to a neurologist if there is no explanation for breathlessness or loss of speech. F2 felt that the hard questions and hard decisions must be addressed and not pushed to one side, by either families or clinicians. F4 was still affected by how his family member with MND had suffered for so long, and despite not liking to admit it, thought it would have been better if his family member had been allowed to die:
F4 (family): Yeah, basically, yes, yes … nine months to a year before she did [died].

4.10. Patient and Family Communication and Understanding

The clinical participants were asked if, in their experience, people with MND and their families were adequately aware of the prognosis and limitations of the disease prior to their involvement. Denial and lack of understanding by some people with MND and their families relating to how quickly the disease can progress, was expressed as a communication barrier by some of the clinician participants. However, it was apparent to clinician participants that some people with MND and their families had experienced limited explanation of the disease:

C2 (palliative care consultant): Some of them have clearly been told but don’t necessarily want to hear, understand or acknowledge, and some clearly have very limited explanation.

C13 (social worker): I don’t think they’ve always understood how quickly things might be changing for them as an individual. And I don’t think they’ve always fully understood the complications that can happen, you know, and the interventions that may or may not be possible.

The majority (n=14) of the respiratory and palliative care specialists and nurses interviewed talked of trying to ascertain the depth of knowledge of the person with MND relating to their diagnosis and what they had previously understood from other health professionals, prior to their own involvement. This was attempted in several ways: repetitive discussions, asking for further clarification on fragments of information from the person with MND, reflective discussion and repetition of what the person with MND and their family had understood. One clinician stated that trying to disentangle where people were in their journey and in their understanding, would “contextualise” any advice that
was then given to them (C11). An example of trying to ascertain what had been understood about all that had been communicated to the person with MND and their family is given by C11:

**C11 (respiratory specialist):** *In a perfect world I use, you know, a lot of communication skills like check back and cross-check, and things like that, to see what they’ve understood. In the real world, I use some of that.*

However, understanding how much a person understood and what they had previously been told was frequently found to be difficult, both to assess and to discuss, as C6 and C2 described:

**C6 (neurological nurse):** *Sometimes you can see that they’re not processing information. You can just see that look of confusion or terror, or blankness on their face. Others, you think that they’ve understood and then you find out later that they haven’t actually understood. They might be repetitive in a question or just say something that makes you realise they haven’t understood’*

**C2 (palliative care specialist):** *We’re palliative care doctors. We like to think that we’re good at communicating, but I think, I acknowledge, that often families need to hear things multiple times.*

The NICE Assessment and Management Guidelines recommend that clinicians ask people with MND how much information they wish to receive and how much they would like to have shared with their families (NICE, 2016). One of the clinical nurse participants described how clinician communication can adversely affect people with MND:

**C16 (MND clinical nurse consultant):** *They’re kind of more psychologically scarred about how diagnosis was given to them, and then how they’ve been sent home to put their stuff in order, and, you know, wait for death to come.*
None of the six people with MND whose families were interviewed had chosen withdrawal of NIV as a method of ending their life. The family member of F3 with MND, a night time user of NIV only, chose instead to refuse food and water to end his life. However, a discussion had taken place with the family, although not documented within his AHD, that NIV could be withdrawn but it would be when the person with MND was ready. However, F3 did state that initially NIV had only been mentioned in passing as a treatment option for later and that the detail of NIV had been communicated by two neurological nurses, not specialists. F4 supposed that NIV may have been mentioned to his family member, but neither the person with MND nor F4 pushed for more information as F4 felt prolonging the illness or suffering was not their preferred option. F5 stated that the NIV was given to them with little alternative, whilst P1 had no real idea or explanation of the implications of NIV:

F5 (family): *It was definitely, “This is what you need because your lungs aren’t working properly.”*

P1 (person with MND): *I think probably I wasn’t aware of the importance with the breathing and I don’t think that that was probably adequately explained. Now, they were saying, “Well, you go and have a sleepover; we’ll test to see what – you know, how you’re sleeping, how you’re breathing,” all those sorts of things. But there wasn’t any sort of gutsy type of verbiage about why … the implications of it.*

F1 and F2 said liquid morphine was suggested to assist their family member to breath but nothing else was suggested until their family member was entering the terminal stage at which point a syringe driver was offered. There was confusion between the siblings in their understanding regarding a syringe driver, as one had understood it to be intravenous delivery of medication (F2) whilst the other had understood subcutaneous infusion (F1); both had attended the same conversation with clinicians:
F2 (family): *I think the reason Mum didn’t want … the IV was because she just didn’t want to accept that she was going to go, and that looked like another step towards the end of life. And having someone in every day.*

The candidate asked if the family member with MND was offered a subcutaneous cannula just under the skin, which is the more normal option with a syringe driver, rather than intravenous. The second family participant (F1) responded that she understood that it was the subcutaneous option offered not intravenous. Both F1 and F2 agreed that the partner of their family member with MND was very hesitant about allowing liquid morphine to be used as he felt his partner could become addicted to the morphine. This perhaps indicates a lack of either communication reassuring the family, or a lack of understanding and health literacy by the partner of the person with MND. Neither F1 nor F2 had any recollection of the burdens of NIV being discussed until such a time when their family member started to get pressure sores on her nose and mouth from the NIV mask. At this point they were informed: “Oh, yes, that happens – that has, you know, been known to happen before” (F1). Apparently, there had not been any communication with F1 and F2 about the potential of withdrawal of NIV when it was no longer effective or the person with MND had decided they had suffered enough. F2 stated that the lack of ability of the person with MND to communicate as the disease progressed was not discussed and that this led to him feeling his family member may have made some ill-defined choices, potentially resulting in increased suffering. F1 described a lack of recognition or at least indication to the family by the clinicians, that the person with MND had reached the terminal stage:

F1 (family): *But still they didn’t think she was at end-of-life stage, but I think she was at end-of-life stage at that stage, and we could have helped.*

F4 was very determined to keep his family member with MND at home and wondered if this may have been why end-of-life choices were not discussed with him. F5 described that the communication surrounding NIV was well
covered particularly by the respiratory technician; however, this was not the case with communication relating to the PEG insertion, which was information F5 found out via the Internet. F3 had no recollection of end-of-life communication in the context of NIV and definitely none from the respiratory clinician. F3 candidly acknowledged that the family had the “intellectual and financial resources” to be able to look after their family member with MND at home:

**F3 (family):** As I say, we were very lucky in the sense that we had the financial, intellectual and emotional resources to be able to manage the situation with assistance at home for most of the time.

As discussed earlier in this chapter, the clinicians interviewed for the study described several reasons for not broaching the difficult conversations surrounding NIV withdrawal and end-of-life choices. These reasons and the barriers for end-of-life communication and the benefits and burdens of symptom control measures offered, are discussed further in Section 4.11 of this chapter.

### 4.10.1. Differentiating between hydration and nutrition

One of the questions asked of the clinician and family participants related to the hydration of the person with MND in the last few days of life, particularly if they were refusing food and water and especially if they had a PEG inserted. The clinicians were asked if, when detailing the consequences of stopping nutrition as a means of ending life, they had described nutrition and hydration separately. The clinicians were also asked if they had experience of people with MND indicating that they were dehydrated or that there was a possibility that they could be suffering as a consequence of dehydration if their fluid intake had been ceased when nutrition was ceased:

**C15 (gastroenterologist):** Potentially, yes.

**C19 (palliative care consultant):** That’s a really good question, isn’t it, hydration – differentiate hydration and nutrition.
Some clinicians stated that they had experience with people with MND apparently indicating symptoms of thirst or dehydration towards the end of life after stopping fluids and nutrition (C7, C8). C11 was unsure if hydration was related to the secretions (excessive or tenacious), and C18 felt nutrition was the most important aspect to discuss and did not really distinguish between nutrition and hydration. C14 stated that generally when the person with MND decided that nutrition should stop, water was continued to prevent dehydration although stopped if clinically indicated. C19 considered the symptoms of hunger and thirst stating that:

**C19 (palliative care consultant):** They may well feel thirsty, and that needs to be attended to, because that’s a terrible symptom if you’re to sit – if you’re lying there and you can barely communicate. So, we need to – I agree, I agree. This [small amount of water via a PEG] is not going to cause any harm to the patient, and it’s not going to elongate life.

C15 said that on consideration of the point that whilst the person with MND may want to stop nutrition, hydration may prevent the feelings of thirst. This was also asked in the context of the AHD and whether the AHD was changed in accordance with the patient’s wishes. The clinician participants were asked whether the differentiation between hydration and nutrition was ever offered, suggested or documented within an AHD:

**C15 (gastroenterologist):** I have to say I haven’t really pushed that differentiation on advance care directives. But I think it is important, and perhaps in future I will.

C1 and C3 stated they would continue with hydration and did differentiate between nutrition and hydration for people with MND and their families. C4 and C2 did not particularly differentiate between food and fluids when discussing PEGs to people with MND but C4 acknowledged that for people using a mask for NIV, thirst and a dry mouth were a potential problem anyway.
The families of people with MND were asked whether those who chose to refuse fluids as well as nutrition were comfortable or whether thirst and dryness may have caused adverse symptoms. F3 was asked whether the oral preparations they used were adequate to alleviate the feeling of dryness in the person with MND following a decision to stop food and water via the PEG/RIG by her family member with MND. F3 stated: “possibly not, you know”. Despite a recommendation of hydration and using various oral rehydration products for the symptoms of dryness for the person with MND, the family member F6 was sure suffering occurred as a consequence:

**F6**: She was constantly, constantly, constantly trying to – yeah, absolutely she did [feel thirsty]. You know, with – it was an overwhelming feeling.

F6 continued to explain how her family member had tenacious secretions and was being inadequately suctioned by nurses in the hospital. F6 suggested that suctioning is “not recommended by MNDA” and the health professionals were reluctant to assist. Apparently, and after much negotiating with the hospital, the partner of the person with MND was allowed to suction their family member. F6 readily admitted that the entire process of suctioning her family member was distressing but did appear to improve her symptoms.

**F2**: So, you know, Mum and Dad got to the point where they were so exhausted they couldn’t make decisions.

To summarise, this research has found that clinicians may not differentiate between nutrition and hydration when communicating end-of-life symptom control to people with MND and their families. Acknowledging that not all people with MND have PEG tubes at the end of their lives, this research has found that some people with MND were described by their families as appearing to suffer from thirst in the terminal stage of their disease.
4.11. Communication Barriers from Clinician and Family Perspective

Both clinician and family participants agreed there were barriers to communicating end-of-life choices, documenting patient wishes (AHDs or clear care plans/goals of care) and the implications of the withdrawal of NIV.

C11 (respiratory specialist): For some other people, you know, an extra minute of life is unbelievably precious regardless of their state of health and ability to interact. I think that that means we need to re-imagine the workforce issues rather than we need to throw our hands in the air and say it can’t be done.

One of the most noted barriers clinician participants was patient denial. When asked how the clinicians dealt with denial:

C4 (respiratory specialist): “Oh, I just don’t want to think about it. I don’t want to know.” So that can be a bit of a challenge that their defence mechanism, their way of coping is one of, “That’s later on,” denial.

C16 (family): I let them be in denial.

Another barrier to communicating NIV withdrawal prior to commencement mentioned by one of the more experienced clinicians (C19 and reiterated by C18, C13, and C4) was the concern that having sensitive end-of-life discussions would upset the patient or the family:

C19 (palliative care consultant): I think one of the barriers is sensitivity or worrying that that’s – even having that conversation is going to upset the patient or the family.

C4 (respiratory specialist): I’ve got one particular lady now that those issues are just too hard for her to discuss. And there’s a lot – even I feel uncomfortable – I feel comfortable about discussing them.
A further barrier to effective communication suggested by a neurological nurse, was if the person with MND perceived themselves as a burden to their family. This was communicated by C7:

**C7 (neurological nurse):** So mostly the reasons have been they don’t want their life prolonged, because that’s how they see it maybe. They don’t want the invasiveness. They don’t want even further carer burden. That seems to be another big thing, the carer burden: “I don’t want to do that to my spouse.”

The information a person with MND and their family may have obtained from the Internet may also pose a barrier to effective communication if not adequately discussed:

**C16 (respiratory nurse consultant):** In many ways we find when patients first come that’s a huge burden that they all have, because they’ve gone on the Internet, they’ve read a lot of stuff about dying and how horrible it is, and, you know – and most patients don’t want to die from that sense – you know, they’ll have that fear of dying from suffocation and pain. And, I guess, you know, having those early interventions – early conversations alleviate that sense.

The following summary (Table 4.5) shows some of the issues which may present as a barrier to clear communication between clinician and people with MND and their families identified by the participants in this study.
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<th>Table 4. 5. Communication Barriers</th>
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<tr>
<td>• Many of the clinicians were reluctant to discuss the poor prognosis of the person with MND too early but admitted that leaving those conversations too late was not a good option for the person with MND either (C6, C4).</td>
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<td>• Worrying that even the conversation will upset the family and person with MND (C19, C6, C7, C14).</td>
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<td>• Sufficient time for detailed communication was a factor mentioned by both clinicians (C2, C3, C4, C11, C12, C19, and C8) and the bereaved family participants (F1, F2).</td>
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<td>• Languages other than English were potential problems with communication, barriers to being able to communicate adequately and misunderstanding, despite the use of interpreters (C12).</td>
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<td>• Communication with a family member because the person with MND does not want to know (C7, C4).</td>
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<td>• Repetition of the end-of-life communication and readiness of the person to want to hear; a person may be told the same thing several times but must be ‘ready to hear it’ (C8, C9).</td>
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<td>• The stigma of MND as becoming a vegetative state or how alive they are until the end (C14).</td>
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<td>• Clinician admitting that a barrier was the uncertainty of the disease and admitting that clinicians did not want to face such uncertainty (C11). Being able to say that not knowing how, when or what will exactly happen and being honest from the beginning (C11) was a barrier.</td>
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<td>• Patients ‘shutting down’ and not wanting to hear or stating they have been told the same thing already (C12).</td>
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<td>• Denial or patient and family reluctance to engage in communicating with their specialists was highlighted by several clinicians (C1, C4, C12 and C16).</td>
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<td>• People with MND need to be ready to listen and hear end-of-life options (C8).</td>
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<td>• Uncertainty to the ethical situation and the law can be a barrier for clinicians to discuss withdrawal of NIV (C5).</td>
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<td>• Time (life) left as a barrier; communication should happen early particularly as families are from all over the world, conflict may be involved and planning for the whole family important (C3, C2).</td>
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<td>• People with MND delaying engaging in end-of-life symptom control choices as they wish to try alternative medicinal cures (C18).</td>
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<td>• People with MND and their families who lack assertiveness and confidence to ask questions (F5, F6, C3).</td>
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<td>• Clinicians just have a snapshot view of the disease and do not look at a trend (F3).</td>
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4.12. Chapter Summary

To answer the research questions, the candidate commenced by undertaking an extensive literature review to ascertain what was already known about the communication surrounding initiation and withdrawal of NIV (Chapter 2). The depth and extent of relevant knowledge determined the independent variables and formulated the questions asked of the participants. The semi-structured interviews with all participants were at least an hour in length and comprehensively covered communication and topics relevant to NIV, MND and the end of life.

Whilst the discussion (Chapter 5) that follows explores in greater depth the areas of interest that have emerged from analysis of the data, a description of the analysis has been provided within this chapter. The research questions explored within this study include how clinicians explained the benefits and burdens of NIV, the continuation of disease progression, referral to palliative care and end-of-life care and choices including NIV withdrawal. The comments and views of the family participants have been included and woven throughout the chapter to add context and perspective to the clinicians’ descriptive narrative. The presented data have been compared against the most recently published recommendations for assessment and care of a person with MND, the use of and potential withdrawal of NIV and, in addition, the understanding and use of PEG tubes in MND. Some of the findings which relate to recommendations from both the family and clinician participants, are included in the recommendations and conclusion chapter, answering the last of the research questions.

This research found that some of the difficult communication such as NIV withdrawal and choices of end-of-life care were either assumed to have occurred with other clinicians, were too difficult for a clinician to fully explain in the time available or left too late for sufficient patient benefit and family understanding. The time and repetition required for ongoing and lengthy
communication relating to end-of-life choices and the removal of NIV was an issue voiced by all the palliative care and respiratory specialists and some of the nurses. Aspects of the most recent recommendations and assessment guidelines (NICE, 2016) which have been referred to throughout this research, have been questioned by both clinicians and family participants. The concept of the need to preserve hope in relation to the difficult discussions and honest communication about end-of-life choices and the potential of NIV withdrawal is considered in the following discussion chapter. A comparison of the findings of this study against recommendations in guidelines is also presented in Chapter 5.

It is hoped that by understanding both the participant clinician comments and the actuality of the bereaved families, particularly the one person with MND who so desired to have his opinions heard, that a greater understanding of MND/NIV/PEG and end-of-life communication has been achieved. The potential for the generation of practice knowledge is more fully discussed in Chapters 5 and 6.
Chapter 5

Discussion

5.1. Chapter Overview

This chapter integrates all that has been studied within this thesis, from the introduction of the subject, the gap identified from the literature and the findings of the participant interviews.

To undertake a detailed discussion of the findings of this research it is important to highlight what is understood to be best practice and patient-centred care in relation to the area under enquiry. The most recent MND recommendations and guidelines have informed the participant questions. Attention to the theoretical perspective, SI, which draws on the shared experiences of the participants, is woven throughout the discussion to make meaning of the findings. Using ID as the methodology to contextualise the findings of this research, the knowledge gained is argued to be relevant and applicable to clinical practice (Thorne, Reimer Kirkham, et al., 2004).

The contribution this research has made to the existing knowledge concerning the communication and symptom control measures for people with MND is explained. Understanding the communication difficulties encountered by people with MND, their families and clinicians alike, formed the principal component of this study. The Dreyfus Model of Skill Acquisition (Dreyfus & Dreyfus, 1986) is used as a concept to understand clinician participant communication in relation to experience.

Identifying where issues arose has provided the opportunity to propose possible solutions and offer some suggestions for further research areas (detailed in Chapter 6).
The final five research questions (the first research question was answered by the literature review) are considered in turn:

1) What is the evidence that the best practice communication content (benefits and burdens of NIV, continued disease progression, explanation and offer of a PEG, end-of-life discussions and understanding potential NIV withdrawal prior to commencement of NIV) is incorporated into the reported communication by clinicians to people with MND and their families at the time of offering NIV?

2) What do MND families/significant others recall about the communication (benefits and burdens, continued disease progression, PEG insertion, potential NIV withdrawal and end-of-life discussions) provided by clinicians at the time of respiratory failure and the offer of NIV?

3) To what degree do MND families/significant others feel that the NIV communication provided by the clinicians at the time of the offer of NIV prepared them for and helped them to understand the implications of potential NIV withdrawal resulting in the death of their family member?

4) What, if any, unmet needs for information, support, palliative care, or other services do the clinicians and the families/significant others (or patients themselves) with experience of MND identify?

5) Following the literature review and clinician and family/significant other interviews regarding NIV communication, what recommendations can be identified that would improve end-of-life communication and process for the patients, families and clinicians involved with the end-of-life care for people with MND?

This chapter commences with a summary of the findings from this research to provide context for the discussion that follows. The chapter is then sub-divided under identified communication issues and discussed within the framework of the research questions which are outlined above.
5.2. Summary of the Key Findings from the Research

The following is a summary of the key findings from this research which will be expanded upon in this chapter:

1) Some of the clinicians are unaware of the specifics of the recommendations contained in the NICE MND Assessment and Management Guidelines (2016).

2) These recommendations are open to clinician interpretation as far as what, when, how and if to communicate end-of-life care options.

3) There is uncertainty around whether the clinicians with experience have the capacity to communicate more clearly with people with MND regarding symptom control and end-of-life choices, but they are known not to follow guideline recommendations about the timing of communication.

4) Communicating end-of-life options and the potential withdrawal of NIV prior to the commencement of NIV is difficult for all involved regardless of a clinician’s years of experience.

5) Clinicians experience limitations on the time available to talk to patients, and so prioritise explaining the technicalities, maintenance and mechanics of NIV to people with NIV and their families. Some of the respiratory clinicians reported that they often feel it is not appropriate to introduce NIV and its benefits and state its burdens and the potential of withdrawal as an end-of-life choice when NIV is no longer of any benefit, at the same time.

6) Many clinicians are reluctant to make an early referral for people with MND to palliative care despite acknowledging this to be best practice.

7) Some palliative care clinicians feel that on referral, people with MND are under-informed about the disease process, prognosis and symptom control options.

8) Families are confused by the need for early involvement of palliative care.
9) Structured MND MDTs were rarely available particularly outside the cities and often within cities. The benefits of an inclusive MND MDT for people with MND are unavailable in many areas (Rooney et al., 2015).

10) NIV and PEGs are often encouraged by clinicians. Respiratory clinicians see NIV as improving the quality of life, perhaps equating quality of life with quantity, which may not be the priority of the people with MND and their caregivers.

11) PEGs are commonly offered to people with MND to prevent or reduce the incidence of aspiration.

12) Communication is often open to misinterpretation such as the word “prognosis” and what the speciality of palliative care can do.

13) Families are confused about the roles and number of health professionals involved with their family member with MND’s care; having to repeat the same information is frustrating and exhausting for them.

14) There is confusion regarding who is involved with the patient’s ongoing care decisions, among both clinicians and family and caregivers. Between clinicians there appears to be confusion regarding which clinician has overall responsibility for care and for communicating and explaining the benefits and burdens of symptom control and end-of-life options. If information relating to symptom control and end-of-life choices is not clearly provided, the person with MND and their family have gaps in their understanding and interpretation of the situation. The family may be confused as to who they ask for clarity.

5.3. Research Question 2:

What is the evidence that the best practice communication content (benefits and burdens of NIV, continued disease progression, explanation and offer of a PEG, end-of-life discussions and understanding potential NIV withdrawal prior to commencement of NIV) is incorporated into the reported communication by clinicians to people with MND and their families at the time of offering NIV?
This research question, concerning clinicians’ communication practice, was answered by asking clinicians rather than through an objective assessment of what clinicians did. This sets the context and limitations of interpretation. Clinicians were asked whether they were aware of aspects of the NICE MND guidelines and if there were specific trigger points they used as an opening for honest discussions including assessing how the person with MND and their family had understood such communication. The clinicians were also asked whether they discussed end-of-life options as recommended by the most recent guidelines, or if they communicated the benefits, burdens and potential withdrawal of NIV when initiating NIV. Their answers reflect their insights into where they do not comply with current best practice as defined in the guidelines, and the barriers they experience in implementing the communication recommendations.

