The Experience of Non-Invasive Ventilation in Motor Neurone Disease: A Qualitative Exploration

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Chapter 9: General Discussion

9.1 Introduction

The research presented in this thesis was undertaken to meet two broad aims. The first aim was to qualitatively explore the lived experience of respiratory impairment in MND, including how symptoms and signs of respiratory insufficiency are interpreted in the wider illness context and how events such as referral and assessment are perceived by patients. The second and more significant aim was to qualitatively explore the lived experience of NIV use in MND. This included exploring how patients approached treatment decision-making, the factors considered important when making choices about NIV, responses to the prospect of imminent treatment initiation, and how patients perceived the experience of treatment implementation. It also included the process of acclimatisation and early-stage use. It was intended that this thesis would investigate the *subjective* experience of NIV use, defining the treatment in terms of process and experience rather than objective outcomes. The ultimate goal was to provide a body of qualitative research to inform the clinicians who provide respiratory care to this unique patient population. This final chapter discusses the results of this research and outlines the significance of the findings in respect to their clinical application. This chapter is intended to summarise and discuss the main conclusions without necessarily reiterating all findings.

9.2 General Reflections

The data presented have demonstrated the highly individual nature of patient and carer experiences in MND. There was no single story that defined respiratory impairment or NIV use in this population but a collection of individual voices all trying to make sense of unique experiences within a personal illness context. This finding itself seemed to justify the use of the idiographic approach that allowed these individual voices to be heard. The diversity of experience reflected in the thematic analysis suggest that it may not be possible to predict responses to NIV from demographic or clinical variables alone, with no discernible 'profile' of patients experiencing 'success' or 'failure' with the treatment emerging from the data. Patients' reactions were often best understood as part of a wider response to the illness, which itself was shaped by complex individual differences, such as personality, coping style, established social networks, and the variable patterns of disability inflicted by the disease. It was, however, possible to identify variables that appeared to be potential determinants of tolerance and compliance. These were 'red flag' issues that should alert clinicians to potential complications and are discussed in this final chapter along with suggestions for clinical changes that might help to improve patients' experiences of referral and NIV use. This is presented in six broad sections: 'The Experience of Respiratory Impairment', 'Response to Non-Invasive

Ventilation', 'Knowledge & Understanding', 'Treatment Decision-Making', 'Defining "Need"' and 'The Experience of Non-Invasive Ventilation'. There then follows some general reflections on the research process.

9.3 The Experience of Respiratory Impairment

As outlined in Chapter 2, research addressing respiratory aspects of MND has relied exclusively on quantitative methods and has generally only charted the physiological outcomes of respiratory compromise. This thesis reports the first qualitative exploration of the subjective experience of respiratory impairment from the patient perspective. Clinicians already have access to objective markers of respiratory function and muscle strength, yet may not be aware of how patients actually experience these changes.

9.3.1 The Emotional Impact of Referral

Clinical referrals are made as pragmatic responses to illness progression, directing patients towards specialist services that can investigate problems and provide solutions; however, for patients in the present study, there appeared to be a significant emotional component to the process of respiratory referral. One of the most significant determinants of negative emotional response was the fact that respiratory involvement was often entirely unexpected. There were clear differences in how much each patient knew and wanted to know about respiratory aspects of the illness at the time that they entered the respiratory service (see Section 6.5). Many patients had been completely unaware that respiratory function might be compromised by MND and even after referral most knew relatively little about this area of the illness. This meant that the majority of patients were making psychological adjustments both to the news that the respiratory system might be compromised by the illness *and* the fact that healthcare professionals suspected that this process had already begun. This may explain why shock, disappointment, fear and anxiety were so commonly reported. It was clear that referral was often an emotionally turbulent event, with patients exposed to uncertainties that challenged existing illness schemas and redefined illness parameters in an unexpected way.

Anxiety also appeared to have been associated with the nature of the referral itself, as most patients attached particular significance to respiratory symptoms over other types of change. This aspect of the illness was afforded special status because of the perceived link between respiration and life *and* because of the specific link between the respiratory system and death in MND. The act of referral made these issues personally relevant to each patient, which meant that even patients who had been aware of the prospect of respiratory compromise were vulnerable to distress. The fact that this area of decline was viewed as conceptually different from decline in other domains moved patients to a

new stage of the illness, one where 'basic' functions were potentially threatened and where life, rather than just *quality* of life, became a relevant outcome.

Section 1.8.2 describes the low levels of anxiety reported in the wider MND literature, yet also notes that anxiety may increase at specific points along the illness trajectory (e.g. towards the end of life; Ganzini et al., 2001). Importantly, it is also noted that vulnerability to anxiety may increase in response to specific events (e.g. diagnosis; Vignola et al., 2008) and when specific symptoms emerge (e.g. bulbar impairment; Goldstein et al., 2006). The data presented in this thesis suggest that respiratory referral is another event that exposes MND patients to a heightened risk of anxiety, particularly where patients have previously been unaware of the risk of respiratory compromise. Respiratory referral introduced the prospect of further decline, yet it also raised questions about the future and the end of life.

Clinicians should be mindful that patients may attach greater significance to respiratory referral over other referrals occurring over the course of the illness and should consider what this referral means to patients. It may even be prudent to ensure that psychological support is available to all patients entering the respiratory service to help in the process of adjusting to this change and making sense of its place in the wider context of the illness.

9.3.2 Evaluating Respiratory Change/Status

Anxiety was presented as a response to the *potential* for respiratory impairment to occur and was not typically linked to current respiratory status, as patients typically presented a positive evaluation of their respiratory function. Many felt that referral was a disproportionate response to their personal situation and some even reported tensions with healthcare professionals over this issue. A number of patients seemed consoled by the idea that referral was merely "routine" rather than responsive. This suggested that not all patients understood why they had been referred in the first place, or at least did not want to accept the reasons. There was a notable tendency for patients to deny that there had been any change at all in respiratory function *despite* many reporting obvious signs of respiratory insufficiency. In some cases it seemed likely that patients were trying to protect themselves from distress; however, many patients intimated that positive evaluations of respiratory status were based on rational appraisals of the situation. Appreciating the strategies that patients used to make sense of respiratory change seemed key to understanding their subjective experiences.

It seemed important to note that patients typically associated respiratory impairment with dyspnoea and when discussing respiratory change most patients were only reflecting on the presence or absence of this specific symptom. This seemed to be an assumption based on existing notions of

respiratory impairment, wherein 'respiratory' was only seen to relate to the subjective experience of breathing. Only a small cluster of patients appreciated that respiratory symptoms might have more subtle or indirect presentation (e.g. as fatigue or waking-headaches). These misconceptions appeared compounded by the fact that many patients and carers assumed that respiratory change would take the same aggressive form as change occurring elsewhere in the body, where it had been observed to quickly deprive patients of functions absolutely. The extreme symptoms that patients predicted seemed to allow them to appraise their own respiratory status as *comparably* good. Many carers shared these ideas about respiratory change, suggesting that these may have been misconceptions that were generated and/or reinforced within the dyad.

Patients are known to use existing illness representations to label and make sense of emerging symptoms within many illness contexts (Baumann, Cameron, Zimmerman, & Leventhal, 1989) and this in turn can both shape emotional responses to change and determine attitudes towards help-seeking behaviours (Cameron, Leventhal, & Lventhal, 1993). Illness expectation has been shown to significantly influence how likely patients are to seek medical attention during acute myocardial infarction (Horne, James, Petrie, Weinman, & Vincent, 2000), after stroke (Zerwic, Hwang, & Tucco, 2007) and in the early stages of cancer (Smith, Pope, & Botha, 2005). In this study, it appears that expectations and pre-established illness representations may have guided MND patients' understanding of respiratory symptoms, emotional responses to these changes, and attitudes towards the offers of help being made through referral. The nature of MND, and past experience of decline in other physical domains, built expectations of more severe respiratory impairment, making referral on the basis of only mild and unproblematic symptoms appear incongruent and unnecessary and helping to appease negative emotional responses. Patients often compared current respiratory status to an imagined extreme rather than comparing it to a time when no symptoms were present. They seemed more likely to compare breathlessness against a standard of "gasping for breath" than against normal respiratory function (see Section 6.7.2.2), which helped in forging the perception of respiratory change as relatively unproblematic. This positive evaluation enabled patients to minimise the significance of respiratory change and decreased the likelihood that they would report symptoms and/or perceive the need for clinical attention.

Many patients had also attributed respiratory symptoms to non-respiratory causes (e.g. general fatigue) and this also allowed them to preserve the belief that the respiratory system remained unaffected by MND. Dyspnoea and fatigue in particular were often framed as secondary consequences of decline in other areas of the body. Attributing respiratory symptoms to non-respiratory causes seemed to protect patients from the idea that they had encountered an *additional* area of decline and also meant that patients did not have to acknowledge decline in a domain that they felt had particularly negative implications. It is likely that some patients were simply resistant to the idea that the respiratory system had been compromised by the illness; however, it may also be

possible that patients without prior knowledge of respiratory aspects of the illness had already integrated respiratory systems into existing illness schemas as secondary symptoms rather than symptoms in their own right. Illness schemas, selective attention and personal interpretation are important in making sense of even normal physiological changes (Pennebaker, 1982; Weinman & Petrie, 1997) and so were likely to be important in this context. Sharpe and Curran (2006) suggest that it can take time for patients to integrate new aspects of illness into existing illness representations and being able to make sense of symptoms is likely to be a particularly complex process in MND, where patients were attempting to understand multiple changes occurring across a number of physical domains.

It was clear that patients and clinicians approached respiratory decline and its consequences differently. Clinicians aimed to identify change and to correct it, whether this change was expressed in subjective impairment (e.g. waking-headaches or fatigue) or where change met objective criteria (e.g. where oximetry showed impaired function). For patients it seemed it was only subjective change that was used to define a need for referral and intervention. Patients did not appear concerned with change per se but with change that was considered problematic according to their own criteria (e.g. where it caused subjective discomfort or functional impairment). These criteria were themselves defined by other complex contextual variables (e.g. impairment in other domains and available coping resources). In interviews, patients seemed to interpret questions about 'change' in respiratory function as enquiries about 'problems' that they were having, which often led them to omit details and, perhaps unintentionally, obscure the true nature of respiratory insufficiency. Where patients felt they were able to cope with changes (i.e. where they were not perceived to be 'problems'), they appeared less likely to report them to clinicians and also expressed less concern. It seemed that patients did not want to have to assign valuable resources – both psychological and practical – to every change that occurred; they wanted to focus on changes that posed immediate challenges and that really needed to be addressed. This is where context appeared most important: patients were often fighting change across multiple physical domains, yet respiratory clinicians were only focusing on respiratory aspects of the illness. This created an implicit conflict of interests and intentions. There appeared to be fundamental differences in the way that clinicians and patients approached the illness: clinicians were proactive, seeking to pre-empt problems to avoid physical suffering and distress; whilst most patients only wanted to address change as it occurred and where it began to pose problems (i.e. where it caused discomfort or imposed functional limitations).

Importantly, most patients felt that the impact of respiratory impairment had been minimal. When discussing respiratory change, patients often cited other symptoms (e.g. pain and sialorrhoea) as being *more* important in the wider context of the illness. This suggested that patients had undertaken a process of symptom prioritisation in order to define a hierarchy of needs. By comparing symptoms in this way, patients were able to focus their limited coping resources on

addressing symptoms that had the biggest impact on their wellbeing. It is perhaps understandable that respiratory aspects of the illness were not prioritised, given that they were not perceived to be having a sustained impact on patients' day-to-day lives. The symptoms that patients did prioritise were often more enduring symptoms that had a continual subjective impact on wellbeing (e.g. pain). For some patients, the fact that clinicians were expressing concern about *nocturnal* respiratory function appeared to add to the sense that there were more important problems to address (i.e. those effecting them during the day when they were awake, subjectively aware and could potentially be physically impeded). Indeed, two patients even suggested that it was preferable that respiratory problems were present at night rather than during the day when they might be active. It was clear that there were significant differences in the importance placed on this new area of decline at the point of referral. Patients naturally prioritised symptoms that caused immediate discomfort and functional impedance; respiratory aspects of the illness did not fall into either category for most patients.

Adding further complication, physical disability also seemed to mask respiratory aspects of the condition. Some patients recognised that not being able to physically exert themselves and place strain on the respiratory system actually meant that there might be respiratory compromise that they were unaware of. However, many patients interpreted this issue *positively*. They suggested that limited mobility *demoted* the importance of respiratory aspects of the condition and hence the importance of referral. A number of patients explained that respiratory symptoms might have been more important had they still been active and had their activity been impeded by respiratory insufficiency. In this way, inactivity seemed to provide protection against respiratory distress. This again reflected the idea that the most important consideration was whether respiratory impairment caused notable *problems* (i.e. distress or impairment), not whether it was simply occurring or not. These complexities added to the sense that clinical attention did not need to be given to respiratory aspects of the illness. Section 2.10 criticises existing quantitative studies for considering respiratory events in isolation, with no attempt to describe or explain how respiratory change fits into the wider experience of MND. Data from the present study confirm the need to look beyond the symptoms themselves to the broader illness context, as patients' experiences of respiratory impairment did not seem to map directly onto the levels of respiratory function or respiratory muscle strength. Patients' subjective experiences were clearly influenced by factors such as existing levels of physical disability.

Most patients also demonstrated a natural propensity to *adapt* to respiratory impairment (e.g. by avoiding specific activities, resting and pacing), which again minimised the subjective impact of symptoms. Despite patients' protestations that they had not experienced respiratory change or else did not find change problematic, it seemed that the need for adaptive strategies provided indirect evidence that respiratory changes had impacted negatively on patients in some way. This may have

been through actual functional impedance or through anxiety about the potential for early changes to develop into more restrictive symptoms. Respiratory change had at the very least been identified as being potentially problematic to many patients and this is why they had made adaptations. Lansing et al. (2009) note that affective responses to respiratory symptoms can prove useful in stimulating learning strategies for the avoidance of potentially threatening sensations (e.g. activity reduction and pacing to avoid respiratory exertion), with breathlessness often acting as the primary cue that something is wrong and needs to be corrected. They suggest that an ability to adapt behaviour to regain respiratory control will lead to minimal psychological consequences, whilst an inability to reduce respiratory distress will incur greater emotional reaction. It seemed likely that initial discomfort instigated the types of behavioural changes observed in the present study. This would also offer explanation as to why dyspnoea at rest was more likely to be seen as distressing, as without an external 'trigger' that can be controlled (e.g. activity), patients were limited in their ability to alleviate or prevent change. Even patients in the present study who were experiencing orthopnea were able to prevent symptoms by not sleeping in a supine position.

Control seemed an important determinant of response. It was clear that MND patients in this study were able to regain respiratory control relatively easily when breathlessness occurred. They adapted to prevent reoccurrence of distress by modifying behaviours; this meant that the emotional strain of respiratory change was low and self-efficacy remained high despite the presence of respiratory impairment. Episodes of notable respiratory change were reported to be uncomfortable rather than distressing and were not felt to compromise wellbeing. Such episodes were also transient rather than sustained, contributing to the notion that they were secondary symptoms rather than being reflective of impairment within the respiratory system itself. This all added to the sense that these changes did not need to be prioritised. Patients seemed to have good control over respiratory disturbance, which they lacked over other symptoms (e.g. pain and sialorrhoea). This indicated that control was another issue that shaped the importance attached to symptoms. A number of patients were aware that they could change behaviours to limit the risk of dyspnoeic responses. However, it is again important to consider context, as many patients had adapted behaviour in response to physical changes in other domains (e.g. limb-weakness or fatigue), with respite from potential respiratory symptoms being a secondary effect or even an incidental consequence. In terms of the *experience* of respiratory change, most patients felt that they were able to cope with symptoms or else were unaware of change and reported low levels of discomfort and distress. The ability to control or avoid respiratory distress is another reason why patients felt that these symptoms did not necessitate clinical attention.

In many ways this situation can be interpreted positively, as it was clear that although healthcare professionals had noted potential signs of respiratory decline, and patients themselves often described symptoms, respiratory impairment did not appear to be impacting on patients' wellbeing.

In respect to the subjective experience of respiratory impairment at the point of referral, most patients seemed content not to focus on this aspect of the illness. It remains unclear to what extent patients truly believed that there had been no change in respiratory function or that these changes somehow did not matter at referral. It seemed that there may have been aspects of denial in patients' dismissal of clinicians' concerns; however, since this strategy protected patients from distress and did not prevent them from complying with clinical tests and procedures, it might be suggested that it did not need challenging. The situation becomes more problematic in instances where respiratory tests provide evidence of respiratory insufficiency and clinicians seek to act. In these instances, patients' subjective appraisal of the situation conflict with the clinical view and this may create a barrier to intervention. Certainly, this conflict appeared to evoke resistance in some patients as they moved onto NIV. Challenging patients' ideas about respiratory status where they were not experiencing subjective physical symptoms seemed to create distress, yet patients also reacted negatively to being "unprepared" for ventilation.

Discrepancies between the views of clinicians and those of patients appeared to be extremely important. There were differences in knowledge and understanding that were to be expected, yet this seemed to create a much wider divide between what each party anticipated in this area of the illness and, as such, how they appraised current status. This determined how patients prioritised respiratory care as a whole. Section 2.10 asks the question of whether patients shared the sense of importance and priority that clinicians assigned to respiratory care and the answers seemed complicated. Patients did share an acknowledgement that respiratory aspects of the illness were important, yet based on their own appraisals of the situation they did not feel that they had experienced sufficient respiratory change to warrant clinical attention at the point of referral.

9.4 Response to Non-Invasive Ventilation

Existing research has focused on the outcomes of NIV use rather than on patient perceptions of the treatment (see Chapter 2). The present study also explored patients' views about ventilation at the point of referral, capturing an initial response to the idea of this intervention. One of the most striking features of patients' responses was that many reacted to what the ventilator represented to them about the illness and its progression, rather than reacting to the ventilator itself. Most patients immediately attended to the psychological challenge of NIV rather than to more practical concerns.

A need for intervention was seen to signal physical decline and illness progression and it was suggested that NIV represented 'defeat'. In this way, ventilation had already been framed as an ending rather than a beginning and represented loss rather than opportunity for many patients. This typically meant that a great deal of negative emotion had been invested in the treatment before

patients had even been invited to trial it. In explaining how this view came to be established, one must consider patients' wider beliefs about respiratory change and the special status that decline in this area held. The ventilator was a means of supporting respiratory function and patients reacted negatively to the idea that they could not sustain this basic function alone. Respiratory decline was perceived by many patients to be the "beginning of the end" and NIV appeared to move patients closer to the final stages of the illness. Interestingly, this was a view that was also held by a number of carers, who displayed significant distress about the wider implications of ventilator use.

Many patients suggested that they considered NIV to be a last resort and an option that would only be considered when they could no longer go on without it. This indicated that there were misconceptions about the role of NIV even at an early stage, with doctors presenting ventilation as a mode of supporting effective respiratory performance, while patients interpreted the treatment as compensating for an almost absolute loss. This seemed to be intrinsically linked to misconceptions about respiratory impairment itself, again emphasising the differences between patients' and clinicians' aims and understanding. Patients frequently described NIV as a necessary intervention for a body that was "shutting down", "giving up" and "struggling" (see Section 6.9.1.1). Clinicians should be mindful of this, as it may be easy for them to lose sight of the magnitude of the intervention for the MND patient, especially when the treatment is so commonly used as an everyday medical aid in the hospital. It is important to note that although NIV is used in a number of chronic illness populations, it is only in MND that it is used to treat progressively degenerative symptoms. It is also significant that MND is a terminal condition and a disease where death is most commonly associated with respiratory failure. It seemed clear that NIV was more than a palliative intervention for these patients; it had far reaching psychological and emotional consequences.

Not all patients viewed NIV in this negative light. A minority of patients presented NIV as an asset in their fight against the illness. These patients perceived NIV as a positive addition to care. The views expressed by these patients are important, as they indicate how clinicians may help to support anxious patients by helping them to reframe NIV in this positive way. Clinicians might usefully frame the ventilator as a way of helping patients to fight the illness by emphasising the supportive nature of the treatment, rather than presenting it as a response to decline and loss of function. Stressing that the treatment is an intervention intended to work collaboratively with patients against the illness, rather than work for them to compensate for physical limitations, may help to shift patients' negative perceptions of the treatment. This may also help those patients who struggle with the idea they do not yet 'need' NIV at a time that doctors feel they would benefit from its use (see Section 7.4.2), as it would allow the treatment to be viewed as supporting remaining function rather than compensating for a lost function.

Patients' ideas about NIV being an extreme measure seemed to interact with their positive appraisals of respiratory status to create and reinforce distance between themselves and the ventilator. All patients were adamant that they did not require NIV, which typically related to the view that there had been insufficient respiratory change to warrant any form of intervention.

The introduction of any new piece of medical equipment (e.g. wheelchair, hoist or PEG/RIG) provided a physical reminder of the illness and served as an indisputable sign that it was progressing. It is unsurprising that patients sought to delay this process and one can see how postponing the initiation of NIV may have been viewed as a means of holding back the progression of the disease. Many patients spoke of attempting to "put off" NIV and of trying to keep going until they could no longer manage without help. This mindset seemed closely linked to the fear of dependence that many patients reported (see Section 7.4.4.1). There was clearly a negative stigma attached to needing a "machine" to support breathing and this was exacerbated by a fear of being "stuck" on the equipment. Again, the nature of the function that this intervention was supporting was important; breathing is an automatic and constant process and patients seemed to fear that where support started off being needed overnight, it might ultimately be needed all of the time. In this way, NIV differed from other interventions (e.g. a PEG/RIG), because there was a risk of it being used to a point of absolute dependence (i.e. on a 24hr basis). Patients were aware that MND was a progressively degenerative illness and that respiratory function could only get worse not better. Given the natural direction of travel of the illness, it seemed logical to assume that increasing dependency on the ventilator would occur and that ultimately there could be a risk that they would come to rely on the machine absolutely. A number of patients were already looking ahead to the prospect of daytime use and being restricted to life on a ventilator. This was received negatively and contributed to patients' attempts to avoid NIV initiation.

Absolute dependency on ventilation in MND has been observed to occur in some countries, namely Japan, where patients are often kept alive via IV in a 'locked in' state (Kawata, Mizguchi, & Hayashi, 2008); however, this is an issue of significant ethical debate in the UK and most clinicians are acutely aware of the need to avoid dependency (Polkey et al., 1999). There are options available to patients to prevent this, including a process of gradually weaning patients off NIV, which often involves increasing the use of pharmacological support to prevent respiratory distress (Chadwick, Nadig, Oscroft, Shneerson & Smith, 2011). It was notable that patients in the cross-sectional study and ventilated patients in the longitudinal study also expressed a fear of dependence, suggesting that this issue is a potential source of anxiety for patients throughout referral, initiation and onto ventilation itself. It was also a concern that Sundling et al. (2009) noted in their cross-sectional investigation. This may be a subject that clinicians should sensitively probe at initial consultation and beyond to assess whether patients would benefit from further discussion and clarification. Discussing practical strategies that can be instigated to avoid dependence may be useful for patients

who have these specific types of anxieties, potentially helping them to address concerns before decisions are being made. However, it is likely that such discussions could increase distress in other patients, making this a difficult issue to approach clinically without a good understanding of the individual patient.

In contrast with initial negativity, most patients responded positively to the idea that there was some form of help available to them. Having been told that the respiratory system might be compromised by the illness, it seemed comforting that there was at least something that could be done to lessen the impact of change if it occurred. This relief was often expressed alongside feelings of reluctance and apprehension, which created inconsistency within narratives. This again appeared a point of difference between patients and clinicians, as it was the positive prospect of help that dominated the clinical picture. Clinicians look at the ventilator as a positive addition to care, yet patients did not seem able to embrace the idea of needing this kind of support. The prospect of actually using NIV appeared to be extremely daunting and any positive responses were only to the abstract notions of help and improvement in an illness that offered few such opportunities. Patients were pleased that they could be helped, yet unenthusiastic about the form that this help took and what the need to be helped in this way meant.

Almost all carers expressed a positive response to the idea of NIV as a form of help for patients and also as a means of helping them to fulfil their own duties as carers. Carers were often more enthusiastic about the treatment than patients, because many seemed to have adopted a pragmatic approach to the illness, as nurtured by the caring role. In some ways carers' views of NIV appeared more aligned with those of clinicians than patients. Carers seemed more optimistic about NIV, often suggesting it should be initiated as soon as it might improve respiratory function, rather than just where there was a tangible problem that could no longer be avoided. This may have been because carers would not be the ones actually using the ventilator, but may also have been encouraged by anxiety about actual or potential respiratory distress in patients and the responsibility that carers expressed in regard to the wellbeing of their loved ones. Patients felt that they were able to cope with respiratory change, yet many carers had greater concerns about respiratory symptoms. The initial reactions of carers are important because these individuals were often living with patients, discussing these issues with them and potentially influencing patient opinion and understanding. The role of carers in decision-making is discussed in detail in Section 8.3.3 and it is important to acknowledge the potential influence of these significant figures from the earliest stage.

Exploration of the views of patients at referral showed conflict and inconsistency in patients' thoughts and feelings about NIV. The tone of patients' psychological responses to the treatment were striking and it is notable that these perspectives are not represented anywhere in the quantitative research base. Indeed, studies tend to report from a point where patients' are already

established on the treatment, entirely bypassing these earlier stages in the process. Although one might predict that treatment initiation would be an emotionally tumultuous time for patients, it was unexpected that even the idea of NIV would evoke such strong responses from patients. These reactions were observed in patients who did and did not go on to be ventilated over the course of the study, suggesting that the daunting prospect of NIV may have remained with selected patients over an extended period. Clinicians should be mindful of these types of response when deciding at what point to introduce the idea of NIV to patients.

9.5 Knowledge & Understanding

Patients presented significant misconceptions about respiratory aspects of the illness, current respiratory status, criteria for NIV initiation and also about how clinicians intended the treatment to be used. In the cross-sectional study it was clear that knowledge and understanding significantly shaped patients' initial responses to ventilation, as patients suggested that not understanding why they needed the intervention increased resistance towards it. This experience was also reported in the longitudinal study; however, what emerged most clearly in the latter phase of the study was not related to knowledge or understanding alone but to broader attitudes towards illness-related information, including motivation for information-seeking *and* information-avoiding in the wider context of the illness. It seems important for clinicians to understand these attitudes, as it was clearly insufficient to give all patients the same information in the same way. Effective provision of information is an important part of the service provided by clinicians in respect to respiratory care and NIV (NICE, 2010). Understanding the reasons why patients displayed different attitudes towards information may enable healthcare professionals to provide information in a more appropriate and accessible way for patients and carers.

9.5.1 Providing Information

Simply providing information does not ensure that it is actually received and retained. Various cognitive and emotional variables can increase or decrease ability and motivation to attend to, process and recall information provided in clinic appointments. In respect to *ability* to retain information, there is substantial evidence to suggest that a patient's memory for medical information provided in consultations is relatively poor, with an estimated 40-80% of information forgotten almost immediately (Ley, 1979; Kessels, 2003). Empirical tests consistently show that patients often misunderstand medical information and are prone to significant inaccuracies in recollection (Anderson, Dodman, Kopelman, & Fleming, 1979). There is also an additional 'age-effect', as older patients have been shown to remember significantly less than younger patients (Morrow, Leirer, Carver, Tanke, & McNally, 1999). This is an important consideration given that MND is an illness of later life. Memory recall for medical information is also known to be poorer where information

defies expectations of the illness in terms of existing illness schemas (Okun & Rice, 2001) and also where patients are anxious or stressed (Shapiro, Boggs, Melamed, & Graham-Pole, 1992). Both of these factors seem important in the context of MND, where most patients reported being unaware that the respiratory system might be affected by the illness and where the primary responses to this news were shock, disappointment, anxiety and fear (see Section 6.6.1). Given the nature of the information being presented to patients, there is good reason to suggest that patients may have found the experience of receiving this information in clinics stressful, making it less likely that they would accurately process and retain it.

It is also important to consider patients' wider attitudes and their *motivation* for engaging with information. Most patients in the present study demonstrated avoidance of information that was punctuated by sporadic bursts of information-seeking when illness progression presented a physical or functional change. In the context of terminal illness, this behaviour mirrors reports of self-censorship in cancer patients, who also appear to both seek and avoid information at different stages of their illness as a means of avoiding distress and preserving hope (Leydon et al. 2000). Other types of information-seeking behaviours were also displayed; a minority of patients sustained avoidance, whilst others were far more active in their pursuits of illness-related information. It is acknowledged that both of these approaches may also serve as effective coping strategies in MND, being used to minimise distress and promote psychological wellbeing (Lee et al., 2001). The three patterns of information-seeking behaviour found in the present study mirror those previously reported by O'Brien (2004), who labelled MND patients as active-seekers, selective-seekers, or information avoiders.

The present study was able to offer insight beyond the descriptive, exploring the reasons why patients avoided and engaged with information at different points in time. Most patients suggested that all information about MND was inherently negative and avoidance was a way of protecting themselves from inevitable psychological distress. In many patients this seemed to be a view that had formed at diagnosis, which was the time that it first became clear that there was no known cure for the illness and when many patients suggested that they began to 'disengage'. The unpredictable illness trajectory also introduced a risk that information obtained in advance would never be relevant, leading patients to avoid information about this 'unknowable' future as a means of circumventing potentially unnecessary upset.

Patients appeared more likely to engage with information when it was seen to be relevant and useful (i.e. when it related to a specific problem that was immediate or likely to occur in the near future). This type of information enabled patients to feel prepared for illness progression in both a practical and psychological sense. Patients did not want extraneous or irrelevant information, yet many also associated anxiety with not having appropriate information in good time. Having

multiple goals (e.g. to be informed ready for change *and* to avoid irrelevant information that may cause distress) left many patients in the precarious position of having to calculate when to switch between strategies. In some cases, this point was usefully signposted by clinicians, yet other relied on their own subjective experiences of physical change to signal the need to learn more. It was clear that most patients were hoping to avoid information for as long as possible, yet were pragmatic about its utility in addressing new challenges that emerged over time.

Patients who were more consistently engaged with the illness reported that they were more active in information-seeking as a way of being prepared for all eventualities; they were cognisant of how the illness could unfold over its full course, rather than just understanding the current situation. These active-seekers tended to express a high need for control across multiple illness domains, suggesting that individual differences in personality also helped to shape information-seeking behaviours. It is important that clinicians are aware of how different patients perceived information and how emotional responses to information might change over the course of the illness.

9.5.2 Respiratory Information

An appreciation of patients' wider attitudes towards information and information-seeking made it easier to explain how patients came to demonstrate such variables levels of understanding about respiratory aspects of the illness and why these levels were often sustained over time. Active-seekers were more likely to have already acquired information about respiratory change and NIV prior to entering the respiratory service and many selective-seekers began to look for information once healthcare professionals initiated the process of referral. This act signalled to these patients that they were entering a new phase of the illness and a number responded by independently looking for information about respiratory change and/or NIV. Overt signals from clinicians that change might be imminent appeared to have been appreciated by these patients.

Most patients did not want to know anything else about respiratory aspects of the illness or NIV at referral. These patients suggested that they did not *need* to know about these aspects of the illness yet and some suggested that they might not ever need that information. Patients who considered referral to be unnecessary or simply "routine" (see Section 6.7.1) often suggested that information about respiratory aspects of the condition – and NIV specifically – was not relevant to them, meaning even motivation to attend in clinics was low. Often knowing that NIV existed was sufficient. These were patients who seemed to rely on subjective appraisals of situations to judge the need for information, which was not always a reliable indicator in the case of respiratory change. A positive appraisal of their own respiratory status seemed to justify a casual approach to the information that clinicians were offering, yet this appraisal already appeared to have been shaped *by*

a lack of information. Patients often dismissed signs of respiratory insufficiency on the grounds that they did not match their expectations of respiratory change in MND and then seemed to use this evaluation to justify not attending to new information about respiratory change and NIV. In some cases the issue of symptom prioritisation was also important, with patients suggesting that they had more important aspects of the illness to attend to and learn about. A preference for dealing with problems rather than changes also seemed influential in demoting the importance of respiratory change and related information. Added to this was the fact that this information was distressing. Respiratory aspects of the illness were particularly upsetting for many patients, which seemed to have encouraged and reinforced avoidance. Some patients seemed to acknowledge the information being presented to them, yet choose not to consciously consider it because it evoked a negative emotional response. This study showed that patients wanted different types of information at different times and for different reasons.

9.5.3 Information & Decision-Making

Knowledge and understanding appeared to be most important during the treatment decision-making process, where it was seen to influence emotional responses to change. Studies exploring information-seeking in cancer populations have reported that patients who have adequate information benefit from increased involvement in decision-making and greater satisfaction with treatment choices (Cawley, Kostic, & Cappello, 1990; Luker et al., 1995), in addition to demonstrating improved coping skills and reduced anxiety (Johnson, Nail, Lauver, King & Keys, 1988; Rees & Bath, 2001). However, this study suggests that satisfaction with decision-making may not necessarily be higher amongst patients who have more information simply because they have more information. One must ask *why* some patients ultimately end up with more information than others. In the present study, patients who were better informed were so because they wanted to be. These patients were more receptive to the information being provided and also invested efforts in acquiring additional information – they were psychologically *ready* to get this information. Information was meeting specific practical and/or psychological needs. Patients who presented stubborn misconceptions had typically chosen this path, electing not to attend to information and even to avoid it. This behaviour was often reflective of avoidance that applied across the wider illness experience. It is recognised that forcibly giving information to patients who do not want it can be just as harmful as allowing patients to proceed with misunderstandings, particularly where this directly challenges a preferred coping strategy (Butow, Maclean, Dunn, Tattersall, & Boyer, 1997).

Information is recognised to be an important component of treatment decision-making in contemporary health services because patients are no longer consigned to merely agreeing or

disagreeing to treatments; it is recognised that they should be given the opportunity to engage in an active dialogue and should be encouraged to voice values and preferences. This is where a distinction between informed consent and informed decision-making arises (Charles, Gafni, & Whelan, 1997; Feste & Anderson, 1995). The NICE guidelines for NIV use in MND stress that patients should take an active role in decisions about ventilation and should be provided with information tailored to their individual needs in order to facilitate this decision-making (NICE, 2010). The idea that information provision should be tailored to meet *individual* needs is crucial, as it was clear in this study that not all patients wanted to be informed or involved at every stage of the referral process.

Clinically, there is a challenge in ensuring that all patients have the information that they want, when they want it, without directly challenging approaches to illness in a way that is unnecessarily distressing for patients. Providing all patients with a verbal overview of respiratory aspects of the illness and NIV during clinic appointments appears problematic because not all patients will attend, even if they might later come to want that information. It is also problematic because even attentive patients might not be able to take in all that is said. Many of these concerns might be addressed by supplementing verbal overviews with written information. This would ensure that all patients had accurate information and could access it in their own time if and when they wanted to. Even avoidant patients who ultimately end up being offered ventilation may wish to have this kind of information available to them outside of the clinic room.

Providing written information would also ensure that patients had appropriate information available to them if at any point in the time between clinic appointments they felt that this information had become more relevant to their circumstance. Most patients were selective information-seekers and one particular difficulty with providing information in clinics is that patients without an incentive to engage at the time of the clinic may fail to attend to information that later does become relevant and wanted. These patients may 'miss' an opportunity to access information in time and thus feel unprepared later, as many patients who were ventilated shortly after referral reported in the present study. If patients develop greater respiratory impairment between clinics, they may wish to have information that they can access themselves rather than having to wait until a next appointment. Clinic appointments are routinely scheduled every 3-4 months, which is a significant time period in the context of MND.

Written information is also useful because patients do not have to read it if they do not want to. This allows them to retain control over information acquisition, which was noted to be an important aspect of information-seeking behaviour. This might be a simple but effective way of improving knowledge and understanding without threatening patients' wider sense of control or challenging their preferred approaches to the illness. Written information-sheets about respiratory

symptoms and NIV are available from the MNDA and could be offered to patients at the point of referral.

One other relevant finding that might prove useful in facilitating effective information provision is the use of patient exemplars. The importance of other patients appeared throughout patient narratives, which might make this a useful vehicle for providing information to even avoidant patients (see Section 6.5.3.2). Peer-influence has been reported to be of significant importance in decision-making in other illness populations, where it is thought to encourage patients to imagine how *they* would cope with treatment options (Morton, Tong, Howard, Snelling, & Webster, 2010). Indeed, in their study of patients with chronic kidney disease, Morton et al. describe this as a "powerful and persuasive method for patients to gain knowledge of their treatment options" (pg. 6). One might suggest that testimonials from patients using NIV might encourage patients to see NIV as a more accessible and manageable intervention; clinically, written materials, audio or DVDs could be made available to patients and carers who would like to access these kinds of resources. Instructional DVDs have been used successfully in other MND services (Pinto et al., 2011) and would also allow patients to see how the ventilator worked. Even patients who are shown NIV or masks in clinics may be overwhelmed by the information being presented to them and so allowing them to re-visit that information in the comfort of their home environment might be useful. Again this also affords control to patients, who can access information in their own time rather than it being presented to them in an all-or-none fashion in time-limited clinic appointments.

Previous studies have shown that peer-support and peer-comparison do have a role in coping in MND, where patients report that practical and physical comparisons allow them to identify coping strategies, derive hope from watching others succeed with the illness, and also to feel better about their own condition by comparing themselves to other patients in a worst physical state (Locock & Brown, 2010). However, it has been suggested that some patients can feel shocked and saddened by seeing other patients with MND, which suggests that having access to patient exemplars may not always be beneficial or useful to patients. These resources should perhaps be *available* to patients, yet access may need to be negotiated through a dialogue with clinicians.

Finally, it is important to consider the role of carers in information provision. This study found that it was often carers rather than patients who wanted and felt they needed most information about respiratory involvement and NIV. This finding has also been reported in studies exploring information-seeking in cancer populations, where carers are often described as being far more proactive in seeking information than patients (James et al., 2007). In the present study this was related to pragmatic considerations, with information being acquired to assist carers in the caring role. In addition, it appeared that carers were often better able to provide information to patients,

suggesting that informing carers might be a route by which clinicians can ensure that patients are also suitably informed. Where carers have access to information, there is always a person to whom patients can turn to safely acquire information as they need it. However, one must ensure that carers are accurately informed, as the present study also noted instances where carers were offering patients inaccurate information or information framed in an inappropriate way.

This study has shown the importance of information and how knowledge and understanding of the illness can shape experiences within the respiratory service. It is essential to allow all patients to have access to the information that they want, when they want it, yet it is also clear that having more information is not always beneficial. It seems important that information provision is tailored to meet differing patient needs. Facilitating *access* to information appears more important than direct provision of information itself.

9.6 Treatment Decision-Making

As outlined in Chapter 2, there has been only one previous study exploring treatment decision-making in respect to NIV use in MND. This study by Young et al. (1994) was a mixed qualitative-quantitative investigation that provided patients with a list of potential reasons for choosing to use mechanical ventilation (both IV and NIV) and asked them to select which reasons they felt were relevant to them. The utility of these findings are assessed in Section 2.8. However, the data reported in this thesis expose a further weakness in that study, as it appeared that Young et al. had presupposed that decision-making would be an explicit process that could be deconstructed into isolated 'reasons' building towards a conclusion. The present study found decision-making to be a far more complex phenomenon. Alongside identifiable 'reasons' were cognitive and emotional factors that subtly shaped the way that patients conceptualised the decision they were being asked to make. Young et al. also focused only on the patient view, neglecting the role of significant others and the dynamic relationships between patients, clinicians and carers, all of which contributed to treatment planning. The present study revealed that patients were 'given' and adopted variables degrees of involvement in the decision-making process.

9.6.1 The Nature of the Decision

A number of patients seemed to approach decision-making already feeling resigned to the treatment. These patients suggested that there was not a genuine 'decision' to be made, because if they needed a ventilator they would simply have to use one, whether they really wanted to or not. For most patients it was not a case of whether NIV should be used, it was just a question of if and when it would come to be needed. There were specific beliefs feeding into this mindset, such as the predicted nature of respiratory decline and the special status that was assigned to respiratory impairment.

Many patients felt that their options were ultimately to either use NIV or risk respiratory distress and it was this that left them feeling that they had "no choice".

Indeed, many patients presented the ventilator as the only method of palliating the signs of respiratory insufficiency and did not mention other methods of treating symptoms. It appeared that many patients had not been educated about the alternatives and this led them to view their choice as NIV or nothing. When viewed in this way, the 'sensible' choice did appear to be ventilation to avoid suffering. There seemed to be an element of fear motivating some patients to accept NIV; these patients appeared anxious about what experiences they would be exposed to if they did *not* accept it. This caused conflict because patients could appreciate that NIV might be helpful at some stage, yet they were still clear that they did not want it.

Although patients were offered a choice about whether or not to accept NIV, it seemed that most felt that it was not a real choice because the option of not using it seemed to involve worse physical consequences. This was a decision that had no positive outcomes; patients' did not want to use a ventilator and yet they did not want to suffer, so they felt they were essentially being asked to choose between two undesirable options. The fact that patients were choosing to use something that they did not want to use seemed to contradict the notion of choice itself. For many patients the ventilator was viewed as something that they would simply have to use if they developed respiratory insufficiency and so it often came to be framed as part of the illness like any other aspect of physical decline. Appreciating patients' negative perceptions of the treatment, their desire to delay its use and the sense of inevitability that was assigned to it, one can better understand how the ventilator came to be a contentious and rather daunting presence.

Where patients felt there was no real choice about NIV they seemed more likely to experience negative emotional responses to the situation. Ventilation was felt to be an issue that, like many aspects of illness progression, was somewhat out of patients' control. If a certain symptom progressed there was nothing patients could do and if they needed NIV they felt they would be similarly unable to do anything about that. Patients suggested that decisions about the ventilator were out of their hands in much the same way that the illness was often felt to be (see Section 6.4). This threat to control was what seemed to create much of the negative feeling surrounding this decision. This was an issue also noted by a number of carers, who felt that it was important that clinicians stressed the fact that patients did have a real choice and remained in control during treatment planning. They felt that this would make patients more amenable to trialling the ventilator and would encourage a more positive response to the transition. Clinicians might actively make decision-making a less stressful experience for patients by reiterating that NIV is not an inevitable component of care, ensuring that all patients are aware of the full range of alternatives available and emphasising the fact that the non-invasive nature of the intervention means that

patients retain choice if they do elect to trial the ventilator. Allowing patients to maintain a sense of control seemed an important means of ensuring better emotional responses to these types of difficult clinical decisions.

9.6.2 Avoidance

Although patients were essentially being asked to make an 'in principal' decision about NIV when they entered the respiratory service, it was clear that many patients did not want to even think about ventilation ahead of time. Most patients were purposefully avoiding the issue of NIV as part of their wider 'one day at a time' approach and also because they felt there were more important issues that warranted their attention. If respiratory symptoms were not prioritised, NIV was not immediately relevant and many patients suggested that affording the ventilator attention before it was needed would cause them only unnecessary distress. A number of patients did not want to commit to the idea that NIV might ever be needed and some appeared hopeful that they might be spared from respiratory impairment itself. Many framed a final decision about NIV as part of the 'unknowable' future that they had already consciously elected to avoid. Patients' avoidance of NIV appeared to be strongly influenced by positive appraisals of their respiratory status at referral and one may again highlight the importance of patients' knowledge and understanding of both respiratory involvement and the role of NIV. Most patients had placed NIV as something that may or may not be necessary but which was definitely something for the future. This should be an important consideration for clinicians who are attempting to engage patients with treatment planning. The idea of 'readiness' to address change and make decisions again appeared important.

9.6.3 Patient Involvement

There is vast corpus of empirical research investigating patient involvement in decision-making, which has become an increasingly important issue in contemporary healthcare policy and provision (Tritter, 2011). In the present study, decision-making was found to be a dynamic process that involved complex interactions between clinicians, patients, and carers. Each individual member of this triad could demonstrate varying degrees of involvement, engagement, and influence, with different patterns of input emerging across cases. Often decisions appeared idiosyncratic and were based on multiple contextual variables, including wider approaches to the illness, appraisal of respiratory status, knowledge of respiratory aspects of the illness, levels of understanding about the role of NIV, and predicted ability to cope with the intervention. All three members of the decision-making triad could have different perspectives on each of these often interconnected issues. For example, in many cases carers expressed significant concern over a patient's respiratory status, whilst the patient did not perceive there to be any problem. The clinicians' insight was again different, as these individuals were using objective test results to appraise the need for intervention.

There were often tensions where one or more members of this triad were not in agreement and ended up being 'led' in a decision that they did not wholeheartedly endorse.

Focusing on the 'reasons' for NIV initiation in some ways distracted from these more important aspects of the decision-making process. Relationship dynamics interacted with clinical variables to define treatment options and motivations. Presenting a list of reasons for using NIV would represent only a small component of this complex process.

9.6.4 The Doctor-Patient Relationship

One of the most important relationships was between the patient and clinician. Over the past thirty years significant effort has been invested in attempts to increase patient participation in healthcare decision-making and the model of care that clinicians aspire to has 'evolved' from a dominating paternalistic model to a more pluralistic model of patient and clinician interaction (see Taylor, 2009). This reflects the rise of the 'collaborative' decision-making process, which Cahill (1996) suggests is characterised by *intellectual* co-operation between clinicians and patients. Shared decision-making should not be confused with the process of obtaining informed consent, which ethically and legally must be established before any procedure or treatment. Shared decision-making goes further than presenting facts and seeking permission; it is a process by which the clinician and patient consider all available information, including treatment options and consequences, and decide together which is the best course of action to ensure the most positive outcomes for the patient in question (Frosch & Kaplan, 1999). Importantly, this is a mutual decision.

There is evidence to suggest that greater patient involvement reduces the risk of misunderstandings and miscommunication (Britten, Stevenson, Barry, Barber & Bradley, 2000), which is likely to relate to the necessary exchange of information that has to occur in order for collaboration to take place. Patients who are involved in shared treatment decision-making have also been shown to demonstrate better treatment compliance (Marinker, 1997) and to report higher levels of satisfaction with processes and procedures (Little et al., 2001). The NICE guidelines for NIV use in MND stress the importance of a collaborative decision-making process, wherein patients should "have the opportunity to make informed decisions about their care and treatment, in partnership with their healthcare professionals" (NICE, 2010; pg. 5). A palliative care context generally increases the importance placed on patient involvement as there is greater scope when planning palliative treatments for patients' wishes, values and aspirations to sit alongside clinical judgements (Gattellari, Voigt, Butow, & Tattersall, 2002); this encourages patients and clinicians to work in partnership to define shared treatment priorities (Kutner, Steiner, Corbett, Jahnigen, & Barton, 1999).

The present study found variable degrees of patient involvement and collaboration; however, it appeared that it was patients themselves who had often engineered a more passive role for themselves by choosing not to engage. It is suggested in the wider literature that not all patients do actually *want* to be involved in treatment-decision making and planning (McKinstry, 2000). A number of specific variables have been associated with the degree of involvement sought. Younger patients are consistently found to have a stronger preference for active involvement (Strull, Lo, & Charles, 1984; Catalan et al., 1994; Stiggelbout & Kiebert, 1997) and higher levels of education are also indicative of a desire to collaborate with physicians (Arora & McHorney, 2000). The type of illness and nature of the problem requiring a decision are also known to be important factors (Arora & McHorney, 2000).

In the present study most patients suggested that they would simply follow whatever advice was given to them by clinicians, even if this conflicted with their own feelings. Indeed, many stressed that they did not want NIV but would accept it if a clinician told them to accept it. In the literature, this attitude has previously been explained in terms of the 'sick role' (Parsons, 1951), which suggests that patients become exempt from normal societal expectations due to their illness and so may look to clinicians to adopt responsibilities on their behalf. Degner and Sloan (1992) tested this notion empirically and reported that while the majority of healthy participants preferred an active role in decision-making, most 'sick' participants (i.e. those with current illness) preferred others to make decisions for them. It is suggested that the 'sick' person is not simply a healthy person with a disease but is qualitatively different in physical, social, emotional and even cognitive terms (Steptoe, Sutcliffe, Allen, & Coombes, 1991). It has been argued that when patients feel physically unwell and/or experience psychological distress caused by illness, they may actually seek a degree of paternalism in decision-making rather than wanting to share in the process (Ingelfinger, 1980; Williamson, 1992). These patients appear to make what Williamson (1992) refers to as "an autonomous choice of dependency", that is, they exercise autonomy by choosing not to be involved in the decision-making process.

It certainly seemed that a number of patients in the present study had made such a choice. Some patients explicitly pointed to the wider illness context to explain that they had "enough to contend with" without having to think about NIV as well. These patients appeared not to want to have to make important treatment decisions, seemingly preferring to be passive recipients of a care plan devised by clinicians. In MND, patients are often coping with significant psychological and physical change and clinicians should perhaps be mindful of how willing and able patients feel they are to contribute to decision-making. This is not to say that patients should be excluded from the process, but that one should allow them to define the level of involvement that they feel comfortable with at any given time.

The idea of specific roles being enacted within the doctor-patient relationship seemed important; patients often took on the 'sick role', yet clinicians were also often assigned the role of incontestable 'expert'. Some patients seemed to offer clinicians control of the decision-making process solely because of this status and position. Where patients focused on the clinician as the expert, they were emphasising that they were *in*expert, highlighting the inequality in knowledge and experience between themselves and clinicians. This created a power differential that seemed to encourage deference in patients. This position was often presented in pragmatic terms (e.g. in respect to clinicians knowing more about the illness), yet it also seemed to reflect an underlying anxiety in many cases. These patients wanted to make sure that the "right" decision was made and seemed nervous about making treatment decision themselves.

It appeared that stepping back from decisions reflected patients' desire to do what they felt *should* be done rather than what they wanted. In addition to having access to all of the necessary information - which often patients themselves did not want - clinicians were also viewed as *benevolent* experts. Clinicians would always make the decision that was in the best interest of the patient and ultimately this is what patients wanted them to do. In some cases, offering decisions to clinicians seemed to be a way that patients could overcome conflict between positive cognitive appraisals of NIV as help and negative emotional responses to the treatment itself. This conflict frequently appeared in patient narratives, with most patients acknowledging that NIV appeared to be a positive source of help but one that they did not want to have to accept. It seemed as if part of the reasoning behind offering clinicians greater control may have been that patients knew that clinicians did not have the same emotional involvement as they did and would make a decision based on objective need alone. This choice would be what was "best" even if it was not what they wanted. It seemed that perhaps some patients did not quite trust themselves to make this decision.

In other cases it seemed that patient involvement was defined by broader role expectations, with patients simply *expecting* doctors to make decisions for them. This view appeared particularly apparent in older patients and may reflect an understanding of the doctor-patient relationship in traditional paternalistic terms developed over the lifetime. If existing schema dictate specific social roles be obeyed within the confines of the consultation room, it may be unlikely that patients will be able to make a transition to a new role without being explicitly guided to it. Some patients did not appear to have been aware that they could or should have been more involved in decision-making. A number of carers also discussed the dynamics of the relationships held with clinicians (see Section 8.3.3.1.4); many explained that the notion that one does not challenge the word of a doctor was seen to suppress expression of patient and carer ideas about treatment. This seemed to be based on expectations about the roles being played and also on factors such as personality. Some patients/carers seemed more confident than others about breaking traditional role boundaries and enforcing their own views and wishes. In the case of carers in particular it seemed that where

clinicians were challenged, it was not based on the understanding of a shared decision-making process but on defying the roles as they were understood for the sake of achieving what was felt to be best for the patient (i.e. in 'rebelling' against the idea that one should not challenge the doctor). A number of carers also spoke of needing to keep doctors 'onside' because they considered them important gatekeepers to help and services, which suggested a further complexity underlying this relationship.

Finally, it also seemed important to consider how patients' attitudes towards treatment decision-making sat alongside their wider approaches to the illness. It has previously been reported that patients with an active approach to the illness are more likely to want a high degree of involvement in decision-making (Arora & McHorney, 2000) and this finding was replicated in the present study. It seemed that patients had already defined in a broader sense how involved with the illness they wanted to be and that this extended to treatment decision-making. Chapter 6 notes that patients preferred to retain a degree of disengagement from the illness until called to re-engage at specific points; it seemed that some patients had been sufficiently motivated to re-engage whilst others had not felt that it was necessary at referral to be increasing their involvement. This appeared, again, to be related to individual differences in patients' subjective appraisal of the situation, their own symptoms and also how much they knew and understood about this aspect of the illness.

9.6.5 Actual Involvement

Most patients contributed to the pre-ventilation theme of 'Doctor Knows Best' (Section 6.10.3); however, when the time came for an actual decision to be made, some patients reported that they had been led by doctors, whilst other patients had taken a far more active role than they had anticipated. One of the key factors that seemed to shape patient involvement was a physical incentive to move towards NIV. A number of patients suggested that it was subjective symptom progression that ultimately led them to seek intervention and engage with the idea of NIV. Patients who reported being more passive in the decision-making process were typically those who had not noted any subjective respiratory change. Importantly, there seemed to be positive and negative consequences to both high and low levels of patient involvement.

For patients who felt that decision-making had been collaborative, clinicians were seen to guide rather than lead, with both parties united in a common purpose. Patients and clinicians shared an incentive to improve a particular problem and it was this that allowed them to work together. Clinicians typically provided information and made the link between symptoms and treatment explicit. For most patients this was a positive experience; they had a specific problem and clinicians explained why NIV was the solution. These patients retained control and were able to move at their own pace towards NIV and this was received positively. However, it is important to note that not all

patients were entirely satisfied with this collaboration when they looked back at the process. Some patients suggested that they had consented to trial NIV because of specific information provided by clinicians, yet retrospectively felt that certain issues were not fully explained or had been presented in a misleading way. This was reflected in the notion that NIV was "sold" to patients, with some expressing disappointment related to expectations built on clinicians' information. How clinicians decide which information they will give to patients and how they will frame treatment may reflect personal values, medical expertise, experience and relationship with the patient (Freedman, 2002). How well the realities of NIV matched the information provided by these collaborators seemed important.

Patients without a physical incentive to trial NIV typically reported that they had been led by clinicians in a more explicit way; these patients did not share the view that something needed to be done. They placed greater weight on their subjective appraisals of respiratory status and so appeared to be in conflict with clinicians working from objective respiratory measures. Interestingly, the restrictions of the traditional doctor-patient relationship seemed to prevent these patients from voicing their concerns. They seemed more confident about expressing their scepticism in interviews. One might suggest that having someone who was independent of the clinical team to discuss decision-making with might have been helpful for some of these patients, particularly as their doubts seemed to negatively influence their early experiences of the treatment and impact on motivation. These patients clearly felt that autonomy and control has been challenged by the move onto NIV. However, it should be stressed that amongst the patients who felt that they had been less involved in the decision-making process, some were accepting of this role and felt content to have let doctors lead, showing again how difficult it was to predict how each individual patient would react. Indeed, one must appreciate that in some cases patients' emotional responses to the decision-making process will have both influenced and been influenced by early experiences of NIV and the degree of 'success' that they experienced on the ventilator. As such, one might suggest that patients with both high and low involvement in decision-making might look back at that time more or less positively depending on how beneficial the treatment ultimately proved to be.

Patients approached treatment decision-making wanting different degrees of involvement and it was difficult to predict how each patient would react to the role that they did actually play. Factors such as how patients' appraised their own respiratory status, how they perceived the need for intervention and what NIV actually meant to patients again seemed to contribute to how involved patients actually became in decision-making. As noted, a number of additional factors may also have shaped how patients retrospectively viewed this process, such as early challenges on NIV and the positive or negative impact of the treatment. It was clear that greater involvement in treatment decision-making did not necessarily equate to greater satisfaction with the process. Some patients who were actively involved were extremely pleased to have been able to work collaboratively with

clinicians, whilst others felt disappointed and misled. Similarly, some patients who felt that they were passive in the move to NIV were content to have taken a back seat, whilst others felt excluded and coerced.

Studies exploring desired and actual involvement in decision making in other illness populations suggest that many patients do not achieve the level of involvement that they want. One study of breast cancer patients found that 64% of patients desired a collaborative role in decision-making, yet only 33% felt that they ultimately had such a role (Keating, Guadanoli, Landrum, Borbas, & Weeks, 2002). Importantly, Keating et al. found that it was patients' whose actual role matched their preferred level of involvement that were more satisfied with treatment decision-making, rather than higher levels of involvement leading to better outcomes. This suggests that it is not involvement per se that increases satisfaction but congruence with a preferred approach to the illness. Clinicians should attempt to define and match the level of involvement that individual patients seek, tailoring care to each patient. Regardless of the degree of involvement that patients want in treatment decision-making, there is robust evidence to suggest that almost all patients want to be given a choice and an opportunity to define their own involvement (Levinson, Kao, Kuby, & Thisted, 2005). This must also be the conclusion drawn in the present study.