The NICE MND Assessment and Management Guidelines (2016) is an extensive, detailed document easily obtainable online and available to download. However, the lengthy document covers an enormous amount of detail and time would be required to fully appreciate its recommendations. Whilst many of the clinicians were aware of the most recent NICE MND Assessment and Management Guidelines (2016) not all were aware of the recommendations on timing and content of NIV related end-of-life communication. Detailed end-of-life communication was assessed to be necessary by the clinicians involved later in the disease, indicating that earlier discussions delivering the diagnosis and general prognosis may not have contained such communication. This research sought evidence for the occurrence of best practice communication relating to the benefits and burdens of NIV, end-of-life choices and potential of NIV withdrawal. This research has found that communication recommended to occur soon after diagnosis and at certain trigger points in the disease process, particularly at the commencement of NIV, frequently does not happen.
Understanding the communication experiences family participants have with clinicians is an important aspect of answering this research question. The family perspective is addressed within this question but covered more comprehensively under the third research question (Section 5.5). This research could not correlate the family participants’ experiences of communication with those of the clinician participants.

The ability to initiate end-of-life communication on patient deterioration and increased reliance on caregivers is extremely difficult for all concerned (Aoun et al., 2012). This difficult judgement of timing appears to be complicated by the clinician perspective of maintaining hope for the person with MND and their family members. Preserving the feeling of hope by clinicians and, therefore, not initiating these discussions was found to be a barrier and a strong theme throughout this research. This finding is mirrored by the results of a study which surveyed neurologists involved with the delivery of an MND diagnosis and found the communication caused much stress for the clinicians for the same reason (Aoun et al., 2016). Maintaining hope, however, may be detrimental to the patient and family if referral to palliative care specialists is delayed until well into the deteriorating phase of the disease. Despite many of the clinicians identifying the barriers, having the confidence to know that the communication is appropriate and the timing correct for the recipient is a challenge. The respiratory clinicians in this research indicated that their priority when initiating NIV was of a more practical nature: explaining the machine and familiarising the family with its use. The respiratory clinicians acknowledged the difficulty they had with introducing end-of-life discussions at the stage of commencing NIV, although they understood it should happen.

When communicating the benefits and burdens of NIV and initiating referral to palliative care, several of the clinicians stated they were reserved in their communication. The time required at appointments to introduce and reiterate the end-of-life options and potential withdrawal of NIV to ensure the person with MND understood, was described as a significant barrier. Both
communication and system barriers were identified in this research. These and other barriers to the incorporation of best practice communication in the management of MND are detailed further below. A table of communication barriers identified by the clinician and family participants in this research is included in Chapter 4 (Table 4.5). The issues found to have most influence on clinicians’ communication with people and families coping with MND were the availability of time and the wish to preserve hope.

5.3.1. System barrier: time allocated for significant communication

The MND Assessment and Management Guideline recommendations (NICE, 2016) emphasise the importance of psychological and caregiver support from the time of diagnosis. Proposed within the European Clinical MND guidelines are two long appointments with a neurologist in order that the diagnosis can be delivered in a sensitive, comprehensible and gentle manner (Andersen et al., 2012). As the delivery of a diagnosis of MND is recognised as requiring lengthy appointment times with the neurologist, so perhaps should the appointment time for communication which incorporates end-of-life choices. Many of the clinician participants within this study reported having insufficient time for these discussions (C2, C3, C4, C8, C10, C11, C12 and C19). A recently published journal article which addressed ethical considerations and palliative care in people with MND, notes the variation in how people process bad news (Danel-Brunaud et al., 2017). Danel-Brunaud et al. (2017, p. 301) reaffirm the need for time to explain and discuss issues with people with MND and their caregivers, suggesting several long interviews to make sure the communication has been understood. A study into the importance of compassion among health professionals from a cancer patient’s perspective found that patients wish to be treated like a person, not the illness they have (Sinclair et al., 2016). Family participants within the study recommended that health professionals understand their patients more holistically. Again, this ability to understand individual patients, their life, and their preferences of care requires time. Building a relationship between patient and clinician by involving palliative care services soon after diagnosis or by having a nurse case
coordinator (neurological or palliative care) to build a trusting relationship may enhance end-of-life communication.

This research has extended existing knowledge on whether the most recent MND Assessment and Management (NICE, 2016) recommendations are being followed and, if not, why not. The candid statements from the clinicians explain the difficulty many find in judging the right time to introduce end-of-life communication without causing the person with MND and their family excessive distress. They also shed light on the time constraints within the medical system which act as barriers to long and repetitive communication. In the Clinical Practice Guidelines for communicating end-of-life issues with adults in the advanced stages of a life limiting illness (Clayton, Tattersall, Currow, & Hancock, 2007), uninterrupted time for such communication is considered essential. Finding the right time and finding sufficient time are often inextricably linked. The lack of time required for such involved, emotional and often repetitive communication with people with MND was a concern voiced by many of the clinician participants (C2, C11 and C12), and reiterated by the families (F5, F6). The clinicians stated that they simply did not have time for the required conversations which included choices of symptom relief end-of-life care and hoped, and in many cases assumed, other health professionals did. As admitted by many of the clinician participants, adequate time to have sensitive discussions relating to the burdens of NIV and end-of-life choices became secondary to addressing the technical aspects of care (C2, C11, C12). Several of the respiratory clinicians explained that they only saw country people with MND when they became symptomatic and required NIV and so explaining the technicalities of the NIV machine took precedence and required explicit training prior to a patients’ return to the country. Adequate time to have the more sensitive communication was simply not factored into the appointments. This research did not delve into the reasons why time could not be allocated for longer specialist appointments to incorporate all aspects of communication surrounding the benefits and burdens of NIV and end-of-life choices, and this may be an area requiring further (political/financial) investigation.
5.3.2. System barrier: delays and poor communication within the health system

The timing of referral to a specialist (neurologist or respiratory specialist particularly) of symptomatic but not necessarily diagnosed people with MND was found to be slow. Families mentioned a delay or hesitancy in being referred on to a specialist for symptoms that had been investigated but not easily explained such as respiratory difficulties (F1, F6 and P1). Comments from some of the bereaved family participants suggested that MND was not often contemplated as a diagnosis by the more generalist doctors (F1, F6, and P1). This may not have been because the diagnosis of MND was not contemplated by clinicians but because they wanted to be sure of a diagnosis given the gravity of the disease. However, the family participants had not understood the delay in reacting to non-specific symptoms. Many and varied exploratory tests proved negative for any specific disease but difficulty breathing continued unexplained (F1, F2). All the family participants understood that there is not a specific diagnostic test for MND and that diagnosis is more a process of eliminating other diseases. However, even once the MND diagnosis had been made one participant family described how their family member was subjected to further uncomfortable and seemingly unnecessary tests to reconfirm the diagnosis (F6). This again reflects the need for improved communication between clinicians and people with MND, particularly relating to potential respiratory issues.

Family participants volunteered information about how the diagnosis of MND was given to their family member. Although this question was not specifically asked, their comments exposed further communication issues. The timing and delivery of the MND diagnosis in each of the participant families and the one person with MND interviewed was poorly conveyed by the specialists. This finding aligns with that of a recent national survey of neurologists undertaken to establish how MND diagnoses are given (Aoun et al., 2016), which found that once diagnosed there was frequently a delay in the results of
tests reaching the person with MND and at times, also in reaching the clinician most closely involved with the patient. By the time the results were made known to the person with MND, an increase in severity of the symptoms had frequently occurred. This finding was consistent with research conducted by McConigley et al. (2014). Their research found that symptom relieving needs had often changed by the time a person with MND had been assessed and subsequently provided with devices to relieve their symptoms (McConigley et al., 2014). Understandably, the difficulty was due to the progressive and often unpredictable course of the disease which can differ according to the variants of MND. The challenge for the clinician was to predict the course of the disease and act on the symptoms without delay. The family participants in this research acknowledged the work and advice given by MNDAust with the supply of equipment, information and support for people with MND and their families. A suggestion from one of the family participants (F5) living in a rural area was the implementation of an online support and discussion forum for people who were also traversing the MND journey so that others could benefit from shared experiences. One family (F6) learnt much of their MND knowledge and gained support from an online social site in the United States, having been unable to find such a resource in Australia.

Practice that aligns with guidelines and recommendations may remain an aspiration: indeed, good intentions to implement these have been demonstrated by the sincere comments from clinician participants within this research (C1-C19). The possibility of clinicians being guided by means other than formal recommendations and MND guidelines was considered a possibility. For example, early palliative care referral for people with MND is a guideline recommendation acknowledged by many of the clinician participants to be important but not necessarily reflected in practice. The clinicians involved prior to palliative care found early communication relating to referral to palliative care and end-of-life choices when these clinicians were offering symptom control measures such as NIV and PEGS could take away hope. This corresponds with
an article by O'Connor (2012) which highlights palliative care intervention occurring too late to be effective. O'Connor suggested that in many instances palliative care recommendations developed by experts were often regarded as optional by clinicians (O'Connor, 2012). The palliative care clinicians interviewed for this study said they were often involved late in the course of the disease and at times stated an earlier referral would have been more beneficial to the person with MND (C1, C2, C3 and C19). This was reiterated by some of the respiratory specialists (C11, C12, and C16) who also felt their input at an earlier stage of MND may improve adverse symptoms. This study found, as other research studies have before, that referral to palliative care in MND often occurs later than that recommended by the most recent guidelines (Bede et al., 2011; Connolly et al., 2015). Referral to a palliative care service is discussed further in Section 5.6.3. When asked how they communicated (i.e., verbal, written document) end-of-life options, including the potential of NIV withdrawal, the specialists involved suggested to patients that they look at the MNDA website. Only in some cases had printed material from MNDA been prepared to give to the people with MND. One family participant stated that following diagnosis she was told not to contact MNDA, as they were described by the clinician as being too negative (F3).

It was evident from the results of this research that when a structured MDT existed, relevant information was provided to the patient when it was considered appropriate after diagnosis, leading to clarity and choice in end-of-life care. A structured MDT also incorporated a palliative care specialist at an earlier stage of the disease, which may have improved patient acceptability of incorporating palliative care earlier into their care.

5.3.3. Communication barriers: clinician knowledge and experience

Within this research the European (Andersen et al., 2012) and U.K. (NICE, 2016) guidelines in particular have been extensively quoted and referred to as the most recently published recommendations for MND. This research into communication surrounding NIV in MND and end-of-life choices sought expert
and experienced clinicians for their views. In order to clarify what may be understood to be an “expert” this research includes a reference to the Dreyfus Model of Skill Acquisition (Dreyfus & Dreyfus, 1986) which has been widely used in research on acquiring skills including the ability to communicate clearly. The Dreyfus model researched how people attained skills and was based on the study of chess players and members of the military, and subsequently used as a model in nursing research (Benner, 1984). The model is based on five levels of skill attainment from novice to expert, with experience and time as the factors for acquiring skills. The skill components include perspective, ability to make decisions and ability to decide relevance of a situation. The expert level suggests a minimal reliance on rules and guidelines as the expert intuitively knows what can be achieved (Dreyfus & Dreyfus, 1986). If the Dreyfus Model of Skill Acquisition is considered in the context of this research then the expert is the clinician (respiratory or palliative care) with experience of communicating with people with MND and who is able to achieve excellence without much difficulty (Benner, 1984). This excellence includes the ability to communicate the difficult end-of-life choices people with MND and their families must make. The findings in this research did not suggest that clinician experience in MND increased the likelihood of following guideline recommended communication, including end-of-life choices and NIV withdrawal, prior to the commencement of NIV.

This research has identified that whilst the guidelines are generally recognised by the clinicians, aspects of their content are not widely followed. The length of time a clinician had been practising in their specialty did not necessarily make the sensitive communication surrounding the end of life and the potential of NIV withdrawal any easier and in some cases did not occur. The idea of acquiring clinical and communication knowledge without necessarily referring to guidelines was explored further in this research.

A study by Gabbay and May (2004) suggested that expertise in clinical decision-making and care was achieved by unconscious assimilation of
knowledge. This knowledge was communally strengthened by collegial discussion and thought sharing (SI) rather than exploring specific clinical guidelines. This concept, known as *mindlines* (Gabbay & May, 2004), revealed that health professionals infrequently acquired guidance through clinical guidelines and research articles; more often they gained their knowledge through discussion with their peers and occasional readings. As identified in Chapter 2, the recommendations in the current MND guidelines are largely based on published studies and expert clinical opinion (Andersen et al., 2012; NICE, 2016). The clinician participants in this research appeared to be basing their communication decisions more on their own personal characteristics, those of the patient and their family, their ability to interact and level of skill in communication rather than reading and following the guideline recommendations. The possibility exists that the further away the clinicians are from their formal education and the more experienced they become, the less likely they are to adhere to guideline recommendations. The personal clinical experiences the participants had with NIV withdrawal in MND were found to have some influence on their communication. The reasons and barriers behind the clinician communication decisions are discussed in this chapter and the implications of their decisions for the families are explored.

### 5.3.4. Communication barrier: preservation of hope

The most recent MND guidelines recommend sensitive communication occurs when the person is ready to receive it and relies on an emotional judgment and assessment of readiness of the patient and family by the clinician (Andersen et al., 2012; NICE, 2016). However, if end-of-life choices have not previously been discussed or understood this communication should occur when the respiratory system fails and NIV/PEGs are offered (Andersen et al., 2012; MNDA, 2014; NICE, 2016). Whilst the guideline recommendations advocate a specific trigger or time for such communication, this research found that sensitive end-of-life communication does not always occur when recommended. The timing of end-of-life communication identified within this research appears to be largely dependent on when a clinician is ready or feels
the person with MND is ready to receive it, concurring with previously published literature (Connolly et al., 2015; Smith, Disler, Jenkins, Ingham, & Davidson, 2017).

Clinicians assess patient and family readiness for end-of-life discussions and respond to patient and family triggering behaviour or emotional indications of wanting to have such communication (Clayton, Hancock, Butow, Tattersall, & Currow, 2007). Clinician knowledge of the guidelines and other literature relating to NIV was found to be variable and, understandably, appears to depend on their clinical specialty and involvement with MND. Any previous experience of NIV withdrawal in MND on the part of the clinician appeared to have little effect in easing difficult early end-of-life communication, particularly in relation to NIV withdrawal.

Coming to terms with the diagnosis and its implications is extremely confronting and may be cloaked with the hope that “they got it wrong”. As this study has found, Blackhall (2012) has suggested that clinicians with little experience of people with MND wait for the patient or family to initiate the discussions relating to the end of life to lessen the sense of hopelessness.

The preservation of hope and the concern that the conversation might upset the family was a barrier for many clinicians to pursuing end-of-life communication (C6, C7, C14 and C19). Clinicians also stated that in their experience many people with MND and/or their families expressed the need to live in the present as the future looked so bleak. However, if the clinicians wait for the person with MND to initiate or ask relevant questions they may be assuming that the person with MND has sufficient health literacy or understanding to approach such difficult discussions. Studies that suggest people with MND would like the clinicians to initiate sensitive communication exist (Anderson et al., 2007; Blackhall, 2012). Commencing end-of-life communication without waiting for the patient to instigate the discussions may help to eliminate the possible difficulties encountered by those less able to ask
pertinent questions. As some of the bereaved family participants acknowledged, traversing the medical system and having the confidence and knowledge to ask the relevant questions required a high level of health literacy (F3, F6). All the bereaved family participants and the person with MND interviewed stated they searched the Internet for information, particularly if they had not understood fully what had been said by the health professionals. Potentially, the assumption that people with MND will turn to the Internet to address their questions and fears may lessen the urgency clinicians feel to initiate such sensitive communication.

If clinicians acknowledge that end-of-life discussions should happen early, but also feel that it takes away hope, when should these discussions occur? A recent German article (Löbbe, 2016) suggests that there are too many guidelines in existence and that they do not always prove useful in complex clinical situations. The article makes the point that evidence-based medicine strives to provide the most advantageous care to patients, but that there are other aspects of a clinician’s expertise which may equally provide the best care. Löbbe (2016. p. 275) suggests that good communication technique, experience and “willingness of the physician to make a decision contrary to the evidence-based guidelines” are important factors in best practice. Importantly, this viewpoint may mean confusion regarding who should undertake the difficult end-of-life discussions relating to NIV and its withdrawal, and when they should or could occur. For the study reported in this research, 19 clinicians (eight of whom were nurses) were interviewed, and the majority (seven out of eight) nurse participants accepted the need to have these discussions and were willing and felt able to do so despite sometimes being “hated” (C8) by the patient for doing so. The nurses felt the person with MND and their family were entitled to be able to plan in the full understanding of their disease potential. This concurs with the findings of the study by Connolly, Galvin and Hardiman (2015) which concluded that families were better able to make plans and symptom-relieving treatment decisions early if given the options for care. The feeling amongst the clinicians in this research who felt able to undertake these discussions early in the diagnosis (C1, C16, C17) was that it alleviated the
worry associated with the unknown course of the disease, particularly the issue of choking to death which may concern them throughout the illness if not tackled early (Borasio, Shaw, et al., 2001).

5.3.5. Communication barrier: frontotemporal dementia

Identifying FTD was difficult for health professionals particularly if they only met the patient once or twice, which was often the case, particularly between country patients and respiratory specialists (C1, C15, C19). The point was made, however, that a person with MND with changes related to FTD may not have a deep enough understanding of their health situation to make decisions, which would affect their caregivers (C13). The family participants in this research were largely unaware of FTD in MND as the condition had not been discussed by their clinicians, although two families had found information on the Internet (F3, F6).

A palliative care specialist (C1) stated concerns that FTD in MND was an area requiring more research and understanding and often a condition going unrecognised by clinicians and families. One of the specialist clinicians had experience with people holding very strong beliefs against nutrition through the insertion of PEGs (C15). Whilst this was certainly presented as a barrier for PEG insertion in some people, the clinician wondered whether perhaps this indicated involvement of FTD affecting decision making. One of the family participants described a situation where their family member with MND was taken shopping and despite not being able to swallow solid food herself, kept collecting chocolate bars into the shopping basket (F1, F2). The family initially thought this was “naughty” behaviour but described how the person with MND was also insistent that her husband’s birthday was imminent and wanted to send a card when the birthday had been some months prior. Despite the family explaining correct dates, their family member with MND persisted. The family concerned were vaguely aware of dementia being associated with MND but had not associated this behaviour with FTD and had, therefore, not discussed it with their palliative care specialists.
The research question addressed in this section asked for evidence that best practice communication is incorporated into communication for people with MND at the time of offering NIV, as reported by clinicians. The clinician participants involved at the stage of required symptom control intervention (NIV and PEG) appeared not to follow the recommended best practice communication and stated their barriers and reasons. In this research, time to discuss and repeat sensitive end-of-life communication was found to be a major barrier to commencing such communication in many instances. This research found that communication is only as useful as the content is unambiguous, the clarity with which it is delivered (in this research communicated by the clinicians), and the willingness and ability to understand and remember by the one who receives the communication. This is particularly relevant when considering the often-subtle changes associated with FTD. The clinician participants in this research admitted that recognising FTD in MND was difficult especially if the family had not communicated personality changes in their family member to the clinician. The often-subtle changes that may indicate FTD could be an aspect of early support and coordination of care involving a case manager soon after diagnosis. The suggestions to improve communication difficulties made by participants in conjunction with the candidates own suggestions are encompassed in Chapter 6.

5.4. Research Question 3:
What do MND families/significant others recall about the communication (benefits and burdens, continued disease progression, PEG insertion, potential NIV withdrawal and end-of-life discussions) provided by clinicians at the time of respiratory failure and the offer of NIV?

This question was answered by the bereaved families of people with MND through their recollections about the communication they had with
clinicians relating to when NIV was discussed, the content of the discussions and what they had understood about end-of-life care options. Their answers show what they understood of their choices and what, in retrospect, they realise they had not fully understood.

The guidelines advise that the benefits and burdens of NIV and PEGs, and the option that NIV can be stopped at any time, should be communicated prior to the commencement of NIV (NICE, 2016. The families were asked about any alternatives to NIV and PEGs offered as symptom control methods. The point of this line of questioning was to ascertain whether the benefits and burdens of NIV and PEGs had been fully described, and then whether pharmaceutical options which may mean a shorter life, were communicated to the person with MND and their families. Of concern was that PEG insertion was described to families as a way to minimise or prevent aspiration pneumonia, for which the research literature shows there is no benefit (F1, F2, F6) (Potack & Chokhavatia, 2008). The families confirmed that much of what they understood about MND, NIV and PEGs was either found or clarified via the Internet. Families also stated that being asked to think about the research questions had made them realise the implications of aspects of NIV and end-of-life communication they had not fully understood at the time; for example, the withdrawal of NIV when a person with MND no longer benefited from it and the option of pharmaceutical alternatives. The content of communication given to the family and person with MND regarding treatment options was found to greatly influence the decisions surrounding symptom intervention and care choices.

It became evident that the family participants were presented with overwhelming and confusing information particularly relating to NIV. This research did not ask about family health literacy, however, many of the participants (family and clinician) spoke about the need for a good understanding of the health system and health literacy. Family participant understanding for the potential of withdrawing NIV when it was no longer
providing symptom relief or at the person’s request, was not clearly understood by the families. Communicating the need to insert a PEG (if the person with MND wants one) prior to any swallowing difficulty, has caused some confusion for people with MND. Understanding that MND causes respiratory weakness which in turn increases a person’s anaesthetic risk for inserting a PEG requires a fair level of health literacy. Clinicians recommending inserting a PEG when the person’s swallowing remains unaffected, the benefits and burdens of such a measure, requires good communication skills and confidence, and patients and families need the knowledge necessary to ask relevant questions. Many of the family participants within this study were confused by the timing of PEG insertion and were unaware of any risks of aspiration pneumonia (F1, F2, F3 and F6) (Potack & Chokhavatia, 2008). Despite guidelines recommending communication of the benefits and burdens of NIV and PEG tubes prior to or, at the latest, at the development of respiratory weakness, clinicians and families in this research state that this did not necessarily occur (NICE, 2016).

Generally, the family participants and the one person interviewed with MND agreed that they understood the workings of the NIV machine as explained by the respiratory clinician. This corresponds with the respiratory clinicians in this research stating that they focus on the workings and benefits of NIV. The families also understood that the NIV did not provide any cessation of the disease process and that they would become more dependent on NIV as the disease progressed. However, the families did not fully appreciate the difficulties they may then encounter such as any influence FTD may have on their family member (not discussed with any of the family participants) and the increase in caregiver involvement to ensure the NIV continued to work (F1, F2, F3). The burdens of wearing and adjusting an NIV mask cannot be fully understood until trialled, and the understanding that whilst the person with MND may be able to initially adjust their own mask, as muscle weakness becomes more pronounced their caregivers will by default become more involved.
The family participants were asked if they were aware of the inevitable disease progression despite NIV and PEG insertion and if this had been communicated to them. The families all understood the severity of the diagnosis but not the speed at which the disease could progress nor the limitations of the interventions available. None of the bereaved families had anticipated how their family member with MND would die even though they all understood respiratory failure to be the main cause of death in MND. In this study one person with MND starved himself to death (F3); another kept trying to pull off her NIV mask for unknown reasons, possibly indicating “enough” (F1, F2); another pulled out her PEG refusing further intervention (F4); and another died of respiratory complications (F6). Of the six family participants interviewed, all but one had a negative experience of the terminal stage of their family members’ life.

Within the sociological framework of SI, the way the family participants interpreted the communication during the social interaction with their clinicians affected the meaning they took from those interactions. The participants identified barriers including being overwhelmed by the concept of an early referral to palliative care. Family participant understanding of the burdens of NIV and benefits of PEGs was unclear from the communication they had with clinicians and was supplemented by their Internet sourced knowledge. Suggestions to improve communication incorporating end-of-life issues and the benefits and burdens of NIV/PEGS by the family participants are considered in Chapter 6, when the final research question is answered.

5.5. Research Question 4:

To what degree do MND families/significant others feel that the NIV communication provided by the clinicians at the time of the offer of NIV prepared them for and helped them to understand the implications of potential NIV withdrawal resulting in the death of their family member?
The family participants in this research were questioned on many aspects of their communication with clinicians and how their resultant knowledge enabled them to adequately make choices. The ability to adequately understand how to navigate an increasingly multifaceted health system requires considerable health literacy. The lack of communication about how the health systems worked and could be accessed, was voiced by the families who were at times overwhelmed. As they endeavoured to navigate the care and services required, the family participants described the difficulties they experienced in understanding why they had to attend different appointments with different clinicians in different locations. In an editorial by O'Connor (2012), discussing palliative care recommendations in Europe, the point is made that some skill or health literacy is required to traverse health systems.

The variability and accessibility of palliative care services for people with MND is apparent within this research. The gap between rural and metropolitan palliative care services and even within the same city appears to be mirrored in some other countries (O'Connor, 2012). This variability makes provision for a specific plan of care for people with MND, which incorporates an MDT difficult to design. There is a need for a more patient-focused plan with emphasis on early recognition of symptoms and information on who to contact locally. Such a plan requires further development and research, but from discussion with participants within this study, this was considered a useful ideal.

Apparent within this study were the emotional consequences for all concerned, particularly the grieving families, when the end-of-life care options had been misunderstood (F1, F2). Whether the option of NIV withdrawal was inadequately communicated, undocumented or not communicated at all to families by the time the person with MND had lost the ability to communicate remains unclear from this relatively small participant group. Generally, the clinicians could see some benefit of early communication about NIV withdrawal, particularly in terms of ensuring transparency of information. However, the majority felt the information was excessively confronting for the person with
MND at the time of offering NIV. If end-of-life choices have not previously been discussed or understood this communication should occur when the respiratory system fails and NIV/PEGs are offered (Andersen et al., 2012; MND Aust, 2014; NICE, 2016).

Previous studies have found that many people with MND and their families would like clinicians to commence end-of-life communication early in the diagnosis in order to organise their lives and make health care choices (Anderson et al., 2007; Blackhall, 2012). Crisis decisions and presentations to hospitals can occur if documented care plans relating to NIV in MND are not in place and understood by families (Connolly et al., 2015). The family participants (F3, F5) within this research, whilst acknowledging a degree of their own denial in some instances, did suggest that in hindsight some of the decisions made would have been different if they had fully understood the choices of symptom control and end-of-life care.

The influences on communication and understanding have been illustrated throughout this research and has demonstrated how denial and health literacy have affected how the family participants communicated with the clinicians (F3, F5, and F6). The perception among some of the clinicians that the preservation of hope outweighs the requirement to fully communicate the end-of-life choices suggests the clinician has assumed the dominant role within the patient/health professional relationship.

From an SI perspective, the role of the clinician and the families and how they interact and communicate can potentially be affected by stress and attitude within the process of role interaction (Hochschild & Machung, 1989). The SI focus on process and the influence of interacting personalities was proposed by Burgess (1931) in relationship to family personalities and roles. Applying the concept introduced by Burgess and identifying family in a broader sense to include the clinicians, person with MND and the family, roles are developed and defined by the interacting personalities (Burgess, 1931). This process may be
fluid, however, values and attitudes are transmitted between all concerned in communicating the prospects of MND, which may ultimately affect the way the patient interprets their options for care.

Research into the dying process in MND in Australia and the United Kingdom, found negative caregiver experiences were mainly due to the unexpected death of their family member from co-morbidities, not respiratory failure as originally anticipated (Ray et al., 2014). The study by Ray, French and Street (2014) indicated that if families and people with MND were fully informed, including about the progressive nature of the disease and how death may eventuate, the family members were found to have had a more positive experience of their family member’s end of life. This finding by Ray et al. correlates with the results of this research where those experiencing ongoing grieving and regrets in the way their family member died, or their symptom control prior to death, were those who felt they either did not have or did not understand the potential complexities of end-of-life choices (F1, F2 and F6). A limitation to answering the fourth research question may be that those families wishing to participate in this study are those who may have ongoing grieving issues related to the way their family member had died.

The difficulties in communicating and encouraging planning in a disease which offers no hope of a cure and only offers symptom control measures is an area that requires further understanding. As some variants of MND may have a life span of 10 years, the concept and treatment of the disease as chronic rather than terminal may be more appropriate in some cases, particularly concerning end-of-life communication (Lerum et al., 2016). None of the participant families within this research experienced the variant of MND which can last for many years. However, despite the length of their family member’s life from diagnosis to death being less than three years, all felt palliative care involvement and end-of-life communication at or soon after diagnosis was too premature. This research found that the family participants had an image of palliative care as terminal care and death-related. This concept may be perpetuated when
clinicians want to preserve hope in people with MND and, therefore, avoid referring to palliative care at a time they consider too early in the disease. Perhaps if the person with MND and their families better understood the significance of any symptoms they were experiencing they may feel more inclined to seek specialist advice. From the time of diagnosis then, the emphasis could be on explaining symptoms and who to contact in the event of such symptoms occurring, instead of focussing on the end of the disease. The timing of end-of-life communication and the involvement of a palliative care service requires further exploration to alleviate the anguish of families planning for their family member’s terminal stage.

The ethical and legal aspects of NIV withdrawal was raised as an issue which could be confusing for families and health professionals alike. When discussing the withdrawal of NIV, clinicians must be able to reassure families that the withdrawal is not euthanasia nor an illegal or immoral act. This communication should acknowledge the grief and sadness the family may experience, but also reiterate that the disease itself will bring the person with MND to their death. The importance of having conversations which include disease progression despite any clinical intervention prior to the commencement of NIV and that NIV withdrawal is a possibility at any time the person with MND wishes is paramount to clear communication. Communication ambiguity was illustrated by a family participant (F1) whose family member was using NIV in her terminal stage and kept pulling the mask off. The family did not know that their family member could have chosen much earlier on, or at that stage, to cease using NIV and have pharmacological assistance for her distressing symptoms (F1, F2). The bereaved participants involved stated that in hindsight, the clinicians may have alluded to such a course of action, but the family had not understood the communication or the implications. The clinicians generally tried to assess how much the person with MND and their family had understood of the communication incorporating end-of-life care by asking what they already knew and repeating the information where necessary. However,
this was often hampered by the level of health literacy of the person with MND and family and where the person was in their disease trajectory.

The family participant component of this study was small, yet the results did concur with the study by Anderson et al. (2007) which found clear communication relating to end-of-life options was welcomed by most people with MND and their families. Previously published literature found that most people with MND and their families would like clinicians to initiate end-of-life communication soon after diagnosis and to further discuss this as deterioration of the disease occurs (Anderson et al., 2007; Astrow et al., 2008).

The “in hindsight” comments of some of the family participants within this research described how, had they understood the course of MND more completely, particularly the symptoms and speed of deterioration of the disease, their end-of-life decisions on behalf of their family member (at the stage when the person with MND was too ill to clearly communicate their wishes) may have been different (F1, F2, P1). Potential withdrawal of NIV prior to the commencement of the symptom relieving device was not clearly communicated to any of the family participants within this study, although two families had discovered this was a possibility from the Internet (F3 and F6). The communication and emotional difficulties surrounding NIV withdrawal faced by clinicians and families are encapsulated within studies (Faull & Oliver, 2016; Faull, Phelps, Regen, Oliver, et al., 2014) and these difficulties were restated by clinician participants within this study (C1, C4, C8, C10, C11 and C19). Some of the complications expressed by the clinicians entailed ensuring the medications were adequate to guarantee comfort on withdrawal of the NIV and coping with the emotions of family members present. This was particularly evident if the family of the person with MND had not fully understood or were not in agreement with the wishes of their family member with MND.
5.6. Research Question 5:

What, if any, unmet needs for information, support, palliative care, or other services do the clinicians and the families/significant others (or patients themselves) with experience of MND identify?

The family participants were asked how satisfied they were with the end-of-life communication delivered by the clinicians, and how this communication enabled them to make their care choices (Appendix E). The family participants were also asked whether there were discussions which they would have liked to occur, but which did not. The following sections are intended to highlight areas as described by the families, which this research has identified as being problematic for them. The communication and logistic issues are presented in the context of areas recommended within the most recent MND Assessment and Management Guidelines (2016) and other substantiating literature.

Faull, Rowe-Haynes et al. (2014) highlighted a need to support all involved with NIV withdrawal in MND and for detailed communication between all parties regarding symptom management and NIV withdrawal. Increased palliative care nurse involvement from diagnosis, perhaps taking the role of a case manager coordinating with an MDT where available, is a recommendation of this study and further examined in Chapter 6. Specific published research into a nurse as an MND coordinator of care could not be found, although this research found two situations in New South Wales where the clinician participants stated this was occurring very efficiently (C14, C16).

5.6.1. Communication identifying differences in the concept of suffering

This research found, as Lemoignan and Ells (2010) have, that quality of life and the concept of suffering (which may equate to living longer with symptom control devices) was poorly understood by those with MND and the families involved with their care. Several of the families (and confirmed by some of the respiratory specialists interviewed) stated that the technical component of
NIV information was prioritised by the clinicians over the understanding of respiratory failure and quality of life. In this research it was queried whether the often-stated clinician view that the suffering of a person with MND would be improved by alleviating respiratory distress with NIV, potentially extending a person’s life (by a few months), was actually increasing their suffering. However, when family participants considered length of survival as a priority over distressing symptoms such as agitation, mask discomfort, frustration and increased caregiver burden it was apparent that an increase in longevity was not a priority, particularly as the disease progressed.

The issues surrounding caregiver burden in MND were not specifically explored within this research as previously noted and has been well researched by others (Aoun et al., 2013). However, it was highlighted as a problem by some families (F1, F4, and F6) and nurses. The interviews from this study revealed that caregiver burden was exacerbated by prolonging their family member’s life when all options for the end of life and symptom control had not been clearly explained. The fatigue, confusion and desperation experienced by the family members F1 and F2 in not understanding that their family member with MND could have had the NIV withdrawn, was profound.

5.6.2. Where a person with MND lives influences communication and care

This research interviewed clinicians from different areas of Australia and found that even within cities it depended on whether a person with MND fell into a certain catchment area as to whether they could access a specialised MND MDT, particularly one incorporating specialist palliative care. People with MND who fell under the umbrella of a city service with a specific MND team, living close enough so that either they could access a comprehensive clinic, or the palliative care specialist/nurse could easily visit appear to be at a huge advantage. The comprehensive clinics are able to assist people with MND and their families to understand their equipment (and back up provision), the
services on offer (including support groups), and to navigate all aspects of allied health (Rooney et al., 2015). Delays in people with MND accessing a specialist MDT (incorporating palliative care), were partly due to the time diagnosis may have taken (during which time the person could have become more symptomatic), and partly due to patient and system barriers. This research concurs with a retrospective study which followed patients from early identification of MND symptoms to the time they were referred to an MDT incorporating palliative care (Galvin et al., 2015). Galvin et al. (2015) found a failure by some clinicians to refer patients to an MDT caused delay in patients having the assistance they may have required.

At the commencement of this study, consideration was given to possible differences in availability of palliative care services and specialists generally between metropolitan and rural areas. A systematic review by Kirby et al., (2016) identified that diagnosis and treatment of people with a terminal illness was less well managed in rural situations when compared to metropolitan areas. The review focused on the caregiver needs of urban and rural patients with a terminal illness and led the authors to suggest further research was needed into how a lack of rural accessibility to palliative care shapes the needs of the person with a terminal illness (Kirby et al., 2016). The difference in availability of specialist palliative care services between rural and metropolitan areas may have played a part in referral time for palliative care, however, this research has also identified the variation of access to palliative care services available within the same city.

5.6.3. Early incorporation of palliative care

Nearly 10 years ago the suggestion was made that in order to refer a patient beneficially and appropriately to a palliative care service the referring clinician could ask themselves a simple question: “Would I be surprised if the patient died within the next twelve months?”, thereby ensuring the referring clinician thought of probable rather than possible death (Moss et al., 2008). In a more recent systematic review, the “surprise question” was found to be an
inadequate prognostic tool particularly with non-cancer conditions (Downar, Goldman, Pinto, Englesakis, & Adhikari, 2017). This has relevance to this research and certainly to further research, as it is unknown how many clinicians may have asked themselves this question and, therefore, not referred people with MND to a palliative care service. This would likely apply more to respiratory specialists than GPs (who may only encounter a few cases of MND in their career) because respiratory clinicians see people with MND as the respiratory muscles are adversely affected. Within the guidelines early referral to palliative care, soon after diagnosis, is recommended for people with MND (Connolly et al., 2015; MNDAust, 2014; NICE, 2016). The bereaved family participants, person with MND and the clinicians all acknowledged that referral to specialists for symptom control measures often happen too late during the disease to be helpful. However, early referral to palliative care was viewed by the families and person with MND and some clinicians (C1, C2, C3) rather differently.

The Australian Institute of Health and Welfare (2014) suggests that specialist palliative care, end-of-life care and palliative care are all terms used interchangeably. The institute suggests that end-of-life care is commonly understood to mean caring for a person with the diagnosis of a life-limiting illness, whilst palliative care is considered to be the approach to terminal care. One of the key areas identified was the need for clarity in understanding terminology among patients, their families and healthcare workers to avoid miscommunication, particularly with terms such as palliative care and advance care planning (Australian Institute of Health and Welfare, 2012). Given the common misunderstanding of the term palliative care - found also within this research (F5, F3) - to mean no hope and death rather than symptom control and support, early referral to the specialist service can be confronting for patients (Galvin et al., 2015). A European study focused on the early referral of people with lung cancer to a palliative care service found that patients do not like being referred to specialist palliative care services (despite available specialist palliative care services) and were, therefore, frequently referred late in the disease process (Charalambous, Pallis, Hasan, & O’Brien, 2014). Despite
an early referral to specialist palliative care being a current recommendation for people with lung cancer (Smith et al., 2012), the study by Charalambous, Pallis, Hasan and O'Brien (2014) found this was not being translated into clinical practice. In a study by Oliver et al. (2011) palliative care specialists have found that people with MND are often referred late in the disease process. Several studies have found clinicians like to preserve hope by not initiating early communication surrounding the end of life (Aoun et al., 2016; Faull, Rowe-Haynes, et al., 2014) and this finding is confirmed and discussed comprehensively within this chapter.

Whilst most of the palliative care clinicians felt they were generally involved later in the course of MND than they would have liked, there was some thought that contrary to the recommended guidelines, referral at diagnosis may be too soon (C1, C2, C3). As MND could in some instances be considered a life-limiting chronic disease, early palliative care may not always be appropriate. This study concurred with the suggestion in an editorial by Burgess, Braunack Mayer, Crawford and Beilby (2013) that people with a life-limiting chronic disease, which may describe some variants of MND, be initiated on patient symptoms and not solely on diagnosis. Most of the palliative care and respiratory clinicians stated that referral at diagnosis of MND was recommended but went on to qualify this by stating it did not often occur as it was too confronting for the person with MND and their families in some instances.

As none of the families with MND were referred to palliative care at diagnosis, the acceptability of this cannot be determined from this study. Overall the families thought that palliative care involvement was appropriate as their family member deteriorated. Conversely, some of the clinicians argued that if palliative care teams were not involved early, symptoms may be missed that could have been ameliorated and prevent unwanted hospital admissions (C1, C11, and C12). The finding in this research that if families (P1) were not fully informed early of their choices of care it may lead to emergency hospital
presentations agreed with Connolly, Galvin and Hardiman (2015). However, the manner in which this communication occurs and whether it necessarily needs to be given by palliative care specialists is highlighted within this study.

The symbolic association of palliative care with no hope emerged from this research, both from the clinicians’ reluctance for early referral to the specialist service, and from the families’ pre-conceived opinions. Certain environments can create feelings of uncertainty and how the individual interacts with that environment may change depending on previous experiences or experiences of others which may have influenced the individual (Blumer, 1969). Palliative care for many people is assumed to be solely involved with the end of life. The individual interpretation and meaning of an environment such as palliative care is not necessarily conforming to a social construct, but more to experiences and individual analysis. This negative image of palliative care has the potential of being reinforced by clinicians who hesitate to explain and communicate the many aspects of the service in the belief any hope a person with MND may have, will be eliminated.

There were also practical reasons mentioned by the clinical participants as to why early referral to palliative care was not happening in their districts. In particular, the unpredictability of the course of MND, the availability, funding rules and budget of the local palliative care teams and in some instances the preferences of those clinicians involved with the diagnosis of MND (C19, C1, and C2). None of the family participants interviewed thought that palliative care from diagnosis was appropriate and felt it to be too confronting.

All the family participants involved with this study were very prepared to be referred and understood the value of referral to a respiratory or gastric specialist as symptoms became evident or worsened. However, the concept of early referral to a palliative care specialist for symptom relief seemed unacceptable. If this is an “image problem” for palliative care, then perhaps there needs to be a subset of the specialty which is called MND symptom
control. The inconsistencies within health areas for the provision of palliative care, particularly in the more urban areas in Australia, the varying levels of palliative care offered and the mixed opinions of when palliative care should be usefully offered, appear diverse from this study.

5.6.4. Multidisciplinary teams

The inclusion of palliative care within the MDT is a current recommendation for the management of MND (NICE, 2016). There is evidence that MDTs can offer health benefits to patients in most areas of disease management (Marsilio, Torbica, & Villa, 2016; Rooney et al., 2015). In Europe, there are MND guidelines which not only recommend referral to a recognised MND Association on diagnosis but also referral to a specialist MND MDT (Andersen et al., 2012). A recent study in Ireland found that people with MND survived approximately seven months longer if under the supervision and care of a MND specific MDT (Rooney et al., 2015). In the study by Rooney et al. (2015) the benefits of a well-structured, centralised MDT were suggested to have the ability to increase the life span of those diagnosed with MND. Rooney et al. explored and compared whether centralised MDT services would have a better survival outcome than community-based care which incorporated a care coordinator. The study concluded that centralised MDT offered a holistic clinical environment for the person with MND and their families, which improved their clinical well-being and survival. The study proposed that this may be due in part to the variation in perspectives and in communicating choices of care between clinicians, patient and caregiver. Rooney et al. imply that this varied input into decision-making can enable clearer understanding of clinical possibilities which can then be incorporated into an AHD.

However, as found in this research, structured MDTs are not always readily available in Australia. There was considerable variation found in the geographical co-location of the health professionals that work together to form an MDT. Ideally the MDT members offer their various skills to improve the disease process of those they help and in doing so share their knowledge with
those within their team. The involvement of multiple organisations; not just those making up the MDT, but those involved as neurological, respiratory or palliative care nurses plus those offering personal care were often uncoordinated and disassociated.

There were comments from all the families regarding the number of people involved with the care of their family member with MND, particularly for those who remained at home. This related to involvement of multiple organisations, even within the MDT, causing confusion for the family and person with MND as to who was responsible for what, and who was organising ongoing care. The MDTs described by family participants appeared, in many instances, to be made up of independent health professionals rather than a formal MDT within one organisation. It is possible that within the more structured, centralised MND MDT this situation does not arise. However, within the small number of family participants, and reiterated by the neurological nurses, confusion does occur. For example, people with MND who lived in country areas were sent home from city hospitals without organised battery backup or contingency plans for power outages or technical problems. Other difficulties voiced by the families included having to attend different specialty appointments in different places, as the MDT was scattered within various organisations. One of the MND nurse specialists involved with trying to coordinate care for people with MND voiced frustration with the lack of feedback she was receiving from within the organisations involved with multidisciplinary care in her area. Another city specialist was frustrated by the lack of cooperation of a country palliative care team post discharge home. It seems that within Australia the level of community palliative care offered varies enormously and depends on the preference of the state or territory health service.

The role of breast cancer nurses has been briefly explored to see whether their model of patient support could be utilised for MND (Jiwa et al., 2013). In the study by Jiwa et al. (2013) the role and availability of the breast cancer nurse negated the need for the person with breast cancer to personally
attend the MDT meetings. The appropriately trained nurse, as case coordinator, was fully cognisant of the health status of the person with breast cancer and was able to report on their behalf to the MDT. The study also found that the sharing of information between health disciplines was challenging, which certainly concurs with the findings from this research (C1, C2, C16). Specialist care nurses, in this case for MND, have been found to play a vital role in the care of people with MND in Scotland (Ferrie, Robertson Rieck, & Watson, 2015). In the comprehensive report by Ferrie, Robertson-Rieck and Watson (2015) into planning care for people living with MND, one of the findings was the importance of the role the specialist care nurses played in coordinating services, care provision and providing advocacy for the person with MND. The nurses, where available, were able to advise availability of local services and provide accurate information which enabled informed decision-making by the person and family coping with MND. However, despite the vital role the specialist nurses played, the report went on to say there were many instances where no one person was identified as taking overall responsibility for assessment and care of the person with MND and their family. Whilst the specialist nurse service is to some extent emulated by the community neurological nurses in Western Australia, their role can overlap with the community palliative care nurses and support workers from MNDAWA. This finding of multiple organisations causing confusion for people with MND was also highlighted in the Scottish MND report by Ferrie et al. (2015) who described the different care and funding systems as a “bewildering array of services and people” (p. 182). Each available service in Australia is extremely valuable but can become confusing for the people with MND and their families.