9.6.6 Carers & Decision-Making

Many carers were aware of respiratory changes that patients were either unwilling or unable to identify and this seemed to offer these carers an incentive to champion NIV from an early stage. Carers appeared influential in the decision-making process; however, it was again notable that they, like patients, designated variable levels of engagement for themselves. Some carers wanted to step back and let patients or clinicians lead, whilst others sought to take control of the process. A number of factors seemed to influence the role that carers played, including how much they knew about respiratory aspects of MND, their appraisal of patients' respiratory status, any emotional response to the changes they had noted, and the type and nature of the dyadic relationship. Younger spousal dyads in particular appeared more likely to approach the decision together and engage in discussions to establish a dyadic consensus. The patient was also an important determinant of carer involvement; some carers seemed to seek greater involvement to compensate for an avoidant attitude in the patient, whilst other carers stepped back to avoid created tension in the dyad. Carers' own approaches to the illness were also important; some carers were themselves more avoidant of the illness and sought to protect themselves from distress by not getting too involved. The role that carers played in decision-making both shaped and was shaped by the roles that patients played.

Carers of patients with advanced cancer have also been observed to take on the role of patient advocate, demanding quality care on behalf of patients (Zhang & Siminoff, 2003) and this was

something that clearly emerged from many carers in the present study as they discussed interactions with clinicians. Carers often expressed a degree of responsibility for patients, which was particularly pronounced where patients were unable to communicate verbally. Bulbar symptoms increased dependency on some carers in a specific way, relating to communicating and negotiating care needs with healthcare professionals. This naturally increased carer involvement in decision-making in these cases. It is also important to note that where some patients adopted the 'sick role' carers were often expected to take on the responsibilities that patients were unwilling or unable to address. It was clearly important to consider and include carers where appropriate, as it was apparent that these individuals were playing an important role in decision-making. Carers often had a better understanding of patients' thoughts and feelings about NIV than clinicians and could appreciate patients' responses in the wider context of the illness.

9.7 Defining "Need"

Most patients repeatedly affirmed that they did not want NIV but would accept it if it was needed. Appraisals of "need" seemed to shape patients' responses to the treatment both before and after initiation. In many ways, this makes the most important question for clinicians one of how patients defined need. At referral, patients appeared to have been assessing a number of different variables when determining whether NIV was needed or not: their subjective appraisal of respiratory symptoms (i.e. whether there were any symptoms to be corrected by treatment and/or how problematic these were deemed to be), how well they felt they could cope with these symptoms without NIV, the positive role ventilation might play in helping them to avoid respiratory distress, the negative consequences of not accepting help, and also the negative impact that NIV might have on wellbeing (e.g. in terms of having to wear the mask each night).

Patients' seemed to perform a 'cost-reward' analysis at various points in time, both before NIV was initiated and after. Prior to initiation the primary 'cost' that was being evaluated appeared to be the severity of symptoms (i.e. subjective impact of respiratory insufficiency or the potential threat to respiratory function outlined in test results), whilst the main 'reward' appeared to be the simple fact that patients were not using a ventilator (i.e. were avoiding the negative experience that they associated with NIV and the negative connotations that came with being ventilated). Crucially, patients seemed to have different thresholds in respect to the costs that they would endure and the rewards that they needed; these were individual differences that appeared difficult to predict and were likely to be shaped by a number of complex variables (e.g. personality, life experience, coping resources, and wider illness context). It seemed that patients placed different weight on different variables and were often relying on subjective interpretations; this meant that there was not a single definition of need that could be isolated.

There has previously been some debate in the literature as to how much weight should be placed on patients' subjective appraisals of need, as compared to objective measures, when making decisions about NIV initiation (see Section 2.5.2.2). Evidence from the present study suggests that patients' subjective appraisals of change did not always match the objective evidence, which some might suggest shows that patients lack the insight to know when intervention is necessary. The present study found that this was an issue confused by misconceptions about the nature of respiratory change and the purpose of NIV, rather than just being a problem of patient awareness of physical change. It was also influenced by patients' illness priorities and wider treatment aspirations and was not based on physical symptoms alone. The clinical criteria may have been purely physical, yet patients' seemed to incorporate wider considerations.

Clinically, the fact that patients' subjective appraisals did not concord with objective indices of change might be taken as evidence that treatment initiation *should* be guided by objective measures, with subjective patient report used to supplement this data in a peripheral way. The present study showed this to be a complicated issue. It was clear in post-ventilation analysis that initiating NIV in response to objective test results alone (i.e. where patients were asymptomatic but where oximetry readings suggested impaired respiratory function) could itself create barriers to compliance, with patients who were unconvinced about the necessity of NIV appearing to lack motivation to succeed. This indicates that some degree of subjective impairment may be necessary to engage certain patients. It should be noted, however, that a number of patients in the cross-sectional study reported that they had not realised how much they needed NIV until they received the physical benefits of use. It is possible that some patients will only ever come to appreciate the need for NIV once they experience the physical changes that it can exert. NIV is a palliative treatment; in cases where patients are asymptomatic and the prospect or experience of using NIV causes them distress, it may become an ethical issue of whether NIV is actually the right option (i.e. whether it is contributing something positive or actually having a deleterious impact on QoL). It seemed that the right compromise might be to encourage all patients to trial NIV when objective indices indicate that they could benefit from it, allowing patients to perform their own cost-reward analysis to determine if the treatment is indeed right for them. Yet trials of NIV should be genuine 'test' periods, rather than being experiences from which it is assumed all patients will emerge as routine NIV users. It seemed important that patients were invested in the treatment to some extent. The issue of motivation is discussed further in Section 9.8.3.

9.8 The Experience of Non-Invasive Ventilation

Clinical concerns typically focus on practical and functional elements of ventilator use, yet it was clear that there were important psychological and emotional dimensions to the treatment that defined both experience and outcome. Even the process of entering a hospital environment to trial

NIV was seen to be traumatic for some patients. Greater attention should be afforded to these aspects of experience, possibly extending home-trial schemes where appropriate. Even routine clinical processes and procedures could be extremely challenging for MND patients. Mindfulness in this respect was needed over the entire course of the treatment, where it seemed that patients' *experiences* of NIV may have been overlooked in favour of addressing practical concerns and charting physical progress.

9.8.1 Masks

Masks emerged as significant determinants of response to NIV both before and after initiation, suggesting that patients did not have to be using a mask to express a negative psychological response to it. The mask was the only aspect of the actual treatment that patients mentioned prior to initiation; all other responses were directed at more abstract notions of what the treatment represented in positive or negative terms (i.e. help or defeat). Many patients presented their aversion to the interface as being instinctive, explaining that it was not necessarily having to wear a mask per se that was perceived negatively but the prospect of having anything on or around the face. Other patients feared that the mask would impact on identity, in respect to how they perceived themselves and how others would view them. These psychological responses are important, yet have not been recognised elsewhere in the literature. It was based on patients' predictions of what it would be like to wear a mask that many wanted to distance themselves from the treatment. A significant component of the desire to 'put off' NIV seemed to be made up of reluctance to have to wear a mask.

When patients came to trial the ventilator, the mask remained the most salient obstacle to tolerance and compliance. Yet, of clinical importance, patients in both the cross-sectional study and the longitudinal phase of the research noted that *changing* the mask had a transformative impact on early experiences. Finding the 'right' mask increased comfort levels to improve the treatment environment *and* allowed patients to feel that they retained choice and control within the broader situation. Being part of a process of defining the treatment experience in this way made patients feel more involved and invested in the treatment and also showed them that there was potential for positive change to occur. A change in mask that resulted in better comfort or function often also increased self-efficacy and transformed attitudes towards the ventilator. It seems important that clinicians should involve patients in a process of open discussion about masks and should remain engaged in consultation throughout the early stages of use.

Existing research has focused on the physical and functional properties of masks, without acknowledging the wider psychological impact of use, yet the present study found that it was often these aspects of response that were most important to patients, particularly during the process of acclimatisation. In terms of the patient experience, the mask created an "alien" environment that was

often distressing. Patients typically needed to acclimatise to the *presence* of the mask, spending a variable period becoming desensitised to the awkward feelings that it created. Patients tended to fare better with smaller masks that were less obtrusive and 'obvious' in the line of vision, yet ultimately responses to the mask "being there" needed to be overcome simply through experience. Preparing patients for this period by forewarning them seemed to be helpful, allowing them to frame these early challenges as part of the normal process, rather than as reflections of failure or inability to tolerate.

There were important psychological dimensions to mask use even where patients were successfully established on the ventilator. In some cases masks impacted on how patients felt about themselves and who they perceived themselves to be when worn; this was as some patients had feared prior to initiation. One can appreciate that wearing a metaphorical 'mask' indicates a concealment of true identity, projecting a specific message about the wearer to the world; it seemed that some patients felt that their own identities might be concealed behind the mask and were distressed by the negative messages that it might send out to others. It is important to note the capacity for masks to evoke psychological reactions not reflective of practical or functional problems but of the presence of the mask per se. The negative impact of masks on self-esteem did not typically affect treatment compliance in a direct way, yet it did colour the subjective treatment experience, which contributed to a negative view of the treatment and increased the chances of non-compliance. Given that treatment is likely to be advised throughout the remaining duration of the illness, it is important that any potential assaults on self-esteem are identified and corrected in the early stages of use. It might be suggested that clinicians should assess *attitudes* towards masks as a separate line of inquiry from practical aspects of treatment, so as to identify patients who may benefit from additional psychological support. Interventions could facilitate adjustment to treatment by normalising masks and cognitively reframing them as positive illness attributes. Some patients were able to undertake this process themselves, framing the mask as a necessary means of accessing positive physical change, yet it was notable that these patients were typically those with strong physical incentives to succeed on NIV from the start (i.e. those with subjective symptoms to address) and those who had already noted positive physical effects of use.

The aesthetic qualities of masks were also important to patients; with the thick straps seen to be unnecessarily intrusive, uncomfortable and visually unappealing, particularly in the cross-sectional study. Calls to improve the subtlety and comfort of masks indicated a desire to minimise their presence. Patients in the longitudinal phase of the study seemed to prefer a smaller, less 'obvious' mask and patients in the cross-sectional study sought masks that had a natural rather than medical aesthetic (defined by the thick strapping and blue plastic components). Masks lacked cosmetic appeal, yet practical steps could easily reduce distress by reconsidering the size, shape and colour of masks and straps (e.g. a more neutral palette, clear plastic, and thinner, softer material for straps to

limit their tactile presence). These steps could have a significant psychological impact by reducing the intrusiveness of masks.

Mask function was of greater concern to patients in the longitudinal phase of the study than in the cross-sectional study, with patients noting that masks could be uncomfortable and prone to movement. It is important to note that in most cases the practical and functional problems outlined by patients did not deter them from using the treatment; however, they did significantly impact on patients' subjective experience of ventilation. One particular concern for patients was control over the treatment environment. Inability to independently remove the mask threatened autonomy and increased the likelihood of anxiety. This was a problem that was related to both the functional challenge of the straps and patients' own waning dexterity. Being unable to release the straps led some patients in the cross-sectional study to feel 'trapped' within the mask; reflected in language that depicted patients as prisoners confined by the treatment. In the longitudinal study threats to control increased frustrations and increased the risk of noncompliance. Patients in the cross-sectional study also reported that anxiety was heightened where masks prohibited communication; these patients appeared comforted by the thought that they could at least raise an alarm if an emergency arose, even if they could not themselves remove the mask.

These factors are particularly important given the progressively degenerative nature of the illness, differentiating NIV use in MND from its use in other patient populations and indicating an evolution in attitudes over time. It could be suggested that both communication failure and loss of strength in the hands should be treated as clinical signposts that mark potential changes in the treatment experience and that healthcare professionals should look for these markers as potential catalysts for change in attitude towards ventilation. There are also practical steps that may be taken to help patients retain a sense of mastery over the treatment environment, such as the introduction of quick release mechanisms to masks. It may also be reassuring for ventilated patients to have an alarm system that enables them to attract assistance should an emergency arise. Currently, machines are fitted with alarms triggered when the ventilator itself detects a functional problem (e.g. an air leak); however, an external system enabling patients to trigger alarms where they appraise a need for help or support may provide further reassurance.

In respect to masks, it appears that clinical attention needs to be afforded to more than simply comfort and efficiency. Clinicians should be mindful of the significant psychological and emotional aspects of mask use. They should also appreciate the way that practical and functional challenges may shape the subjective treatment experience and influence patients' attitudes towards the intervention.

9.8.2 Outcomes & Responses

The vast majority of research addressing NIV use in MND populations has focused on physiological outcomes, defining treatment success by the degree to which it meets clinical objectives. This thesis aimed to explore what meaning patients attached to these outcomes and how patients responded to them; this was felt to be important given that QoL in MND is reportedly unrelated to physical aspects of the illness, being defined instead by subjective elements of experience (see Section 1.8.5). It was clear from the post-ventilation analysis that many patients benefitted physically from the treatment and felt that ventilation contributed something positive to their care. The qualitative results mirror the quantitative findings in the range of positive physical effects noted, including relief from dyspnoea, improved sleep and reduced fatigue/somnolence. There were also wider psychosocial changes, including improved mood and greater social interaction. These results emerged both in the longitudinal and cross-sectional data. There were individual cases that seemed to exemplify 'success' on NIV; for example, one patient felt that the physical transformation that he had experienced radically changed his perception of the whole illness, allowing him to rediscover much of the life he thought he had lost. The positive physical effects of NIV often validated patients' decision to trial the ventilator and motivated them to continue using the intervention.

However, it was clear that these positive physical changes did not tell the whole story. Even patients who had noted positive changes were acutely aware of the limits of NIV. These patients were also conscious of the fact that the illness continued to progress in other domains. Existing research has focused on isolated clinical outcomes whilst ignoring the wider clinical context, failing to recognise that improvement in one symptom (e.g. somnolence) might be overshadowed by a lack of improvement or decline in another symptom (e.g. limb-weakness or fatigue). Positive emotional responses to post-NIV changes were often embedded within negative responses to other aspects of illness progression, so that satisfaction with the effects of NIV sat alongside frustrations directed towards other areas of continued decline and changes were seen to improve aspects of experience rather than to radically transform lives. It was clear that meeting a single clinical objective did not always result in treatment being deemed a success by patients.

There were also patients who felt disappointed in the changes that they had noted, this was often related to expectation. Quantitative studies mapping only physical improvement after ventilation may have reported the presence of a physical improvement in these cases, yet would not have detected this type of emotional response to change. In an illness where there are so few opportunities for improvement, it seemed natural that patients would invest hope in NIV. One might suggest that managing expectations should form a vital component of clinical care in the period prior to initiation, as it was clear that unrealistic expectations put patients at greater risk of a negative emotional response to the treatment. Alongside wider expectations, a number of patients

had specific treatment aspirations that were not met, which again resulted in disappointment. It seemed likely that a patient desperate for respite from a specific symptom (e.g. debilitating fatigue) who did not see the expected results in that domain would not receive improvements in other symptoms as positively as a patient who was not so emotionally invested in making a specific change. There are cognitive and emotional components to the hopes attached to NIV and these kinds of contextual factors were found to be important in the way that patients responded to change. In these cases, significant clinical improvements were being registered, yet patients' responses were not always correspondingly positive. There is general consensus that rates of depression in MND are low but that periods of low mood can be triggered by significant clinical changes (Section 1.8.1.1), it may be that disappointment caused by unfulfilled expectations with NIV is a trigger that clinicians should be aware of.

It is important to acknowledge that some patients felt that the changes they had noted were insufficient to justify the use of the machine and other patients had not noticed any change at all. Indeed, a number of patients felt that the ventilator could actually be counterproductive at times, making the situation appear worse rather than better. The fact that this range of experiences was identified seemed to justify a qualitative approach; one can see how aggregating sample data in quantitative analysis would have masked many of these differences, with significant positive and negative changes potentially cancelling each other out. Existing quantitative studies have also failed to show the apparent mismatch between outcomes and response, defining clinical changes but not how patients actually receive them. This is an important consideration given that NIV is a palliative treatment, aiming to improve illness experiences rather than to cure or to solely meet clinical targets.

Looking across individual cases, it seemed that patients who demonstrated the greatest satisfaction with NIV were often those who had been experiencing the most significant respiratory symptoms prior to initiation. Many of these patients moved onto NIV because they felt they had little to lose. Patients with the greatest scope for change had clearly defined problems and were able to observe tangible physical improvements. Treatment had the greatest impact because it led to the greatest change. In patients who were asymptomatic prior to initiation, and who typically moved onto NIV in response to overnight oximetry readings alone, there was often simply less potential for noticeable change to occur. This naturally impacted on patients' perceptions of the treatment and what each patient felt they were taking from the experience of ventilation. Generally, patients who noted significant subjective change had evidence of positive effects and so had reward for the efforts invested, yet patients whose physical improvements were only noted by physicians in objective tests did not have the same evidence and often appeared unconvinced by the necessity and value of the treatment.

Some patients were able to rationalise that improvements in oximetry readings represented sufficient return from NIV, yet this typically depended on knowledge and understanding of what improved overnight oximetry meant for patients in the context of MND and also the wider goals that patients held for the illness. The existing illness context (i.e. what other symptoms patients were dealing with) also played a part in this appraisal. Most patients were not looking ahead to how the illness would develop and were not looking to extend life; they were concerned with the 'here and now' and what impact NIV was having on their subjective illness experiences at that particular time. For some patients NIV did not appear to be making the kind of difference to their lives that would allow them to perceive the treatment as 'worth' the efforts invested and the hardships endured were not suitably compensated for by improvements that they could not subjectively perceive. It was clear that there was not a direct relationship between treatment outcomes and patients' responses to change and there appeared to be no reliable way to predict how patients would appraise the treatment based on physiological outcomes alone.

9.8.3 Tolerance and Compliance

This study found that the definitions of tolerance and compliance traditionally used in a research context (see Section 2.7) did not usefully contribute to an understanding of patients' experiences or map effectively onto patient ratings of success or failure. For example, the number of hours that patients spent on the ventilator did not correspond to how positively the treatment was received; some patients were routinely using the ventilator all night, yet still reported significant challenges. It was also clear that tolerance did not necessarily lead to compliance, as a number of patients who were able to use the ventilator often elected not to.

In the cross-sectional study all patients were using NIV in the same way (i.e. all night, every night); however, in the longitudinal study, patients displayed variable patterns of use. This is likely to reflect differences in the time taken to acclimatise to the treatment and the fact that some patients in the longitudinal study never attained routine use before death whilst patients in the cross-sectional study were those who had – by definition – already established good compliance. The variability in patterns of use has important research implications. Most quantitative studies group patients in analysis at an arbitrary point post-initiation (e.g. 3-4 months after starting on the ventilator), yet the results of the present study suggest that this kind of aggregation might again mask significant individual differences in response, this time due to patterns of use.

In respect to the factors that determined both tolerance and compliance, there were no particular experiences or events that applied across cases. It was not possible to say that any one factor was ever singularly 'responsible' for defining success or failure, as different patients often interpreted the same challenges in different ways. To restate, multiple variables, such as personality, preferred

coping strategies, motivation, and type/severity of physical disability in other domains, all shaped patients' responses to hardships encountered on the ventilator. This is important as most previous studies have attempted to isolate individual variables that explain tolerance and compliance (see Section 2.7). In this study, each patient presented a varied combination of variables and contexts that influenced attitudes towards treatment.

Tolerance was most often defined by *practical* aspects of the treatment, such as mask discomfort or feeling overwhelmed by the force of air being delivered, which were more generally observed in patients with significant bulbar impairment. This has previously been highlighted as a risk to tolerance in the literature (e.g. Aboussouan et al., 2001), yet the present study was able to identify additional psychological aspects to these problems. For example, one patient reported experiencing anxiety in anticipation of the volume of air being delivered by the ventilator, such that her heart-rate increased and she felt overwhelmed by the experience. One could see that her emotional response to the treatment caused a physical response that exacerbated asynchrony with the ventilator. It is noted in Table 2.2, Section 2.2.2, that asynchrony between patient and ventilator can actually cause dyspnoea; this study identified how emotional responses to the treatment could also feed into this process. Practical difficulties could also significantly shape self-efficacy in other ways, such as where mask discomfort led patients to question whether they could physically manage the experience. These trials often led patients to believe that they *could* not use the ventilator. In this way, practical difficulties and their psychological consequences could interact to create a self-fulfilling prophecy.

Compliance was a far more complex phenomenon. Again there were a multitude of factors put forth to explain why patients did not use the ventilator as prescribed, yet these were both practical and psychological in nature. Practical reasons for non-compliance typically related to problems caused by the mask, including discomfort, abrasions, and air-leaks. Yet the presence of these challenges did not necessarily challenge compliance in all cases; some patients simply accepted them as part of the treatment. This was also true where concerned tolerance, as many of the practical issues presented as barriers to tolerance were often challenges that other patients had faced but overcome through experience or perseverance. It seemed that motivation to succeed was a crucial factor in determining tolerance and compliance where there were practical challenges associated with ventilation. Although difficult treatment environments themselves depleted motivation, patients who seemed unable to tolerate were - exclusively - those who repeatedly suggested they did not the need the treatment. It also seemed that patients who were motivated to use the ventilator were more likely to persevere in the face of early challenges and to comply despite hardships.

Patients were typically motivated where they had a physical incentive to use NIV. There seemed to be a cycle of success that was initiated by the presence of subjective respiratory symptoms: these symptoms provided a reason for patients to support the initiation of NIV and also seemed to

motivate patients to want to succeed with the treatment. This in turn made patients more likely to persevere in the face of early hardships. In persevering, these patients appeared more likely to see physical improvement and this positive change was seen to reinforce motivation to comply over time. It also seemed easier to attain positive effects for these patients as there were clear problems to address. When patients were appraising the significance of practical problems associated with NIV use, it seemed that they were performing the same kind of cost-reward analysis that patients undertook during decision-making. Compliant patients ultimately perceived the rewards of using NIV (e.g. positive physical changes) as being greater than the costs of use (e.g. mask discomfort), one might also suggest that the cost of *not* using the treatment might be viewed as higher for these patients.

In contrast, one can also look at the interpretation of practical difficulties from the perspective of patients who did not feel incentivised and did not see physical rewards. These patients tended to be those who had not noted any subjective respiratory change prior to initiation and who reported being passive during the decision-making process. They had not determined their own reasons for using the ventilator and in some cases appeared reluctant and resistant from the start. This is not to say that all of these patients lacked any form of motivation, it just tended to appear as more abstract and less personally relevant (e.g. being motivated to help doctors or placate carers' concerns). Where patients had less motivation, or fewer personally relevant reasons for initiating treatment, it seemed that they were less forgiving of early challenges. This typically meant that these patients failed to accrue the necessary time on the ventilator to see physical improvements. This was a situation exacerbated by the fact that the absence of subjective respiratory symptoms already meant that there was less scope for significant – or even noticeable – physical improvement after ventilation. These patients, in performing a 'cost-reward' analysis of the situation, appeared more likely to suggest that the results of NIV were not 'worth' the effort that they were investing and this seemed to encourage non-compliance. There was also seen to be less risk involved in noncompliance for these patients, as often they did not perceive there to have been any problem for NIV to address in the first place.

Other contextual factors were also important in defining tolerance and compliance; for example, the level of physical disability that patients experienced in other domains. It was noted that some patients were already experiencing great discomfort, particularly at night, and in these situations any irritation that NIV additionally contributed was seen to be amplified. In these cases, treatment was often assessed in terms of the total discomfort experienced rather than just the contribution made by the ventilator. Although NIV is used in many other chronic conditions, MND represents an almost unique treatment environment because patients were often attempting to acclimatise to NIV whilst simultaneously coping with other significant challenges. These challenges also differ significantly between patients, making it difficult to predict who might struggle and why.

Patients who failed to comply seemed to focus more on the immediate situation than they did on the future and the potential benefits that they might have accrued by using the ventilator. If the ventilator did not lead to subjective improvement, these patients were left querying its necessity. This again emphasised the individual differences that existed in illness priorities and treatment aspirations, as other patients who had not noted subjective change managed to reconstruct new treatment goals and to sustain motivation by suggesting that the ventilator was probably helping them in ways they were not aware of. The fact that there were patients who found motivation in improved overnight oximetry readings alone and did not need to experience subjective physical change indicated that there was an element of subjective interpretation in these responses. These responses were also likely to have been shaped by natural personality variables (e.g. optimism), as it was clear that some patients were able to respond more positively to disappointment that others across multiple illness domains. This type of individual perspective is more readily captured within qualitative analysis, particularly within an approach that seeks an idiographic perspective, yet it is still a complex quality to capture in thematic analysis.

In the general nursing literature there are two types of noncompliance identified: 'intentional', where patients make a conscious choice to use the treatment in an alternative way or not to use it at all; and 'unintentional', which results from inadequate understanding of the condition or treatment (Hussey & Gilliland, 1989). In the present study, both forms of noncompliance seemed to be evident. A number of patients suggested that they would have used the ventilator more had they been *able* to. It seemed striking, given the complex psychological responses that often shaped compliance, that some patients were actually being held back by a simple lack of practical skills (e.g. being able to attach the mask interface properly). This type of noncompliance is easily rectifiable by the provision of information and training, yet might have been avoided completely had sufficient guidance been offered prior to discharge from hospital. In this situation, as outlined in Section 9.5.1, healthcare professionals overseeing inpatient trials needed to be aware that providing information to patients verbally was not a guarantee that patients had received and retained that information. This appeared as another instance where provision of written information might have benefitted patients. These patients reported that they had tried to use NIV as directed but had abandoned efforts when this was too challenging, suggesting again that patients could only invest so much effort before the treatment no longer seemed 'worth' it. It might be argued that more determined patients would have made a greater effort to 'figure out' the straps, which were challenging but not impossible.

The present study found tolerance and compliance to be complex issues that were best understood in the context of the individual patient. There were no consistent or predictable factors that shaped outcomes, merely a collection of potentially relevant variables for clinicians to consider.

9.8.4 Patient Experience

This thesis aimed to go beyond objective clinical outcomes and explore the *experience* of NIV use in MND. It was apparent that positive clinical/physiological outcomes did not always go hand in hand with positive experiences on the ventilator and compliance did not mean that the treatment was problem-free. Indeed, some patients were routinely using the ventilator all night, yet still reported significant challenges. Difficulties that patients were often prepared to endure still coloured the overall treatment experience, which should be of equal importance to clinicians.

Many patients reported notable frustrations caused by ventilator malfunction. This included masks that moved during the nights, straps that seemed to require constant adjustment, and air-leaks that created noise and physical discomfort. Even patients who reported positive clinical outcomes noted ongoing frustrations with the ventilator, which created conflicting emotional responses. In these cases, the clinical outcomes of NIV were positive, yet the experience itself was generally negative. Patients who were experiencing positive physical effects of treatment typically tolerated negative aspects of use because they felt they were sufficiently rewarded for the efforts that they were investing. In this way, patients endured the negative *experience* of NIV to access the positive outcomes. Of clinical importance, one patient appeared so highly motivated to achieve physical benefit from NIV that he was endured significant discomfort and painful skin abrasions caused by the mask. Clinicians should perhaps be wary of just how much highly motivated patients will tolerate to keep using the treatment; intervention to change the mask may have enabled this patient to avoid the physical injuries that he sustained. There should be close monitoring and open channels of communication between clinicians and patients, particularly during the early stages of use, as some patients clearly needed clinicians to intervene to set reasonable clinical limits to the experiences that patients have to endure to achieve routine use of the ventilator and associated respiratory benefits. Where this patient had to stop using the ventilator to let mask-related injuries heal, the option of NIV was taken away, which threatened his sense of control.