5.6.5. The usefulness of the advance health directive (AHD) or documented care plan

A documented advance care plan is a recommendation of the most recent MND Assessment and Management Guidelines (2016) and it is suggested that it is discussed prior to the person with MND losing the ability to clearly communicate. Detailed exploration of the content of an AHD was not a
priority of this study; however, the participants were asked at what stage they were discussed and whether they had proved useful. The palliative care specialists generally initiated the AHD. Considering how late in the disease they were involved, perhaps the contents of the AHD may not have assisted the patient appropriately in enabling choices of care. Where the palliative care clinicians had experience with following AHDs the documents were generally vague and inadequate (C11, C15, C18). Conditions such as aspiration pneumonia were not covered, despite recommendation in the current MND guidelines to detail how concurrent infections should be treated (or not) (NICE, 2016). Rather than writing a formal documented care plan, the families seemed to have informal inter-family agreements about what the person with MND would have wanted as they entered the terminal stage of their disease. None of the families interviewed were encouraged to write AHDs early in the disease, but most admitted they probably would not have wanted to do so. The AHD appeared to be as confrontational as the early referral to palliative care. This made treatment choices difficult for clinicians having to rely on the family making difficult decisions, such as to withhold feeding and hydration to enable one person with MND to starve himself to death (C3). The clinician participants were asked if they ever re-visited the AHDs with a patient when a symptom control measure was initiated or if a decision to withdraw NIV under certain circumstances (such as not being able to communicate) was made. Few clinicians other than palliative care specialists involved themselves with AHDs and fewer yet re-visited them to make relevant additions or adjustments, although all thought this a good idea (C11, C15, C18). The American Academy of Neurology (2012) recommends annual communication and documentation of a patient’s preference for continuation and potential withdrawal of NIV and any reasons for not having those discussions to also be clearly documented. Annual discussions, and documentation of these discussion, with patients about continuation of NIV and related symptom control preferences could be an inclusion into an MND specific pathway, was one to be developed (further discussed in Chapter 6).
A recently published journal article which explored NIV in patients’ (with varying diagnoses) perspectives on advance care planning or AHDs found evidence that discussing advance care may increase hope rather than diminish it in people with respiratory failure (Smith et al., 2017). As found in this research, having end-of-life communication and clear understanding on the part of the patient and family may have alleviated some of the concerns and distress leading towards the terminal stage of life (F1, F2, F3, and C7).

5.6.6. Barriers to clinician and family communication

In summary, there were many barriers to effectively communicating symptom control and end-of-life to people with MND and their families. The points below encapsulate the barriers as described by the participants in this research:

- Many of the clinicians were reluctant to discuss the poor prognosis of the person with MND too early but admitted that leaving those conversations too late was not a good option for the person with MND either (C6, C4)
- Clinicians worried that even the conversation will upset the family and person with MND and take away hope (C19, C6, C7, C14)
- Insufficient time available for detailed communication was mentioned by clinicians (C2, C3, C4, C11, C12, C19, and C8) and family participants (F1, F2)
- Time (life) left - communication should happen early, particularly as families are from all over the world, conflict may be involved and planning for the whole family is important (C3, C2)
- A person may be told the same thing several times but must be “ready to hear it” (denial) (C8, C9)
- The uncertainty of the disease and clinicians who did not want to face such uncertainty (C11); clinicians reticent to say they do not know how, when or what will exactly happen and not being honest from the beginning (C11)
Denial about the disease or patient and family reluctance to engage in communicating with their specialists was highlighted by several clinicians (C1, C4, C12 and C16)

Uncertainty around the ethical situation and the law can be a barrier for clinicians to discuss withdrawal of NIV (C1, C5)

People with MND and their families who lack assertiveness, adequate health literacy and the confidence to ask questions (F5, F6, C3).

5.7. Research question 6:

Following the literature review and clinician and family/significant other interviews regarding NIV communication, what recommendations can be identified that would improve end-of-life communication and process for the patients, families and clinicians involved with the end-of-life care of people with MND?

The final research question is answered in the final chapter of this thesis and encapsulates the recommendations of all the participants in this research, and those recommendations suggested by the candidate. Further areas for research are identified, and improvements for the vision of care and communication are suggested.

5.8. Chapter Summary

At the time of writing 50 years have passed (1967-2017) since the founding of modern palliative and hospice care at St Christopher’s Hospice, and in that time there has been much progress towards improving all aspects of end-of-life care including communication. The importance of allowing time to ensure clarity in communicating and documenting the patient’s end-of-life choices irrespective of the diagnosis has been identified.

Within this research there have been three main perspectives to explore, and these were identified by gaps in the literature reviewed (January 1990 to
July 2016). The most recent MND guidelines recommended communication relating to NIV initiation, potential withdrawal and the end of life were compared to the clinician participant interviews (NICE, 2016). The bereaved family participants, who were also the caregivers for the person with MND, were asked what they remembered being communicated to them and what they understood about NIV and end-of-life. The comparison of all material derived from participant interviews to the most recent guidelines has continued throughout the results and the discussion chapters.

Complex processes are at play between those delivering and those receiving communication encompassing end-of-life choices, yet in many instances it is the clinician who decides whether the person with MND is ready to hear such communication. The theoretical approach which underpins this study reveals how varied people are in making sense of their own and others’ behaviours and communication. Symbolic Interactionism has identified how the same experience can have different meanings for everyone. This aspect of the study could be explored further to understand how communication can be delivered in various ways to minimise misapprehension and confusion with end-of-life choices.

The potential burdens of NIV and PEG tubes should be communicated early in the disease process to prevent futile treatment which may prolong suffering. Time to reinforce or repeat end-of-life choices to ensure patient and family comprehension as well as patient and family willingness to participate in such communication are all factors considered by clinicians before commencing sensitive discussions. Patient initiation of sensitive communication and the ability to ask relevant questions in a stressful situation may depend on the health literacy and assertiveness of the person with MND and their family. This was recognised to be the case by some of the family participants within this study, who tended to turn to the Internet for their information. Again, insufficient time is a factor in commencing sensitive discussions and was reiterated by clinicians of all specialties interviewed in this research.
Communication between people with MND, their families and health professionals can be clouded by a mixture of technical jargon, insufficient time to have insightful conversations, the barriers of patient denial and perceived lack of empathy in the clinician. Whilst this is not unique to MND communication (Humphris, 2015), clinician avoidance of difficult discussions has been identified by Faull, Rowe-Haynes and Oliver (2014) in MND and seen again in this research.

The clinicians may find clear direction for end-of-life care in well written AHDs; however, the point has been made by participants within this study that few directives are written down, sufficiently explicit or readily available. None of the clinician or family participants re-visited their AHDs (or where there were no written documents, re-discussed with family members) when situations changed, or the disease progressed. Denial of the incurable nature of the disease is a barrier to clear written treatment choices but even such a barrier may only affect formality in documentation; this research has shown that the preferred end-of-life patient choices often appear to be known by the clinicians either by discussions with family members, or remarks made to the specialist palliative care team at some stage of the disease.

Finally, the recommendations which all participants proposed and those recommendations which the candidate has suggested based on the completed research are encapsulated in the last chapter of this thesis, answering the last research question.
Chapter 6

Recommendations and Conclusion

6.1. Chapter Overview

This study has explored the relevant literature extensively and involved 26 participants in detailed interviews exposing the experience and difficulties encountered with communication surrounding the initiation and withdrawal of non-invasive ventilation (NIV) and end-of-life discussions in MND. The clinical participants have provided insight into barriers to sensitive communication into end-of-life choices and the benefits and burdens of NIV. Improving communication and managing end-of-life in MND and minimising undue prolonging of suffering are the objects of the recommendations derived from this research. The bereaved family participants, who have been involved with a family member with MND, have provided a glimpse into their experiences and understandings of the options for symptom relief as communicated to them. The family participants have also described the health systems they learnt to navigate, any frustrations they encountered and made some suggestions for other families negotiating the MND path.

This concluding chapter commences with the key issues and recognition of what this research has contributed to the knowledge and understanding of end-of-life choices and communication in people with MND. The recommendations from all the participants who were involved in the study cover aspects of MND which they felt could be improved upon. Whilst some of the suggestions are not directly related to the original research topic, the candidate felt these valuable comments should be included as they may indicate areas of potential research and avenues for improvement in caring for people with MND and their families.

The final chapter answers the last research question posed by the candidate by providing recommendations for improvement in communicating
the difficult, sensitive discussions surrounding the end of life. The final and sixth research question is:

Following the literature review and clinician and family/significant other interviews regarding NIV communication, what recommendations can be identified that would improve end-of-life communication and process for the patients, families and clinicians involved with the end-of-life care for people with MND?

6.2. Key Issues Identified from this Study

The following findings were identified as issues within this study, and recommendations regarding these are found in Section 6.5:

1. Communication is adversely affected by specialists having insufficient time to ensure people with MND and their families are sufficiently well informed about their end-of-life choices. Specialists, particularly respiratory clinicians, have difficulty having the necessary sensitive communication about treatment choices and the progressive nature of the disease due to limited consultation time.

2. There appears to be some disparity between what the MND guidelines recommend regarding early referral to a palliative care service and what families stated was appropriate for them. From this study it is also apparent that clinicians do not always refer newly diagnosed people with MND to palliative care. This study has highlighted that the bereaved families of people with MND have not understood the benefits and capabilities of palliative care as explained to them and perceive palliative care, with some fear, to mean death. Clinicians and families agree that the optimal time to refer people with MND to a palliative care service is challenging.
3. Families reported that too many agencies/health professionals are involved with general and specialist care for people with MND. Families spoke of the need to repeat the same issues to several different people, which consumed valuable time and effort. The families were very aware that time was not on their side and resented the intrusion from so many different services. This potentially affected end-of-life care and symptom control as stated by one family (F1, F2).

4. Communication barriers exist for people with MND and their families particularly when discussing end-of-life care and particularly when there is no structured MDT or single individual responsible for patient care involved. Families with little experience or knowledge of the health care system and who have lower levels of health literacy often do not have the skills to ask relevant questions related to symptom control and end-of-life issues.

5. Complex communication barriers have been identified in this research when clinicians initiate NIV in people with MND. Trigger points have been suggested in the literature as a time when communication regarding end-of-life choices be commenced; the offering of NIV is one such trigger point. However, this research has found that in many instances, insufficient time and a fear by clinicians of taking away hope often prevents the necessary NIV and end-of-life conversations from taking place.

6. The clinicians acknowledged that advance end-of-life care planning was not as honest as it might be because doctors are reluctant to say they do not know what will happen or how long until death. Most clinicians did not re-visit AHDS once written despite changes in symptom control such as a PEG use which might cause an adverse health situation necessitating an instant decision for ongoing care.
7. This study found that some of the families would have liked more understanding of the disease trajectory and prognosis so they could make plans and organise their lives. Whilst all the families acknowledged the variability of the disease and its progression, comment was made that when certain symptoms developed, a better idea of the remaining duration of time might be given.

6.2.1. How this research contributes to knowledge

The issues identified from this research as listed above have provided a platform for making improvements for people with MND and the communication of their end-of-life choices particularly surrounding the initiation and withdrawal of NIV. The communication surrounding death and palliative care remains offensive to many, clinicians and patients alike. This research revealed the gap between clinicians’ understandings of MND communication recommendations and guidelines on the one hand, and the communication they consider appropriate on the other. Valuable insights to the barriers preventing such communication between clinicians and people with MND and their families have been provided (Chapter 4; Table 4.5). Family participants stated that referral to palliative care at or soon after diagnosis was confronting, despite it being a guideline recommendation. This research suggests the possibility that the further away from a clinician’s original formal medical education and, therefore, the more experienced they are in their field, the less likely they may be to read and comply with MND recommendations and guidelines.

This research provides a much-needed addition to the literature on the experiences of caregivers and families who have had a family member die from MND and had experience with, or refusal of, NIV as a palliation measure. The research also adds valuable insight into the clinician’s perspective of what, when and how communication is delivered and the barriers they encounter.
6.3. Participant Recommendations for Improved Communication

The family participants were asked to “please comment on any discussions which you would have liked to have taken place, with whom and how would you have liked those discussions?” (Question 21; Appendix E).

Some of the families (F2, F5, and P1) within this research expressed a lack of understanding of the distressing symptoms and available options for care particularly at the terminal stage of the disease. Whilst all had some knowledge of what could occur, and that death would very likely be from respiratory distress, some of the families appeared unprepared for what eventuated. There was general lack of understanding that the person with MND would find increasing difficulty in communicating clearly and, therefore, a poor comprehension as to why early discussions of end-of-life choices were necessary. There was confusion about some clinical aspects of the end of life which had not been discussed either by clinicians or between family members during the disease.

The families made many valuable points and recommendations within this study. First, the person with MND interviewed (P1) suggested that clinicians be more proactive and able to anticipate potential issues instead of responding to a situation. Two family members (F1 and F2) recommended that families could be more in control of their situation by asking the difficult questions of the clinicians to understand what could eventuate within the disease progression. However, that involves a level of health literacy and confidence, plus the emotional capacity to cope with the potentially confronting answers. Several family participants made the point that in hindsight, they would have asked different questions because now they knew what was important to ask. Some families may have required guidance to ask the right questions, as levels of health literacy varied. One family member (F4) stated that if he did not understand what a clinician had said, he turned to the Internet to find answers, whilst other families used the Internet for information as soon as their family member had been diagnosed (F3, F5, F6). The families were asked whether it
would have been useful to be provided with more written information on the relevance of the progressive symptoms in relation to the relevant specialist intervention. All suggested it would have been helpful as negotiating the health system was confusing and very time consuming. The family participants (F3, F4) and the person with MND (P1) said they would have liked an idea of the progression timeline of the disease to plan their lives better. All acknowledged specific prognostic timing was difficult with MND but suggested an indication of where the disease was heading when a particular symptom occurred would have helped them manage their lives.

The family participants recommended earlier referral to specialists once a symptom was evident but not necessarily explained clinically. Importantly, the more rural families felt they should have the same access to services as metropolitan families, although the specifics of those comments were not advanced further. For example, F5 stated that there was a lack of informed MND caregivers in her rural area and it was the ability to talk to others in a similar caring situation that she longed for. Mentoring at an early stage of MND from others who have gone through the MND experience before was recommended by F5.

One family (F3) would have liked information about genetic testing and its availability communicated to them by the clinicians. The content of communication which one person with MND and their family may require may, however, be too confronting for another. Better communication between a clinician and those involved with the person with MND, may mean that many of these concerns could be explored and discussed.

In summary family participants recommended that:

- There is better explanation from clinicians about the symptoms which may occur in MND and the meaning of those symptoms in relation to the progression of the disease.
• Involvement of a palliative care specialist does not occur until symptoms became more obvious and symptom control is required.

• A mentoring (online or in person) system is established to enable regular contact with others going through the MND experience for more rural families.

• There is a single agency (or care coordinator) appointed instead of multiple agency involvement.

The clinician participants were asked two questions (Questions 35 and 36; Appendix D) which yielded recommendations for communication and care for people with MND: “What are the barriers to discussion between clinician and patient, significant others and caregivers about commencement and subsequent withdrawal of NIV?” and “Anything else you think I should know about your experience in communicating about the MND experience for patients?”

Various themes emerged from the clinician participant interviews and one of the most common was the need to be able to allocate sufficient time for communicating all aspects of symptom relief and end-of-life choices. Informed communication, developing a rapport and establishing trust with the person with MND and their family were considered important, but these involve a considerable amount of time spread over several occasions. The frustration voiced by the clinicians was the difficulty in allocating the amount of time required to ensure understanding and being able to repeat the communication. As stated by C2 (palliative care specialist) doctors don’t know where they can find this time. None of the clinicians could give a satisfactory answer as to how to solve this issue, only that they had to try to make time for those discussions. Many of the specialists felt that they were rushed and probably inadequately communicating end-of-life care options. People with MND from the country, who only came to see the MND MDT for symptom management interventions such
as NIV, had informative discussions about NIV but without follow up to address the more sensitive issues. This was frustrating for the clinicians and unsatisfactory for the person with MND and their family (C13).

Some of the clinicians admitted difficulties with having end-of-life discussions when initiating NIV: although they felt they were offering something which may help the patient they did not want to remove hope by discussing potential withdrawal. The point was made that some people with MND and their families do not want to have those discussions, despite the clinician trying to do so. Denial, the understanding held by the person with MND and/or their family plus variable levels of health literacy were barriers to end-of-life communication occurring at the time the current guidelines recommend (NICE, 2016).

Frontotemporal dementia was found in this research to be a symptom of MND which some clinicians are not fully conversant with or is not easily recognised. Frontotemporal dementia could be affecting decisions made by some people with MND but, again, time is required for the clinicians to be able to listen to and understand the individual to recognise the FD behavioural nuances.

The sharing of relevant information between clinicians and closer collaboration within MDTs and with their patients was found to be an area which can be improved, concurring with previous studies (Danel-Brunaud et al., 2017). In areas where the palliative care specialist, respiratory specialist and allied health professionals were not part of a formalised MND MDT, communication between all the clinicians involved was described as particularly poor at times. Differing opinions can exist within clinical teams which could confuse a person with MND and their family, or communication may be assumed to have occurred with the patient when it hasn’t. These problems may well be eliminated if there was a person coordinating and responsible for the care of the person with MND and their family.
In summary clinician participants recommended that:

- There is better explanation for people with MND and their families about the emotional end-of-life issues and not just the practical symptom control issues such as the workings of NIV.
- More appointment time is allocated to enable a rapport to develop with the patient and to ensure clear communication.
- Improved communication between health care providers, and even within the MDT.

6.4. Recommendations for Further Research and Suggestions for Policy and Funding Reform

There is a need for further research to assist people with MND and their families to understand the implications of the course of this devastating disease. In light of the research findings, the following section considers how changes to policy and funding may be achieved.

6.4.1. Coordination of care incorporating sensitive end-of-life communication

Specialist clinician time needs to be allocated for sensitive communication so that people with MND and their families better understand the benefits and burdens of symptom control measures and end-of-life choices. Communication may be improved if there was an overall coordinator responsible for the needs of the person with MND and their family. As discussed in Chapter 5 (Section 5.6.4), other health disciplines incorporate a specialist nurse to guide a person through the complexities of decision-making at diagnosis, and onwards with symptom control and either disease progression or cure. MND is different from most other diseases in that there is no hope of a cure and such hope that there is remains within the realms of symptom control and the assurance of a comfortable death.
Throughout Chapter 5 barriers have been identified relating to clear delivery of information from clinicians, and the understanding of important issues by people with MND and their families. Also revealed was an apparent lack of overall responsibility for the person with MND, unless in a highly specialised MND MDT. Allocating a health professional to take overall responsibility within the MDT is not always practical if the MDT is fractured and disparately located. However, ensuring one health professional is responsible for coordinating the care and changing requirements for the person with MND and their family is needed. The recommendation that sensitive end-of-life choices and potential NIV withdrawal is communicated to the person with MND prior to its commencement suggest that a case manager, possibly an MDT member, is allocated at the time of diagnosis. Allocating responsibility for planning ongoing care with the family of a person with MND and taking the lead in communication with them is a recommendation of this study and is in line with current MND recommendations (Faull, de Caestecker, Nicholson, & Black, 2012; NICE, 2016). One of the recommendations of this research is for a multidiscipline (trained in palliative care and neurological diseases) nurse case manager to be appointed from diagnosis, and preferably to be one of several nurses in the role. The case manager would be involved from diagnosis until either hospital admission or death at home. To avoid coordinator nurse “burn out”, it may be preferable to have a small team who can organise the early requirements and support for the person with MND and their family. The case manager would assist people with MND to navigate their local specialists for symptom control, allied health services and general entitlements to community care. The case manager/team would also ensure that the patient’s GP was kept informed with all the specialist interventions and recommendations. Many of the long and sensitive discussions relating to end-of-life choices can be undertaken or reiterated by the nurse, thereby alleviating the need for patients (and specialists) to have repetitive time-consuming communication. This would facilitate continuity of care and trust and may avoid duplication of services in some areas.
An area for further research is to develop and trial the recommended model of allocating a palliative care/neurological nurse case manager from the diagnosis of MND. Of interest would be exploring the communication with the patient’s GP and how well the GP is kept informed of their patient’s progress and any interventions. This research found that GPs remain involved with the day-to-day wellbeing and support of people with MND and their families. An individualised care plan could be developed to assist sharing of information between GPs and local specialists, (such as when symptoms begin), and specialists who are willing to bulk-bill (direct billing to Medicare). The case manager would also report back to the MDT for ongoing advice and support, eliminating the need for the person with MND to be present and thereby reserving precious personal time for the person with MND and their family.

This model allows for continuity of care, development of trust and, therefore, the ability to engage in sensitive communication relating to end-of-life choices and the useful aspects of early palliative care. An audit of the efficacy of a case manager can measure the savings made in terms of specialist time, multiple lengthy appointments, and associated financial costs. The case manager would attend the first specialist appointment and then be able to reiterate and clarify communication involving all aspects of MND care to the person with MND and their family. The possibility for funding a case manager might be investigated under the National Disability Insurance Scheme in conjunction with state health departments.

6.4.2. Reducing the number of people involved with patient care

This research recommends trying to minimise the number of agencies involved with long-term people with MND under palliative care. The possibility of coordinating home care within one organisation, may alleviate some of the issues of too many people being involved, as identified by the families within this study. This may require further research into policy and funding to determine the feasibility of allocating one agency to provide community palliative care (including all aspects of nursing requirements such as dressings,
urinary catheters and equipment), personal and home care. An issue only touched upon in the current study was the variability in funding in relation to accessing long-term specialist palliative care. This was identified as a significant barrier for people living with MND and other potentially long-term neurological diseases.

6.4.3. Addressing the communication barriers for people with MND, their families and clinicians

This research did not study how health literacy affects communication and choices of symptom control in people with MND. However, during the interviews the family and clinician participants recognised that inadequate health literacy may affect how some families understand health issues and their ability or confidence to communicate concerns. Ideally health literacy can be assessed by the clinician, so they can ensure end-of-life choices have been understood. However, the subtle changes which can be associated with FTD can mask patient choices for care which could ultimately adversely affect caregivers (caregiver burden). A trial of an online discussion group or mentoring facility is recommended to investigate the value or providing the opportunity for caregivers and people with MND talk to others who are also going through complex MND experiences and decisions. Such a mentoring facility may also inform and encourage people with MND and their families to have the courage to ask questions of their clinicians and may assist in overcoming some of the communication barriers.

Disease specific guidelines have a very detailed purpose in providing evidence of certain treatments and ongoing care advice for a person with that specific disease. Most diagnosed illnesses offer an element of hope of cure and, therefore, the guidelines relating to that illness assist clinicians to offer current best practice for finding a cure. However, in many of the neurological diseases, MND in particular, there is neither a specific diagnostic test nor any hope of a cure. Therefore, the present guidelines offer recommendations for symptom control, general management and support which, whilst generally
known by clinicians, are not always followed. Whilst communication recommendations are incorporated into the most recent MND guidelines, there is little specific guidance on best practice communication for the clinicians to follow. This research has highlighted the area of recommended communication surrounding the initiation of NIV and PEG tube insertion and found the comprehensive recommended end-of-life communication infrequently occurs. A further recommendation of this study is for research involving families and people with MND so that they can give their views on when the sensitive communication relating to the end of life should occur, and with whom they may prefer those conversations. This would lead to greater involvement of bereaved MND families in the writing of recommendations.

A palliative care nurse (or a case manager) who liaises with the respiratory clinician at commencement of NIV (if the person with MND has not previously been involved with a palliative care team), would be advantageous for rural patients particularly, who have not had the benefits of a structured MDT. This would facilitate continuation and elaboration by the nurse of any communication tentatively commenced by the respiratory clinician. The palliative care nurse also offers a conduit between the deteriorating patient, symptom control via NIV and PEGs and the palliative care specialist team (which includes all members of the MDT). This would be the equivalent of a breast care/cancer nurse being included in the specialist appointments to support and coordinate ongoing care for people with breast cancer. This role could be either a neurological nurse upskilled to palliation or a palliative care nurse trained more comprehensively in neurological conditions, as previously recommended in this section.