Control emerged as an important factor in determining the quality of patients' treatment experiences. Many patients had expressed a desire to stay in control as the illness progressed and being able to retain mastery over illness experiences was an important determinant of wellbeing in many different domains. Not being able to exert control over the treatment seemed to have a negative impact on patients' emotional responses to it and also created anxieties that negatively coloured the experience of ventilation. One particular source of distress appeared where patients could not move the mask themselves to make adjustments and also where the mask itself could not be removed independently, as discussed in Section 9.8.1. Patients in the cross-sectional study also spoke of anxiety developing as the illness progressed and it became harder to physically manipulate the interface. In a progressively degenerative illness like MND, this is a significant consideration; it

suggests that even where the clinical outcomes of NIV remain constant, the experience of ventilation might change as the illness progresses. A number of patients reported anxieties about the prospect of losing control over the treatment environment over time, which showed that this was an issue that patients were often aware of and which seemed to be making a contribution to experiences even before any inhibitory physical progression started.

As has been stressed throughout this chapter, the link between subjective experience and tolerance/compliance was not direct; it appeared that factors such as motivation, need, and outcome were able to modify this relationship. Where motivation to succeed was high and/or results positive, patients were more likely to tolerate a poor treatment environment, yet where motivation was low and/or results poor, patients appeared less tolerant of negative experiences. This again was part of the cost-reward analysis that seemed to shape patients attitudes towards the treatment and its use.

9.9 Final Conclusions

It was intended that this research would qualitatively explore the lived experience of respiratory impairment in MND, establishing how patients make sense of respiratory change in the wider illness context. This was part of a wider goal of exploring the experience of ventilator use in MND, including how patients perceive the intervention and subsequently approach decisions about its implementation. This extended to explore the patient experience of actually being ventilated, focusing on subjective responses to experience, process and outcome rather than the objective clinical consequences of its use. These aims have generated a body of qualitative evidence to inform the clinicians providing respiratory care to this population and thus to maximise the likelihood that each MND patient considered for ventilation will receive the best possible experience within a given respiratory service. This section reaffirms the key messages from the thesis and brings together the practical clinical advice that has been drawn from it.

9.9.1 Key Themes and Ideas

In respect to the lived experience of *respiratory* change in MND, the most important conclusions can be summarised in the following points of note:

- **The objective physiological profile of patients is not always indicative of their subjective experience or perceptions of respiratory decline.**

Clinicians should not assume that the objective physical status of patients maps directly onto their subjective experience of respiratory decline; respiratory function tests and measures of respiratory

muscle strength offer invaluable insight into the physiological picture but do not necessarily correlate with patient perceptions of change, even in cases where the degree of respiratory impairment is judged to be clinically significant. Indeed, insight into respiratory aspects of the illness may still be minimal in patients presenting with advanced symptoms.

Alongside the physical confounds that appear to mask patient insight into respiratory change in MND (i.e. disability that limits mobility and reduces the opportunity for exertion, protecting the respiratory system by no longer overtly 'testing' its function), there may also be a number of important psychological mechanisms at work, shaping the way that patients experience and report respiratory change. Unique patterns of physical impairment may encourage rather idiosyncratic patterns of symptom prioritisation, with patients having to differentially assign finite physical and psychological resources to address successive and cumulative losses across multiple areas of physical function. Symptoms such as pain, sialorrhea, dysarthria or limb-weakness may be considered of greater importance to some patients, relegating respiratory change in respect to the attention it demands. Patients may also be able to consciously or unconsciously adapt to respiratory change to limit its impact, changing routines and behaviours to allow them to circumvent any need to seek external help. Existing illness schemas may contribute additional confusion, encouraging patients to attribute respiratory symptoms to non-respiratory causes (e.g. general fatigue) and reducing their awareness of the impact of MND on the respiratory system. Potential misunderstandings may be corrected with education in those who are willing and able to accept illness-related information, yet it should also be considered that some patients may not want to acknowledge or attend to respiratory change. These patients may not be receptive to learning about and/or accepting the presence of respiratory impairment or the clinical attention being afforded to it.

Clinicians should aim to determine the *subjective* experience of respiratory decline and the *perceived* impact that this has on life for each patient. Failure to do so may result in conflicts of intention that prevent patients from engaging effectively with clinicians/services and may potentially generate psychological resistance and/or distress in patients.

- **Respiratory decline may take on a wider meaning in the context of MND, representing illness progression and being strongly associated with the end of life.**

It is important that clinicians appreciate how patients might come to conceptualise respiratory change in the wider context of MND. The patient experience of respiratory change extends beyond the physical realm, including emotional appraisals of events and psychological interpretations of potential change. Patients appear to incorporate ideas about what decline in this area of the body signifies and symbolises in respect to an overall illness timeline. Lay perceptions implicitly connect

breathing to life and death; the added context of a progressive terminal illness may afford ominous gravitas to this conceptual bond. It is essential that clinicians are sensitive to the wider implications of acknowledging respiratory decline from the patient perspective. In many cases procedures and outcomes that are clinically commonplace may be perceived as somewhat sinister in an illness already known to be progressing towards death. Although referral into a respiratory service is a routine step for clinicians, it may carry significant psychological weight for patients; clinicians should attempt to understand what this event means for patients, as this is likely to be a time of active sense-making for many. Patients may feel the need to protect positive ideas about their physical status in order to maintain psychological wellbeing or, similarly, to minimise negative ideas in the same way to avoid distress. This again has implications for service engagement.

- **Incongruence between clinician and patient perceptions of respiratory status may lead to conflicting interpretations of 'need' both for attention and intervention.**

At referral, respiratory change that may be considered a clinical 'problem' may not necessarily be considered a 'problem' by patients. Where patient priorities do not mirror those of involved healthcare professionals, it is possible that conflicting clinical agendas and goals will develop and create tension. For patients, 'problems' might be more readily defined as factors causing tangible physical discomfort or functional impairment at a specific time. As noted above, this can be understood as a response to the unique nature of the illness and may be interpreted as an attempt to allocate limited coping resources to the changes that are having an immediate impact on the 'here and now', rather than potentially wasting energy and efforts addressing – or even worrying about – all of the physical changes that are or may be occurring at any point. It is recognised that MND assaults physical function across many different domains and that patients are likely to establish their own ideas about what is most important to them at any given time. This idea of living *with* change and impairment rather than addressing it immediately does not fit with the clinical ideal of circumventing distress by maximising physical function, yet this may be the patient inclination. Patients may appreciate the significance of respiratory involvement and the importance of monitoring respiratory function, whilst still not identifying these things as being personally relevant to them at that time. It is perhaps unnecessary for patients to prioritise respiratory change if an attuned respiratory clinician is able to monitor and guide patient input and insight in an appropriate way; however, clinicians must acknowledge the notion of 'readiness' to address change and should recognise that patients may not instinctively share clinical priorities.

There were complicated emotional and psychological components to the experience of *ventilation* that must also be identified and acknowledged clinically. In respect to the experience of ventilation in MND, the most important conclusions can be summarised in the following points of note:

- **Ventilation may take on a wider symbolic meaning in the context of MND, coming to be viewed as a marker of illness progression and even a signal of impending death.**

Patients' perceptions of respiratory decline and its consequences appear to colour and shape their early reactions to ventilation. Although clinicians may present ventilation as a positive and pragmatic response to respiratory insufficiency, patients may still interpret it as a sign of illness progression that has extremely negative connotations, potentially bringing a sense of defeat and foreboding about the nature of imminent decline. Although non-invasive ventilation is widely used to treat a range of conditions, it seems open to different interpretation in the distinctive context of a progressively degenerative illness; it may be viewed as both an intrinsic part of the physical breakdown of the body and as an ending in itself. MND patients are in the unique position of knowing that their condition will only worsen and this makes the real possibility of increasing physical dependence a daunting prospect. The perceived permanence of ventilation, and the idea that once it has been initiated it will become a fixed component of care, is likely to motivate the desire to postpone the treatment. This might also reflect a desire to delay the perceived progression of the illness; patients may be keen to maintain a sense of 'normality' that ventilation overtly negates and eager to sustain independent function for as long as they can. MND patients already know that they are going to die; however, whilst this outcome is assured, the route to this destination remains unclear. Introducing the idea of respiratory insufficiency and the prospect of a ventilator into this future may have understandably negative psychological ramifications for patients who had been previously unaware.

- **Positive cognitive appraisals of ventilation can exist alongside negative emotional responses to the treatment: patients may welcome the prospect of 'help' whilst finding its form and meaning anxiety provoking.**

There may appear distinct contradiction in patient narratives, wherein patients are pleased that help is available to them should respiratory insufficiency occur, yet simultaneously disparaging about the form that this help takes. There may be clear apprehensions about using a ventilator and wearing a mask that encourage patients to maintain a distance from the treatment, even where they welcome the presence of an intervention that might help them to avoid the physical distress of respiratory impairment. This contradiction appears to represent a divide between cognitive appraisals of the treatment and emotional responses to it. A consenting and compliant patient may not necessarily be

an emotionally engaged or even accepting patient; patients might well appreciate the logic and sense in using ventilation to support them as the illness progresses, yet also fear and resent any need for it.

Sensitivity to the complexity of this response may help to support patients during the process of treatment decision-making. Patients are likely to be implicitly aware that clinicians view decision-making in a more logical and problem-focused way, yet whilst it may be easier for some patients to separate their own emotional responses from the process, others might naturally respond more to how they *feel* about ventilation and struggle to see past this immediate emotional pull to align their views with those of clinicians. Patients may *think* that ventilation is a good idea, yet *feel* apprehensive about its use to such a degree that they are willing to forego its use. Teasing apart these apparently paradoxical elements of response may require clinicians to explicitly probe both the 'thinking' and 'feeling' responses to gauge where patients stand.

- **Ventilation may come to be viewed as a 'crisis' intervention, encouraging and justifying active attempts to avoid or postpone use.**

Ventilation may be seen by patients as an intervention to be welcomed at crisis point, yet actively avoided otherwise. Patients may be unwilling or unable to identify a clinically appropriate time for its implementation and might be willing to tolerate significant impairment before they concede that help is required. Clear and apparent physical need (e.g. discomfort or functional impairment) seems to engage patients with the idea of ventilation and increase readiness to consider the treatment; however, interactions between knowledge and understanding may mean that information and education might also be used to encourage interest. Although clinicians can appreciate that ventilation is intended to support respiratory function and to maximise respiratory efficiency, patients may still prefer to maintain distance from the treatment by seeing it as an intervention that will only be used to address a loss in function that they can no longer live with or compensate for. There are likely to be interactions between how/when patients are willing to use ventilation and the contradictory responses to the treatment described above; patients who both appreciate that ventilation offers a sensible solution to respiratory problems *and* predict that emotional responses to using it will be negative may be more likely to accept it in principal whilst trying to postpone its use until the last opportunity. It arguably suits these patients to conceptualise the ventilator as a treatment that will only be implemented when they can no longer go on without it.

- **Ventilation may be more 'acceptable' to patients where they have identified their own physical incentive to use it. Success can sustain motivation and may be more readily appreciated where the effects of intervention are subjectively perceived.**

Patients may accept trials of ventilation without necessarily being fully 'invested' and using a ventilator does not necessarily reflect a belief in the necessity of the treatment or its utility; patients may be driven by factors such as anxiety, duty/obligation or even simple expectation. Routes onto ventilation may be crucial determinants of response and simply partaking in trials does not ensure that patients are fully committed to establishing a routine of successful use. Patients who are able to identify their own reasons for initiating the treatment and who can relate to a clear need for the ventilator are likely to be more motivated and committed; this does not ensure success but does seem to increase the likelihood that patients will overcome early challenges. Psychologically, these patients may have a greater sense of there being tangible consequences to non-tolerance. The presence of overt physical symptoms (i.e. disturbed sleep, fatigue, waking headaches, dyspnoea) may also provide scope for a degree of physical improvement that sustains motivation over time. Patients are more likely to regard the ventilator as a useful addition to care where they notice clear physical improvements that can be directly linked to its use. Patients who are initiated onto the treatment in response to oximetry readings alone – and who perhaps do not perceive any real 'need' for intervention – are less likely to invest in the treatment and may be less motivated to persist when faced with challenges. There may be cycles of success and failure that are triggered and maintained by perceptions of need, potential for change, motivation to succeed, difficulties encountered and actual consequences of use. Clinicians should consider these factors in their planning and response.

- **Objective clinical outcomes do not offer information about the subjective experience of ventilation, nor are they always indicative of a patient's satisfaction with the treatment.**

Patients may use ventilation in different ways and for different reasons and are likely to define their own concept of treatment 'success'. This may appear to be independent of the clinical direction offered to patients and the goals defined by clinicians. It is possible for a patient to use the ventilator in an idiosyncratic way (i.e. sporadically and only to address dyspnoea) and be highly satisfied with the addition to their care or for a patient to be fully tolerant/compliant in nocturnal use and report extremely negative appraisals of the treatment. Patients' ability to tolerate the ventilator and their willingness to comply with use as directed does not tell the whole story; attitudes appear just as important as actions and clinicians should enquire about both when trying to establish how effective and appropriate the treatment is. Subjective interpretation of results and the realisation of expectations – or, conversely, the failure to have hopes sustained – may be viewed as important components of the post-ventilation experience.

Importantly, complex psychological responses to ventilation can exist alongside positive clinical outcomes; for example, there may be significant disappointment that specific hopes are not realised, even where the treatment has improved areas of physical function. In the context of MND, ventilation is a palliative intervention and one should seek an indication of how and why ventilation is adding to or subtracting from the overall *quality* of life for patients; this should involve more than a measure of physical change, extending to the psychological and emotional consequences of use. Patients may remain acutely aware of what it means to be ventilated, with a fear of dependence on the ventilator not necessarily waning after initiation and an anxious attachment to the treatment potentially merging with this sense of uncertainty about the future. There is a significant potential for ventilation to be positively received and to make a significant contribution to improving life with MND, yet one must acknowledge and address the psychological as well as physical challenges that it represents.

Finally, as a more general finding from this thesis, one that is central to the qualitative philosophy that informs it, it must also be noted that:

- **All patients are individuals living with the same disease; the varied and unpredictable physical presentation of MND creates a unique illness experience and this in turn may be interpreted differently by each person attempting to derive meaning from it.**

All patients arrive into respiratory services with a history that extends beyond the illness; they are potentially very different people with ideas and understanding about the world and their own place within it. Yet their attitudes may also be shaped by their illness experiences and the ways that they choose to 'cope' with the life that MND creates. They come with variable levels of knowledge and understanding about their illness and may or may not wish to learn more about the physical reality of their condition. The act of providing information to a patient does not mean that the patient necessarily understands or retains that information. Similarly, these individuals may wish to take an active role in determining the direction of their care or they may prefer to be led by medical professionals in the direction that these trusted others perceive to be 'best' for them. Collaborative decision-making may be essential for engaging some patients, whilst others may actually want clinicians to lead treatment planning according to their own clinical judgements. One size does not fit all and it would be unfortunate for clinicians to interpret clinical guidelines as suggesting that there is a single pathway towards ventilation decision-making and implementation that will work for all MND patients. Clinical guidelines that promote best practice are indubitably useful, yet they must be applied with a degree of caution; the individual responses of patients and their unique circumstances and perspectives should encourage clinicians to work within an agreed framework but to be flexible and responsive to the *person* who arrives into the clinic on any given day.

9.9.2 Clinical Practice

Although this thesis was primarily intended to increase awareness amongst healthcare professionals, a number of practical steps have emerged to guide best-practice in respiratory services. The process of disseminating the key messages and clinical outcomes of the research has commenced and activities are outlined in Appendix 7; this section concludes the thesis by highlighting the most significant contributions made to routine clinical practice. It does not provide an exhaustive list of procedures or offer definitive clinical guidelines, it outlines the changes that have been implemented or suggested at a clinical level within the research team at WCNN and UHA and through communication with the MNDA as the funding body of this research. External grant reports may be available on request from this organisation (see Appendix 7).

9.9.2.1 Defining the Respiratory Service

As a note on the way that respiratory services are perceived and presented, it is acknowledged that caution should be applied when defining a 'place' for ventilation within the care pathway. There is a risk that patients will interpret ventilation as being an 'end point' within respiratory services, with monitoring seen to be leading up to this intervention. It is suggested that explicit efforts should be made to emphasise the role of the respiratory service as monitoring and supporting respiratory function throughout the duration of the illness, with ventilation being only one treatment option that might be explored along the way. It is important that ventilation is presented as a choice within the service and not an evitable conclusion to the care being offered. Consultant neurologists are often in the best position to explain the role of the service at the point of referral.

9.9.2.2 First Appointments

Routes onto ventilation may not always be clear to patients and the reasons for referral may appear equally confused. It should be made clear to patients why they have been referred. Clinicians should ask patients to offer their own reasons at the first appointment so that a level of understanding can be established. This may also offer insight into how much insight and information patients have about respiratory aspects of the illness and also how much they want at that stage. This should be documented in medical case notes.

Patients may misinterpret questions aimed at establishing the impact of respiratory change on day-to-day living; where clinicians focus on defining whether any 'change' in respiratory function has occurred, patients may interpret this as an enquiry about *problematic* change. It appears that patients

often answer questions about symptoms by responding to whether they are causing physical discomfort or impedance rather than whether they are present or not. Clinicians can usefully provide patients with lists of specific respiratory symptoms with graded responses (i.e. levels of breathlessness or fatigue) to ascertain presence and severity. This also encourages information to be collected about symptoms that patients may not identify as having respiratory origin (i.e. disturbed sleep or changes in appetite). This can then be used as a baseline against which change can be compared over time.

Carers often have insight into respiratory change that patients are unable or unwilling to provide. Family members can present as useful sources of corroborative evidence; where patients consent, carers' testimonies should be sought to substantiate and verify the accounts provided by patients.

9.9.2.3 Introducing the Idea of Ventilation

It is suggested that longer initial appointments should be offered to MND patients to allow an appropriate time for them to process information and ask questions about ventilation when the idea of this intervention is first being presented. When communicating information about NIV, clinicians should be sensitive to the gravitas of the information being conveyed; this means attending to *how* information is communicated as much as to the content. Clinicians should aim to be empathic communicators who understand that the ventilator is more than a routine intervention to patients; they should be ready to listen to patients' concerns and there should be time and space allotted to do this in clinic appointments.

As a means of reducing the stress associated with ventilation, clinicians may wish to emphasise its non-invasive nature and the fact that choice is retained throughout its use. They should also highlight the alternatives available to patients (e.g. pharmacological support), so that they do not feel that ventilation is the only option and can make an informed choice. Clinicians should include a summary of the evidence base when they inform patients about the treatment and there should be an open and honest exchange of ideas. Asking patients to feed information back is a simple but useful way of establishing their level of understanding.

9.9.2.4 Information Provision

Clinicians should appreciate that the same information and advice may be received and interpreted differently by different patients. Over time, the primary focus should be on ensuring that information is *available* if and when it is wanted; where patients appear unable or unwilling to receive or retain information, it is suggested that it could be provided to a willing carer or family

member and that written information should be available to patients so that it can be sourced as needed. The information made available should also offer more than the simple mechanics of the treatment. When weighing up whether to trial the ventilator it is important for patients to know what it might be like to actually *live* with a ventilator, rather than just how it worked. The testimonies of other patients may prove particularly useful in this respect. These points have been incorporated into updated information sheets provided by the MNDA, all of which are downloadable from their website (see www.mndassociation.org).

9.9.2.5 Decision-Making

It seems likely that having people other than clinicians to discuss treatment decision-making with may be useful to patients; in some cases there may be implicit barriers to effective communication within the doctor-patient relationship that prevents a completely open exchange of idea in the formal setting of the consultant's clinic room. This does not necessarily reflect poorly on clinicians themselves, as this is likely to be influenced by preconceived ideas about the status of doctors and the nature of the relationship between the two parties. Physiotherapists within the respiratory team or MND nurse-specialists within general MDTs appear to be particularly effective in this role, allowing patients to discuss thoughts and feelings in more informal settings. These are key workers already situated at the heart of the clinical teams at WCNN and UHA and are familiar with and to patients; they can also be contacted by telephone or email, making them accessible to patients.

Where the symptoms that signal a need for ventilation are not overtly 'respiratory' (e.g. fatigue), and particularly where patients are not dyspnoeic, doctors should invest greater efforts in clarifying why intervention might be useful and in engaging the patient in a dialogue to ensure shared understanding. Special care should be taken in cases where ventilation initiation is sought in response to oximetry readings alone (i.e. in the absence of subjective need being expressed by patients).

9.9.2.6 Treatment Initiation

Initial treatment trials are important in terms of defining competence and establishing a relationship with the ventilator. The process of acclimatisation appears complex, containing both physical and psychological components that include issues of trust, control, acceptance and adjustment. Negative early experiences may lower self-efficacy and so greater attention should be afforded to this experience to enable patients the best possible chance of success. At a practical level it is suggested that patients should commence ventilation on lower settings and build up air-pressure gradually to allow confidence to grow as they learn. This may be particularly important for patients with marked

bulbar involvement, which appears to increase the risk that the ventilator might feel physically overwhelming.

The hospital experience is itself an important factor. There should be a degree of preparatory work before patients are admitted, with efforts made to establish specific care needs/preferences. Hospital staff should be informed and sensitive to the unique demands of MND, particularly where patients have bulbar impairment that reduces their ability to communicate verbally. One must appreciate that what might appear trivial and/or tolerable for some patients may represent significant challenges for the MND patient and minor occurrences might become significant events in the hospital. The trial environment is also important; sensitivity should be afforded to the physical location, which should be as quiet, private and relaxed as possible. At UHA patients are now invited to trial ventilation in their own room rather than on general medical wards. There is also accommodation for carers to stay in the hospital where patients/carers feel this would be beneficial. Continuity of care in hospital and having the same key workers should also be encouraged. Community out-reach teams that allow patients to trial ventilation at home should also be considered as part of the wider respiratory service.

Healthcare professionals might facilitate successful acclimatisation by helping patients to frame early challenges as part of the process of 'getting used to it', i.e. as expected complications that can be overcome in time. Normalising in this way may help patients to attribute challenges to the treatment or process (external causes) rather than to themselves or their abilities (internal causes), and may also help them to frame the problem as temporary rather than permanent. It is likely that this will limit assaults on self-efficacy, help to reduce negative emotional responses to early challenges and allow patients to retain a sense that the process of acclimatisation and ventilator use is manageable and achievable. However, trials should be framed as ongoing periods of use rather than being temporally constrained; it may take a number of weeks or months to determine whether ventilation is an appropriate choice and patients should not be given the impression that a trial begins and ends in the hospital environment or over the space of only a few days. Trials of ventilation should represent genuine test periods that allow patients and clinicians to come to conclusions about whether NIV is appropriate and useful. This is a decision that patients may not wish to base on clinical criteria alone and clinicians should be mindful of this. Patients should be allowed to decline ventilation after a suitable trial without feeling that the endeavour has ended in 'failure'.

When planning treatment initiation, careful consideration should also be given to the availability and potential competence of carers. It is important to ensure that the support network around the patient is sufficiently educated and motivated to enable ventilation to be used. Carers may themselves require psychological support to prepare for the role that they might play in helping

patients to acclimatise to the treatment and facilitating its successful use at home thereafter. On a practical note, carers and/or patients should also have the ability to manipulate the mask interface and operate the machine before they are allowed to take it away from the hospital and this should be demonstrated and recorded in medical case notes at discharge. Written information should be provided and should include, if possible, diagrams of even basic points of operation (e.g. how the straps fit together and how the equipment is to be cleaned and maintained).

Managing expectations may be an important part of the pre-initiation work that clinicians must do with patients. Clinicians should appreciate that in an illness where there is scant opportunity to improve physical status, patients may invest significant hope in any form of treatment. Clinicians should be mindful of the possibilities of hope, expectation and potential disappointment and should informally monitor these constructs in discussions with patients/carers throughout the early stages of use.

9.9.2.7 Masks

There should be a period of consultation prior to initial trials in which patients are able to select the mask that they would like to start with and there should be an ongoing dialogue as to the appropriateness of the mask throughout initiation and early stage use. Clinicians should be aware that masks have the ability to significantly deplete self-efficacy and motivation in the early stages.

At an industry level, the data presented in this thesis suggest that masks can still be improved for use with this population. The medical aesthetics of masks (i.e. the use of distinctive blue plastic components; see Appendix 1) is appropriate for use in hospitals, yet patients may not always respond well to this in the home. The respiratory masks used with the ventilator originate from use in acute medical settings where specific designs and marks signal quickly and easily to medics the type and function of masks; this may not be as applicable to patients using them in the context of MND, where use is planned, prolonged and may extend over a number of years. Work is now being disseminated to convey the views of patients about these aspects of the treatment (see Appendix 7). More neutral colours and naturalistic materials may help patients to psychologically acclimatise to the masks and increase how acceptable masks are deemed at an intuitive level. The size, shape and physical appearance of masks may convey an intimidating message to patients.

9.9.2.8 Long-Term Ventilator Use

One of the key factors differentiating ventilator use in MND patients from other illness populations is the progressively degenerative nature of the illness. MND patients are uniquely aware that

dexterity will wane, verbal communication will gradually deteriorate, and that the respiratory problems that the treatment aims to address will also increase in severity. Patients arrive at ventilation in different physical states and may also see their clinical circumstances change significantly over the course of its use. Clinicians should monitor change in both outcome and process carefully. They should aim to sustain effective ventilator function (e.g. optimal oxygen levels) by modifying pressure settings and should also seek to maintain satisfaction with the experience by investigating changes in the treatment environment (e.g. communicative ability, ability to independently maneuverer the mask interface). Not being able to raise an alarm or remove the mask may have significant psychological ramifications; these problems may be addressed by having quick-release mechanisms incorporated into mask designs and alarms fitted to enable patients to attract attention in instances where they feel unsure or unsafe when using the ventilator.

Patients who are highly motivated to succeed may be willing to pay a high price to do so. Clinicians should be aware that some patients might simply accept discomfort and even painful abrasions caused by masks if it means that they are able to continue using the ventilator. Clinically, it may be necessary to monitor this closely, particularly in the early stages of use. It might be suggested that all healthcare professionals that have contact with patients (e.g. GPs, nurses, occupational therapists, dieticians) should enquire about basic aspects of ventilator use (e.g. mask function and comfort), to ensure that problems are not missed between appointments with the respiratory service. Clinicians should also maintain an interest in patient's attitudes towards the treatment over time. They should be conscious that patients may develop an anxious attachment to the treatment both in the early stages of use and over time; dependency is an important clinical issue and one that should be explored in respect to both its physical and psychological dimensions. Establishing patients' views from the start of treatment provides a baseline of feelings and wishes that can be used to check against for changes in view over time and as the illness progresses. Continuity of care is important as relationships and understandings can be developed between clinicians and patients over time; there should be open channels of communication that allow honest appraisals of the situation as the illness progresses.

9.9.2.9 The Role of Psychology

It is suggested that a psychologist should be working in some capacity within the multidisciplinary team. This role should entail one-to-one work with patients *and* the provision of consultation/education to other team members. In respect to how and why additional psychological support might be necessitated, a number of key circumstances and events have been highlighted in this thesis. For example, all patients may potentially experience significant anxiety at referral, regardless of actual respiratory status or routes into the services. In some instances the degree of anxiety experienced may escalate to produce hypersensitivity to potential symptoms, excessive

monitoring and behavioural changes aimed at limiting the impact of respiratory change; psychological support may help patients to address this anxiety and limit its intrusion into day-to-day living.

The period of monitoring and assessment may also trigger psychological distress; formal tests may be perceived as stressful, yet may also offer reassurance that there are no significant problems. It seems likely that a cycle of anxiety/relief might be sustained and re-lived and that over time this may have an accumulatively negative impact on psychological health. Patients might reasonably be monitored for many months or even years before being offered ventilation and it seems prudent to be offering support to patients who are not using ventilation as well as to those who are. There is also a great deal of psychological work that might be done with patients who want to use the ventilator yet perceive psychological barriers to use. For example, patients who feel claustrophobic in the mask may be offered support and graded tolerance/exposure work to address their concerns and psychologists may also be able to facilitate cognitive reframing to enable the treatment to be viewed as a positive addition to care, rather than a defeat or sign of illness progression. At UHA a psychologist in now included as a key worker within the respiratory team and this has been evaluated as an effective addition to care.