6.4.5. Establishing a progressive timeline of information

The development of a written and/or an electronic instrument which describes the various symptoms people with MND may experience (rather than describing what is happening within the disease process), who to question (which health professional) about any symptom and some of the alternative
symptom-relieving choices available (including their benefits and burdens) is recommended. This would also incorporate a timeline which gives families and people with MND an idea whether they need to act fast. For example, if the patient is experiencing headaches on waking or generally feeling breathless, it is time to speak to a doctor about a referral to a respiratory specialist. Symptom relieving options offered by a respiratory specialist would be identified, and some relevant questions listed to assist the person with MND know what to ask the specialist. The timeline would include a list of the specialists in the patient’s area (linked into Google Maps) and which specialists will bulk-bill (Medicare fund) for appointments.

An instrument such as this may prove useful for people with MND to pre-empt their needs, understand their choices and be more proactive with implementing their symptom relieving requirements. This could be developed as an interactive app for smart phones, and/or as a written hand-held document for people with MND. The instrument would include instructions, comments and recommendations inserted by specialists (or case manager) at the time of their appointment. This tool may assist people with MND and their families to remember, understand and revisit sensitive communication. It would also incorporate advice on when to consider writing an AHD and outline the advantages for optimising care when patient preferences are clearly known and documented. This would be suggested to occur at trigger points identified in the literature (NICE, 2016; Rafiq et al., 2012). An electronic timeline would enable up-to-date care wishes concerning end-of-life care to be identified and changed as required by the person with MND. The instrument would also incorporate the ability to SKYPE (or similar) with specialists at an allocated appointment time, so any updated information would be shared between clinicians, patient and family plus the case manager. This would minimise the need for the person to travel which becomes difficult as the disease progresses.
6.4.6. Study the timing of preferred referral to a specialist palliative care service

The recommendation in the NICE MND Assessment and Management Guidelines (2016) for early referral to palliative care, whilst known by clinicians, does not translate into clinical practice. The barriers and clinician concerns for early palliative care referral have been identified throughout this research. However, it is the family participants who clearly stated their reluctance for early referral and were shocked by the suggestion of palliative care when their family member with MND was relatively asymptomatic. The clinicians have identified that family members may not be ready for this discussion early on. This research recommends that palliative care is introduced and described to people with MND after they have identified a symptom requiring treatment or control, using the symptom as a reason to explain the benefits of palliative care involvement. Greater involvement of MND families regarding explanation, timing and level of involvement of palliative care, timing of end-of-life communication would be provided by the case manager as proposed in Section 6.4.1.

6.4.7. Funding of palliative care services as a barrier to ongoing care

This study has encountered several clinicians who have described access to a specialist palliative care service (in their area) for people with MND to be limited to their last three months of life. Others described the palliative care service not being able to keep people with MND on their books long-term due to the palliative care funding in their area. This finding neither complies with the MND guideline recommendation of referral to palliative care at or soon after diagnosis, nor makes the time of referral any easier to determine for clinicians. The recommendation of this research is intermittent referral to a palliative care service which would provide advice on symptom control, without a specific prerequisite for survival time or diagnosis for admission to the service. This model of intermittent palliative care, which has been discussed in Chapter 5 (Section 5.6.3), may encourage recognition of symptoms and provide ongoing high-level care. An intermittent palliative care service is recommended in
conjunction with an MND case manager. The cost of an intermittent palliative care service may be offset by fewer emergency admissions to hospital which occur particularly when people with MND have symptom complications.

6.6. Limitations

This study has covered many aspects of communicating end-of-life choices focusing particularly on the stage of MND when the respiratory muscles weaken, and the effects of respiratory difficulty become apparent to the person with MND. Several limitations are identified within this study. Importantly and unfortunately there were no neurologists available or willing to participate in this study. The neurologist viewpoint would have added greater depth of understanding of both communication content and responsibility for overall care of the person with MND. The lack of neurologists may also have influenced the recruitment strategy of family participants. As GPs are involved with the ongoing general health care of people with MND, it may have been relevant to have included their perspective and this may be considered a limitation of the study. However, the focus of the research questions was on those participants with considerable clinical experience of MND, and GPs may not have such experience.

The clinician participants have given examples of how they communicate and candid examples of when they found it difficult. As this study could not link the family participants with any clinicians interviewed, the study was limited in being able to verify how clearly each clinician communicated end-of-life options. However, the bereaved family participants were clearly confused on many aspects of the terminal stage of their family member’s MND, although this confusion may have been a symptom of their grief. Unfortunately, a relatively small number of bereaved families applied to participate (six) and this is recognised as a limitation of the study. This may be due to participant fatigue as there have been various MND research projects in Western Australia involving
bereaved families undertaken recently, or simply that as MND is a rare disease there were few families available.

A further limitation identified is the interview process which involves retrospective recall as well as aspects of life which impact on memory, which in turn may affect how their reality is recalled. In considering the methodology, research questions and participant semi-structured questions, the research method was developed to obtain the most accurate and dependable information. There were some disadvantages to using semi-structured questions to interview all the participants. The main disadvantage was the extended time of the interviews which meant that the analysis of the data was also time consuming. In analysing the data from the in-depth interviews, generalisation of the information was not always obvious and at times, difficult. The possibility exists that the data collected for this study may have been different if a method other than the one chosen was selected. The candidate did, however, take trustworthiness measures to ensure objectivity was maintained as much as possible (Chapter 3, Section 3.13.1).

6.7. Conclusion

Whilst acknowledging that MND offers no hope of a cure and early end-of-life communication is often laden with emotional difficulties, it nonetheless remains vital for the person with MND and their family to be provided with the information they need to be able to make informed choices. The research questions within this study aimed to determine whether the international and national guideline recommendations into NIV communication which included the benefits and burdens of NIV, end-of-life choices and the potential of being able to withdraw NIV were being integrated into clinical practice. The significance of whether this sensitive communication was occurring had implications for the management of how people with MND planned their end of life and the choices they could make for their care. The questions sought to determine whether the guideline recommendations were being followed, and if the people with MND
and their family members understood their care options. The research also explored the clinician and family view of the timing of referral to a palliative care service, and the barriers to implementing the recommendations for communication.

Despite the bereaved family participant cohort of this study being small (six participants of which two were siblings interviewed together), their experiences provided valuable information. Of particular interest was the family perspective on the value of early referral to specialist palliative care. The most recent guidelines and recommendations advise referral to palliative care soon after diagnosis (NICE, 2016). However, none of the family participants nor the one person with MND interviewed, stated that palliative care was a service they required soon after diagnosis when they had so much to come to terms with in their lives already. Their reluctance could be due to the influence of a community-wide view of palliative care as a service associated with death, or that the services that palliative care provides were not adequately explained. In this research, those who were referred early felt it too confronting to contemplate at that time (F3, F5), particularly as the person with MND had minimal symptoms. The person interviewed with MND and three other families experienced late palliative care referrals when the symptoms had significantly worsened. Whilst beneficial to them for symptom control at that point, it was too late to adequately commence end-of-life care communication, fully understand the potential of NIV withdrawal and be able to plan for what was left of their lives. The understanding amongst the family participants of what palliative care could offer and how it can improve the experience for people with MND and their families appears to not be communicated by some clinicians. This finding was readily admitted to by many of the clinicians, particularly those involved early in the disease trajectory. The rationale for not referring or discussing specialist palliative care was stated to be a combination of the time required to have adequate end-of-life communication and the sense of taking away any hope the person with MND may have. There was also the barrier of the access
to palliative care services which differed geographically, in many instances determined according to a patient’s time left to live (in months).

A nurse case manager involved soon after diagnosis of MND would offer continuity of service, facilitate collaboration between specialties and act as a conduit between clinicians, patient and their family. Rather than clinical speciality boundaries for care and communication, collaboration and sharing the cost of a nurse coordinator would not only save the specialists time (and money) but improve the experience for the person with MND and their family. The idea of a progressive timeline document for the patient, either hardcopy or an electronic app, that provides clear information about symptom description and significance, was informally discussed with the family participants and person with MND and was welcomed as a helpful idea to improve understanding and planning. However, further research into the design and structure of such an instrument is required.

The opposition by families for multiple organisation involvement with patient care and the complaint of valuable time being taken by repetition of issues and perceived personal intrusion was a valuable finding of this thesis. However, it is a difficult issue to solve. Presently, Australia is very variable in how MDTs and community services are structured and offered to patients and families. Even within cities it depends on which area a person with MND lives as to how the services are structured and how accessible they are. This study recommends one single local organisation being charged with providing community nursing, palliative care, personal care, home help as well as a structured MDT to optimise patient care, efficiency and cost. Within this structure a nurse case manager would manage the patient requirements, assist with appointments and communicate between all health professionals involved. Such a model of care ought to be trialled and evaluated.

As more research is undertaken into MND, hopefully all aspects of communication will be improved. In a very recent systematic review of
evidence-based studies into all aspects of multidisciplinary care in MND/ALS, timely symptom management and end-of-life communication are highlighted as areas needing improvement (Hogden, Foley, Henderson, James, & Aoun, 2017). The review incorporates many of the published research articles which broadly make up the most recent MND management recommendations (Andersen et al., 2012; National Institute for Health and Care Excellence, 2016). Hogden et al. (2017) discuss optimal management of palliative care as well as the effectiveness of MDTs for people with MND. Hogden et al. recommend the development of further guidelines to improve communication between clinicians to provide coordinated care for people with MND and their families. The research reported in this thesis echoes the findings and suggested further work by Hogden et al. (2017) and offers additional recommendations which may help to provide solutions to the barriers encountered in communicating end-of-life options in MND.

Communication surrounding the initiation and withdrawal of NIV was the principle issue for this research. The experiences of the families and person with MND showed that despite the guidelines recommending communication of potential NIV withdrawal prior to initiation, this seldom occurred. This was reiterated by the health professionals responsible for NIV initiation and verified by the palliative care clinicians who in many cases felt they were the first to suggest this option. The reluctance to communicate NIV withdrawal and by default, end-of-life options prior to the terminal stage, had emotional consequences for the families and clinicians alike. This study established that whilst many of the clinicians involved with the early symptoms of MND vaguely knew the content of the most recent MND guidelines regarding communicating NIV withdrawal and end-of-life options, few followed the recommended timing of such communication. Some of the clinicians were not fully aware of the recommendations, and some decided to do what they considered best practice rather than follow the recommendation. The incorporation of an appropriately trained nurse case manager from diagnosis, in cooperation with the specialist clinicians, would help develop rapport with the patient and family, encouraging
such communication. In conjunction with either an electronic or hand-held progressive timeline instrument, end-of-life and symptom control options and communication would be documented for the family and patient to revisit and reacquaint themselves with their choices. By exploring family, patient and health professional perspectives on NIV and end-of-life communication, opportunities for further research have been identified from this study.

This research adds valuable information and expands on the most recent literature with insights into family and patient preferred timing of palliative care and why communication between organisations and between clinicians and people with MND and their families is frequently difficult. MND is a progressively debilitating disease with some appalling symptoms and complex tangential dimensions such as FD. MND is also a complex disease, varied in its course and timing of symptoms and its rarity makes clinician experience in coping with those involved difficult. This research has taken a distinctive in-depth approach, covered many areas of importance and taken the time to extensively interview and listen to clinicians and families involved with MND.

Whilst understanding that recall and retrospective memories can be confusing and influenced by situational variables, this thesis has provided insightful and valuable descriptive information which, it is hoped, may improve how people with MND are helped to understand and communicate their end-of-life care.

The epilogue which follows explores the personal growth the candidate experienced whilst undertaking this study and the candidate’s appreciation of the enormity of receiving a diagnosis of MND for the person and family involved.
Epilogue

Thesis Reflections

Exploring the communication surrounding the initiation and potential withdrawal of NIV in people with MND has provided the candidate with a profound understanding not just of the people with MND but their families, caregivers and clinicians. The participants spoke with sincerity and reflection which provided insightful information both for this study and to enable the candidate to improve her nursing and communication skills.

All the participants involved in this study were English-speaking as a first language. However, communication between clinicians and those with MND was often reported as confused, unclear and sometimes involved avoidance of the inevitable outcome of MND. The emotional impact and difficulty of delivering such devastating news to people with MND and their caregivers created communication barriers. This was clearly stated by C3: “So, I think it’s all about the quality of the communication leading up to that process (symptom relief, NIV withdrawal and end-of-life communication) and investing the time in the process.”

Despite the English language being understood by all concerned, what was reported to be said and what was understood often appeared at odds. Perhaps the English language itself is partly to blame, with many words having different meaning to different people. An example of this was the word “prognosis” whose dictionary definition is “A medical prediction of the future course of a disease and the chance for recovery. Note: Prognosis is often used as a general term for predicting the unfolding of events” (www.dictionary.com/browse/prognosis). This term was most frequently used by clinicians to mean a prediction of length of time a person had left to live. However, in their conversations with clinicians many caregivers and people with MND appeared to understand the term to mean "what to expect; symptoms and control methods, benefits and burdens of NIV and gastrostomy tubes"
Therefore, conversations appeared to disengage at that point unless the person with MND or their caregiver had the confidence or sufficient health literacy to ask further questions about symptom control or other aspects of prognosis. It would be unreasonable for a clinician to ensure that those involved with MND had the same literal meaning to every word used, but it might be an indication that verbal communication maybe insufficient to ensure understanding of the vital aspects of MND.

By the completion of data collection, no participant had had to cease the interview or request emotional support, and all sent affirming post-interview emails to the researcher. Most offered to answer more questions or be re-interviewed should further information be required. All the bereaved participants stated that the interview was a cathartic experience for them and hoped the information they provided might enhance communication for those people with MND and their families in the future. The enthusiasm, insight and thought with which the family participants and the one participant with MND spoke meant the experience was as enlightening and profound for the candidate as it was a therapeutic for the participants.

As an investigator into understanding clinician’s delivery of end-of-life and symptom control communication and having listened to the experiences of the bereaved families, the candidate is left with an overwhelming feeling that clinicians are communicating as time dictates. There are too many variable influences at play to completely remove ambiguity and guarantee that crucial information is always fully understood by the recipient via verbal communication alone. It is hoped that some of the recommendations of this study will lead to further research and improved communication between clinicians and people with MND. Some of the clinical participants provided feedback either during or after the interviews which may indicate potential change, and these comments have been included within this thesis and epilogue. Time for professional reflection whilst answering the study questions appeared to be appreciated by all the clinicians interviewed:
C19: I think that set of questions is an excellent set of – if I may say so, Lottie, because you’ve covered so much there, and very astringently. You know, you’ve gone into depth with each of those areas. So, I think that that’s excellent. No, that’s really good. Yeah, yeah, I think that that’s good.

C18: No, no, look, this is very important. You’re doing great work.

C15: That’s a very good question, Lottie, and I have to say I haven’t really pushed that differentiation on advance care directives. But I think it is important, and perhaps in future I will.

I am immensely grateful for the opportunity to have undertaken this study and to have enhanced my own comprehension of the importance of clear communication not only with those involved with MND but generally in palliative care. It is my sincere desire that this study will lead to further investigation into the delivery of end-of-life communication and the understanding of the measures required by people with MND to ensure choices of symptom control are explained and understood.
References


Munn, Z., Moola, S., Riitano, D., & Lisy, K. (2014). The development of a critical appraisal tool for use in systematic reviews addressing questions of


Oishi, A., & Murtagh, F. (2014). The challenges of uncertainty and interprofessional collaboration in palliative care for non-cancer patients in the community: A systematic review of views from patients, carers and health-care professionals,. *Palliative Medicine, 28*(9), 1081-1098. doi:10.1177/0269216314531999


Phelps, K., Regen, E., Oliver, D., McDermott, C., & Faull, C. (2015). Withdrawal of ventilation at the patient’s request in MND: A retrospective exploration of the ethical and legal issues that have arisen for doctors in the UK. *British Medical Journal Supportive & Palliative Care, 7*(2). doi:10.1136/bmjspcare-2014-000826


Steinhauser, K., Clipp, E., McNeilly, M., Christakis, N., McIntyre, L., & Tulsky, J. (2000). In search of a good death: Observations of patients, families, and providers. *Annals of Internal Medicine, 132*(10), 825-832.