9.10 Reflections on the Study

The unique and variable nature of patient and carer experiences reported in this thesis seemed to validate the decision to adopt an idiographic approach to this research. Existing quantitative studies have treated samples as homogenous groups and have aggregated and summed responses in an attempt to define the experience of ventilation. The data presented in this thesis suggest that this approach is likely to obscure important individual differences and stifle the patient voice. Implicitly, clinicians understand that all patients are individuals, arriving at clinics with different physical profiles *and* different psychological strategies, yet existing quantitative research has still attempted to impose a generalisable framework onto the patient experience. This study has formally presented the patient perspective so that the subjective and individual nature of the patient experience is evidenced within the wider literature. The fact that experiences differed so radically is one of the most important findings in this study. It is not possible to identify a 'typical' response to respiratory impairment or to predict which patients will 'succeed' on NIV; there appeared to be a complex combination of variables interacting to define these outcomes. This thesis has provided insight into the nature of these variables so that clinicians can better understand patients as they pass through the respiratory service and onto NIV.

One of the most important observations made in the present study was the impact that patients' wider approaches to the illness were seen to exert throughout. Existing studies have addressed responses to NIV in isolation, seemingly assuming that this aspect of the illness experience takes place in a contextual vacuum. The present study has shown how previous illness experiences can influence patients' interpretations of subsequent events and also how they react to them. The way that patients chose to 'deal' with the illness (e.g. by being actively involved or by trying to avoid the illness altogether) influenced important clinical variables; for example, these attitudes defined information-seeking behaviours, which then determined levels of knowledge and understanding. Patients' interpretations of events, symptoms, clinical experiences and responses to treatment were also influenced by how much they knew and wanted to know about aspects of the illness. By exploring the wider illness context, the present study was able to offer potential explanations for patients' responses to treatment. The one other qualitative study in this area (Sundling et al., 2009) only described responses to ventilation without suggesting how wider illness experiences may have informed these reactions.

This research has highlighted a number of important issues that clinicians should be aware of and has also proposed practical interventions that might help to improve patients' experiences of referral and ventilation. However, it is acknowledged that there are limitations to this research that should be considered when evaluating the results. One criticism often waged against qualitative studies using smaller samples is that the results may not be generalisable in the same way that larger quantitative studies can claim to be. This study elected not to use a grounded theory approach and so did not attempt to undertake theoretical sampling to achieve theme saturation. Had this technique been adopted, one might have had greater confidence that the data reported could be applied more generally to MND patients in the same position. Indeed, although there was a significant degree of repetition and overlap in the themes generated from ventilated patients in the cross-sectional study and those in the post-ventilation stage of the longitudinal study, it was clear that new and unique themes did emerge from the larger sample. It is possible that had more patients been recruited, additional themes would have come to light. However, the replication of important themes served to validate what had been found and it is important to remember that this study did not aim to produce generalisable theories. This was an exploratory study that sought to raise clinical awareness rather than to offer a definitive or prescriptive guide to patients' experiences; it was the *nature* of the experiences reported that were important in this respect. This study elected to use IPA because of its idiographic principals, encouraging greater depth to the analysis undertaken for each patient. Qualitative research rightly focuses on the quality and not the quantity of data and how well the research aims have been met; in this case, it was not deemed appropriate to speak in terms of absolute generalisability.

It should also be noted that even if this study had sought to expand recruitment, it was subject to practical restrictions imposed by the population sampled. MND is a rare illness and even amongst MND patients not all go on to trial NIV. As such, it was not feasible to recruit significantly larger numbers of patients. It is noted that the rate of recruitment for this study was high, with almost all consecutive patients referred into the respiratory service during the recruitment period agreeing to take part.

The fact that recruitment took place at only one centre might also be identified as a potential limitation of this study, as one might question whether the results can be applied to other MND centres and respiratory services in the UK. It is true that results may have been shaped by the organisation, philosophy, resources and personnel at the hospitals involved in the care of these patients. However, it should be noted that the centre selected is one of only a small number of specialist MND centres in the UK. It covers a wide geographical area and the patient population itself was observed to be representative of the wider UK population in terms of clinical and demographic profile (see section 5.2.3.1, Table 5.2). The fact that the sample was clinically representative and reflected the natural flow of MND patients into clinics may actually be seen as a key strength. The care pathways operating within this centre adhere to recommendations set out in the NICE guidelines for NIV use in MND (NICE, 2010), which are national guidelines. This suggests that both the patients and the service should be considered inclusive and representative, increasing the likelihood that the experiences reported in this study are also representative.

As noted, IPA was selected for use in this study for its idiographic focus, yet a sample of twenty-six patients was actually large for a qualitative study of this kind. One of the major challenges faced over the course of data analysis was balancing the wider thematic analysis with an appropriate level of idiographic detail. On reflection, it might have been easier to achieve this balance with a smaller sample; however, when considering what the sample might have looked like had it been reduced to fifteen or even ten patients (a more typical IPA sample size), it was clear that much of the richness that was apparent in the resultant analysis would have been lost. It was useful to have sufficient numbers to clearly identify cases of convergence and divergence within the sample. One must also consider the variable contribution that patients were able to make in respect to interview data, as some patients were more physically limited, potentially influencing the insight that they could offer. Conducting qualitative research with patients who had impaired communication was also a significant challenge and some patients in this large sample were not able to provide the quantity of data that non-bulbar patients could offer. This is not to discredit their contribution; indeed, it may be considered a significant strength of this study that it sought to include all patients entering the respiratory service and not just those who could easily communicate their experiences.

In the planning stages of the research, it was intended that conducting a longitudinal study would circumvent the possibility that patients' experiences would be biased by subsequent experiences or that memories would become distorted over time. However, it was clear that interviewing patients periodically did not capture all events in 'real time'. Patient accounts of events such as decision-making were ultimately retrospective and it was difficult, for example, to decipher whether patients who reported negative emotional responses to decision-making were more likely to encounter problems on the treatment or whether early problems led patients to retrospectively describe decision-making in a more negative light. This criticism may be true of all research that does not collect data in precise temporal alignment with the events or experiences being explored. Many qualitative and quantitative studies involve a recall period of many months and even years, which is a logistic necessity in this type of research. One might suggest that patients could have been interviewed more frequently (e.g. every month); however, it is important not to cross the line into intrusion. These patients were faced with extreme physical and psychological strain in simply living with MND and placing extra demands on them may have increased distress and made them less likely to cooperate with the research. Patients were already generous in the time and efforts that they afforded to the study and this should be recognised.

It should also be noted that there did seem to be advantages to using this longitudinal design, as it was able to capture other aspects of patient's experiences as they were unfolding (e.g. referral and early stage use of the ventilator). This method seemed to have captured greater variety in the descriptions of these types of experiences and one can see that it is likely that the accounts of patients in the cross-sectional study may have been coloured by the fact that all had emerged from the process of acclimatisation and were established in routine use of the ventilator. There appeared far greater negative emotional response reported in the longitudinal phase of the research, which was exploring these experiences with patients as they occurred and as the challenges of early use were still being addressed. This data seemed to be of greater clinical utility. Retrospectively it is possible to identify processes and procedures that could have been improved over the course of the research; however, it is important to note that the research aims were still successfully fulfilled.

9.11 Personal Reflections

An IPA approach emphasises the importance of the researcher as an integral part of the research process and researchers are encouraged to reflect on their own preconceptions both during and after analysis. This section details my own thoughts on the research process and how my ideas about this subject area changed over its course.

9.11.1 Goals & Application

In order to identify where gaps existed in the literature, it was necessary to examine the established research base. This process itself shaped my own perceptions of NIV as I embarked on this study. Existing quantitative research focuses on clinical outcomes and presents a positive picture of the potential of NIV to help patients. In retrospect, I can see that I expected the results of this enquiry to be used to change clinical practice to ensure that every patient could use and benefit from NIV. I did not consider the possibility that NIV might not be right for each patient. In seeking to explore tolerance and compliance in particular, I had perhaps assumed that gaining insight into the reasons why patents failed to tolerate or comply would allow clinicians to devise methods of ensuring that more or even *all* patients tolerated and complied. Over the course of the research, my views changed.

Spending time with patients and witnessing firsthand the global challenges that MND presents, I came to appreciate the significance of the barriers to NIV use in this population. I could also see how different physical and psychological profiles meant that NIV might not be a useful or appropriate treatment for all patients. Although I might initially have thought that the best option for these patients would have been clinical intervention to try to facilitate greater success on the treatment, I latterly came to respect that patients should be allowed to 'fail' on NIV if they do not want to use the treatment. Indeed, my notions of success and failure on NIV were transformed by patients who seemed convinced that they would be better off without the ventilator. NIV is palliative treatment and patients' should be given autonomy to make choices based on their personal wishes and values, rather than what is clinically possible. My hopes for this thesis now relate to helping clinicians to ensure that all patients who *want* to succeed on NIV are given the best opportunity to do so and that patients' experiences of NIV are as positive as they can be.

9.11.2 Methods & Process

At the start of this research process I felt confident that a qualitative methodology would allow insight into individual differences in experience that were absent from the quantitative data, I did not anticipate that there would be quite as much variation as there ultimately was and in many ways this limited the direct clinical utility that I had intended to provide. Often the practical advice that emerged from the data could offer only the importance of clinicians being 'mindful' of specific issues or experiences. There appeared to be few reliably predictable patterns of behaviour that emerged. For example, it was not possible to identify, using qualitative methods alone, which patients would experience greater or lesser success on NIV. I can now appreciate that the framework that is imposed on the data by quantitative methods may sometimes be useful for the results to have meaningful clinical application. For example, using standardised scales may have revealed that patients who did not want to be involved in decision-making or who were less tolerant of the practical difficulties involved in NIV were more anxious or more depressed. I still believe that using quantitative methods alone would have missed the rich detail of individual experiences that

emerged; however, I would now suggest that there may be utility in performing mixed qualitative/quantitative research in this field. If it were found that patterns of standardised scale scores were in some way associated with or predictive of certain responses to MND or NIV, these scales could be integrated into routine clinical care to act as an heuristic guide for clinicians.

Over the process of interviewing patients, and particularly where patients were interviewed on a number of occasions (some patients were interviewed regularly for eighteen months), I also became more aware of how the research process itself may have shaped experiences. By engaging patients in interviews, I was asking them to consciously reflect on their experiences in a way that they might not have otherwise done. A number of patients indicated that the process of being interviewed had encouraged them to be more reflective in their experiences; some suggested that they had been thinking of issues in advance to discuss during interviews. This may have meant that certain emotional responses were bought into conscious awareness and given attention *because* of the research process. It may also have helped patients to address and resolve particular problems.

A number of patients and carers suggested that the interview process had been a positive experience because it allowed them to talk about issues with an impartial person. This appeared to have had some therapeutic benefit and so it may be suggested that the process itself helped to ameliorate certain negative responses to situations. As a researcher, I am encouraged to be careful not to change or influence the things I am seeking to describe, yet it seemed interesting in itself that having an independent person to talk to may have helped these patients to make sense of their experiences. This is perhaps something that might be considered within the wider MND service, as currently the multidisciplinary care team does not include psychological support. Carers in particular seemed to benefit from this process and it was noted on a number of occasions that they were relieved to be able to talk to somebody because patients did not want to discuss the illness. Carers are important figures within the healthcare system and one might suggest that they would also benefit from greater psychological support or counselling services.

Over the course of the research, I also became increasingly conscious of how patients perceived the study itself and how the subject of the research may have influenced responses to events and experiences. All participants were aware that this research was interested in exploring their experiences of respiratory aspects of the illness and NIV use and this may have elevated the importance of these experiences. In some cases it seemed likely that directing patients to these issues had increased their salience in a way that encouraged patients to attend more closely to what was happening and how they felt about it. I was also aware as a researcher that I was asking patients to consciously reflect on their wider illness experiences where they might not otherwise have done so. A number of patients preferred to remain detached from the illness and to avoid addressing specific issues, yet the interview process encouraged them to explore their experiences in a way that may

have changed their feelings. My own role in this process was difficult; I both wanted to extract as much detail and perspective from patients as I could, yet was also aware of the need for care and to avoid exposing patients to unnecessary distress. The way that patients approached the interview situation informed my impression of how they were coping with the illness. Many patients felt able to discuss issues in a composed and rational way, whilst others quickly became distressed, sought to avoid questions, or used humour to resolve tensions. This was a form of 'soft' data that influenced my interpretation of the interview transcripts.

9.11.3 Data Interpretation

It is important to appreciate that the analysis presented in this thesis is my own subjective interpretation of interview transcripts. As noted in Section 3.5, IPA researchers are not attempting to define an objective 'truth' but to offer a coherent and credible account of the data collected; this is how validity is assessed in IPA. As such, I feel it is important to acknowledge my experiences with patients and how this informed the analysis undertaken. During data analysis I was able to interpret the words on the page within a wider context, based on my impression of patients and the atmosphere that was created in interviews. I had been aware of facial expressions, body language, eye-contact and subtle changes in tone and intonation that were not easy to convey through transcription yet shaped my understanding of the text. My interpretation was also shaped by a more sustained relationship over time, where it became possible to consider wider context and previous interactions. I feel that this connection with the patients and with the data strengthened the reliability and validity of the analysis presented in this thesis, placing me in a unique position to credibly interpret patients' accounts. This was a significant strength in this study.

Working with MND patients was often emotionally demanding. Building the rapport necessary to conduct productive interviews meant establishing trusting and empathic relationships with patients and carers and it was difficult not to be affected by physical and emotional changes over time and in many cases by a patient's death. The nature of the relationship between the qualitative researcher and participant is arguably of a different kind to that of the quantitative researcher, simply by virtue of the type of data exchange that takes place and the way that the researcher themselves becomes an intrinsic part of the research process. An appreciation of these differences in roles has been an important part of my learning.

I feel that my own perspective on the research process has changed over the course of this study. I now have a more realistic view of how research aspirations are translated into research achievements and the challenges and compromises that are made along the way. Conducting applied research presented its own trials and it was necessary to constantly adjust the research process to accommodate clinical realities. Although there was a finite time frame in which to capture the experiences of these patients due to the aggressive nature of illness progression, I am aware that being able to spend relatively extensive periods of time talking to patients and really exploring their

experiences is a luxury that clinicians simply do not have in a busy health service. The breadth and depth of the interpretative analysis reported in this thesis and the narrative style adopted to convey the results were attempts to offer the kind of intensive insight that can only be achieved through this type of comprehensive research process. Being able to translate the results of analysis into meaningful clinical advice and intervention was also challenging, yet ultimately I feel this has been provided an effective focus for the thesis. Indeed, I feel this venture has reinforced the value of qualitative research in medical setting and within the health service more broadly.

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Appendix 1

Pictures of Non-Invasive Ventilation Respiratory Masks

Figure A1. Mask interface: Nasal Mask.

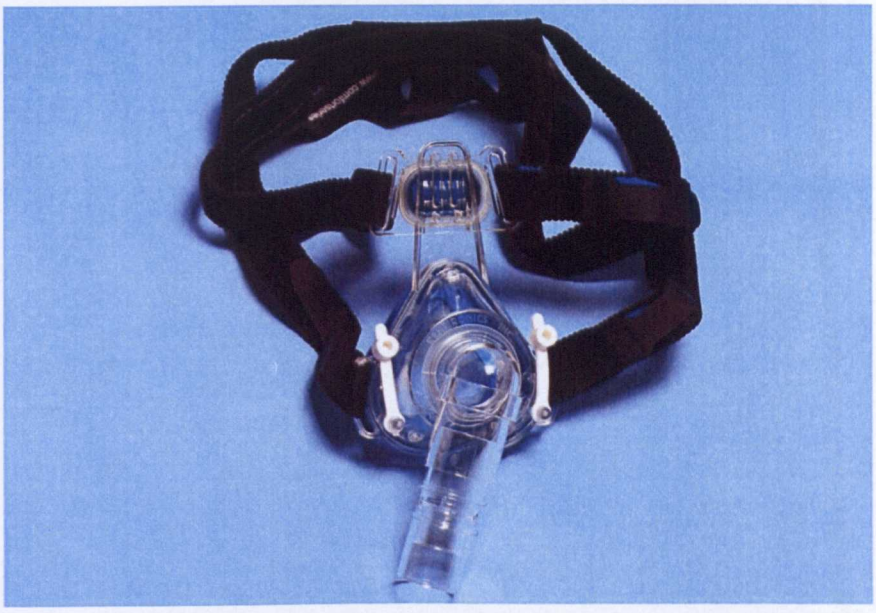


Figure A2. Mask interface: Oro-Nasal ("Full Face") Mask.

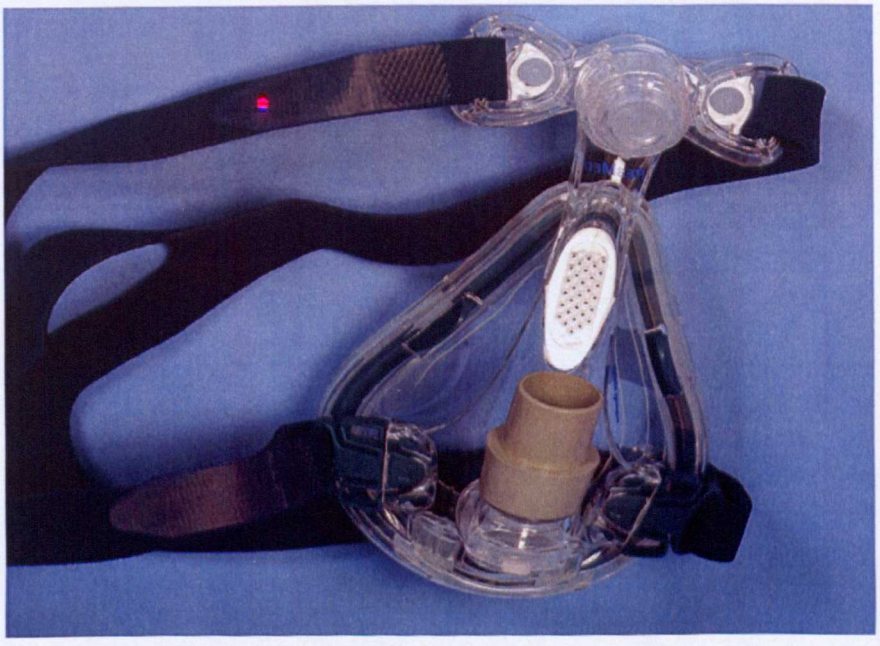


Figure A3. Mask interface: Nasal Pillows.

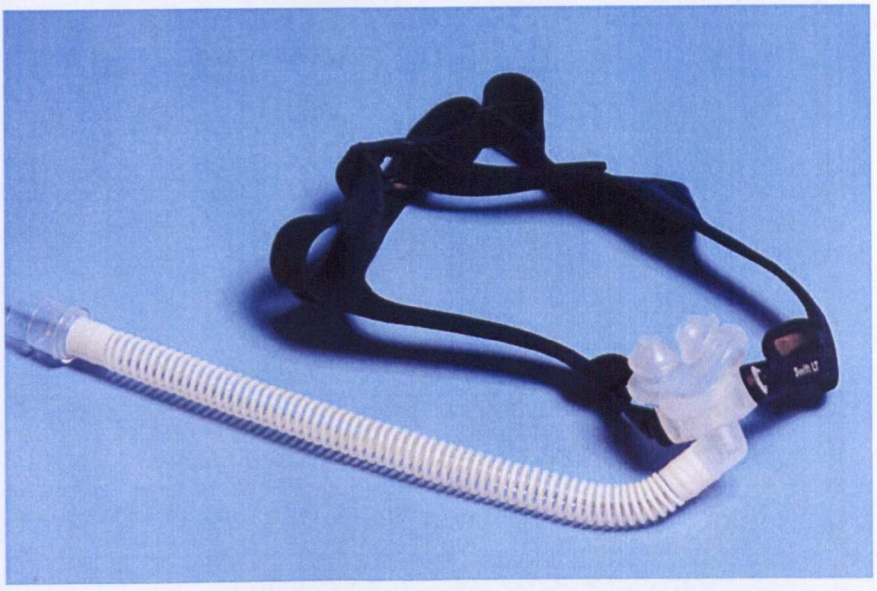
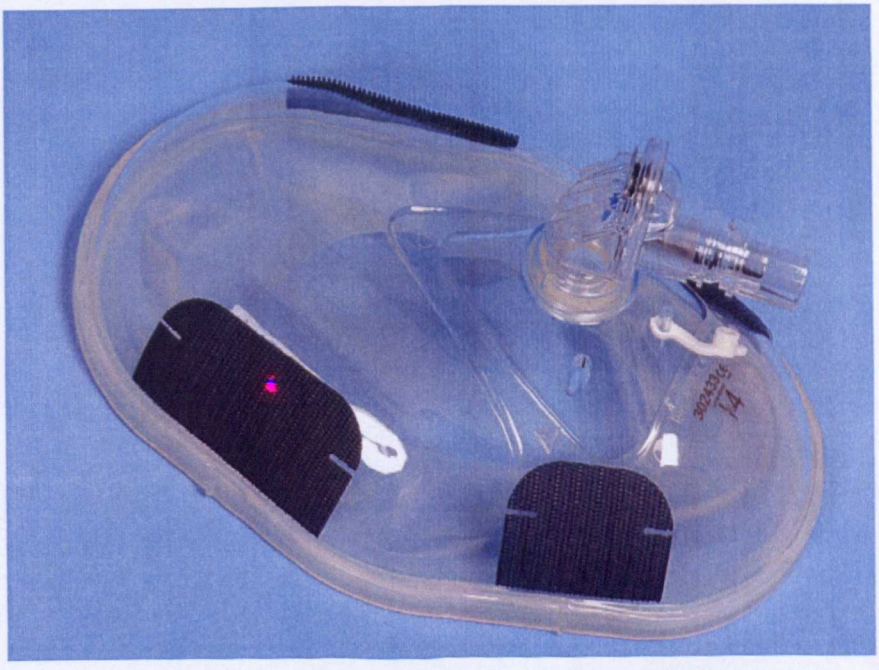


Figure A4. Mask interface: Total Face Mask.



Appendix 2:

Interview Transcript, Sample Analysis, Theme Construction

Interview Transcript

P7 Interview1, Pre-Ventilation. Notes: Female. Patient's speech significantly dysarthric, slow and became increasingly slurred as the interview progressed. It took a long time to complete interview, yet the patient appeared keen to contribute. In good spirits, though became noticeably fatigued over time.

OK, well, as this is our first interview what I'd like to do is to get some background information from you first. So, to start could you tell me when you were first diagnosed with motor neurone disease?

Yeah, last August.

Last August, ok, and what can you tell me about that time?

For about twelve months prior, I'd had various things that I'd gone to the doctor with, and at one time I couldn't barely speak - my tongue was too big and I couldn't... I had to really form the words, you know, it was bad. And another time it was breathing was really bad, you know, I took the dog out and I was gasping. Another one was I had a hot pain in my, well, not a pain, a hot sensation in the same area in my head, that was sort of one. And then I found I was dropping things, you know, you were thinking you'd picked something up and you hadn't, you know? And, I know I... Oh and my throat; I felt from my nose to my throat was thick and like when you have a bad cold and I knew there was a problem there and that's when my doctor finally, well, one of the doctors - I saw different ones - sent me to the ear, nose, and throat guy and he put the camera down to see if there was anything, you know, stopping me talking or, you know? And there was nothing down there but he saw my tongue wobbling, 'fasciculation' they call it, but it tremors and so he sent me to Mr... Oh, what's his name? [Mr] – [doctor1] – and then I went into hospital for four days and he said I had that, you know? So - But it had been coming on for, obviously now looking back, for twelve months prior, December prior. But, I still had all these things that, they weren't normal things that were cured. Well, ok, I got a nasal spray to cure this but there was nothing any of them came up with to until I went in hospital and then he spotted it. And that was it then! I got the diagnosis.

And when you were finally given that diagnosis, what kinds of thoughts and feelings did you have at that time?

I thought 'How long have I got?' Really, that's it, 'Crikey!' You know, 'How long?' Because you hear people with different time scales and I was, well, my daughter was with me and it's the shock I think, you don't think that much. You sort of - he's telling you and I had an idea it would be something like that - it wasn't anything normal, you know? It's just a case of 'Oh golly, how long have I got now' that's it really. It's 'Oh gosh'. Especially when [doctor1] said to me... I was going to New York with my daughter, she'd paid for me to go for Christmas - before anything - and we were going in January and this was in August, so it was only four or five months, and I said to him 'Well, will I be able to go?' You know, with like the plane and the pressure and that, but [doctor2] said that wasn't a problem, but *he* [doctor1] said to me 'I'd better go sooner rather than later' [looks shocked] and I mean I thought 'Blimey!' you know? That wasn't very good was it? So we did go, we went in September, but when they say something like that you think 'Blimey, I must only have about six months' and then he said, the tablet I'm on, this riluzole, he said 'But it's not a cure, it gives you an extra three to six months' I mean he's not - and you can put this down - he's not got any empathy really. He's quite sort of, matter of fact, whereas [doctor2] is lovely, totally different. It's just that [doctor1] told you, you know, 'You've had the tests and that's what you've got and I'll see you every three months and test your liver once a month' but they don't even organise that for me. So, I've gone to my own doctor and had it done in between a couple of time, but I thought I was going to be able to... I don't know whether to go to my doctor and say 'Look, can you give me a monthly appointment?' because I want to keep on top of it because if the liver's been damaged, it could... Well, I said 'What happens if it does, if they find it?' and he said 'Stop taking the tablets.' So, I think it is important. So, I've got an appointment on the 29th, so, I'm waiting. I get it done while I'm at the

hospital, you know? I say and my blood tests as well, and well, if they say it's important they should keep a check on it.

Yes, absolutely.

But other than that, I've seen him once, [doctor1], since then and seen a registrar and I don't know who I'll see at [hospital] at the end of this month – the 29th. So, I'll just wait and see then. Ooh I hate that.

So, you don't know who you're going to see them?

To be honest with you, because I saw the registrar I thought 'Oh he [doctor1] can't be that bothered with you'. You feel that if it had been something he wants to deal with, he would see me, but because I saw the registrar, you know? It really isn't very good. You have to deal with so much, you see, there's really a lot to keep on top of and it isn't really, helpful I suppose, that you have to retell your story and... Well, it would, I think, be better to know that these people actually do care and they do want to help and you're not passed around from pillar to post, or whatever. But that's it really, I just wait for appointments.

Yes.

But at least I'm keeping on top of them. *I'm* able to do that but I don't know how others, well... I just wait for appointments and there you go.

Yes, so you're keeping on top of those, which is good.

Yes.

Ok, so, if I take you back to the time that you were first diagnosed, how much did you actually know about the illness at *that* time?

Well, I knew that it was terminal and I knew that it was a deterioration of various parts of your body, so I knew that much about it, but other than that... Oh no, a friend's brother-in-law had it and he went very quickly. He was in a wheelchair quite quickly and he lost his speech quite quickly. So, I got the information from the MND [Association] and it's very, well, it depends on the actual – I don't know how to put it – there's three different kinds of it, but, it's only a small word but I can't remember. It's affected my chest muscles and my tongue as well, and buttons, I can't do buttons or pull zips up. I can do this [holds pen] and I can write but sewing, no.

The finer movements are difficult?

Yes, yes, you can't - there's just no pressure, no strength, and that's the first thing that I really noticed. So, at times, well, I don't know a lot about it but I just know that it's terminal but you don't know when. It depends on your deterioration and that's about it really. I don't think you can really pre-empt it, you know, what's going to happen? You think 'Ok, well, I'll just wait and see'. So that's been my attitude. I'll just keep going.

And when you got the diagnosis, did you want to get more information about the illness? Did you go off then and look for more information?

I didn't really, because I just accepted it. Yeah, ok, I've got that now but at the time I thought 'I'm not going to go into...' I'm quite positive you see and so I just try and find ways around doing things, take it easy, you know, don't do this and... I just really thought, well, adjust, basically, adjust. Eating and swallowing: I'm alright eating, drinking sometimes is a bit difficult, in the sense that I've got to time it and if I don't time it right, it goes down the wrong way, and, well, I just time it. But no, I didn't bother, but my daughter did, I think she went on the internet, but I didn't, I thought, well, I think you can get depressed if you learn too much. I knew enough about it to know that it was serious and there's no cure and blah, blah, blah. I knew enough; I didn't want to go into all the details that the internet can give you, it's depressing I think.

There's a lot of information there?

Well, yeah, and some of it you think 'Oh God!' you know? I'd just rather wait and see. I know I'll deteriorate – but in saying that, in the magazine that you get from the [MNDA] there are people

who have various - some people have seven years and there's a chap in the magazine from Australia and he's had twenty-six years and they've give him three, you know, you just don't know so what's the point of going into it? Just take it and deal with it as it comes. That's what I thought, just deal with it. That's it really.