doi:10.1177/0269216315583032


doi:10.1080/10640266.2015.1013393


**Appendix A: 1) Summary of Peer Reviewed Articles**

<table>
<thead>
<tr>
<th>Author and year</th>
<th>Research design: sample, size</th>
<th>Comments and key findings</th>
<th>Limitations</th>
<th>Implications of findings for thesis research and questions</th>
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<tr>
<td>Astrow, A., Sood, J., Nolan, M., Terry, P., Clawson, L., Kub, J., Sulmasy, D. (2008).</td>
<td>Decision-making in patients with advanced cancer compared with amyotrophic lateral sclerosis. Long-term longitudinal study comparing some cancers with ALS relating to decision making and AHD's.</td>
<td>Despite shorter survival rates, cancer patients were less likely to have AHD and to have made decisions relating to their end-of-life care compared to ALS patients.</td>
<td>*Method used to gather information: oncologists may have relevant conversations but not document. *Relatively small group of cancer patients.</td>
<td>ALS has predictable outcome, whereas cancer varies. The few options for ALS symptom control may prompt end-of-life discussions.</td>
</tr>
<tr>
<td>Baxter, S., Baird, W., Thompson, S., Bianchi, S., Walters, S., Lee, E., McDermott, C. (2013).</td>
<td>The use of non-invasive ventilation at end of life in patients with motor neurone disease: A qualitative exploration of family caregiver and health professional experience. Thematic analysis of in-depth interviews.</td>
<td>Death with MND usually peaceful. NIV was not perceived to have had an adverse end-of-life impact. Initiation of NIV should be seen as an opportunity for end-of-life discussions and clarity.</td>
<td>*A relatively small sample study of NIV MND users and health care professionals. Sample contained mainly limb onset patients; authors acknowledge results may differ in bulbar affected patients.</td>
<td>The study confirms the importance of communicating end-of-life wishes to healthcare professionals and family, plus clarity in end-of-life discussions. Identifies patient and clinician issues with withdrawing NIV.</td>
</tr>
<tr>
<td>Bede, P., Oliver, D., Stodart, J., van de Berg, L., Simmons, Z., Brannagain, D., Hardiman, O. (2011).</td>
<td>Palliative care in amyotrophic lateral sclerosis: a review of current international guidelines and initiatives: a systematic analysis of type and timing of palliative care in ALS</td>
<td>Quality palliative care can improve quality of life for ALS patients and families: development of international guidelines and framework recommended</td>
<td>Although considering all palliative care, appropriate and timely, clear communication recommended to be vital to patient choices</td>
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<td>Author and year</td>
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<td>Belkora, J. (2003).</td>
<td>Use of the Decision Process for Improving the Quality of Medical Decisions; a discussion paper.</td>
<td>A theory of using a discussion dialogue process to enable patient/care provider to set a treatment strategy to enable clarity when making medical treatment decisions. Acknowledges a gap between communicating medical evidence and patient preferences.</td>
<td>+ Suggested theory used specifically trained facilitators, not clinicians specialising in MND; a generalised approach to medical decision making.</td>
<td>By identifying an information gap between patient and clinicians, particularly relating to a terminal diagnosis, this paper suggests delegation of decisions from patient to care provider due to overwhelming emotion. This may lead to patient/family dissatisfaction and regret.</td>
</tr>
<tr>
<td>Bourke, S., Tomlinson, M., Williams, T., Bullock, R., Shaw, P., &amp; Gibson, G. (2006).</td>
<td>Effects of non-invasive ventilation on survival and quality of life in patients with amyotrophic lateral sclerosis: a randomised control trial.</td>
<td>Study showed NIV can maintain quality of life and offer some increase in survival, but not in people with MND with bulbar dysfunction.</td>
<td>Identified NIV as a symptom control measure in some variants of MND. This study tries to understand which people with MND are offered and benefit from NIV.</td>
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<td>Clinch, A., &amp; Le, B. (2011).</td>
<td>Withdrawal of mechanical ventilation in the home: A case report and review of the literature.</td>
<td>Identified the complexities when medical treatment is no longer useful and is withdrawn.</td>
<td>+This study was based on a single case of mechanical ventilation.</td>
<td>Identifies issues surrounding the withdrawal of treatment and the extensive communication required to ensure barriers removed.</td>
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<tr>
<td>Connolly, S., Galvin, M, &amp; Hardiman, O. (2015).</td>
<td>End of life management in patients with amyotrophic lateral sclerosis: a review.</td>
<td>This review encourages clinicians to ensure people with MND are prepared and appropriately supported for end-of-life.</td>
<td></td>
<td>The review recommends a greater emphasis on health professionals’ communication skills and palliative care approaches.</td>
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<tr>
<td>Dharmadasa, T., Matamala, J., &amp; Kiernan, M. (2016).</td>
<td>Treatment approaches in MND: a review of health care professional’s attitudes to treatments/symptom control.</td>
<td>The end-of-life phase is poorly defined, but the review suggests that symptoms of MND can be effective.</td>
<td></td>
<td>This review finds the end of life phase is poorly defined. A well-structured MDT represents optimal care.</td>
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<tr>
<td>Eng, D. (2006).</td>
<td>Literature review1966-2004; Cochrane library of systemic reviews, gray literature through Caresearch. Management Guidelines for people with motor neurone disease on non-invasive ventilation at home.</td>
<td>Exploration of issues surrounding the use of NIV with the objective of suggesting guidelines to ensure appropriate, structured and coordinated approach to NIV use in MND with a palliative care focus.</td>
<td></td>
<td>Highlighted the need for good, structured communication for NIV/MND and the ethical dilemma when NIV is withdrawn.</td>
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<tr>
<td>European Federation of Neurological Societies and European Association of Palliative Care. (2014).</td>
<td>New European Consensus on Palliative Care in Neuro Disease.</td>
<td>A plan for palliative care medicine and neurology that includes advance health planning, family/caregiver support and end-of-life care.</td>
<td></td>
<td>Recommends early planning and open communication to enable patient involvement and goal setting.</td>
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<tr>
<td>Author and year</td>
<td>Research design; sample size</td>
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<td>Faull, C., Phelps, K., Regen, E., Oliver, D., McDermott, C., &amp; Rowe-Haynes, C. (2014).</td>
<td>Withdrawal of NIV at the patient's request in MND: exploration of the issues related to communication. A retrospective qualitative study involving 20 specialist doctors.</td>
<td>Poster presentation: Withdrawal of NIV closely related to loss of patient ability to communicate. Clinicians require skills in communicating to ensure patient wishes and comfort.</td>
<td>+ Study did not appear to include doctors involved with the initiation of NIV and the communication they provided to patients regarding withdrawal.</td>
<td>Highlights the time-consuming and worrying aspect of communicating withdrawal of NIV, with attention drawn to the frequent lack of knowledge about patients’ wishes prior to their loss of ability to communicate clearly.</td>
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<tr>
<td>Faull, C., Rowe-Haynes, C., &amp; Oliver, D. (2014).</td>
<td>Issues for palliative care doctors surrounding the withdrawal of non-invasive ventilation at the request of a patient with motor neurone disease: a scoping study: electronic questionnaire to palliative care doctors.</td>
<td>Withdrawal of NIV involves many challenges for palliative care doctors including emotionally, practically and ethically.</td>
<td>*Possible that only doctor who had experienced problems was more inclined to respond to survey.</td>
<td>NIV withdrawal guidelines suggested which incorporate an ethical statement. The study identified a better understanding of the difficulties of NIV withdrawal.</td>
</tr>
<tr>
<td>Foley, G., Timonen, V., &amp; Hardiman, O. (2014).</td>
<td>Understanding psycho-social processes underpinning engagement with services in motor neurone disease: A qualitative study.</td>
<td>Identified control, reassurance and trust vital for people with MND to communicate and engage with healthcare professionals.</td>
<td>* Authors suggest findings may not be fully representative globally for people with MND.</td>
<td>Findings suggest study group did not prioritise life-prolonging measures and there was some confusion in relation to the understanding of the role of PEGs.</td>
</tr>
<tr>
<td>Greenaway, L., Martin, N., Lawrence, V., Janssen, A., Al-Chalabi, A., Leigh, P. N., &amp; Goldstein, L. H. (2015).</td>
<td>Accepting or declining non-invasive ventilation or gastrostomy in amyotrophic lateral sclerosis: patients' perspectives. Patient sample size of 78 offered PEG and/or NIV. None of the 21 interviewed had refused NIV.</td>
<td>Individualised support is suggested to be preferable to responding to treatment guidelines, as this may be a too pressured approach for some patients.</td>
<td>* Few NIV patients involved and no interviews with patients who had refused NIV. * Caregivers spoke on behalf of people with MND.</td>
<td>This study recommended clear and high-quality information be provided for people with MND and their caregivers and suggested those with less understanding of their illness were potentially more likely to refuse an intervention.</td>
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<td>Author and year</td>
<td>Research design: sample, size</td>
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<td>Hardiman, O. (2011).</td>
<td>Management of respiratory symptoms in ALS; clinician’s discussion incorporating recent literature.</td>
<td>Advance care directives should be suggested with respiratory failure to prevent emergency mechanical ventilation.</td>
<td>Implications of NIV and respiratory failure be discussed early in disease; no suggestion as to content of discussions and by whom.</td>
<td></td>
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<tr>
<td>Jenkins, M, Hiollinger, H, &amp; McDermott, C. (2014).</td>
<td>The evidence for symptomatic treatments in amyotrophic lateral sclerosis; a discussion paper.</td>
<td>NIV can improve quality of life however unclear if same applies to gastrostomy.</td>
<td>+ Identifies symptoms of respiratory failure and implementation of NIV, but timing of discussions relating to end-of-life unclear.</td>
<td>Suggests good evidence for NIV and some evidence for MDT involvement. Emphasis’s needs of the caregivers.</td>
</tr>
<tr>
<td>Lemoignan, J., &amp; Ells, C., (2010).</td>
<td>ALS and assisted ventilation: How patients decide: 10 semi-structured patient interviews (phenomenology).</td>
<td>Discussions relating to NIV should be individualised and allow for patient autonomy.</td>
<td>Interviewer knew participants prior to research, which the author identifies, may have influenced the findings. * A small study.</td>
<td>Further research necessary which explores ALS patient decision making.</td>
</tr>
<tr>
<td>Lerum, S., Solbrække, K., Nyheim, H., &amp; Frich, J. (2016).</td>
<td>Family caregivers’ accounts of caring for a family member with motor neurone disease in Norway. 25 participants; 17 active and 8 bereaved caregivers. A qualitative study</td>
<td>People with MND with cognitive impairment may not have the concept of burden of care by their caregivers.</td>
<td>Researchers identified possible skewed information by family members when communicating on behalf of their family member with MND.</td>
<td>Rapidly changing situation may compromise caregiver ability to access adequate help from health professionals.</td>
</tr>
<tr>
<td>Lerum, S., Solbrække, K., Nyheim, H., &amp; Frich, Jan C. (2015).</td>
<td>Unstable terminality: negotiating the meaning of chronicity and terminality in motor neurone disease. A systematic review.</td>
<td>Limiting futile palliating treatments, good symptom management, respect and dignity remain key aspects of patient centered care.</td>
<td>Effective communication and shared decisions of care remain the most important palliative care requirements found in people with MND.</td>
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<td>Martin, N., Landau, S., Janssen, A., Lyall, R., Higginson, I., Burman, R., Goldstein, L. (2014).</td>
<td>Psychological as well as illness factors influence acceptance of non-invasive ventilation (NIV) and gastrostomy in amyotrophic lateral sclerosis (ALS): a prospective population study in 78 ALS and 50 caregivers.</td>
<td>Provides insight into complexity of patient factors to be considered when providing patients with information relating to NIV and PEG use e.g. IQ, education level.</td>
<td>* Follow-up data was limited particularly relating to patients refusing NIV. Decision making about NIV and PEG grouped together.</td>
<td>Study suggests how further work should try to clarify that MND/ALS patients are not either denied or pressured into treatment and decisions; and that cognitive and psychosocial factors need to be considered.</td>
</tr>
<tr>
<td>McConigley, R., Kristjanson, L. J., Aoun, S. M., Oldham, L., Currow, D. C., O’Connor, M., &amp; Holloway, K. (2014).</td>
<td>Staying just one step ahead: providing care for patients with motor neurone disease. Interviews and focus groups with 31 specialists.</td>
<td>Highlights the need for further education for clinicians relating to MND and the complex communication issues.</td>
<td>*Results limited by small number of participants. Focus group limited as run during national conference.</td>
<td>Introducing palliative care to people with MND/family provided communication difficulties for clinicians.</td>
</tr>
<tr>
<td>Mitsumoto, H., &amp; Rabkin, J.G. (2007). JAMA, 298(2), 207-216.</td>
<td>Palliative Care for Patients with Amyotrophic Lateral Sclerosis: &quot;Prepare for the Worst and Hope for the Best&quot;. A clinician’s personal perspective.</td>
<td>Fast deterioration phase, impairing communication and respiration. Burden for clinician to preserve hope whilst discussing AHD’s.</td>
<td>The author acknowledges that the terminal phase of MND is often too fast for caregivers and patient to adjust to changes. Clinician must have communication early in the disease process to enable adjustment.</td>
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<th>Implications of findings for thesis research and questions</th>
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<tr>
<td>Motor Neurone Disease Australia. (2012).</td>
<td>Palliative Care in Australia; senate inquiry.</td>
<td>Efficient use of palliative care resources and access to MND specific palliative care information and funding for research to provide a national framework.</td>
<td></td>
<td>Promotes timely and targeted information and benefits of palliative care for people with MND.</td>
</tr>
<tr>
<td>National Institute for Health and Care Excellence. (2014).</td>
<td>Treatment with NIV for patients with Motor Neurone Disease; a pathway designed to assist clinicians inform people with MND of their choices of symptom management.</td>
<td>An interactive pathway to assist clinicians guide people with MND; section on NIV use and recommendations.</td>
<td>+ designed for the UK with slightly different availability of specialists and proximity to hospitals.</td>
<td>Reiterates discussion regarding the benefits and burdens of NIV and regular opportunities to discuss continuing or withdrawal of NIV.</td>
</tr>
<tr>
<td>Neudert, C., Oliver, D., Wasner, M., &amp; Borasio, G. (2001).</td>
<td>Telephone interviews with 121 caregivers, 8 nurses and 3 physicians present at the time of death of people with MND.</td>
<td>The course of the terminal phase in patients with amyotrophic lateral sclerosis is generally a peaceful one.</td>
<td>* German/UK data which may not compare with if patient groups different. * Also, a third of caregivers could not be contacted.</td>
<td>NIV reported by caregivers to be beneficial but no report on the communication surrounding the initiation or withdrawal.</td>
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<tr>
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<td>Oliver, D., Campbell, C., Sykes, N., Tallon, C., &amp; Edwards, A. (2011).</td>
<td>Decision-making for gastrostomy and ventilator support for people with motor neurone disease: variations across UK hospices: a telephone audit of 22 palliative care specialists.</td>
<td>Identifies a need for greater application of guidelines for NIV and PEG; also identifies combined approach inclusive of other services.</td>
<td>Whilst contact is made between specialists, communication could be improved by clear, known and utilised guidelines. The aim of study was to encourage discussion.</td>
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<td>Oliver, D, &amp; Faull, C. (2013).</td>
<td>Non-invasive ventilation in amyotrophic lateral sclerosis/motor neurone disease; a discussion paper.</td>
<td>Careful discussions essential prior to commencement of NIV so patients are aware of benefits and burdens.</td>
<td>States the importance of communicating the benefits and risks associated with NIV to people with MND prior to initiation. Recommends further research on impact of withdrawal on families.</td>
<td></td>
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<tr>
<td>Pagnini, F., Banfi, P., Rossi, G., Castelnuovo, G., Marconi, A., Fossati, F., Molinari, E. (2012).</td>
<td>Respiratory function of people with amyotrophic lateral sclerosis and caregiver distress level: a correlation study using questionnaires.</td>
<td>Correlations between a caregiver’s level of distress and a patient’s respiratory function; shows importance of caregiver’s function. *No contributory inferences can be made as this is a cross-sectional study *small study sample.</td>
<td>If a caregiver has the potential to influence the person with MND’s respiratory function, then the relevance of complete understanding of the benefits and burdens of NIV may influence not only the patient choices but quality of the remaining life as well as the quality of life of their caregivers.</td>
<td></td>
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<tr>
<td>Phelps, K, Regen, E, Oliver, D, McDermott, C, &amp; Faull, C. (2015).</td>
<td>Withdrawal of ventilation at the patient's request in MND: a retrospective exploration of the ethical and legal issues that have arisen for doctors in the UK. Qualitative research: 24 interviews with doctors.</td>
<td>Ethical and legal advice and guidance required to support clinicians in NIV withdrawal in MND.</td>
<td>The ethical and moral dimensions for communication surrounding the initiation and withdrawal of NIV are discussed with doctors. Study suggests greater integration of neurology and palliative care specialties.</td>
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<tr>
<td>Phukan, J., &amp; Hardiman, O. (2009). <em>Journal of Neurology</em>, 256, 176-186.</td>
<td>The management of amyotrophic lateral sclerosis; a review of current practice.</td>
<td>Recognises strategies in cognitive management, caregiver burden and depression guided by the American Academy Practice Parameter.</td>
<td>Symptom relief and slight increase in survival, caregiver burden should be discussed early in diagnosis, and discussions relating to the withdrawal of NIV should be included.</td>
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<td>Phukan, J, Elamin, M, Bede, P, Jordan, N, Gallagher, L, Byrne, S, . . . Hardiman, O. (2012).</td>
<td>The syndrome of cognitive impairment in amyotrophic lateral sclerosis: A prospective population-based study in 160 ALS patients and 110 control patients.</td>
<td>Co-morbid dementia found to occur in 14% of newly diagnosed ALS patients and some cognitive impairment in 40 % of ALS. *Study limited to analysis of cognitive dementia; subtle behavioral changes did not fulfill study criteria.</td>
<td>Cognitive impairment may occur in 40% of ALS patients; the implications of cognitive impairment may affect communication and understanding skills?</td>
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<tr>
<td>Preston, H., Fineberg, I., Callagher, P., &amp; Mitchell, D. (2011).</td>
<td>The preferred priorities for care document in motor neurone disease: Views of 11 bereaved relatives and caregivers; semi-structured interviews.</td>
<td>Discusses the use of a document which describes a patient’s priority of care (PPC). Document seen as a good prompt for difficult decisions between clinicians and patients. *Due to limited resources, a small study the findings of which cannot be generalized. *Possible unintentional male gender bias.</td>
<td>Used as a communication aide; A document of this type may help to improve or offer triggers to clinicians to initiate end-of-life discussions. Participants felt the document had little impact on actual end-of-life care.</td>
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<tr>
<td>Rafiq, M., Proctor, A., McDermott, C., &amp; Shaw, P. (2012).</td>
<td>Respiratory management of motor neurone disease: a review of the current literature and discussion paper.</td>
<td>The impact of the use of NIV for treating respiratory weakness. Palliative care needs and stopping NIV support /end-of-life issues should be regularly discussed with people with MND.</td>
<td>Outlines the problems which can be associated with NIV plus the benefits if patients are able to tolerate it. Discusses palliative care strategies as alternatives to NIV and reiterates the importance of ascertaining the patient’s wishes regarding their end-of-life care at the time of or soon after the initiation of NIV.</td>
<td></td>
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<tr>
<td>Author and year</td>
<td>Research design: sample, size</td>
<td>Comments and key findings</td>
<td>Limitations</td>
<td>Implications of findings for thesis research and questions</td>
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<tr>
<td>Radunovic, A., Annane, D., Rafiq, M., &amp; Mustfa, N. (2013).</td>
<td>Mechanical ventilation for amyotrophic lateral sclerosis / motor neurone disease. Search and review of Cochrane Neuromuscular Disease Group literature regarding NIV use.</td>
<td>Review suggests that NIV can increase and improve the life of people with ALS.</td>
<td>Suggests further research to understand the influences which affect access to NIV, the cost personally and economically for patients and their families.</td>
<td></td>
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<tr>
<td>Ray, R., Brown, J., &amp; Street, A. (2014).</td>
<td>Dying with motor neurone disease, what can we learn from family caregivers? Secondary analysis from two data sets employing similar data collection and analysis.</td>
<td>Recommends effective planning for death; normalise the dying process in MND from caregiver perspective.</td>
<td>*Sample size limited despite data from two countries. Identifies the need for strategies to encourage and normalise end-of-life discussions and AHDs.</td>
<td></td>
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<tr>
<td>Virdun, C., Luckett, T., Davidson, P. M., &amp; Phillips, J. (2015).</td>
<td>Dying in the hospital setting: A systematic review of quantitative studies identifying the elements of end-of-life care patients and their families rank important.</td>
<td>This review reiterates the domains of palliative care most important to patients and their families.</td>
<td>Effective communication and shared decisions regarding treatment are fundamentally important at the end-of-life particularly relating to the hospital environment.</td>
<td></td>
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<tr>
<td>Wood-Allum, C., &amp; Shaw, P. (2010).</td>
<td>Motor neurone disease: a practical update on diagnosis and management based on a lecture given by author.</td>
<td>A general update on MND care, reinforcing the need for discussions with patients and families relating to NIV ahead of need.</td>
<td>Due to proven prolonged survival, clear discussions must be held with patients and families well ahead of commencement of NIV.</td>
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*Note: Symbols used within table:  * Authors identified limitation  + Candidates suggested limitation*
### Appendix A: 2) Summary of Additional Literature Relevant to Study: July 2016-July 2017

<table>
<thead>
<tr>
<th>Author and year</th>
<th>Research design</th>
<th>Comments and key findings</th>
<th>Limitations</th>
<th>Implications of findings for thesis research and questions</th>
</tr>
</thead>
<tbody>
<tr>
<td>Smith, T., Disler, R., Jenkins, C., Ingham, J., &amp; Davidson, P. (2017).</td>
<td>A qualitative study using thematic analysis.</td>
<td>Perspectives on advance care planning among patients recently requiring non-invasive ventilation for acute respiratory failure.</td>
<td>Explains the complexities and perspective of NIV use in acute respiratory failure illustrating similar ethical and communication difficulties as in MND.</td>
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Appendix B: Ethics ECU Approval

13 July 2015

Ms Charlotte Sabrina CHAPMAN
C/o Post Office
BOYALUP WA 6237

Dear Ms Chapman,

I am pleased to write on behalf of the Research Students and Scholarships Committee who have approved your PhD research proposal: A retrospective study into the communication surrounding the initiation and withdrawal of non-invasive ventilation (NIV) in patients with motor neurone.

I also wish to confirm that your proposal complies with the provisions contained in the University’s policy for the conduct of ethical research, and your application for ethics has been approved. Your ethics approval number is 12099 and the period of approval from 13 July 2015 to 31 May 2017.

Approval is given for your supervisory team to consist of:

Principal Supervisor: Professor Anne Wilkinson – ECU
Associate Supervisor: Professor Moira Sim – ECU
Associate Supervisor: Dr. Sara Bayes – ECU

The examination requirements on completion are laid down in Part VI of The University (Admissions, Enrolment and Academic Progress) Rules for Courses Requiring the Submission of Theses available at: http://www.ecu.edu.au/GPS/legaL_logis/uni_rules.html

Additional information and documentation relating to the examination process can be found at the Graduate Research School website: http://research.ecu.edu.au/prs/

Please note: the Research Students and Scholarship Committee has resolved to restrict doctoral theses to a maximum of 100,000 words with a provision that under special circumstances a candidate may seek approval from the Faculty Research and Higher Degrees Committee for an extension to the word length. (RSSC 96/2).

I would like to take this opportunity to offer you our best wishes for your research and the development of your thesis.

Yours sincerely

Shelley Huts
Senior Student Progress Officer
Research Assessments
Student Services Centre
Phone: 08 6304 8770
Email: researchassessments@ecu.edu.au

Principal Supervisor: Prof Anne Wilkinson – ECU
Associate Supervisor: Prof Moira Sim – ECU
Associate Supervisor: Dr. Sara Bayes – ECU
HDR: SIO HES
Appendix C: Ethics ECU Amendments

Hi Lottie

**Project: 12099 CHAPMAN**  
**Project Name: A retrospective study into the communication surrounding the initiation and withdrawal of non-invasive ventilation (NIV) in patients with motor neurone disease**

Thank you for your email requesting amendment to your ethics application. The following amendment has been reviewed by members of the Human Research Ethics Committee:

- Inclusion of the stories of people with MND who spontaneously contacted the researcher to participate in the project. The researcher will not be recruiting people with MND however has requested to include these people as part of the project. The Information Letter and Consent Form for family members (already approved) will be used for people with MND to provide consent for inclusion in the project.

Ethics approval has been granted for this amendment.

Kind Regards

*Faye*

**Faye Walmsley**  
Ethics Support Officer  
Office of Research & Innovation, Edith Cowan University,  
270 Joondalup Drive, Joondalup, WA 6027  
Tel: +61 08 6304 5032 | Fax: +61 08 6304 5044 | CRICOS IPC 00279B  
Email: research.ethics@ecu.edu.au
Hi Lottie

Project: 12099 CHAPMAN
Project Name: A retrospective study into the communication surrounding the initiation and withdrawal of non-invasive ventilation (NIV) in patients with motor neurone disease

Thank you for your email requesting an amendment to your ethics application. The following amendment has been reviewed by members of the Human Research Ethics Committee:

- Advertise the project through interstate contacts via palliative, MNDA (general) websites or word of mouth

Ethics approval has been granted for this amendment.

Kind Regards

Faye

Faye Walmsley
Ethics Support Officer
Office of Research & Innovation, Edith Cowan University,
270 Joondalup Drive, Joondalup, WA 6027
Tel: +61 08 6304 5032 | Fax: +61 08 6304 5044 | CRICOS IPC 00279B
Email: research.ethics@ecu.edu.au
Appendix D: Clinician Participant Questions

Semi structured questions and prompts for clinicians

Target group Clinicians

Clinicians involved/experienced with patients with MND who are:
- At the stage of respiratory decline and NIV initiation, and /or
- Involved with the terminal stage of the disease process when the withdrawal of the NIV is considered.

These may be general practitioners, respiratory physicians, neurologists, palliative care specialists, palliative care nurses (Registered and Enrolled Nurses) and allied health professionals.

Questions are prompts only and will not be asked if they have been answered previously.

Introduction

- Thank you for making the time to help with this research.
- As you know my name is and my background is....

Aim

- As you know from the information sheet, the aim of this research is to understand the communication surrounding the initiation and withdrawal of non-invasive ventilation in patients with MND, including the benefits, burdens and limitations in order to optimise future health care provided.

Consent & control of the interview and support if required

- Everything you tell me is confidential. I will not reveal your identity to anyone and I will de-identify the information you give me by using codes known only to me. If you give me permission I may use some of your quotes but these comments will remain anonymous. I'd like to thank you in advance for your contributions to this research, as it will help improve care provided to people with MND and their families.

- I would like to tape the interview so that I can concentrate on listening to you rather than writing.

- Is that okay? YES  NO

- It is up to you how much you want to say and if there's a question you would rather not answer, you can just say you don't want to answer it. If at any point you want to stop, pause for thought or reschedule the interview, you simply have to say so.

- (I would like to remind you of your employer assistance program (EAP) should you feel you would like to discuss any issues that have
adversely affected you, and the availability of a Lifeline Counsellor (92614444) or Lifeline Crisis Line (131114)

I will turn the tape on now.

General introduction open-ended questions which may negate the need for some of the question prompts
1. Before we start on the more structured questions, do you mind my asking general questions relating to your area of expertise and how long have you been practicing in your specific area?

2. What is your ‘role’ here at [NAME OF CLINIC, HOSPITAL, GP PRACTICE]?

3. Have you undertaken any ‘specialty’ training either related to or not directly related to your own discipline? What/Where/When?

4. Can you provide an estimate of your current practice in terms of percentage of people with MND (e.g., 80%)?

5. Understanding that MND is a rare disease, how much experience in terms of years, have you had working with patients diagnosed with MND?

6. Do you work within a multidisciplinary team? YES NO

IF NO: Do you refer the MND patient at diagnosis to the multidisciplinary team associated with you?

7. How involved are the MND patient’s general practitioners with ongoing, day-to-day care?

Next I’d like to discuss what information you provide to people with MND and their families:

8. The NICE MND NIV guidelines and much of the recent literature, recommends various trigger points for ‘honest’ communication with patients and families relating to end-of-life care and the benefits and burdens of NIV. With that in mind;
   • 8.1 Please can you tell me when in the patient’s journey you usually have these conversations?
   • 8.2 At which stage are end-of-life issues addressed by your team, and is there standardised information offered to patients and families?
   • 8.3 Are there specific trigger points within the disease trajectory that you use as an opening for these honest discussions?
   • 8.4 Is the patient’s prognosis included in these ‘honest’ discussions?
8.5 Is there anything else you would normally include in ‘honest’ communication?
8.6 Is there usually only one “honest” communication session or are there usually more than one, and do these conversations include family members?
8.6 How do you assess whether the MND patient and family have fully comprehended the information regarding prognosis, end-of-life and NIV implications communicated by you?
8.7 In your experience are people with MND, families and significant others adequately aware of the prognosis and limitations of treatment options for the disease prior to your involvement with them?
8.8 If the patient with MND declined to have family members present at the time of sensitive discussions, how is medical and general support information provided to the families/caregivers?

9. Thinking how you approach the discussion with patients with MND at the start of NIV, what information do you usually give?
   9.1 Is end-of-life care discussed at this time?
   9.2 What are the specifics of the benefits, burdens (including carer burden) and limitations of NIV that you always discuss with people with MND?
   9.3 Is the potential of increased dependency on NIV discussed?
   9.4 Is hydration and nutrition mentioned i.e. PEG/RIG insertion?
   9.5 Do you usually include information about physical support i.e. potential equipment needs, and psychological support for both patient and family when NIV is commenced?