And has that been helpful for you, that strategy?

Oh yes. I think I would be a nervous wreck if I knew half the things out there! Far better to deal with what you know, what you can see, what's happening to you really. You just don't know so there's no point worrying about it. And maybe it's different because I'm the one with it. But, there you go – it's each to their own, isn't it?

Absolutely. Ok, well, we're talking a little about what you know. If we talk about a more specific area of motor neurone disease, which is the changes that can occur in breathing with the illness, at what stage did you find out that people with motor neurone disease can develop these kinds of problems?

I don't really know actually. I think I only really know about it because of what the specialist has told me is happening to me.

So it was when you were developing those problems yourself?

Yeah, because I was getting out of breath and my speech... People thought I'd had a stroke, a lot of people thought I'd had a stroke, but nobody said anything to me, but I mean I can hear that I'm not talking properly. Some days it's better than others - some days it's *clearer* than others. People just think I've had a stroke, you know, I have to just say. You know, actually, *I* thought I'd had a TIA on one of my visits to the doctor, I thought I had just because of the speech. I thought it was a stroke as well. But, no, I think it's more when the specialist explains what's happening, you know, that's about it really. One thing after another!

How did you feel when you were told about these kinds of changes?

Oh, just not looking forward to it, thinking 'Oh god', that's it really. Having to be fed and different tubes and machines for breathing... Oh my god. But it might not come to that, you never know. I mean, this guy, as I say, in the magazine, he lost his voice and he's in a chair but he can still eat, smoke, and drink, you know? So, you know, the voice bit I'm not bothered about, if I'm feeling ok, you know? And if the voice goes, I suppose, it goes and that's it. Not much you can do. I mean, my idea is that there's not much I can do about it, 'cause as they say, there's no cure for it, so it's just waiting for god really, isn't it? I mean, yes, I take on board what they're saying and you must be sensible, but really, oh I dread it really. But, as I say, I'm [pause]... Well, what can you do? It's out of my hands.

Yes. Right, well if we stay just talking about your breathing for a moment, what are the main changes that you noticed in your breathing?

Just getting out of breath, basically, walking up a slope here [her home being on a hill], you know, or if you walk too far. I don't find a breathing problem just sitting like this now - I don't find a problem in *breathing*.

So, it's just during these activities – if you exert yourself?

Just during activities, yeah, that's it. So I just don't do things [laughs]. A bit restricting.

So do you feel it has restricted you a bit and the things you can do?

Oh yes, yes. Well, I could drive until up to last year. My partner and I used to do real rock 'n roll driving, you know? We were good, but I've had to stop driving because I know, well, I did notice a little bit probably about, well, before I went to hospital, I thought I didn't have as much 'go' in me. I still did it and I still do it occasionally. I go if it's not too fast, but I don't feel like it, I'm like 'Oh, what's the point?' you know? But, that was the first sign that it sort of [sighs] takes it out of you. I mean we did twirling round and all sorts and I was doing alright up until last year.

Yeah...

But I think once I knew I had it, it altered my thinking and I sort of thought 'No point doing that because it'll get me tired' and like the physio said 'If you're going out at night, have a rest in the day'

and sometimes I have a sleep, you know? I just pace myself. I'm pacing myself. I mean I'm seventy anyway! I don't need to go charging round, I've done it all [laughs]. But I'm not bothered, you know, it doesn't depress me. I just think, you know, I used to make cards, decorate goose eggs, but I can't do it as much anymore, you know, because I haven't got the dexterity and the feeling, you know? But other than that, I haven't got any problems. I don't think about it – I'm fine, really, well, when I say I don't think about it, I don't *dwell* on it. I do have the odd nap, yeah, I can sleep a bit during the day. I can sleep for an hour, you know, I might have a rest and I'll sit there sort of resting, you know, not budging myself. I don't do energetic stuff anymore unless I can't help it; unless I have to. So I just pace it, no point in pushing yourself. You know, what's the point?

Yeah, and -

Oh, has your tea gone cold?

Oh no, don't worry about that, it's fine. I'd forgotten it was there...

It's wet and warm as my mother used to say.

Absolutely, no it's fine, thank you.

Oh good.

Ok, well, if we move now to talk about the issue of the ventilator, which you did mention to me earlier. Could you tell me about how you came to find out about that?

It was [doctor2] he told me about it and that. It was the first time I'd seen him; I've only seen him once and he bought it up then.

Ok, it wasn't something that had been mentioned earlier – with [doctor1] perhaps?

No, no he just... [Pulls a face]

No? Oh ok...

Oh, he just, well, how do I say? He's just so matter of fact about things, I mean, I told him that I had cramps and I was getting all sorts of pain in my legs and he just went 'Oh, well, you will do.' You know, there was no 'Well, why don't you do this or do that' you know? Just 'Oh, well, you will do'. So matter of fact - cold really. I'm really not very keen on him to be honest – don't put that in your report! [Laughs]

Oh, don't you worry, it's all completely anonymous, don't worry about that at all...

Good! So, no it was just [doctor2].

And you much prefer the way he approaches things?

Oh yes. It's serious stuff, you know, give me some credit. [Doctor2], you know, a lovely man - took the time really, so, yeah, it was [doctor2] who talked to me about that. You just want someone to at least act like they care! I'll listen then... [Laughs] Well, these things that I'm dealing with here – in this – they're serious matters and you want someone to sit with you as a person and explain to you and show that they understand that it's not just a machine or whatever, it's my life here and these are, well, significant matters, you know? But [doctor2] he, I felt, he did take some time to talk about the ventilating machine. So, yes...

Good. Ok, so you talked to [doctor2] about it. How did feel about that, as an initial reaction?

Well, I guess he just wanted to let me know about this new thing that's giving people an extra year of life, he said. He said, and, 'It's non-invasive' he said, and 'It gives you even more time than you initially might have had'. And he just wanted to... Well, he got the nurse, his head physio girl I think, to show me the mask and explained it to me, so that's how I knew about it.

And how did you feel about that?

I actually said 'Wow, well, if that's on your bedside...' but of course the cord was only like that [demonstrates short distance with hands] I said 'If you turn over, it'll be in bed with you!' [Laughs] But she said, you know 'It'll be a longer pipe'. And, I just said 'Well, if I'll need it then I'll need it', you know? Not much I could say about it. But, it did look horrible though; the straps to keep it on your head - like the old fashioned gasmasks, you know? All those big straps! The actual thing itself was quite neat, but it's all this... I mean, taking it to bed with you? I won't be able to tap off with anyone! [Laughs] Not if you've got to wear that, will you? [Laughs] Anyway, that's' all I know about it. I don't think about – why should I think about it now? I'll think about it when I need to think about. No use in worrying yourself, so, back of the mind with it! But I gave it a go, you know, in the clinic like?

And, when you tried that, how was it?

Well, she just put it over me to show me where it went. I just thought 'Well, if I've got to have that kind of thing, I have to have it' but I wasn't sort of overjoyed about having something on your face, because that's just not a pleasant thought. You know, it's not a pleasant thought, thinking I've got to have... You know, 'cause you see people who are on oxygen all of the time – mind you, they have that little pipe, which is not too bad, but even that I think 'If you've got to have it, you've got to have it'. To me, that's it, you know? Not really a choice is it – you wouldn't *choose* it? But if that's what they say, that's what you have. But, I'll put it off for as long as possible.

Looking forward, it sounds like you don't feel you'd be *happy* using it when that time comes...

No, no. And I don't think about it; because I don't have to at the moment, so there's no point in thinking about it. That's it really; I just keep thinking 'No, I'm alright at the moment'.

And how did you feel after that meeting with [doctor2], did you feel that you were given all of the information that you wanted?

Yeah, yeah, he explained, I mean I said to him – oh another thing that happened - I keep remembering what happened, sorry!

No, that's fine, go ahead.

Well, it was my breathing at night, I used to wake up and I used to put my head out of the window, you know? To get air in, you know? I was like 'Oh!' I was thinking 'I can't get enough air' and I told him about that and I said 'I'm sleeping with the window open to make sure. And he said 'Oh you don't need to do that' he said 'There's enough oxygen in the air, you don't have to sleep with your window open' [laughs] But, that was another symptom – I keep remembering different things that I've had. But, no, that was, well, I don't do that now. That's levelled out or whatever, but that must have been an indication, so...

And so you're sleeping better now, not waking in the night anymore?

No, I go for a wee and then go back to sleep again [laughs], I seem to be, at the moment, ok. But I mentioned it to him, you know, just that was one thing. So, yeah, I did feel better after seeing him about that. But, maybe the machine was like, well, as I say, I know now but I don't need to think about it at the moment.

Ok, well if I ask you about your thoughts on the future and about using the machine here at home. What are your thoughts there now?

Well, if I had to, yeah. Obviously, if it's going to make me ok, you know, not have a problem during the night, than obviously, yeah I'd have to have it. I mean really, you know, there's no question. There's no point in distressing yourself; waking up not breathing or not waking up. So, if it was *necessary* I probably would.

And what are your thoughts about monitoring how your breathing is by going and having the respiratory tests and the overnight tests?

Well, I don't like the respiratory tests. Well, they're all ok apart from one where you have to blow, well, the one for home where you blow into it and the needle goes up, I can't do them. I cannot do them. [Doctor1] tried on the first appointment I had at the hospital and he gave up in the end

because I tried ten times. I go 'huh' and I make a noise but it's just a noise. The others weren't too bad actually. [Doctor2] said my results were quite good, you know, when I went in for two hours. Apart from that first one that they gave up on, and even that one they gave up on, but the others were all ok. I don't know why I can't do that one but I've got one at home where I go to like 150, you know? But that's when I'm left alone, and when the doctor showed me what to do, it went up to 600 for him. You know? [Laughs] But that's basically the one, the rest of them are alright, pinch my nose and like 'Keep going! Keep going!' Have you ever tried to keep going when you've no breath 'left? They're not easy! And after two hours of doing that when you have a problem breathing, I mean, come on, you know? I just think the tests are a bit much, I do really. So, I don't know if I'm having it or when, another one of those, but I didn't like doing that. I mean, I don't mind them monitoring it or whatever, but, I feel that I know when about my breathing.

You think it would be better to rely on your own judgement...?

Well, yes! [Laughs] I just think they should consider my illness, you know, making me do all these tests and what not. It's just so tiring. You think, there must be another way? So anyway, you do what you have to, but there's nothing to say I have to be happy about it [laughs].

[Patient appeared to be becoming increasingly tired, speech becoming more slurred over time, gets harder to understand].

Ok, well, I think that's all of my initial questions asked now – I'm getting a feel for what your views on the ventilator are at this stage - but do you have anything else that you'd like to say now?

No, no, everything's ok. Well, I've nothing to compare with so, as far as I'm aware I was seen very quickly and it was very, very thorough in the hospital. It was absolutely, you know, everything was done so I'm quite happy basically with what they've been doing. You know, I'm... Well, I can keep going you know what I mean, I can drive still, I can drive, I can still talk, so at the moment...

So you feel at the moment that – in terms of your breathing - you don't need any help or support at the moment?

Yes, I'm happy not to use it at the moment. I want to put it off for as long as possible. It's a shame really, but, I don't know, I think what one would like to know why it happens. Why's it gone wrong? I know it's something to do with these cells, the muscles they support, you know the jargon but 'Why me?' you know, 'What did I do?' I never smoked, I never drank, I didn't take drugs. You think, 'What went wrong?' You know? I take vitamins; I've taken vitamins for donkey's years, you know, so you think 'Well, what can you do?' It's been a reasonably healthy lifestyle, but if they could tell you why it happens, that would be interesting. If they knew *that*, they could probably find a cure then. You know, they'd be able to work on something. It would be nice to find out. In fact, what did I read recently? I think it might have been in that Reader's Digest, it was about Alzheimer's, it's something to do with, was it stem cells? Oh, erm... Well, I've given it to my daughter, I meant to keep it out, because it's something that they're looking at in Alzheimer's and they reckon it's a similar type - it's something gone wrong in the brain.

So you do like to get some information – on the research? Anything on the ventilators and other such things, or just...

Oh no, just the nice things [laughs] – the research to cure me! Yes, I wish I had that article now... But the stem cells, or was it... Oh, I'll have to get it, but I think if they keep working on that, that'll be good. Like that other magazine you get, you know? And if they found the cause, that would be good wouldn't it?

It would, yes, it would.

It's something to do with the cell and the support – something round the cell that's going wrong. That's what I read recently. I re-read the symposium with all the people, you know, the big doctors and professors and their research. And, I hope they do find something. Well, yeah... [pause] Oh never mind! Well, I guess they might find something before - in the next year or two, while I'm still here. But hey, you get what you're given. But yeah, I've no other questions I don't think, there's nothing I *need* to know. All I know is, I'll take it easy and I'll do my best.

Yes...

I'm going to these motor-cise beds.

Oh right...?

I do them because I feel that that's my muscles working without me having to do it, you know? If you go the gym you're knackered, you know what I mean?

Yes...

And you don't need to, you go for half an hour and all these machines are working all your muscles. So, I try that, you know? Because if you don't it's like, it atrophies doesn't it, if your muscles don't work. So I'm giving them something to do, keeping them all going, you know? And that's the kind of thing – it's all you can do. So there you go. So I'm looking after myself [laughs].

Yes, good! I'm very glad to hear it... [pause – patient clearly tired]. Ok, well unless there's anything else you'd like to say, I can stop the recorder there, I know we've been talking for some time now, but is there anything else that you would like to add or clarify?

No, no, I'm very happy.

Great, ok, well we'll stop there then. Thank you very much.

[END]

Example of Data Analysis Process

Emergent Themes	Original Transcript	Initial Thoughts & Exploratory Comments
	And how did you feel about that [see the ventilator in clinic]?	
Negative Initial Response Use of Humour/Defence Anxiety **Reluctance** **Restricted Choice** **NIV as Necessary** **Negative Reaction to Mask** **Avoiding NIV (Thoughts)** **Disengaging**	I actually said 'Wow, well, if that's on your bedside…' but of course the cord was only like that [demonstrates short distance with hands] I said 'If you turn over, it'll be in bed with you!' [Laughs] But she said, you know 'It'll be a longer pipe'. And, I just said 'Well, if I'll need it then I'll need it', you know? Not much I could say about it. But, it did look horrible though; the straps to keep it on your head – like the old fashioned gasmasks, you know? All those big straps! The actual thing itself was quite neat, but it's all this… I mean, taking it to bed with you? I won't be able to tap off with anyone! [Laughs] Not if you've got to wear that, will you? [Laughs] Anyway, that's all I know about it. I don't think about – why should I think about it now? I'll think about it when I need to think about. No use in worrying yourself, so, back of the mind with it! But I gave it a go, you know, in the clinic like?	Surprise at physical dimensions of machine, use of humour to diffuse tensions? Apprehension, attempts to think about what it might be like to use the ventilator in her home. Already resigned to use, immediate response – if 'needed', not much she can do to change the situation. Restricted choice – not voicing own views, NIV as something necessary. Negative emotional response to the physical qualities of the mask. Mask as distinct from rest of the machine, different emotional responses. Mask as barrier to intimacy ("tap off": kiss) Dismissive, changing the subject. Does not want to think about it, avoidance? Avoiding the subject as a means of avoiding distress. It is a worry to her – if she thinks about it, she will worry. "Back of the mind" a purposeful avoidance strategy. Still willing to try it despite reluctance, a show of compliance in clinics despite reservations.
	And, when you tried that, how was it?	
Restricted Choice **NIV as Necessary** **Negative Reaction to Mask** **Self-Reassurance** **Avoiding NIV (Thoughts)** **Avoiding NIV (Use)**	Well, she just put it over me to show me where it went. I just thought 'Well, if I've got to have that kind of thing, I have to have it' but I wasn't sort of overjoyed about having something on your face, because that's just not a pleasant thought. You know, it's not a pleasant thought, thinking I've got to have -You know, 'cause you see people who are on oxygen all of the time – mind you, they have that little pipe, which is not too bad, but even that I think 'If you've got to have it, you've got to have it'. To me, that's it, you know? Not really a choice is it – you wouldn't *choose* it? But if that's what they say, that's what you have. But, I'll put it off for as long as possible.	Resigned to its use, not presented as a decision to be made but an inevitability that if needed it will be used. Negative emotional reaction to mask. Anything on the face as unpleasant, even the thought of it creates a negative response; linked to her decision to not think about because of worry? Comparison to other patients and treatments. Reinforcing the necessity of the treatment; is she trying to convince herself of this? Is this a prepared mantra that is used to quell negative responses? She thinks these things to herself to try to disengage from potential distress. Not a choice – NIV not something you would choose – it is a need not a want. It's what "they say" that matters, you have it if "they" tell you that you need it. Not in her hands. Impotent? Wanting to delay NIV, reflects negative attitudes, suggests purposeful attempts to avoid. Patient attitude as reluctant and resistant to treatment. Keeps repeating 'If you've got to have it…' – a mantra to reassure? Does this help her to keep a distance, alleviate anxiety?
	Looking forward, it sounds like you don't feel you'd be *happy* using it when that time comes?	
Avoiding NIV (Thoughts)	No, no. And I don't think about it; because I don't have to at the moment, so there's no point in thinking about it. That's it really; I just keep thinking 'No, I'm alright at the moment'.	Avoidance, even of thinking about it – because it will cause her distress. Not something that she is currently thinking about. "No point" thinking about it suggests only think about what you need to, prioritising, what is useful to be thinking about and what is just harmful and pointless. Yet, she does think about being alright and not needing NIV, so maybe it is on her mind even though she

Avoiding NIV (Use)	**And how did you feel after that meeting with [doctor2], did you feel that you were given all of the information that you wanted?**	tries not to think about? The need for this strategy of self-reassurance is interesting. A necessary strategy for reducing anxiety? A response to high anxiety?
	Yeah, yeah, he explained, I mean I said to him – oh another thing that happened – I keep remembering what happened, sorry!	
	No, that's fine, go ahead!	
Respiratory Symptoms **Symptom Distress** **Clinician Reassurance** **Dismissing Change**	Well, it was my breathing at night, I used to wake up and I used to put my head out of the window, you know? To get air in, you know? I was like 'Oh!' I was thinking 'I can't get enough air' and I told him about that and I said 'I'm sleeping with the window open to make sure. And he said 'Oh you don't need to do that' he said 'There's enough oxygen in the air, you don't have to sleep with your window open' [laughs] But, that was another symptom – I keep remembering different things that I've had. But, no, that was, well, I don't do that now. That's levelled out or whatever.	Clear signs of respiratory insufficiency – air hunger, yet had previously stated no change in respiratory function and no need for NIV. Contradiction? Taking action to try to address respiratory distress – showed emotional response and physical discomfort. Panic – not enough air? Feeling not enough air – overt respiratory impairment. Reassured by doctors – misleading her perceptions? Could latterly laugh at opening window to get more air, but at the time this was probably quite a serious concern. The fact that this symptom had gone seemed to influence her view that she did not have a respiratory problem. There have been other problems, but she judges her status on the immediate situation and not what has gone before. Phasic nature to change and symptoms. Can now be dismissed.
	And so you're sleeping better now, not waking in the night anymore?	
Clinician Reassurance **Avoiding NIV (Thoughts)**	No, I go for a wee and then go back to sleep again [laughs], I seem to be, at the moment, ok. But I mentioned it to him, you know, just that was one thing. So, yeah, I did feel better after seeing him about that. But, maybe the machine was like, well, as I say, I know now but I don't need to think about it at the moment.	Reassured by doctors, helped her to dismiss the signs of respiratory impairment? Despite change in oximetry that suggested she did need ventilation, she felt she did not, casual attitude of doctor used to confirm her own views and justify her avoidance. This comfort encouraged her to sustain her avoidance of NIV? Again stressing no *need* to think about it yet. Not prepared for NIV only weeks later? Concept of readiness important here, she does not perceive the need to be considering NIV and seems to find the subject itself distressing.

This table offers an example of how raw transcripts (middle column) were initially analysed, with initial ideas being noted down as an immediate reaction to the data (right hand column). This then led to ideas being encapsulated in the emerging themes (left hand column). See 'Reading, Re-reading & Note-Making' (pg. 96).

Theme Construction

Example of emergent themes leading to the development of superordinate themes and subthemes through the process of abstraction described in Section 4.3 (pg. 97).

Immediate Responses to Ventilation	
Superordinate: Treatment Apprehension	Superordinate: Treatment Apprehension
Subtheme: Ventilator as Defeat Resistance Fear Dismissive Fighting Need/Initiation Struggle Preconceptions Body Giving Up Shutting Down Physical Weakness Negative Implications Putting it Off	**Subtheme: Fear of Dependence** Anxiety Risk/Threat Resistance Relying on a Machine Loss of Independence Progression of Need Succumbing to the Illness Being Stuck/Trapped Restriction Permanence Constance
Superordinate: Apprehension Treatment	Superordinate: Apprehension Treatment
Subtheme: Mask: Fearful Presence Scary/Frightening Foreboding/Predicted Distress Claustrophobia Presence Discomfort (Physical) Discomfort (Psychological) Importance of the Face Universal Understanding of Fear	**Subtheme: Mask: Threat to Identity** Negative Appearance of Mask Immediate Reactions Challenge to Self-Concepts Incongruence with Self-Image Reactions in Others, Fear Becoming Scary Barrier to Intimacy Restriction
Superordinate: Relief/Help	Superordinate: The Nature of Choice
Subtheme: Ventilator as Help Hope Good Idea Pragmatic Solution, Problems Solving Relief Comforting Help as Intrinsically 'Good' Gratitude Reassuring Positivity Safety	**Subtheme: 'No Choice'** Essential/Necessary Reluctance/Resistance Obligation Defensive Responses Fear of Consequences Fear of the Unknown Stifled Voice Loss of Control/Autonomy Helplessness/Hopelessness Anxiety Submission/Passivity

Appendix 3

Information Sheet: Cross-Sectional Study

The Walton Centre for Neurology and Neurosurgery NHS Trust

Aintree University Hospitals NHS Foundation Trust

Patient Information Sheet

'Health benefits of Non-Invasive Ventilation in MND: the Psychosocial and Physical Impact on People with MND and their Carers'

Thank you for your interest in our research. This leaflet will explain the background behind our research, why we are doing this research, and how you would be involved in the research. Please read the information carefully. If you are in any way unsure or have any questions, please do not hesitate to ask.

Research Background

Motor Neurone Disease (MND) is a condition that leads to progressive muscle weakness in different areas of the body. Depending on which muscles are affected, MND can lead to a range of different symptoms, such as loss of mobility in the limbs or difficulty with speech and swallowing. Where muscle weakness progresses to the muscles of the chest, breathing problems may develop which may lead to greater breathlessness, more chest infections, and a weaker cough. It is important to identify when patients develop breathing problems in MND. Nowadays, these problems are often treated with a machine to help breathing (a ventilator). This machine is connected to a well-fitted facemask that delivers air to the patient at high pressure, enabling them to breathe more comfortably. We call this 'Non-Invasive Ventilation' and we now know that this can benefit people who have MND.

What is the research about?

We wish to understand what our patients (and those who care for them) think about Non-Invasive Ventilation. We would like to understand both the positive and negative aspects of life with Non-Invasive Ventilation for patients and carers. We hope that by understanding the attitudes of our patients and their carers towards Non-Invasive Ventilation, we can improve the service that we deliver to our patients. We also aim to see how opinions and attitudes towards Non-Invasive Ventilation relate to the quality of life of our patients and carers and to how patients' lungs may be working. We are interested in your views and experiences with Non-Invasive Ventilation.

Do I have to take part?

No. It is entirely up to you to decide whether you would like to be involved in our research. There is no penalty for not taking part and if you do agree to take part you can still withdraw at any time and do not need to give a reason for doing so. Taking part is voluntary and you will receive no payment. However, if you do wish to join the study any costs incurred will be met by the researchers.

What does my participation involve?

If you do agree to take part in the study, you will first be requested to sign a consent form. After this you will be asked to take part in an informal interview. On the day of the interview you will be invited to come to a

location at University Hospital Aintree. However, we would be happy to visit you at home if you would prefer this. You have the right to choose where you would like the interview to take place (hospital or home). We will arrange all transport and will pay for any expenses incurred as a result of your involvement in the research.

In the interview we will ask you about your views, opinions and experiences of MND and Non-Invasive Ventilation. This interview will be recorded on a tape recorder but your name and personal details will be kept anonymous. The interview itself will be conducted by one researcher, who will ask you around 10-12 questions. You are free to say whatever you like, as much or as little as you like, or not to answer questions if you do not feel comfortable with them.

If you choose to be interviewed at the hospital we will arrange transport to take you home (covering all costs incurred). You will be invited to take as many breaks as you require and refreshments will be available at all times for yourself and those who accompany you.

Will my treatment be affected by my participation?

No, not in anyway. Whether you choose to take part or not, your care in hospital will not be affected now or at any time in the future.

Will a doctor be available during the visit?

Yes. Two of the senior researchers are medical doctors, one of whom will always be available at the hospital if required for consultation. A doctor will not be available if you are visited at home.

Will my taking part be kept confidential?

Yes. Your participation is strictly confidential. If you consent to take part in the research, only the named researchers will have access to your medical records. Your name will not be disclosed to anyone outside of the research team in the hospital nor will it be possible to identify you in any way in any written reports. All information will be stored safely and securely.

What are the benefits of taking part?

This research will not directly influence the care and services that you will receive, however, we hope that the information that you give to us will help us to understand MND and Non-Invasive Ventilation better and to improve services for patients and their families in the future. We also hope that being able to talk to us about your experiences may be useful to you.

What are the risks of taking part?

We aim to understand your views on the impact of breathing difficulties in MND and the treatments we offer. We fully understand and appreciate that discussing certain things related to this may be sensitive or upsetting. We would like to reassure you that you do not have to answer any question or talk about anything that you do not feel comfortable with. We would also stress that medical and nursing support will be available throughout. If you feel that you need extra help or advice, we can give you information about support services that are available to you.

Can I withdraw from the study?

Yes. You are free to withdraw from the study at any time and do not need to provide any reason for doing so. This will not affect your treatment in any way, now or in the future.

Who is doing the study?

The senior researchers are doctors from the Respiratory Department at University Hospital Aintree (Dr [redacted]), the Neurology Department (Professor [redacted]) at the Walton Centre, and from the Psychology Department at the University of Liverpool (Dr [redacted]). Interviews will be conducted by a trained researcher (Lucy Piggin).

Who has reviewed the study?

This study has been favourably reviewed by Liverpool Paediatric Research Ethics Committee.

What if something goes wrong?

If you have any questions or concerns at any stage of your involvement in this research project, please feel free to discuss these with the research team. We will do our best to resolve any problems quickly. If you are still unhappy and wish to complain about any aspect of the way you have been approached or treated during the course of this study, the normal National Health Service (NHS) complaints mechanisms are available to you. Further details and advice can be found on the NHS website www.nhs.uk or by phoning NHS Direct on 0845 4647. You may also discuss the complaints process with the Patient Advice and Liaison Service (PALS) at the Walton centre by calling 0151 529 6100.

We do not foresee any risk of harm to you during this study. However, if you are harmed in any way by taking part in this research project you should be aware that there are no special compensation arrangements. If you are harmed due to someone else's negligence, you may have grounds for legal action but you may have to pay for this yourself.

Where can I get more information from?

If you have any further questions or would like some more information, please feel free to contact Lucy Piggin by telephone (07[redacted]5) or by email ([redacted]liv.ac.uk). Lucy will be happy to discuss any concerns and to answer your questions.

Thank you for taking the time to read this information sheet.

If you agree to take part, you will be given a copy of this document and a signed consent form to keep. If you have any questions please feel free to contact Lucy.

Information Sheet: Longitudinal Study

The Walton Centre NHS for Neurology and Neurosurgery NHS Trust

Aintree University Hospitals NHS NHS Foundation Trust

Patient Information Sheet

'Health benefits of Non-Invasive Ventilation in MND: the Psychosocial and Physical Impact on People with MND and their Carers'

Thank you for your interest in our research. This leaflet will explain the background behind our research, why we are doing this research, and how you would be involved in the research. Please read the information carefully. If you are in any way unsure or have any questions, please do not hesitate to ask.