10. What, in your opinion, do people with MND need to know to be able to make an informed decision regarding the use of NIV?

11. Do you feel that the communication between you and other healthcare providers relating to MND patient treatment and/or medical care is adequate to ensure patient and family understanding of:
   11.1 Symptom relief options i.e. PEG/NIV
   11.2 What to expect relating to disease progression and death
   11.3 Psychological support for patients

12. Do you consider NIV a palliating measure in people with MND, and if so, do you describe it as such?

13. When in your opinion, should people with MND become involved with the palliative care team?

14. Have you had experience of any people with MND declining the use of NIV at the stage when the respiratory muscles and diaphragm start to fail and if so what were the MND patient’s reasons and what alternatives did you give (palliative care options?)
15. When do you usually discuss the potential/eventual withdrawal of NIV with the patient/family: is there a trigger point or indication that this conversation should occur?

16. Were discussions relating to possible NIV withdrawal reiterated during the course of the MND patient’s use of NIV?

17. How is the specific information given; verbal and written form? Who by?

**Now I’d like to discuss the communication you may have with people with MND and their families around the actual terminal phase of the disease, or the end-of-life issues:**

18. As the patient’s swallow is adversely affected (or prior to), is the use of PEG/RIG tube discussed with the family and patient in relation to prolonged survival? When would this be discussed?

19. Do you differentiate between nutrition and hydration in the context of ongoing patient survival, particularly if the patient has indicated/documented a desire not to be kept alive?

20. Do you mention hydration via a PEG/RIG for patient comfort of alleviating feelings of dehydration?

21. Do you weigh your MND patient prior to the PEG insertion, in order to monitor weight changes?

22. Have you ever had issues with people with MND in the end-of-life stage indicating they were dry or dehydrated? YES/NO:

23. Do you think dehydration is a problem for some people with MND at the end of life?

24. When NIV and PEG/RIG’s are initiated, what if any further directions are incorporated into the AHD? *(Are nutrition and hydration discussed in relation to the end-of-life stage and as separate entities, and how are these documented?)*

25. When do you discuss Advance Health Directives with people with MND?

26. When an exacerbation of the disease occurs, are the Advance Health Directives re discussed and clarified? If so by whom?

27. Understanding that dementia/personality changes can be subtle and can affect people with MND decision making, have you had changes to the patient’s wishes/AHD which have surprised you i.e. not as understood by family and yourself to be the wishes of that patient or lack of
understanding of the importance? (*should AHD’s be discussed at diagnosis and at revisited at various trigger points onwards?*)

28. Are the family incorporated into discussions relating to the Advance Heath Directives? If not, when are the patient’s wishes discussed with families or significant others?

29. Generally, in your experience, are the Advance Care Directives clearly detailed enough to ensure the wishes of the patient are upheld?

30. Are potential emergency presentations to hospital with shortness of breath discussed in relation to tracheostomy (discussions which include family/significant others?)?

31. Is the difference between NIV and tracheostomy (invasive ventilation) explained?

32. Have you ever been involved with the withdrawal of NIV in an MND patient and if so can you describe the situation(s)?

33. Do you have any other comments about the withdrawal of NIV?

34. Do you have any comments relating to the communication surrounding NIV initiation or withdrawal?

- Anything about the issues relating to NIV use?
- Anything about the terminal stage of the disease?

35. What are the barriers to discussion between clinician and patient, significant others and caregivers about commencement and subsequent withdrawal of NIV?

36. Anything else you think I should know about your experience in communicating about the MND experience for patients?

*Thank you for participating in this research*
Appendix E: Family Participant Questions

The following questions and statements are considered prompts for interviews with family members and significant others of people who have lived with Motor Neurone Disease. Questions will not be asked if they have been answered previously.

Introduction

- Thank you for making the time to help with this research.
- As you know my name is and my background is....

Aim

- As you know from the information sheet, the aim of this research is to understand the communication surrounding the initiation and withdrawal of non-invasive ventilation in patients with MND, including the benefits, burdens and limitations in order to optimise future health care provided.

Consent & control of the interview and support if required

- Everything you tell me is confidential. I will not reveal your identity to anyone and I will de-identify the information you give me by using codes known only to me. If you give me permission I may use some of your quotes but these comments will remain anonymous. I’d like to thank you in advance for your contributions to this research, as it will help improve care provided to people with MND and their families.
- I would like to tape the interview so that I can concentrate on listening to you rather than writing.
- Is that okay? YES NO
- It is up to you how much you want to say and if there’s a question you would rather not answer, you can just say you don’t want to answer it. If at any point you want to stop, pause for thought or reschedule the interview, you simply have to say so.
- Details of a free counselling service and the MNDAWA Care Advisory team offering emotional support and advocacy (Motor Neurone Disease Association of WA Inc Phone: 9346 7355) are supplied to you should you wish to discuss any issues which may arise from the research interview.

Individuals who may become upset may choose to cease the interview or continue. I will provide information about support available through the Motor Neurone Disease Association of Western Australia (MNDA [WA]) emotional support program which they may choose to access soon after or at a later date.
I will also offer a written handout with details on Lifeline Counsellor (92614444) or Lifeline Crisis Line (131114). Individual participants may elect not to answer any of the questions given as prompts.

I will turn the tape on now.

General introduction open ended questions which may negate the need for some of the question prompts

- If you feel comfortable with discussing your family member, (may I call him/her by his/her Christian name?) perhaps you would like to tell me something about his/her life prior to the diagnosis on MND?
- Are you able to tell me about how the diagnosis was made and how you both came to understand that he/she had MND?
- When you heard the diagnosis, did you have any concept of MND and the implications of the disease?
- May I ask how or whether you searched for further information and if so from where did you seek this information?
- As time and the disease progressed, how did this affect you both and those within your extended families and friends?
- May I ask how long from the time of a definite diagnosis until your family member died?
- Do you mind my asking your family member’s age on diagnosis and in which area were you living? This is just for general demographic information and anonymity by using coding as previously described, is assured.

Question prompts

1. Who initially discussed NIV with you and your family member with MND?  
   (prompt: GP, Respiratory specialist, Neurologist)

2. It is a recommendation of MND guidelines and some journal articles that 'honest' communication occurs between doctors and people with MND and their families. Do you consider that your conversations with the doctors contained the 'honest' information you required?

3. Thinking back to when non-invasive ventilation (NIV) was first discussed, what information was given to you at that time about it? (prompt: I will explain about NIV if this is required). What information was provided about:

   - the benefits,
   - the burdens and
   - the limitations?
4. When NIV was first discussed, was there information given about the possible need of a feeding tube to be inserted into the abdomen? *(prompt: PEG/RIG)*
   - Did you understand that your family member’s ability to swallow could be affected by the disease?
   - Was there a discussion about nutrition and hydration via the feeding tube?

5. What information was given to assist you and your family member make a decision to start or decline NIV?
   - For example any paperwork relating to support groups in your area, palliative care or equipment availability?
   - Were any alternatives offered such as general support and palliating medications?

6. Do you recall mention of NIV eventually no longer being useful and having to be withdrawn?
   - Please tell me what you understood then about what “withdrawal” meant?
   - Did you understand that the disease would still progress even with NIV?

7. Can you remember the clinician who initiated NIV having conversations with you about the end-of-life of your family member with MND?
   - Can you recall how you understood what was discussed?
   - Can you remember when an Advance Health Directive (AHD) was first discussed?
   - Do you recall what was included within the AHD?

8. Was positive invasive ventilation (tracheostomy) mentioned or explained? *(prompt: sometimes, patients have an emergency tracheostomy unless they have made it very clear they do not want this kind of emergency treatment)*
9. Knowing what you know now:

- Is there anything you did not understand about NIV?
- Is there anything you wish you had known?
- Is there anything that would have changed the decisions or choices relating to NIV made by your family member with MND or yourself?
- Who would you like to have explained those choices to you?
- When do you think would have been the best time to explain this?

10. What do you think are the important things to know in order to be able to make an informed decision about using NIV?

- In hindsight, how satisfied are you with the use of NIV and the end-of-life process?
- Did the communication prior to the commencement (or refusal) of NIV cover the relevant information on the benefits, burdens and limitations?

11. In the case of the person you knew who had MND:

- How informed do you think their decision (commencement or refusal) regarding the use of NIV was?
- Was the withdrawal of NIV discussed and if so when and by whom?
- Were you present when this discussion took place?
- Was the Advance Health Directive re discussed during the progression of the illness?
- At what stage was this discussed?
- Did this AHD include the commencement and withdrawal of NIV?

12. If the patient with MND declined to have family members present at the time of discussions about NIV, were you (as a family member or significant other) able to obtain information relating to ongoing patient care?

13. During the last few days of your family member’s life, are you aware if they experienced any feelings of dehydration or dryness?

- If so was it felt that to offer water via the feeding tube (if they had one) would prolong their life?
- Was this potential issue discussed clearly with you by the doctors?
14. If the NIV was withdrawn can you describe what happened and whether you were fully informed for the sequence of events before the withdrawal?

15. Who explained what was going to happen?

16. Was this the same as the explanation given before the NIV was commenced?

17. Would you have liked more information before the NIV was withdrawn?
   - From whom?
   - When?

18. Do you recall if the person you knew who had MND had involvement with a multidisciplinary team; specialists to cover all different areas of their care?
   - Speech Pathologist
   - Respiratory Physician
   - Social Worker
   - Palliative Care
   - Occupational Therapist
   - Physiotherapist
   - General Practitioner
   - Neurologist
   - 18. B) of all the specialists involved, which of the health professionals had overall responsibility for your family member’s ongoing care?

19. If the MND patient you knew decided not to commence on NIV, what options were suggested i.e. palliative involvement/symptom control and support?

20. How satisfied were you with your family member with MND end-of-life care and the communication between the patient, family/significant other and the clinicians?
21. Please comment on any discussions which you would have liked to have taken place, with whom and how would you have liked those discussions?

22. Please add any comments relating to your understanding of the person you knew that had MND and their choices of treatment.

Thank you for participating in this research
A retrospective study into the communication surrounding the initiation and withdrawal of non-invasive ventilation (NIV) in patients with motor neurone disease

This study has been approved by the Edith Cowan University Human Research Ethics Committee.

- I have been provided with a copy of the “Participant Information Letter”, explaining the research project.

- I have read and understood the information provided.

- I have been given the opportunity to ask questions and have had any questions answered to my satisfaction.

- I am aware that if I have any additional questions I can contact the research team.

- I understand that participation in the research project will involve participating in a face to face or telephone interview.

- I understand that the information provided will be kept confidential; the identity of participants will not be disclosed; and that all information will be securely stored.

- I understand the information provided will be used for the purposes of this research and may be included in future research projects, and I understand how the information is to be used.

- I understand the research content will be presented for publication and I will be given the opportunity to read the research prior to publication

- I understand that my involvement is voluntary and I can withdraw at any time without an explanation or penalty.

- I understand that the content of this research may cause some possible sadness and that professional counsellor information and contact details will be offered to all participants

____________________  ____________________  ________________
Signature                  Print name                Date
Appendix G: Clinician Participant Information

A retrospective study into the communication surrounding the initiation and withdrawal of non-invasive ventilation (NIV) in patients with motor neurone disease

Dear ..........(doctor or nurse)

My name is Lottie Chapman, a Registered Nurse, and Ph.D. candidate in the School of Nursing and Midwifery, Edith Cowan University. I am writing to you to ask for your participation in the research I am conducting for my Ph.D.

The aim of my research is to explore the communication process that occurs between clinicians, patients and significant others regarding the initiation (or refusal) and withdrawal of non-invasive ventilation (NIV) for people diagnosed with Motor Neurone Disease (MND). Communication around NIV has been identified as an area needing further research (National Institute for Health and Clinical Excellence, 2010). In this study, I am looking specifically at how clinicians, patients and families make treatment decisions and how this decision-making can be improved. This study has been approved by the Edith Cowan University Human Research Ethics Committee as research for a PhD.

You are invited to discuss your experiences with MND and specifically NIV, contributing to recommended research into NIV communication. Participation involves one face-to-face or telephone interview with me, a nurse researcher, which will last 20-30 minutes and will occur at a time and place most convenient for you. Your participation is entirely voluntary and you may cease or withdraw from the interview at any time.

The interview will be audio taped for accuracy. All information will be kept private and confidential, and no identifying information will be released to any source except where required by law. A research identification number will be assigned to the interview and associated transcripts and only I will know the identity of the respondent. All data will be stored on password-protected computers in the researcher’s office at ECU. Only authorised persons, who understand that this information must be kept confidential, will have access to it. When the study has been completed, all anonymous recordings and written documentation will be archived and stored in securely locked files for a minimum period of five years, as required by law. Your answers will be combined with the answers from other people for analysis and reporting purposes. Findings will be reported in my dissertation and may be published in relevant health related peer-reviewed journals. No personal or identifying information will be conveyed in any of the publications arising from this research. By taking part in this study you agree not to restrict the use of any data, even if you withdraw.

Although this study might not benefit you or your patients directly, this study should contribute to a better understanding of the complex issues involved in treatment decision-making for seriously ill patients. Should this research raise any emotional concerns or questions, and you wish to have someone to talk with you, support information from your free employer counselling scheme (EAP) is available or information for Lifeline counsellors will be supplied to you.

All clinical participants (respiratory, neurologists, palliative doctors and nurses) must have experience with MND discussions particularly surrounding the initiation/refusal and/or withdrawal of NIV or personal experience of being present at the initiation or withdrawal of NIV, and be able to converse in English in order to be eligible to participate in this research.

To take part in the interview or obtain further information, please contact me: Phone: 0417960430   cschapma@our.ecu.edu.au. You may also speak to my Ph.D. Supervisor about this study, Professor Anne Wilkinson at (08) 6304-3540 or email her at: anne.wilkinson@ecu.edu.au. If you have any concerns about the research project or wish to talk to an independent person, you may contact the Research Ethics Officer at Edith Cowan University, 270 Joondalup Drive, Joondalup WA 6027 Phone: 08 6304 2170 or email research.ethics@ecu.edu.au.

Many thanks for your interest and assistance,

Sincerely,

Lottie Chapman, PhD Candidate, Edith Cowan University
Opportunity to participate in Research

A retrospective study into the communication surrounding the initiation and withdrawal of non-invasive ventilation (NIV) in patients with motor neurone disease

The aim of my research is to explore the communication process that occurs between clinicians, patients and significant others regarding the initiation (or refusal) and withdrawal of non-invasive ventilation (NIV) for people diagnosed with Motor Neurone Disease (MND). Communication around NIV has been identified as an area needing further research (National Institute for Health and Clinical Excellence, 2010). In this study, I am looking specifically at how clinicians, patients and families make treatment decisions and how this decision-making can be improved. This study has been approved by the Edith Cowan University Human Research Ethics Committee as research for a PhD, and has the support of MNDA WA/NSW.

You are invited to participate and discuss your experiences in this area, contributing to much needed research into NIV communication. All family/significant other participants must have been closely involved with the MND patient particularly if the patient used or refused NIV.

Participation involves one face-to-face or telephone interview with me, a nurse researcher, which will last only 30-40 minutes and will take place at a time and place most convenient to you. Your participation is entirely voluntary and you may cease or withdraw from the interview at any time.

Although this study might not benefit you or your family directly, this study should contribute to a better understanding of the complex issues involved in treatment decision-making for seriously ill patients.

To be included in this research, participants must speak English and be over eighteen years, be a family member or significant other of a deceased MND patient, be able to give informed consent, and will be assessed on these criteria if interested in participating, by the researcher at the stage of the return of the consent form prior to the interview commencing.

If you are interested in assisting this research, further information can be obtained by contacting the researcher Lottie Chapman RN., M. Palliative Care at: cschapma@our.ecu.edu.au. Or by phone

You may also speak to my Ph.D. Supervisor about this study, Professor Anne Wilkinson at (08) 6304-3540 or email her at: anne.wilkinson@ecu.edu.au.
Appendix I: Silver Chain Notice of Research

Approved Research Project for Palliative Care, Respiratory, Neurology Doctors and Nurses or Allied Health professionals with experience with MND and NIV

A retrospective study into the communication surrounding the initiation and withdrawal of non-invasive ventilation (NIV) in patients with motor neurone disease

The aim of my research is to explore the communication process that occurs between clinicians, patients and significant others regarding the initiation (or refusal) and withdrawal of non-invasive ventilation (NIV) for people diagnosed with Motor Neurone Disease (MND). Communication surrounding NIV has been identified as an area causing some confusion and needing further research (National Institute for Health and Clinical Excellence, 2010).

In this study, I am looking specifically at how clinicians, patients and families make treatment decisions and how communication influencing decision-making can be enhanced. The PhD research also incorporates a small study interviewing clinicians at a UK hospice as a comparison with Western Australia. This project has been approved by the Edith Cowan University Human Research Ethics Committee as research for a PhD and by the Silver Chain Human Research Ethics Committee.

You are invited to discuss your experiences with MND, and specifically NIV, contributing to recommended research into NIV communication. Participation involves one face-to-face or telephone interview with me, a nurse researcher, which will last 20-30 minutes and will occur at a time and place most convenient for you. Your participation is entirely voluntary and you may cease or withdraw from the interview at any time.

To take part in the interview or obtain further information, please contact me: Lottie.Chapman,RN.,M.PalliativeCare: cschapma@our.ecu.edu.au. Phone:

You may also speak to my Ph.D. Supervisor about this study, Professor Anne Wilkinson at (08) 6304-3540 or 075 or email her at: anne.wilkinson@ecu.edu.au
Appendix J: Family Participant Information Letter

A retrospective study into the communication surrounding the initiation and withdrawal of non-invasive ventilation (NIV) in patients with motor neurone disease

Dear Family Member:

My name is Lottie Chapman, Registered Nurse and Ph.D. Candidate in the School of Nursing and Midwifery, Edith Cowan University. I am writing to you to ask for your participation in the research I am conducting for my Ph.D. The aim of my research is to explore the communication process that occurs between clinicians, patients and significant others regarding the initiation (or refusal) and withdrawal of non-invasive ventilation (NIV) for people diagnosed with Motor Neurone Disease (MND). Communication around NIV has been identified as an area needing further research (National Institute for Health and Clinical Excellence, 2010)(National Institute for Health and Care Excellence, 2016).

In this study, I am looking specifically at how clinicians, patients and families make treatment decisions and how this decision-making can be improved. This study has been approved by the Edith Cowan University Human Research Ethics Committee as research for a PhD.

You are invited to discuss your experiences in this area and contributing to much needed research into NIV communication. Participation involves one face-to-face or telephone interview with me, a nurse researcher, which will last only 30-40 minutes and will take place at a time and place most convenient to you. Your participation is entirely voluntary and you may cease or withdraw from the interview at any time.

The interview will be audio taped for accuracy. All information will be kept private and confidential, and no identifying information will be released to any source except where required by law. A research identification number will be assigned to the interview and associated transcripts and only I will know the identity of the respondent. All data will be stored on password-protected computers in the researcher’s computer at ECU. Only authorised persons, who understand that this information must be kept confidential, will have access to it. When the study has been completed, all anonymous recordings and written documentation will be archived and stored in securely locked files for a minimum period of five years, as required by law. Your answers will be combined with the answers of other people for analysis and reporting purposes. Findings will be reported in the researcher’s dissertation and may be published in relevant health related peer-reviewed journals. No personal or identifying information will be conveyed in any of the publications arising from this research. By taking part in this study you agree not to restrict the use of any data, even if you withdraw.

Although this study might not benefit you or your family directly, this study should contribute to a better understanding of the complex issues involved in treatment decision-making for seriously ill patients. Discussing medical decision-making for your family member is not expected to create any
emotional distress for you, the participant, but it may raise minor concerns for some individuals. If you would like to have a support person present during the interview that may be very helpful for your support. Details of a free counselling service and the MNDA Care Advisory team offering emotional support and advocacy (Motor Neurone Disease Association of WA Inc Phone: (08 9346 7355) and MND NSW (02 8877 0999) are supplied to you should you wish to discuss any issues which may arise from the research interview. If you have any questions raised about this survey or to have someone to talk with you, support information and contact information for counsellors will be supplied to you.

The following information provides the specifications required for participation in this research:

**Inclusion criteria:**
1) All family/significant other participants, must have either been closely involved with the MND patient particularly if the patient used or refused NIV
2) All participants must speak English and be over eighteen years, be able to give informed consent, and will be assessed on these criteria if interested in participating, by the researcher at the stage of the return of the consent form which includes a description of these criteria.

**Exclusion criteria:**
1) Anyone under 18 years
2) Anyone unable to give consent
3) Anyone unable to communicate in English
4) Anyone with no connection to people with MND
5) Any family previously associated with the researcher as a nurse who cared for their family member with MND
6) A person with MND (unless the person with MND specifically wishes to tell their story)

To take part in the interview or obtain further information, please contact me at: cschapma@our.ecu.edu.au. You may also speak to my Ph.D. Supervisor about this study, Professor Anne Wilkinson at (08) 6304-3540 or email her at: anne.wilkinson@ecu.edu.au. If you have any concerns about the research project or wish to talk to an independent person, you may contact the Research Ethics Officer at Edith Cowan University, 270 Joondalup Drive, Joondalup WA 6027: Phone: 08 6304 2170 or Email: research.ethics@ecu.edu.au.

I really appreciate your interest and time in reading this letter,

Sincerely,

Charlotte (Lottie) Chapman, PhD Candidate, Edith Cowan University
### Appendix K: NVivo

#### Node Screen Shot

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<th>Sources</th>
<th>References</th>
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on their condition, one GP in particular, or doctor, was very inappropriate about discussions over mum’s bed, like it’s that not including the client as such, to say that “she would need to be out of here in a nursing home.” And that was particularly upsetting for mum and dad.

Reference 6: 0.68% Coverage

there seems to be sort of stages with MND. You see – well, with mum’s case. So there was a significant change and drop. That was probably when I stopped work and came down, and, you know, was there too. So that was a very inappropriate and very upsetting type of conversation, and it happens all the time with the medical model,

Reference 7: 1.84% Coverage

F6: Because these people are not suitable for a nursing home. I mean, to
Appendix M: 1) Example of Initial Interview Coding

being on the phone to me every – you know, virtually every hour or so, changing medications and changing things, and it’s just enormously difficult.

And, you know, when do you decide that, you know, today they’ve been on their NIV for 23 and a half hours; is today the day, or was yesterday the day, or is tomorrow the day, or are we not going to do this? It’s just – oh, difficult every time.

INTERVIEWER: Okay. So that’s as far as you’re concerned. Obviously the nurses would probably share that too. What about the families? Have you – –

RS1: I think it’s difficult from their point of view as well, because they, you know, look to us for guidance, and particularly to the doctors for guidance. And I’m more thinking about my palliative care experience rather than my respiratory experience, because as a respiratory physician I rarely get involved in MS and motor neuron disease, but sometimes. I suspect it’s enormously difficult from the family’s point of view unless the conversations leading up to it have been really good and really clear, and they feel like they – when it works well, they say, you know, “Patient X said when it got to this point that, you know, he/she didn’t want to be on NIV any more, and the plan was always going to be that we were going to, you know, focus on medicines or focus on keeping them comfortable.”

Or, you know, “They said that they didn’t care if they died on the machine just so long as they were comfortable.” When the conversations have been clear and their directives are clear upfront it’s easier, but that’s not as common as it should be.

INTERVIEWER: Well, I guess that’s what this is all about, trying to find a process, or something, that’s going to make – because this is my experience too, and I guess that’s what I’m trying to probe all about. Question 34: Do you have any comments relating to communication surrounding NIV initiation or withdrawal? Anything that you think could help; something that we can learn from, me in particular; the issues relating to NIV use and withdrawal?

RS1: I think the more conversations the better. I think that those – that you can’t have too many of them in one way, though you don’t want to, you know, berate people and beat them over the head with the idea that they’re going to die. So there is a balance in that to be struck. I think that very often – I’m going to be a little bit controversial, and intentionally so. I think very often
### Appendix M: 2) Secondary Random Participant Coding

<table>
<thead>
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<th>F1:</th>
<th>F2:</th>
</tr>
</thead>
<tbody>
<tr>
<td>Yeah, MNDAWA. They offered – we ended up getting Silver Chain in, who helped out with getting some home modifications. Mum and dad already had electric beds, but dad had already bought mum an electric recliner chair, and she pretty much slept on that for the last year and a half before we knew she was sick, because she said she can’t sleep in the bed. <strong>INTERVIEWER:</strong> But did they offer you alternatives to actually – even starting on the ventilation? I mean, they could have simply offered you medications to help with her comfort and things. Was she quite happy to start on the - - - -</td>
<td>She just didn’t want the one over the nose and the mouth. <strong>F1:</strong> Yeah, you could have the one that went up the nose, and she just flatly refused, because mum slept with her mouth open, which stopped the sucking. So she only got the one on the nose. So we tried to get the one on the nose and the mouth, but she wouldn’t have it. She felt, I guess, claustrophobic.</td>
</tr>
<tr>
<td><strong>F1:</strong> We just – you mean the - - - -</td>
<td><strong>F2:</strong> She just didn’t want the one over the nose and the mouth. <strong>F1:</strong> Yeah, you could have the one that went up the nose, and she just flatly refused, because mum slept with her mouth open, which stopped the sucking. So she only got the one on the nose. So we tried to get the one on the nose and the mouth, but she wouldn’t have it. She felt, I guess, claustrophobic. At that stage she wasn’t talking very well, and she was a little bit cranky at us because we kept trying to offer her more things and she just didn’t want it.</td>
</tr>
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Mum and dad are probably just of that generation that, “The doctors told us not to –” So I’m going to do it. But then mum started saying no to things, which frustrated dad and caused some issues, but - - - -

Do you ever recall mention anywhere along the line of when the ventilation was being offered of it no longer being useful and may have to be withdrawn, or stopped or that – the other part of this question is did you all understand that it wouldn’t halt the progression of the disease?

F2: Yeah.

---

4) To what degree do MND families/significant others feel that the NIV communication provided by the clinicians at the time of the offer of NIV prepared them for and helped them to understand the implications of potential NIV withdrawal resulting in the death of their loved one?

F1: Mum and dad are probably just of that generation that, “The doctors told us not to –” So I’m going to do it. But then mum started saying no to things, which frustrated dad and caused some issues, but - - - -
Appendix N: Silver Chain Ethics Approval

19 May 2015

Charlotte (Lottie) Chapman

Dear Lottie,

Ethics Application 101 – A retrospective study into the communication surrounding the initiation and withdrawal of non-invasive ventilation (NIV) in patients with Motor Neurone Disease (Staff Interviews)

Thank you for your application to the Silver Chain Human Research Ethics Committee in relation to the above study.

I am pleased to advise that the above application (staff interviews only component) is approved, after the following documents were reviewed:

- Research Application Coversheet dated 15 April 2015
- Ethics Application Form
- Project Proposal (PhD Candidacy) and associated documents

Silver Chain ethics approval is valid from 18 May 2015 to 18 May 2018 (3 years).

It is a condition of approval that a report be provided to the Committee at least annually (reports are due on 18 May each year) and on completion of the study. Any adverse experiences associated with the study should be reported to the Committee as they occur, and any changes to the project require prior approval.

The Silver Chain Human Research Ethics Committee is constituted and functions in accordance with NHMRC National Statement on Ethical Conduct in Human Research (March 2007).

Please quote EC App 101 on all future correspondence relating to the study.

I wish you every success for the conduct of the study.

Sincerely,

Dawn Woods
Executive Officer
Silver Chain Human Ethics Committee
Appendix O: St John of God Hospitals Ethics Approval

13 August 2015

Ms Lottie Chapman

Dear Ms Chapman,

Re: A retrospective study into the communication surrounding the initiation and withdrawal of non-invasive ventilation (NIV) in patients with motor neurone disease (Our ref No: 814)

I refer to the letter of 12 June 2015, advising of ethical approval of the above study, as granted by the St John of God Health Care (SJGHC) Human Research Ethics Committee (HREC).

I am now in receipt of the Participating Site Operational Approval Form for the St John of God Murdoch Hospice ("the participating site"). Accordingly, I now confirm final approval for your study to be conducted at the participating site.

I wish you well with your research.

Yours sincerely

Dr Mark Lubliner
Group Director Medical Services
St John of God Health Care

cc. Ms Sarah Hession, Acting Director of Nursing, SJG Murdoch Hospital (via email)
cc. Professor Leanne Monterosso, Chair Nursing Research, SJG Murdoch Hospital (via email)
Appendix P: Research Approval Neurological Council of WA

27 July 2015

To ECU Ethics Committee

In regards to:

**Project Number:** 12099 CHAPMAN

**Project Name:** A retrospective study into the communication surrounding the initiation and withdrawal of non-invasive ventilation (NIV) in patients with motor neurone disease

**Student Number:** 3025013

I would like to provide support to this project on behalf of the Neurological Council of Western Australia (NCWA) and the Community Neurological Nurses who provide support to this client group, in collaboration with other relevant agencies in this regional area.

I will copy the Executive Officer of the MND association into this letter and the local NCWA team.

Please let me know if you require any further information.

Yours sincerely

Kathy McCoy, RN, BPSM, MTA, Dip Hed (Neuro), PGCert (Riv), BSc (Hons), MSc (Neuro), GAICD

Executive Director / Neurological Council of Western Australia

Professional Affiliate and Honorary Senior Professional Fellow: WA Neuroscience Research Institute (WANRI)

Cc: Lisa Lambert Advanced Community Neurological Nurse

Cc: Maeve Egan Executive Officer MNOWA

---

THE NEUROLOGICAL COUNCIL OF WA INC

Perth
Centre for Neurological Support
Suite B, The Niche
11 Abdiham Road
Necklands WA 6009
Phone: 1800 665 771
or (08) 9364 7533
Fax: (08) 9346 7534
Email: admin@ncwa.com.au

Bunbury
Lotteries House Building
90-103 Victoria Street
Bunbury WA 6230
Phone: (08) 9792 4073
Fax: (08) 9791 7414
Email: admin@ncwa.com.au

Albany
Lotteries House Building
35-47 North Road
Albany WA 6330
Phone: (08) 9841 5433
Fax: (08) 9841 5433
Email: admin@ncwa.com.au

Geraldton
Lotteries House Building
114 Sanford Street
Geraldton WA 6530
Phone: (08) 9938 0760
Fax: (08) 9946 8481
Email: admin@ncwa.com.au

Mandurah
102, 1st Floor
117 South Street
Mandurah WA 6210
Phone: (08) 9595 6324
Fax: (08) 9595 6325
Email: ncwaadministrations@gmail.com

320
CONFIDENTIALITY AGREEMENT between:
LOTTIE CHAPMAN
AND
MARILYN BECK

This is to certify that I, Marilyn Beck (transcription subcontractor), agree to be bound by a confidentiality agreement with regard to transcription work undertaken on behalf of Lottie Chapman.

I agree that no reproduction of any audio or transcriptions undertaken by me on behalf of Lottie Chapman will occur, and the contents of the audio and typed transcripts will remain confidential and not divulged to another person or entity.

Upon total completion of the above work, and as requested by Lottie Chapman, all audio and transcript copies shall be destroyed by me, Marilyn Beck.

DATED: To commence September 2015

SIGNED:

Marilyn Beck
Appendix R: Ethics Approval Wisdom Hospice

Miss Charlotte Chapaman

6th July 2015

Dear Miss Chapman,

Letter of access for research

A retrospective study into the communication surrounding the initiation and withdrawal of non-invasive ventilation (NIV) in patients with motor neurone disease (15-054)

This letter should be presented to each participating organisation or organisations listed below, before you commence your research at that site.

**NHS organisations and locations:**

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<thead>
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<th>Organisation giving permission</th>
<th>Date of Permission</th>
<th>Site or sites to which permission applies</th>
</tr>
</thead>
<tbody>
<tr>
<td>Medway Community Healthcare CIC</td>
<td>6/7/15</td>
<td>Wisdom Hospice</td>
</tr>
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In accepting this letter, each participating organisation confirms your right of access to conduct research through their organisation for the purpose and on the terms and conditions set out below. This right of access commences on 06/07/2015 and ends on 31/08/2015 unless terminated earlier in accordance with the clauses below.

You have a right of access to conduct such research as confirmed in writing in the letter of permission for research from The RM&G Consortium for Kent and Medway acting on behalf of KCHT. Please note that you cannot start the research until the Principal Investigator for the research project has received a letter from us giving confirmation from the individual organisation of their agreement to the research being carried out.

The information supplied about your role in research at the organisation or organisations has been reviewed and you do not require an honorary research contract with the organisation or organisations. We are satisfied that such pre-engagement checks as we consider necessary have been carried out. Evidence of checks should be available to the organisation or organisations on request.

You are considered to be a legal visitor to the premises of the organisation or organisations. You are not entitled to any form of payment or access to other benefits provided by the organisation (or organisations) to its employees and this letter does not give rise to any other relationship between you and the organisation (or organisations), in particular that of an employee.

While undertaking research through the organisation (or organisations) you will remain accountable to your substantive employer but you are required to follow the reasonable instructions of the organisation (or organisations) or those instructions given on their behalf in relation to the terms of this right of access.

---

*The RM&G Consortium for Kent & Medway provides services to independent primary care contractors in Kent and Medway, Kent Community Health NHS Trust, Medway Community Healthcare CIC, Kent & Medway NHS & Social Care Partnership Trust and South East Coast Ambulance NHS Trust.*
Where any third party claim is made, whether or not legal proceedings are issued, arising out of or in connection with your right of access, you are required to co-operate fully with any investigation by the organisation in connection with any such claim and to give all such assistance as may reasonably be required regarding the conduct of any legal proceedings.

You must act in accordance with the organisation's policies and procedures, which are available to you upon request, and the Research Governance Framework.

You are required to co-operate with the organisation or organisations in discharging their duties under the Health and Safety at Work etc. Act 1974 and other health and safety legislation and to take reasonable care for the health and safety of yourself and others while on their premises. You must observe the same standards of care and propriety in dealing with patients, staff, visitors, equipment and premises as is expected of any other contract holder and you must act appropriately, responsibly and professionally at all times.

If you have a physical or mental health condition or disability which may affect your research role and which might require special adjustments to your role, if you have not already done so, you must notify your employer and the Occupational Health department (Jane Hitchins) at each participating NHS organisation prior to commencing your research role at each site.

You are required to ensure that all information regarding patients or staff remains secure and strictly confidential at all times. You must ensure that you understand and comply with the requirements of the NHS Confidentiality Code of Practice and the Data Protection Act 1998. Furthermore you should be aware that, under the Act, unauthorised disclosure of information is an offence and such disclosures may lead to prosecution.

You should ensure that, where you are issued with an identity or security card, a bleep number, email or library account, keys or protective clothing, these are returned upon termination of this arrangement. Please also ensure that while on the organisation's or organisations' premises you wear your ID badge at all times, or are able to prove your identity if challenged. Please note that the organisation or organisations accept no responsibility for damage to or loss of personal property.

This letter may be revoked and we may terminate your right to attend the NHS organisation or organisations at any time either by giving seven days' written notice to you or immediately without any notice if you are in breach of any of the terms or conditions described in this letter or if you commit any act that we reasonably consider to amount to serious misconduct or to be disruptive or prejudicial to the interests or business of the organisation or if you are convicted of any criminal offence. You must not undertake regulated activity if you are barred from such work. If you are barred from working with adults or children this letter of access is immediately terminated. Your employer will immediately withdraw you from undertaking this or any other regulated activity and you MUST stop undertaking any regulated activity immediately.

Your substantive employer is responsible for your conduct during this research project and may in the circumstances described above initiates disciplinary action against you.

No organisation will indemnify you against any liability incurred as a result of any breach of confidentiality or breach of the Data Protection Act 1998. Any breach of the Data Protection Act 1998 may result in legal action against you or your substantive employer or both.

If your current role or involvement in research changes, or if any of the information provided in your Research Passport changes, you must inform your employer through their normal procedures. You must also inform each participating organisation and the RMG Consortium for Kent and Medway.

Yours sincerely

Richard Collins
RMG Manager, RMG Consortium for Kent and Medway
Copies to: Fiona Stephens, Sandra Sanchez (MCH)
Appendix S: PRISMA Checklist

TITLE
Title 1 Identify the report as a systematic review, meta-analysis, or both.

ABSTRACT
Structured summary 2 Provide a structured summary including, as applicable: background; objectives; data sources; study eligibility criteria, participants, and interventions; study appraisal and synthesis methods; results; limitations; conclusions and implications of key findings; systematic review registration number.

INTRODUCTION
Rationale 3 Describe the rationale for the review in the context of what is already known. Objectives 4 Provide an explicit statement of questions being addressed with reference to participants, interventions, comparisons, outcomes, and study design (PICOS).

METHODS
Protocol and registration 5 Indicate if a review protocol exists, if and where it can be accessed (e.g., Web address), and, if available, provide registration information including registration number.
Eligibility criteria 6 Specify study characteristics (e.g., PICOS, length of follow-up) and report characteristics (e.g., years considered, language, publication status) used as criteria for eligibility, giving rationale.
Information sources 7 Describe all information sources (e.g., databases with dates of coverage, contact with study authors to identify additional studies) in the search and date last searched.
Search 8 Present full electronic search strategy for at least one database, including any limits used, such that it could be repeated.
Study selection 9 State the process for selecting studies (i.e., screening, eligibility, included in systematic review, and, if applicable, included in the meta-analysis).
Data collection process 10 Describe method of data extraction from reports (e.g., piloted forms, independently, in duplicate) and any processes for obtaining and confirming data from investigators.
Data items 11 List and define all variables for which data were sought (e.g., PICOS, funding sources) and any assumptions and simplifications made.
Risk of bias in individual studies 12 Describe methods used for assessing risk of bias of individual studies (including specification of whether this was done at the study or outcome level), and how this information is to be used in any data synthesis.
Summary measures 13 State the principal summary measures (e.g., risk ratio, difference in means).
Synthesis of results 14 Describe the methods of handling data and combining results of studies, if done, including measures of consistency (e.g., I2 ) for each meta-analysis.
Risk of bias across studies 15 Specify any assessment of risk of bias that may affect the cumulative evidence (e.g., publication bias, selective reporting within studies).
Additional analyses 16 Describe methods of additional analyses (e.g., sensitivity or subgroup analyses, meta-regression), if done, indicating which were pre-specified.

RESULTS
Study selection 17 Give numbers of studies screened, assessed for eligibility, and included in the review, with reasons for exclusions at each stage, ideally with a flow diagram.
Study characteristics 18 For each study, present characteristics for which data were extracted (e.g., study size, PICOS, follow-up period) and provide the citations.
Risk of bias within studies 19 Present data on risk of bias of each study and, if available, any outcome level assessment (see item 12).
Results of individual studies 20 For all outcomes considered (benefits or harms), present, for each study: (a) simple summary data for each intervention group (b) effect estimates and confidence intervals, ideally with a forest plot.
Synthesis of results 21 Present results of each meta-analysis done, including confidence intervals and measures of consistency.
Risk of bias across studies 22 Present results of any assessment of risk of bias across studies (see Item 15).
Additional analysis 23 Give results of additional analyses, if done (e.g., sensitivity or subgroup analyses, meta-regression [see Item 16]).

DISCUSSION
Summary of evidence 24 Summarize the main findings including the strength of evidence for each main outcome; consider their relevance to key groups (e.g., healthcare providers, users, and policy makers).
Limitations 25 Discuss limitations at study and outcome level (e.g., risk of bias), and at review-level (e.g., incomplete retrieval of identified research, reporting bias).
Conclusions 26 Provide a general interpretation of the results in the context of other evidence, and implications for future research.

FUNDING
Funding 27 Describe sources of funding for the systematic review and other support (e.g., supply of data); role of funders for the systematic review.

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For more information, visit: www.prisma-statement.org.
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### Appendix: T: COREQ checklist

Consolidated criteria for reporting qualitative studies (COREQ): 32-item checklist

<table>
<thead>
<tr>
<th>No</th>
<th>Item</th>
<th>Guide questions/description</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Domain 1: Research team and reflexivity</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Personal Characteristics</td>
<td></td>
</tr>
<tr>
<td>1.</td>
<td>Interviewer/facilitator</td>
<td>Which author/s conducted the interview or focus group?</td>
</tr>
<tr>
<td>2.</td>
<td>Credentials</td>
<td>What were the researcher’s credentials? <em>e.g. PhD, MD</em></td>
</tr>
<tr>
<td>3.</td>
<td>Occupation</td>
<td>What was their occupation at the time of the study?</td>
</tr>
<tr>
<td>4.</td>
<td>Gender</td>
<td>Was the researcher male or female?</td>
</tr>
<tr>
<td>5.</td>
<td>Experience and training</td>
<td>What experience or training did the researcher have?</td>
</tr>
<tr>
<td></td>
<td>Relationship with participants</td>
<td></td>
</tr>
<tr>
<td>6.</td>
<td>Relationship established</td>
<td>Was a relationship established prior to study commencement?</td>
</tr>
<tr>
<td>7.</td>
<td>Participant knowledge of the interviewer</td>
<td>What did the participants know about the researcher? <em>e.g. personal goals, reasons for doing the research</em></td>
</tr>
<tr>
<td>No</td>
<td>Item</td>
<td>Guide questions/description</td>
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<tr>
<td>8.</td>
<td>Interviewer characteristics</td>
<td>What characteristics were reported about the interviewer/facilitator? e.g. Bias, assumptions, reasons and interests in the research topic. Trustworthiness is reported in Chapter 3 of this thesis. No issues reported by the candidates.</td>
</tr>
<tr>
<td>Domain 2: study design</td>
<td>9.</td>
<td>Methodological orientation and Theory</td>
</tr>
<tr>
<td>10.</td>
<td>Sampling</td>
<td>How were participants selected? e.g. purposive, convenience, consecutive, snowball. Purposive and snowball.</td>
</tr>
<tr>
<td>11.</td>
<td>Method of approach</td>
<td>How were participants approached? e.g. face-to-face, telephone, mail, email. Initially the family participants applied to partake in the research having read an advertisement in the MND Australia newsletter. The clinicians were approached via their places of work who sent out an email advertising the research, and interested clinicians applied. This was snowballed by word of mouth between clinicians.</td>
</tr>
<tr>
<td>12.</td>
<td>Sample size</td>
<td>How many participants were in the study? Twenty-six.</td>
</tr>
<tr>
<td>13.</td>
<td>Non-participation</td>
<td>How many people refused to participate or dropped out? Reasons? None.</td>
</tr>
<tr>
<td>No</td>
<td>Item</td>
<td>Guide questions/description</td>
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<td>-----------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------</td>
</tr>
<tr>
<td>14</td>
<td>Setting of data collection</td>
<td>Where was the data collected? e.g. home, clinic, workplace</td>
</tr>
<tr>
<td>15</td>
<td>Presence of non-participants</td>
<td>Was anyone else present besides the participants and researchers?</td>
</tr>
<tr>
<td>16</td>
<td>Description of sample</td>
<td>What are the important characteristics of the sample? e.g. demographic data, date</td>
</tr>
<tr>
<td>17</td>
<td>Interview guide</td>
<td>Were questions, prompts, guides provided by the authors? Was it pilot tested?</td>
</tr>
<tr>
<td>18</td>
<td>Repeat interviews</td>
<td>Were repeat interviews carried out? If yes, how many?</td>
</tr>
<tr>
<td>19</td>
<td>Audio/visual recording</td>
<td>Did the research use audio or visual recording to collect the data?</td>
</tr>
<tr>
<td>20</td>
<td>Field notes</td>
<td>Were field notes made during and/or after the interview or focus group?</td>
</tr>
<tr>
<td>21</td>
<td>Duration</td>
<td>What was the duration of the interviews or focus group?</td>
</tr>
<tr>
<td>22</td>
<td>Data saturation</td>
<td>Was data saturation discussed?</td>
</tr>
<tr>
<td>23</td>
<td>Transcripts returned</td>
<td>Were transcripts returned to participants for comment and/or correction?</td>
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</table>

**Domain 3: analysis and findings**
<table>
<thead>
<tr>
<th>No</th>
<th>Item</th>
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<tr>
<td>Data analysis</td>
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<tr>
<td>24.</td>
<td>Number of data coders</td>
<td>How many data coders coded the data?</td>
</tr>
<tr>
<td>25.</td>
<td>Description of the coding tree</td>
<td>Did authors provide a description of the coding tree?</td>
</tr>
<tr>
<td>26.</td>
<td>Derivation of themes</td>
<td>Were themes identified in advance or derived from the data?</td>
</tr>
<tr>
<td>27.</td>
<td>Software</td>
<td>What software, if applicable, was used to manage the data?</td>
</tr>
<tr>
<td>28.</td>
<td>Participant checking</td>
<td>Did participants provide feedback on the findings?</td>
</tr>
<tr>
<td>Reporting</td>
<td></td>
<td></td>
</tr>
<tr>
<td>29.</td>
<td>Quotations presented</td>
<td>Were participant quotations presented to illustrate the themes / findings? Was each quotation identified? e.g. participant number</td>
</tr>
<tr>
<td>30.</td>
<td>Data and findings consistent</td>
<td>Was there consistency between the data presented and the findings?</td>
</tr>
<tr>
<td>31.</td>
<td>Clarity of major themes</td>
<td>Were major themes clearly presented in the findings?</td>
</tr>
<tr>
<td>32.</td>
<td>Clarity of minor themes</td>
<td>Is there a description of diverse cases or discussion of minor themes?</td>
</tr>
</tbody>
</table>