Research Background

Motor Neurone Disease (MND) is a condition that leads to progressive muscle weakness in different areas of the body. Depending on which muscles are affected, MND can lead to a range of different symptoms, such as loss of mobility in the limbs or difficulty with speech and swallowing. Where muscle weakness progresses to the muscles of the chest, breathing problems may develop which may lead to greater breathlessness, more chest infections, and a weaker cough. It is important to identify when patients develop breathing problems in MND. Nowadays, these problems are often treated with a machine to help breathing (a ventilator). This machine is connected to a well-fitted facemask that delivers air to the patient at high pressure, enabling them to breathe more comfortably. We call this 'Non-Invasive Ventilation' and we now know that this can benefit people who have MND.

What is the research about?

We wish to understand what our patients (and those who care for them) think about Non-Invasive Ventilation. We would like to understand both the positive and negative aspects of life with Non-Invasive Ventilation for patients and carers. We hope that by understanding the attitudes of our patients and their carers towards Non-Invasive Ventilation, we can improve the service that we deliver to our patients. We also aim to see how opinions and attitudes towards Non-Invasive Ventilation relate to the quality of life of our patients and carers and to how patients' lungs may be working. We are interested in your views and experiences with Non-Invasive Ventilation.

Do I have to take part?

No. It is entirely up to you to decide whether you would like to be involved in our research. There is no penalty for not taking part and if you do agree to take part you can still withdraw at any time and do not need to give a reason for doing so. Taking part is voluntary and you will receive no payment. However, if you do wish to join the study any costs incurred will be met by the researchers.

What does my participation involve?

If you do agree to take part in the study, you will first be requested to sign a consent form.

After this, you will be asked to take part in a series of interviews over time, with a maximum of four interviews per year for a period of up to three years (or until you no longer wish to take part in the study). These interviews are separate from your clinic appointments and are in addition to your routine visits to the hospital.

However, if you would prefer to combine some interviews with clinic appointments (so that that they happen on the same day) we may be able to arrange this for you.

On the day of each interview you will be invited to come to a location at University Hospital Aintree. However, we would be happy to visit you at home if you would prefer this. You have the right to choose where you would like the interview to take place (hospital or home). We will arrange all transport and will pay for any expenses incurred as a result of your involvement in the research.

In the interview we will ask you about your views, opinions and experiences of MND, respiratory aspects of the illness and non-invasive ventilation. Each interview will be recorded on a tape recorder but your name and personal details will be kept anonymous. Interviews itself will be conducted by one researcher, who will ask you around 10-12 questions. You are free to say whatever you like, as much or as little as you like, or not to answer questions if you do not feel comfortable with them.

If you choose to be interviewed at the hospital we will arrange transport to take you home (covering all costs incurred). You will be invited to take as many breaks as you require and refreshments will be available at all times for yourself and those who accompany you.

Will my treatment be affected by my participation?

No, not in anyway. Whether you choose to take part or not, your care in hospital will not be affected now or at any time in the future.

Will a doctor be available during the visit?

Yes. Two of the senior researchers are medical doctors, one of whom will always be available if required. If we visit you at home, a trained medical or healthcare professional will always accompany the researcher.

Will my taking part be kept confidential?

Yes. Your participation is strictly confidential. If you consent to take part in the research, only the named researchers will have access to your medical records. Your name will not be disclosed to anyone outside of the research team in the hospital nor will it be possible to identify you in any way in any written reports. All information will be stored safely and securely.

What are the benefits of taking part?

This research will not directly influence the care and services that you will receive, however, we hope that the information that you give to us will help us to understand MND and Non-Invasive Ventilation better and to improve services for patients and their families in the future. We also hope that being able to talk to us about your experiences may be useful to you.

What are the risks of taking part?

We aim to understand your views on the impact of breathing difficulties in MND and the treatments we offer. We fully understand and appreciate that discussing certain things related to this may be sensitive or upsetting. We would like to reassure you that you do not have to answer any question or talk about anything that you do not feel comfortable with. We would also stress that medical and nursing support will be available throughout. If you feel that you need extra help or advice, we can give you information about support services that are available to you.

Can I withdraw from the study?

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Who is doing the study?

The senior researchers are doctors from the Respiratory Department at University Hospital Aintree (Dr [redacted]), the Neurology Department (Professor [redacted]) at the Walton Centre, and from the Psychology Department at the University of Liverpool (Dr [redacted]). Interviews will be conducted by a trained researcher (Lucy Piggin).

Who has reviewed the study?

This study has been favourably reviewed by Liverpool Paediatric Research Ethics Committee.

What if something goes wrong?

If you have any questions or concerns at any stage of your involvement in this research project, please feel free to discuss these with the research team. We will do our best to resolve any problems quickly. If you are still unhappy and wish to complain about any aspect of the way you have been approached or treated during the course of this study, the normal National Health Service (NHS) complaints mechanisms are available to you. Further details and advice can be found on the NHS website www.nhs.uk or by phoning NHS Direct on 0845 4647. You may also discuss the complaints process with the Patient Advice and Liaison Service (PALS) at the Walton centre by calling 0151 529 6100.

We do not foresee any risk of harm to you during this study. However, if you are harmed in any way by taking part in this research project you should be aware that there are no special compensation arrangements. If you are harmed due to someone else's negligence, you may have grounds for legal action but you may have to pay for this yourself.

Where can I get more information from?

If you have any further questions or would like some more information, please feel free to contact Lucy Piggin by telephone ([redacted]) or by email ([redacted]). Lucy will be happy to discuss any concerns and to answer your questions.

Thank you for taking the time to read this information sheet.

If you agree to take part, you will be given a copy of this document and a signed consent form to keep. If you have any questions please feel free to contact Lucy.

The Walton Centre NHS for Neurology and Neurosurgery NHS Trust

Aintree University Hospitals NHS NHS Foundation Trust

'Health benefits of Non-Invasive Ventilation in MND: the Psychosocial and Physical Impact on People with MND and their Carers'

Patient Consent Form

(Please read each statement carefully and initial each box)

- I agree to take part in this study. ☐
- I have read and understood the information leaflet for this study and have had the opportunity to ask about anything that I do not understand. ☐
- I understand that I am free to withdraw from the study at any time and this will not affect my future treatment. ☐
- I understand that my medical notes may be looked at by a researcher and I give permission for this. ☐
- I understand that participating in this study involves taking part in interviews that will be tape-recorded. ☐

_____	_____	_____
Name of Patient	Date	Signature
_____	_____	_____
Name of Researcher	Date	Signature

The Walton Centre NHS
for Neurology and Neurosurgery
NHS Trust

Aintree University Hospitals NHS
NHS Foundation Trust

'Health benefits of Non-Invasive Ventilation in MND: the Psychosocial and Physical Impact on People with MND and their Carers'

Carer Consent Form
(Please read each statement carefully and initial each box)

- **I agree to take part in this study.** ☐
- **I have read and understood the information leaflet for this study and have had the opportunity to ask about anything that I do not understand.** ☐
- **I understand that I am free to withdraw from the study at any time and this will not affect the continuation of the study.** ☐
- **I understand that participating in this study involves taking part in interviews that will be tape-recorded.** ☐

Name of Carer	Date	Signature
Name of Researcher	Date	Signature

Appendix 5:

Interview Schedules

It should be noted that the following provides a *guide* to the interview process that was not intended to be prescriptive. Interviews using an IPA approach allow patients and interviewers to pursue their own interesting and important lines of thought over the course of the interview. What is presented here is a general overview of topics for inclusion and not a definitive structure or script. Primary questions appear underlined, with prompts being employed as and when necessary and depending on the nature of patients' responses and degree of engagement.

Contained within Appendix 5 are:

1) The cross-sectional interview schedule for patients (and carers)
2) The longitudinal interview schedule for patients pre-ventilation.
3) The longitudinal interview schedule for patients post-ventilation.
4) The longitudinal interview schedule for carers pre-ventilation.

Pre-Interview Preparation

- Provide an initial expression of thanks for participating in the study.
- Recap on the purpose of the study and what will be involved in the interview, including showing the patient the Dictaphone. Emphasise that the research is interested in their own experiences and there are no right or wrong answers.
- Define the roles: interviewer will ask questions but patients should be encouraged to talk about what they feel is important and if topics that patients feel are relevant are not raised, they should feel free to raise them.
- Provide an assurance of confidentiality and remind patients that they can withdraw at any time.

Post-Interview Closure

- Thank patient for their contribution.
- Assess mood and allow time for general conversation to diffuse any potential distress.
- Provide the opportunity for patients to ask questions and ensure they have contact details if questions arise later.

Cross-Sectional Interview Schedule (Patients/Carers)

Respiratory Aspect of the Illness: Before we start to talk about the ventilator, perhaps we could start off by talking a little bit about respiratory aspects of the illness more generally (clarify 'respiratory' as breathing related). Could you tell me about when you first found out that your breathing might be affected by the MND? Prompts: How was this found out? How much (if any) known before noted changes? What was initial reaction to that news? What were first thoughts? How did you feel about it? Respiratory experiences?

Respiratory Symptoms/Service: Could you tell me about the time that you were told you might need some support or help with your breathing? Prompts: Thoughts and feelings? What problems were you experiencing? How did you feel about them? Meaning? What knowledge did you have about possible interventions? Did you feel prepared? What was the process like for you?

Treatment Options: What do you remember about being told about the options that were available to you? Prompts: Who mentioned what and when? What were thoughts and feelings about that? Do you feel that you were told enough about the help that you could have for your chest and your breathing? How did you feel about these options?

Treatment Information: Could you tell me about the decision to trial a ventilator? Prompts: What was the process of making a decision like for you? Can you think of any other things that would have been useful in helping you to decide what to do about your chest or breathing? Is there any other information that you feel you could have had that would have helped? Is there anything that would have changed your mind? Did anything helpful or unhelpful happen at this time that we should know about? What was important in helping to make decisions?

Ventilator Experiences: Could you tell me a little about your first experience on the ventilator? Prompts: How did you come to see/use the ventilator? When you were shown the ventilator, what were your thoughts then? Could you tell me a little about your experience of using the ventilator – how was it initially? And now? Good and bad points for you?

Carer Experiences: We've talked about *your* experiences and I wonder, how do you think the experience has been for ⌜carer⌝? What do you think their thoughts and feelings are about the ventilator? Good and bad points?

Future: What are your thoughts about moving forward with the ventilator, into the future? Prompts: Now that you know what it's like to use the ventilator, would you go for this treatment option again if you had the choice? If you were to get a concurrent illness (another illness alongside the MND) and your chest became worse (if you were seriously ill), how would you like to be cared for? Would you want to stay on the ventilator? At what point do you feel you may want to come off the ventilator?

Opportunity to add, clarify or raise other issues.

Longitudinal Interview Schedule for Patients (Pre-NIV)

Starter Question: To start with, perhaps you could tell me a little bit about the time that you received your diagnosis? Prompts: Could you tell me more about the process of diagnosis? What symptoms did you have at that time? Where? When? How?

Reaction to Diagnosis/Illness: Could you tell me more about finding out that you had MND; what were your initial thoughts and feelings when you were told? Prompts: What were your first thoughts – what came to mind when they first said that it was MND? How did you feel about the news? If they 'can't remember' – ask what is the first thing they *remember* thinking and feeling at that time? Why did they think/feel that way? What were your main concerns?

Understanding of the Illness: At the time that you were told that it was MND, how much did you actually know about the condition? Prompts: Had you heard of the condition before? Did you feel you had enough information? What effect did the extent of your knowledge have on how you felt at that time? Did you want to find out more about it (or did you feel you had sufficient information)? Why?

Respiratory Aspect of the Illness: If we could move now to talk about a more specific aspect of the condition, and the changes that can occur in your breathing. Could you tell me about when you first found out that your breathing might be affected by the MND? Prompts: Reactions to this news, thoughts feelings?

Experience of Respiratory Change: You've been referred into the respiratory service, how did that come about? Prompts: What was your initial reaction to that news? What were your first thoughts? How did you feel about it? Could you tell me a little bit about how you feel your breathing is at the moment? Feelings about being assessed at the hospital?

Treatment Options: What do you remember about being told about the help that's available to you if you did need help or support with your breathing? Prompts: What options were presented? Thoughts and feelings about these? Do you feel that you were told enough about the help that you could have for your chest and your breathing? How did you feel about these options?

Ventilation: How did you come to find out about the ventilator? What were your first thoughts? How did you feel? What were you actually told? Prompts: Knowledge and understanding of the treatment? And now, how do you feel about the ventilator? What are your thoughts about using the ventilator? If you did go on to use a ventilator, what would you hope it would do for you? Do you think you would benefit from using a ventilator now? Do you feel you know enough to make a decision?

Decision-Making: What are your thoughts and feelings about making a decision about using a ventilator? Prompts: What kind of things are you thinking about there? Are there specific factors that are important to you at this time?

Future: What are your thoughts about moving forward in respect to the illness? What are your thoughts about respiratory aspects of the illness as you look ahead? Are there any specific concerns? General feelings?

Opportunity to add, clarify or raise other issues.

Longitudinal Interview Schedule for Patients (Post-NIV)

Starter Question: So perhaps to start with you could tell me how things have been since I last saw you? Prompts: Physical changes? How do you feel about these? How are you feeling about things as they stand at the moment?

Respiratory Status: [If not already stated] Before we start talking about the ventilator, perhaps you could tell me how things have been in respect to your breathing and that side of things? Prompts: Change? How do you feel about this aspect of things? [If a notably absent] Last time we spoke about your respiratory symptoms and you reported [INSERT], how are things there now? How do you feel about that? What have been the implications of any change in day to day life?

Ventilation: Now, since I last saw you, you've started to use a ventilator; how did that come about? Prompts: What were your first thoughts when you were told you might benefit from NIV? How did you feel at that time? What motivated the move: reasons for trialling the ventilator?

Decision-Making: Could you tell me a bit about the decision for you to start using the ventilator? Prompts: Thoughts and feelings about that time and process? What were you hoping would be the result of that process? Reasons/motivations/aspirations? Did you feel you knew enough to make the decision?

Experiences: Could you tell me about when you first trialled the ventilator, what happened there? Prompts: Thoughts and feelings? How would you describe your first experience of NIV? What's it like to use the ventilator now? Prompts: Thoughts and feelings? Good points and bad points? Has the experience changed? Reasons/motivations/aspirations?

Reflections: As you look back at the decision to start using the ventilator, how did you feel? What are your thoughts on that now? Would you make the same decision again?

Future: What are your thoughts about moving forward in respect to the illness? What are your thoughts about respiratory aspects of the illness as you look ahead? What are your thoughts about using the ventilator as you move forward?

Opportunity to add, clarify or raise other issues.

Longitudinal Interview Schedule for Carers (Pre-NIV)

Starter Question: To start with, perhaps you could tell me a little bit about the time that [patient] received his/her diagnosis? Prompts: Could you tell me more about the process of diagnosis? What symptoms did you have at that time? Where? When? How? Thought/feelings?

Reaction to Diagnosis/Illness: Could you tell me more about finding out that [patient] had MND; what were your initial thoughts and feelings when you were told? Prompts: What were your first thoughts – what came to mind when they first said that it was MND? How did you feel about the news? If they 'can't remember' – ask what is the first thing they *remember* thinking and feeling at that time? Why did they think/feel that way? What were your main concerns?

Understanding of the Illness: And at the time that you were told that it was MND, how much did you actually know about the condition? Prompts: Had you heard of the condition before? Did you feel you had enough information? What effect did the extent of your knowledge have on how you felt at that time? Did you want to find out more about it (or did you feel you had sufficient information)? Why?

Carer Role: Could you tell me about your role, in supporting [patient] with the illness? Prompts: What kind of support is offered and why? How? Thoughts, feelings, coping? Relationship; has this changed? Links between illness response and caring response?

Respiratory Aspect of the Illness: If we could move now to talk about a more specific aspect of the condition, and the changes that can occur in your breathing. Could you tell me about when you first found out that [patient]'s breathing might be affected by the MND? Prompts: Reactions to this news, thoughts feelings?

Experience of Respiratory Change: [Patient]'s been referred into the respiratory service, how did that come about? Prompts: What was your initial reaction to that news? What were your first thoughts? How did you feel about it? Could you tell me a little bit about how you feel his/her breathing is at the moment? Feelings about being assessed at the hospital?

Treatment Options: What do you remember about being told about the help that's available to [patient] if they did need help or support with their breathing? Prompts: What options were presented? Thoughts and feelings about these? Do you feel that you were told enough about the help that you could have for your chest and your breathing? How did you/patient feel about these options?

Ventilation: How did you come to find out about the ventilator? What were your first thoughts? How did you feel? What were you actually told? Prompts: Knowledge and understanding of the treatment? And now, how do you feel about the ventilator? What are your thoughts about [patient] using the ventilator? If [patient] did go on to use a ventilator, what would you hope it would do for him/her? Do you think [patient] would benefit from using a ventilator now? Do you feel you/patient know enough to make a decision?

Decision-Making: What are your thoughts and feelings about making a decision about using a ventilator? Prompts: What kind of things are you thinking about there? Specific factors that are important to you at this time?

Future: What are your thoughts about moving forward in respect to the illness? What are your thoughts about respiratory aspects of the illness as you look ahead? Prompts: Tell me how you feel looking ahead? Are there any specific concerns? General feelings?

Opportunity to add, clarify or raise other issues.

Appendix 6:

Supplementary Analysis from Patients' Pre-Ventilation Interviews

This appendix contains the full thematic analysis from 'The Experience of Diagnosis' (pg 156) and 'Attitudes & Coping Strategies' (pg 161).

The Experience of Diagnosis

'The Experience of Diagnosis' explores patients' early illness experiences; it provides insight into the psychological impact of diagnosis and how lasting attitudes towards the illness began to form at an early stage. Table A6.1 provides a summary of all subsections, superordinate themes and subthemes contained within this section.

Table A6.1

An overview of the subsections, superordinate themes and subthemes contained within 'The Experience of Diagnosis'.

Section	Subsection	Superordinate Theme	Subtheme
The Experience of Diagnosis	Pre-Diagnosis	Uncertainty: Fear & Anxiety	Time & Delay Exclusion: Withheld Information Back & Forth
	The 'Actual' Diagnosis	Affective Impact	Shock & Devastation Relief Delay, Avoid, Deny
		Expectation & Disappointment	Nothing You Can Do Redefining the Future
		Sense-Making: Why Me?	

Pre-diagnosis (Sub-Section)

Patients experiences of the pre-diagnosis phase are encapsulated within the single super-ordinate theme 'Uncertainty: Fear & Anxiety'. Within this superordinate theme there are three subthemes: 'Time & Delay' describes the impact of the time spent awaiting diagnosis and the tendency for patients to interpret this as *avoidable* delay; 'Exclusion: Withheld Information' reflects how patients' perceived the reluctance of healthcare professionals to divulge medical suspicions and the impact that this had on their own ideas about the illness; and 'Back & Forth' discusses the psychological impact of misdiagnoses and the emotional highs and lows that patients endured during this period.

Uncertainty: Fear & Anxiety (Superordinate Theme)

The period immediately prior to diagnosis was characterised by confusion and uncertainty and many patients reported feeling significantly anxious and isolated. This was a time of both knowing and not knowing, as patients had a clear idea that there was "something" wrong but were in the dark as to what exactly this was. Most patients could recollect specific instances or patterns of unexplainable behaviour that sparked concern; P15 recalled:

> He [doctor] couldn't find out what was wrong and this all started. It could have been longer than months, but I was seeing the doctor and just collapsing. I was just collapsing and I didn't know why... I come out the kitchen and 'Bang!' I hit the deck. I didn't know anything about it, and our [son] get's me up and I says 'I don't know, I didn't trip over nothing, it's flat as hell' and then I did the same by the shop – that's not normal that. (P15)

Although doctors were often themselves confused by symptoms and events, patients were clear that things were "not normal", with even simple routines being threatened by an increasingly unreliable and unpredictable body. P13 recalled: "I've always been a quite active person, and when I tripped I couldn't control myself, I just went down. ... I didn't seem to have control" Most patients recounted similar stories; P19 explained:

> At first it was just falling over... I was falling over a lot more and plus I had arthritis in my right knee so I got myself a walking stick to help prevent me from falling over and that did help a bit, but then I was falling over more and more, I mean, I wasn't hurting myself, you know, I was just falling over. But I got worried... so I went to the doctor and he said go to A&E – and I'd been going to my doctor practically every week with this drop foot and he said to me 'Oh go on holiday!' and I won't tell you everything he said, but he didn't know what it was, so he examined my leg and he couldn't think of what it might be; couldn't see anything wrong with it. So I went to another doctor, because I was really, really worried, you know? (P19)

An increasing catalogue of atypical symptoms and experiences heightened confusion and often resulted in spiralling anxiety; indeed, although life "went on" during this period, the illness was a lingering presence that distorted normality and cast a threatening shadow over the future. The fact that doctors could not offer any immediate answers and seemed to share patients' uncertainty only served to exacerbate their concerns.

The uncertainty that dominated this period threatened autonomy and control, leaving patients feeling physically and psychologically vulnerable. Retrospectively, P18 felt that this pre-diagnosis phase had been the hardest of all, explaining:

> ...for a long time that I didn't know what it was, I went through more emotions being really down before the diagnosis than after... The waiting, you know, was worse. And I always look back at that time as being far worse than this, you know, knowing what I've got. It's that fear of the unknown and even though this might be the worst thing it could have been, at least I know... (P18)

It was the "fear of the unknown" that evoked the most significant distress; patients did not understand the situation and so felt unable to accurately appraise their own cognitive and emotional resources for coping with it. This was reflected in P18's assertion that it was better to "know" even if knowledge provided a poor prognosis. Patients appeared to have been desperate to establish certainty; yet without a diagnosis on which to focus, they were left in a state of emotional flux.

Time & Delay (Subtheme)

Most patients focused on the length of time that they had to wait for diagnosis and were keen to emphasise how time interacted with uncertainty to intensify distress; the process was often felt to have been *unnecessarily* protracted and a sense of delay evoked strong negative emotional reactions. Many patients spent months and years waiting for diagnosis; P4 explained:

> Three years it was; three years waiting up in the air. Just up in the air and waiting and not knowing, and just - nobody told me nothing for three years.... they kept saying there was nothing wrong... passing me on to other people. (P4)

It was clear that P4 felt isolated and abandoned during this time. The notion of being "up in the air" exemplified the uncertainty that many patients felt during this period; the diagnostic process often seemed to lack direction and much of life appeared to be put 'on hold' as patients awaited conclusions. Being 'passed on' to different services was also a common occurrence and seemed to intensify the sense of delay; P12 recalled: "Oh I knew very little until I seen [doctor, at diagnosis], to be honest, I seen so many doctors, like, doctor after doctor..." For some patients, this was a clear frustration; they felt that healthcare professionals were not listening and were dismissing their concerns as unimportant. P22 explained:

> ...they sent me with exercises and things – nothing serious, go home. On and on and on – nothing wrong, go home. ...they still weren't even listening to me still. [...] Waiting to know is hard, that was hard. It took a long time. It did take them a very long time to say MND... (P22)

The process of 'waiting' was clearly interpreted as delay; patients often felt that the diagnosis could have been made sooner if they had been listened to or if healthcare professionals had applied themselves with greater commitment to the problem. A number of patients were adamant that diagnosis could and should have been made earlier, leaving them frustrated. P19 was angry that her legitimate concerns had been dismissed by healthcare professionals; she felt that this had both held-up her diagnosis *and* increased distress:

> You know, and I was going all the time to my own doctor, and I felt he was just fobbing me off. Because I'm sure they do have people who just go to their doctor every week and they must just think 'Oh, hypochondriac', you know? [...] ... I felt [pause] very, very frustrated and just so worried. It is frustrating when people don't listen to you. But I thought, with me going

> backwards and forwards to him, I was still falling over and having the drop foot – as I thought – he still should have referred me earlier to the hospital. But he didn't. (P19)

Some patients clearly believed that healthcare professionals had not been efficient or focused enough during this period. Even many months after her diagnosis, P19 reported feeling "...just very angry that I wasn't listened to, because you know your own body, don't you? So, yeah, it does upset me now to think back about that time and that doctor not listening to me." It was not uncommon for patients to retain negative feelings about this time and about medical professionals, even months and years after diagnosis. P19 felt that her concerns were undermined by her doctor; many patients expressed the idea that *they* had a far clearer (physical) insight into problems than doctors and that, as a result, their own views should have taken precedent. This was a belief that also emerged later when respiratory 'problems' were highlighted. During this pre-diagnosis phase, overlooking patients' own insights was seen to increase delay.

The notions of time and delay were also important given the *progressive* nature of symptoms; patients recalled an inclination to dismiss or adapt to early symptoms, yet they still became increasingly anxious over time. There was a temporal nature to uncertainty and anxiety, with patients acutely aware that "...things were going wrong and things were getting worse." (P3). As time lapsed and explanations were not forthcoming, uncertainty took a progressively sinister form. Symptoms were spreading and the *need* for a diagnosis became ever more urgent, with patients aware that they were in a process of decline. P20 explained:

> ...we had to wait and we waited [but] they couldn't tell us. I mean, [pause] we understood that there wasn't a test that says 'This is what it is'. So we just waited and eventually they told us it was MND, but we had to wait for things to get a <u>lot</u> worse before they could finally tell us that it <u>was</u> MND. (P20)

In respect to physical presentation, patients needed to 'become' MND patients before they could be recognised as such. The medical picture became clearer over time, but for patients there was felt to be a worrying delay as symptoms escalated and they merely "waited". This delay created a sense of powerlessness in many patients. As investigations moved forward and patients passed through a hierarchy of services/assessments, many began to frame problems as increasingly "serious" over time. P5 explained:

> I went to stick my thumb up to the kids... ...my thumb wouldn't go up and I thought 'That's a bit odd'. So I went in about that to see my doctor, and I was referred to the orthopaedic people first and they said 'Oh no, I don't think it's orthopaedic' so they referred me to neurologists and I had the MRI scan, and I thought 'Oh this getting a bit serious now' and I went into Walton for the week to have the, um, I had a lumbar puncture and all the EMG tests... (P5)

Time and process were important determinants of response; P5's symptoms went from being "a bit odd" to "a bit serious" as she passed through different services. Negative emotional responses to the situation appeared to have been slowing accumulating and anxiety was clearly building as time passed and answers were still not forthcoming.

Exclusion: Withheld Information (Subtheme)

The abject nature of uncertainty exaggerated the lack of information being offered by healthcare professionals, which a number of patients explicitly linked to feelings of exclusion. Often patients saw numerous specialists before being referred to the neurology service and some suggested that doctors should have told them more during this phase. When P4's diagnosis was finally communicated, he felt disappointed in the doctors that had assessed him, revealing:

> Well, when I first heard that [diagnosis], I felt a bit let down. I felt let down for them three years [unclear] and let down by all them doctors. Why hadn't they told me it might be that, you know? [...] I was scared [long pause] 'cause I didn't know what it was. And nobody would tell me nothing, all them doctors I seen and they told me nothing... (P4)

P4 tellingly stated that doctors *would* not, rather than *could* not give him information during this period, even if only to tell him what the illness "might" be; he believed that information was being withheld. A number of patients suggested that there was a lack of engagement whilst investigations were ongoing; they felt excluded from the process and this was interpreted by some as clinicians being disinterested or uncaring. P12 recalled:

> Ah now it was interesting, I mean, when we went to see [doctor], 'cause I mean I'd seen that, er, what's his name? I paid to see him - I'd seen that [doctor] and he didn't tell me a thing about it – not a damn thing. He just referred me on to somebody else. [...] ...[diagnosis] was like my first thing of finding out what it was all about really. Just like, what it is, 'cause no one really explained that to me proper before... Finally, someone to tell me what was going on with it. (P12)

P12 had not felt part of the process; he felt excluded until the point that diagnosis was finally confirmed. He adopted an incredulous tone as he explained how his doctor failed to tell him "a damn thing" about what was going on.

The fear that patients associated with uncertainty appeared to have been exacerbated by the seemingly non-committal and often secretive nature of medical opinion that surrounded investigations. As P25 stated:

> ...nobody said at that point that it was what it was [pause] and actually that was the thing; because if it were something simple, not very bad, they just tell you off the bat don't they? You know, 'Oh we think it might be this; we're not sure, we'll do tests.' So actually, the way the mind works, if people withhold information, you think 'Well, there's got to be a reason; this is something big, bad, or ugly'. And actually as it happens it's all three... (P25)

Patients did not have information *and* they suspected that information was being kept from them; this was distressing. The fact that healthcare professionals would not tell patients what they thought the illness *might*

be was interpreted by some as being duplicitous. Formal diagnosis requires a cautious approach and healthcare professionals were clearly encouraged to protect patients from potentially unnecessary distress by concealing suspicions until there was clear evidence. This approach is consistent with best practice and follows national care guidelines (Andersen et al., 2005), yet it appears to have been misconstrued by some patients. P25's statement reflected the risk involved in this strategy; he felt that not knowing was an ominous sign, one that arguably caused as much distress as knowing that MND was a possibility. It seemed that, for most patients, no news was not good news and the cloak of uncertainty surrounding assessments fuelled distress. Information at this time was considered to have been important to patients.

Back & Forth (Subtheme)

Uncertainty during the pre-diagnosis phase was often punctuated by a number of 'false dawns' and there were emotional highs and lows as potential conditions were suggested and then dismissed. A number of patients (P1, P7, P10, P14, P15, P19, P25) were offered a less significant diagnosis prior to being told that they had MND; these patients were often offered a glimpse of certainty that was snatched away again leaving them back where they started. P10 recalled:

> I think I had physio about six times in that year, in that autumn, because they weren't sure if I'd damaged my back or something like that, and I think I was referred on to the physio here at [hospital], she then referred me on to another consultant and I think I went to three consultants who were not sure and they were very good and very kind or whatever, but [pause] I think one thought I'd cracked a part of my spine or did something with my back and that's about it. But I had about twenty or thirty falls; just collapsed or my knee would give way... That's how we got to the diagnosis: a long time; a long time of not knowing, thinking you know, then not knowing again. (P10)

Uncertainty fluctuated, with patients seeing themselves as passing into and out of the diagnostic process until the definitive diagnosis was achieved. Indeed, it may have been the temporary nature of previous diagnoses that led some patients to question the final diagnosis of MND. Misdiagnoses heightened uncertainty and reinforced the rather desperate sense that there was limited progress being made. P15 described herself as being trapped in "no man's land":

> Anyway, as I say, the leg came off worse and worse and they said it was sciatica and it stayed as that for about nine months and then we started getting referred to the ozzy [hospital] see if there's anything else wrong; we had x-rays, scans, but nothing wrong with the hips, nothing wrong with anything else, couldn't find nothing... ...And then it started in my arms and they said it was carpel tunnel. I said 'Carpel tunnel? Oh, okay.' I came home and I said 'What the fucking hell is carpel tunnel?' Got the medical dictionary out: carpel tunnel; 'Oh that's not a complicated explanation' I got that in the first five minutes, think nothing of it. So we got that one, but we couldn't find nothing for the pain in the right leg and occasionally my back, and now it's in my left leg too. It was like being in no man's land [pause] and although we'd been going the ozzy [hospital] at this time for six months, we were no further on. Then

> it went to twelve months and we still weren't sure, because I wasn't bringing out normal symptoms of motor neurones, even the early symptoms said I didn't have it as it was written down. (P15)

P15's attempts to understand the differing diagnoses that were offered to her showed her desire to establish certainty; it was clear that patients urgently wanted to be able to make sense of the situation and of the changes that were occurring within their own bodies. New and evolving symptoms seemed to quickly challenge early diagnoses and patients often had to rebuild dashed hopes. This seemed to add to the confusion and frustration that already existed. Patients suggested that it did not feel helpful that other illnesses were being 'discounted', as the correct diagnosis still eluded them. Discarding less serious conditions also increased the possibility that the illness was something significant. Retrospectively, most patients felt that they understood the difficulty that clinicians faced and only a minority retained ill-feeling; however, it was evident that alternative explanations and misdiagnoses added to the turbulent nature of this period. At the time, finding the *correct* diagnosis was the most important thing and most patients had remained focused on this goal.

The 'Actual' Diagnosis (Sub-Section)

When the diagnosis was finally confirmed, reactions were predictably negative; patients were "devastated" (P10, P14, P17, P19, P21, P22), "shocked" (P7, P9, P13, P14, P17, P18), "stunned" (P2), "scared" (P21, P22), "gutted" (P26), "crushed" (P26) "angry" (P4, P19), "upset" (P22), "frustrated" (P22), and even experienced a sense of "dread" (P14). All patients shared this immediate emotional impact; the diagnosis was depicted as a cruel end to the uncertainty that had defined the pre-diagnosis phase and a number of patients (P13, P18, P19, P24) suggested that MND was the "worst" diagnosis they could have received. Three superordinate themes emerged from patients descriptions of this event. The first, 'Affective Impact' encapsulates the immediate *emotional* response to diagnosis; the second superordinate theme 'Expectation and Disappointment' focuses on the *cognitive* consequences of the news; and the third superordinate theme, 'Sense-Making: Why Me?' explores patients' attempts to derive meaning from early post-diagnosis experiences.

Affective Impact (Superordinate Theme)

The first superordinate theme, 'Affective Impact' presents the immediate *emotional* response to diagnosis; it contains three subthemes: the first, 'Shock & Devastation', outlines a universal response shared by all patients; the remaining two subthemes describe how smaller clusters of patients reacted with either a positive sense of 'Relief' or a more destructive tendency towards a 'Delay, Avoid & Deny' response. The latter two subthemes demonstrate somewhat antithetical responses to the shared sense of shock described in the first subtheme.

Shock & Devastation (Subtheme)

Almost all patients reported "shock" and/or "devastation"; these terms expressed the magnitude of the immediate emotional impact of diagnosis and also conveyed the lasting effects, as P14 explained: "It was devastating news. I was in shock. I'm still in shock. I still don't want to believe it. ... I don't want it to be true."

A number of patients struggled in this way to come to terms with the news, leaving them in a prolonged state of disbelief. An extended pre-diagnosis period did not appear to have prepared patients, even though a number indicated that they believed the illness to be serious from the time it was taking to make the diagnosis. Most patients had avoided contemplating the actual diagnosis until it could be confirmed; however, even the small number of patients who had "suspected" MND (P13, P18, P22, P24) expressed surprise at the news. Despite patients stressing how long they had waited for a diagnosis, the notion that the news occurred "suddenly" was apparent. Diagnosis was frequently presented as a dramatic end to the enduring uncertainty that had preceded it. P18 explained:

> Well, I remember it clearly. [Doctor] told us virtually straight away, you know, there was no slow build up or anything – she did it right, if you know what I'm saying. It was just suddenly out there [pause] and I felt like all of these months of not knowing and not knowing and struggling with it had suddenly crashed into a wall in that room. (P18)

The point that patients were finally made aware of the diagnosis marked the end of a specific period of anxiety and carried the weight of all of the months of "struggling" with uncertainty that had gone before. This sudden stop – the "crash" described by P18 – seemed to be part of the shock that patients experienced. Patients seemed to have gone from knowing nothing, to knowing too much.

The enormity of diagnosis emerged in affective displays as well as in narrative form, with many patients becoming visibly distressed as they relived the event. Diagnosis imposed new 'existential' anxiety', as patients' whole lives were suddenly and dramatically redefined. The future that MND represented was extremely daunting. P19 recalled:

> Oh it was devastating really, I don't think I can say it any other way than that; yeah, just so devastating. I was a bit, inside, I was a bit overwhelmed, you know, it was that sort of overwhelming feeling of it and shocking as well; it was a shock because you just don't think something like that will ever happen to you, you just don't think it. ... You know, to think of what was to come with it all, to think of what was going to happen to me – all of the awful things that were going to happen – that happen to people with MND. I thought about those things for a long time; the course that the illness would take, you know. Even when I was being told what it was, those thoughts were already there in my mind. (P19)

These reactions were immediate, which concords with the idea of a sudden and intense impact; diagnosis was often a *traumatic* event. This created a strong sense of foreboding in P19 as she began to realise that she had become one of the "people with MND" to whom she referred. The sense of shock was occasionally so overwhelming that it appeared to have blunted emotional and cognitive responses; P10 quietly and contemplatively described his memory of diagnosis as "a bit of a blur" He was unable to recall many details of the actual event, though did remember moving acquiescently through the day in a calm and compliant manner:

> ...I suppose I thought 'Well, if there's nothing else that can be done at the hospital I might as well go home' and so that's what I did. [Long pause] Yes, I went home. I suppose, you don't

> really think at a time like that, well, I didn't that I remember. And after, I mean, [nurse] was there and she was very good and she was very supportive and very kind; very supportive and attentive and kind and then, do you know, it's a bit of a blur quite honestly, everything just, well... **...there must have been a lot for you to take on board at that time...** Well, exactly, that's exactly right I think. (P10)

P10 appeared dazed in his recollection; seeming to have been unable to fully absorb what was going on around him. P7 also reported of this time: "...it's the shock I think, you don't think that much." For many patients, the experience of diagnosis was *emotionally* overpowering and although patients had widely anticipated the event, it seemed that once the news had been communicated, patients were actually unsure of what to do with themselves.

Relief (Subtheme)

Alongside negative emotional reactions, a small number of patients described diagnosis as bringing a sense of emotional relief (P3, P12, P18, P22). Diagnosis freed these patients from the uncertainty that had characterised the pre-diagnosis phase and marked a clear psychological endpoint to a long period of increasing confusion. It seemed that diagnosis appeased distress because it meant that the situation was no longer escalating out of control in an unpredictable way. P18 recalled:

> I mean, in one way it was a shock when she told me and yet I was relieved because, um, things had been going wrong for a long time and I'd been deteriorating. [...] Knowing is much better than not knowing. I'd rather not have this but at least I know what I'm working with now. ...I'm back in control of things a bit more – I know what I'm up against and I can get on with it and deal with it. (P18)

Relief reflected the perception that diagnosis had given back some of the control lost during the uncertain pre-diagnosis phase. Knowledge itself was a comfort, giving the situation stable parameters and providing these patients with focus; even if the diagnosis was disappointing, it had at least afforded clarity and reintroduced certainty. P3 concurred:

> I felt relieved at the time, because it was taking so long for someone to say what I had and I realise now why, but I felt relieved at the time and thought 'Well, I've got it and there's nothing I can do about it' I just got on with life the best I could. I thought 'At least you know now what it is'. (P3)

Diagnosis defined the situation so that patients like P18 and P3 could appropriately appraise their own practical and emotional resources for coping with the illness; this afforded them a greater sense that it was not completely out of their grasp, which allowed them to 'get on with life'. Diagnosis enabled them to move forward and gave them somewhere to direct positive efforts; it was seen as an opportunity to begin actively coping and engaging.

In a different context, P22 had suspected his illness was MND for many years prior to diagnosis and had asserted his suspicions throughout the diagnostic process. At diagnosis, P22 was relieved to have been vindicated. Although he conceded he had been "upset" at the news, he explained:

> Waiting to know is hard, that was hard. It took a long time. It did take them a very long time to say MND when I already knew. [...] I knew you die from it. I knew it kills you. I already knew that. And when they finally said it was MND, I'd already been living with it for years, so I could see it happening already... when I knew what it was, when they said, in a way it was better that they said I was right, and then everyone knew. I knew then, so I wasn't so frustrated. (P22)

Again, it was the removal of uncertainty and the gift of 'knowing' that allowed P22 to move on from the negative emotions that had defined the pre-diagnosis phase. Diagnosis connected patients with appropriate services and personnel, and it was reassuring to know that the situation was mutually understood; the label 'MND' facilitated communication and enabled *shared* knowledge and understanding to emerge. In a similar scenario, P12 had been "unofficially" diagnosed many years earlier. When MND was finally confirmed it was a definite relief for him and also bought a sense of purpose to his illness experience. P12's diagnosis was a sign that he and his experiences mattered and that he was being "cared" for; he explained: "...it was like she [doctor] cared enough to say it, you know? So it makes you feel like this person's looking out for you a bit more. So, yeah, I was glad like we was getting somewhere." (P12). For these patients, diagnosis represented progress and there was a clear feeling that whilst MND had not been the diagnosis that any patient had been hoping for, it at least meant that the diagnostic uncertainty was removed and things could move on in a practical, as well as psychological, sense.

Delay, Avoid, Deny (Subtheme)

A further subset of patients explained that although shocked by the news, they did not fully address the psychological ramifications of diagnosis until after the event, choosing instead to delay, avoid and/or deny the situation (P10, P15, P16, P20, P19, P24, P25). This behaviour tended to be prevalent in patients with lower levels of physical disability, who reported that they tried to immerse themselves in the 'everyday' to distract from the illness. As a result, a small number of patients suggested that much of their reaction to diagnosis was initially cushioned by circumstance. For P10, work had been a distraction that kept him from really processing the news of his illness. He explained:

> I think because it was only last year that I gave up work you know? I think perhaps, well, perhaps I was working all the time and not thinking – working and not really thinking [pause]. It almost didn't need thinking about at first, it was there and mostly I was just carrying on. ...life had gone on, and so, life went on after this. (P10)

P10 felt that the diagnosis did not "need" thinking about in the early stages, suggesting that it was later physical impairment and functional incapacity that provided the necessary impetus to face his illness. It seemed that a combination of not *needing* to address the consequences of the illness and not *wanting* to think

about it, allowed P10 to "carry on" without having to really accept the diagnosis. Ultimately, it seemed to have been illness progression that vitiated this avoidance; P20 explained that his initial symptoms had not been "serious" enough to fully engage his attention and it was only later, as the illness began to progress and to intrude on everyday life that he really connected with the diagnosis:

> Well, at first I didn't think too much of it, 'cause I felt okay, you see? I still felt okay. So, they told me I had MND and I thought [shrugs] – I just thought 'Oh fine'. But it was only when three months later we went over to Ireland I really started to notice it, and my speech had got a lot worse – a lot worse – and I fell over. That was when I started really to pay attention to it more then [unclear], because when they actually told me what it was, I didn't know anything about MND and because I felt okay, I just thought, 'Oh right' and it didn't bother me. I had no idea. I just thought I'd get on with things as they were, just carry on with it all as normal but with MND. (P20)

As the illness progressed P20 was confronted with the reality of the situation: "It's a delayed reaction – a delayed reaction. I'm more upset now than I was when I found out what it was..." P20 had been able to live without thinking about the diagnosis because it was not perceived to have significantly changed his life; an appreciation of the diagnosis and a more overt emotional reaction came later, when illness progression necessitated information-seeking and he discovered the true implications of the condition. He explained that his initial shock at diagnosis was related more specifically to the news that doctors could not do anything to improve his failing speech and not to the wider consequences of the illness.

In other patients (P15, P16, P25) avoidance was far more significant and patients admitted that they had initially tried to deny the illness altogether. P15 recalled: "[after diagnosis] I was still not interested – in denial. I said 'Motor neurone disease; motor neurones not getting to where they're supposed to be? I'll leave that one. Not interested'...". P15 did not *want* to acknowledge the diagnosis, and so simply chose not to. Her casual attitude in stating that she would "leave that one" and that she was "not interested" appears a startling response to such a dramatic diagnosis; but can readily be understood as an attempt to protect herself from the distress that might otherwise have been incurred. As in the cases of P10 and P20, the progressive nature of MND was such that it was not possible for patients like P15 to adopt denial as a long-term strategy; the rapid pace of physical degeneration provided irrefutable evidence of the illness and physical symptoms forced these patients to acknowledge the diagnosis. P15 continued: "I denied it for twelve months and if I could have I would have denied it permanently, but you can't deny this, can you? [Laughs] [...] ...you sort of like, resign to the fact." This view was echoed by P25, who also looked back at his early attempts to deny the illness as somewhat futile:

> I have come a very long way since that early time of not wanting to know. That's trying to deny it or avoid it; but you can't, it's real. Good luck trying to deny this illness, because it soon makes its presence known. (P25)

P19 concurred; she had initially sought to protect herself from the distress of the "difficult" diagnosis, but ultimately had to confront the reality that the illness itself presented to her. She explained:

> ....it was just difficult to accept it at first... [...] ...then of course the MND gets worse in you and then you really can't deny it to yourself anymore. So actually the MND makes you accept it because it's all of a sudden there and real – it's really in your face very quickly [laughs]. (P19)

Denial was clearly a comfort in the early stages, yet became untenable as symptoms progressed. These patients experienced further negative emotional reactions to diagnosis many months after the event.

Expectation & Disappointment (Superordinate Theme)

The second super-ordinate theme 'Expectation & Disappointment' focuses on the *cognitive* consequences of diagnosis and contains two subthemes. The first subtheme 'Nothing You Can Do' expresses how hopes and expectations of treatment were dashed by diagnosis and explores the impact that this had on patients' engagement with the illness situation; the second subtheme 'Redefining the Future' addresses the realignment of expectations and the process of reconstructing new futures that incorporated the illness.

Diagnosis was a cataclysmic event that initiated a chain of fundamental life changes. Patients arrived at diagnosis with a number of implicit and explicit hopes and expectations – predominantly in respect to treatment and restored health - and this meant that they subsequently experienced a profound sense of disappointment. This led many to disengage from the situation and marked the start of a more cautious and controlled approach to the illness. These themes appeared to represent the more considered cognitive consequences of diagnosis; responses that were experienced at the time, yet perhaps only acknowledged and assimilated on reflection.

Nothing You Can Do (Subtheme)

All patients seemed to have endured the uncertainty of the pre-diagnosis phase buoyed by hopes that the illness would be an acute rather than chronic state and that diagnosis would mark the beginning of its end. This made diagnosis a point at which patients had to adjust specific expectations, seeing further illness and decline where they had hoped to see treatment and improvement. P18 recalled:

> Like it was, um, like when you're going through the diagnosis bit it's frustrating and that, but there's always that bit of hope that they'll say 'Oh yeah, it's that' and then they can sort you out. But not with MND; there's no hope at all of that. ...that's the big thing you latch onto – there is no cure. (P18)

Naturally, patients' attention was immediately consumed by the lack of any curative treatment and the shock that patients experienced in part reflected the fact that the entire illness context had to be suddenly redefined. P21 described diagnosis as a "big shock" and explained: "I didn't think it would be serious. [I] thought they would be able to do something for me. I was very disappointed." Determining *how* – rather than *if* - the illness could be treated seemed to have been the most significantly anticipated consequence of diagnosis and when it

became clear that no opportunity for improvement was forthcoming, the negative and avoidant tone of patients' enduring attitudes appeared set. P5 explained:

> I think if you get any diagnosis, the first thing you want to know is 'What will it do to me and how can you treat it?' and with MND it means you are going to die and they can't do anything about it. So when I found out that, I thought 'Well, what more is there?' I just knew enough to know I didn't want to know anything else [laughs]. Once you know it's final and there's nothing you can do, there's not a lot of point knowing anything else. (P5)

P5 appeared to have become apathetic once it was established that she could take no positive action against the illness and many patients similarly seemed to have disengaged at that point. P22 recalled:

> I did listen to them, yeah. But the most important thing was there was nothing they could do. [...] ...once you hear that, you switch off a bit. [...] Once you know it kills you, you're just sort of waiting [lights a cigarette]. You go along with what happens... (P22)

KV delivered these statements in a calm and detached manner, which seemed incongruent with the distressing content of what he was saying – essentially explaining that he was "waiting" to die. This may be interpreted as reflecting the disengagement that the news evoked, with a strong sense of distance and defeat audible in the quiet resignation of the delivery. P14 echoed these sentiments, describing how he also began to distance himself from the illness situation once it was determined that there was no treatment available:

> I didn't really *want* to know it really. I knew enough then. I mean, once you know that, well, there's nothing that can be done [long pause] I think I felt that there was very little point in finding anything out. What good would that do? That was my feeling. (P14)

In the early stages, many patients felt there was little "point" engaging with the illness and the lack of a treatment led a number to frame the situation as hopeless. This is not to say that there was no hope to be found in life with MND, but that patients arrived at diagnosis with specific hopes and expectations that could not be sustained.

The role that patients had themselves expected to step into was often undermined by the lack of treatment; most patients had expected to play an active part in determining the course of the illness, yet in most cases, diagnosis enforced passivity onto them. P5 explained: "It was like, that's the end now; there is nothing left for you. So, it was just go home and wait for it to happen to you." In a similarly pessimistic appraisal, P18 recalled: "...my first thought was, I thought 'Am I going to just sit here at home and wait for it to get worse, worse, worse?'..." Patients seemed to leave diagnosis feeling unsure about both the future and their place within it. Initially, a number of patients expressed a sense of abandonment and it was clear that patients had been susceptible to feelings of powerlessness and 'pointlessness' during the early stages of the illness.

Redefining the Future (Subtheme)

Fundamental life-goals and wider expectations about the future were challenged by diagnosis. Some patients (P5, P11, P14, P18, P22) reported that they initially struggled with the idea that life would go on; P5 expressed an initial sense of defeat as she realised the magnitude of change she would endure:

> ...that was it - I'd had it. It was like, that's the end now; there is nothing left for you. [...] I just thought it was all over for me as soon as I heard them say what it was. And that's the way it has been in lots of ways, you know, I'm glad I'm still here for the kids and that, but life has never been like it was and I know it never will be again and sometimes that does get me a bit, you know, I have days even now where I feel down. (P5)

Many patients, like P5, grieved for the life that they had lost *and* for the loss of the life that they had expected to live. This was a particular form of enduring disappointment; a bereavement that was shown in anger, sadness and frustration. The disappointment that stemmed from patients having had their expected futures so brutally distorted was seen particularly clearly in female patients with younger children; P18, P5, and P15 all spoke of painful changes within the family structure and of trying to sustain elements of the life that they had planned before diagnosis, such as being there for children as they started school or as teenage children took important exams. The threat to these futures also motivated patients as they moved forward with the illness; P18 spoke movingly of her desire to 'keep going' for her two children. In her description of diagnosis, she recalled:

> ...one of the things that went through my mind is that one of my goals is to live to see [long pause – becomes upset] is – [long pause] is to live to see my children grow-up. It keeps me going. [...] I think if you didn't have something like that, something to look forward to and to really live for, you'd just lie down and give up. As soon as you found out like there was no cure for it, you'd give up. (P18)

Although the future had been threatened and this was a devastating blow, it was important that there was still a future there for P18. Identifying things to "look forward to" and to "live for" was an important psychological strategy; performing this kind of cognitive restructuring enabled new or redefined futures to emerge. Families often provided continuity, serving as a common thread between the futures that patients had expected to have and the new futures that MND defined. Although P4 himself clearly loathed talking about the future, he spoke earnestly of attempts to prepare his family for a future without him. He explained:

> ..I have to start helping them to do the things I would have done, because it's getting harder for me to do everything. ...I want them to be alright. I want them to be alright when I'm not around. When I'm not around I won't be there to do things, so I want her [wife] to be able to do them now. I need to pass it all on. I need to prepare the family for when I'm not around [becomes upset]. [...] Yeah. I want them to be okay without me. I mean I'm helping them to do it now. We do it together at the moment [...] I'm just hoping that's going in there [taps head] and staying in there. Then when I'm not around she can do it for herself. (P4)

In this way, patients needed to be able to redefine the future for those around them as well. P4 was trying to actively shape a better future for his family, compensating for the loss that MND would inflict on the futures of his wife and two teenage daughters, as well as his own.

Negative responses to the changes incurred to future plans were not restricted to younger patients or those with dependents; comparably older patients also lamented the changes that deprived them of the things they had expected to enjoy in their later years, such as an active retirement and time spent with loved ones. It was difficult for patients who felt that they had contributed a lifetime of work so that they could enjoy retirement, to have the future they had planned snatched away from them. In some cases, this disappointment cast a lasting shadow over life. Whilst P18 had been able to redefine her future positively, P11 felt that:

> ...[it] feels like life's finished [laughs] [...] There's nothing good much to look forward to, like no more holidays now, can't go nowhere, can't do nothing. And I worked hard so I could enjoy my retirement. I feel like, what a waste of time that was. (P11)

Although he laughed dismissively, P11 appeared visibly low in mood and spoke in a despondent tone. The important things that defined his life had been taken by the illness, which did not feel fair. He had expected to enjoy these things as reward for his hard work and the disappointment that he felt left him frustrated and prone to expressions of hopelessness. It seemed that P11 had not been able to redefine his future in a more positive light, still feeling the disappointment of all that he had been deprived of and still working to accept the implications of his diagnosis.

Older age did not prevent patients' from grieving lost lives; however, there was evidence to suggest that older age was a comfort to some patients, as in P12's pragmatic suggestion that "When you get to my age, you just gotta take each day as it comes... [...] I'm not a young man, you know? I'm just living out me days really, so I'm not stressing over nothing." and in P16's later musings that

> ...it must be terrible for the very young to get this, 'cause their life is ruined then. I mean, I know my life is ruined, but I've had a good life; a full life and with a family I love. That's what I was thinking. (P16)

All patients had to realign expectations when the diagnosis of MND was finally confirmed, even if the impact of this process differed between patients. There were clear individual differences; P11 responded to diagnosis by suggesting that his life was "finished", whilst P12 used humour in recounting:

> ...after all the testing and testing and what not, he just went 'Oh yeah, you got it'. So I packed in playing professional football [laughs]... to be honest it don't cause me no bother really, I wasn't planning on running no marathons before I got it, you know, so, yeah. (P12)

Clearly, patients' expectations of the future had differed even before diagnosis, which meant that some patients felt loss and disappointment more acutely than others and the ways that they coped with loss also differed.

Although diagnosis changed 'life' to varying degrees, all patients seemed aware that the illness had redefined the future. P15 explained:

> I've give up now. Old and decrepit – just a bit quicker than I planned to be, I mean, I'm fifty this year, I was hoping not to get it until I was seventy-odd, but it's come on a bit quicker than I expected. [...] I'm just hoping, I keep calm and do what I'm doing and I might last a bit longer. (P15)

The physical degeneration of the body left P15 feeling old before her time; her fundamental expectations about the future had shifted significantly and this left her somewhat despondent. Hopes and expectations came to focus on things that one might ordinarily take for granted, such as survival and being able to "last a bit longer". P25 explained:

> I can't change things; I've still got a life to live, even if it might not be as long a life as I thought or as full a life as I thought. It's a different future that I've been given now, but it is a future still. [...] Looking forward, yes, looking forward - [long pause] People talk to you sometimes about the future and what will happen to you like it's hopeless. I don't think it's hopeless, although... [long pause]... I don't <u>think</u> I'm mentally blocking it out. I'm looking forward to the future as much as I possibly can. (P25)

It seemed that patients wrestled with a rational/pragmatic appraisal of the future as extremely negative – perhaps even hopeless - and a more positive view of life going on and still being fulfilling. This second view was arguably necessary to maintain psychological wellbeing. The self-doubt in P25's suggestion that he did not *think* he was mentally "blocking" out what others perceived to be hopeless in his future showed the difficulty that patients themselves had in making sense of the future after diagnosis. P25 paused on a number of occasions to collect his thoughts and to choose his words carefully; this was of the few times during his long interview that P25 seemed to falter over what he was saying or to be constructing his ideas as he spoke. The new and somewhat uncertain future that awaited patients after diagnosis did not need to be considered "hopeless", yet it was a *different* future and patients reported a need to redefine and adjust to this.

Sense-Making: Why Me? (Superordinate Theme)

This superordinate theme stood alone as the sense-making question 'Why Me?' This theme describes attempts to make sense of the illness and explain why patients' had been afflicted, a process often conjuring strong feelings of frustration and injustice. All patients responded negatively to the lack of specific information available about the aetiology and retrospective/prospective timeline of the illness. The limited information that is available to MND patients about their condition is descriptive rather than explanatory; it provides an outline of the potential physical changes that lie ahead, but offers no tailored prognosis and no causal reasoning. This is instantly problematic given that being able to both explain and find meaning in illness is acknowledged to be an important part of the experience. The lack of information accompanying diagnosis was an important determinant of response, particularly as it seemed that a diagnosis of MND created as many questions as it answered. Indeed, P25 recalled: "...generally, my reaction to this illness was, well, not really a reaction at all, just a lot of questions; that's how I would say it was."

There is an established literature that highlights the psychological importance of understanding and finding meaning in adverse or threatening situations (Pakenham, Sofronoff, & Samios, 2003). It has been suggested that searching for meaning is a crucial part of the coping process and that being able to integrate the meaning of illness into everyday schemas plays an important role in determining emotional status (see Fife, 1994). There are also implications for long-term adjustment, with successful adaptation to adversity thought to involve a two-step process of first making sense of the situation and then finding benefit or drawing some positive implication from it (Janoff-Bulman & Frantz, 1997). It seemed that patients desperately wanted to make sense of the illness after diagnosis, with the lack of information about a *cause* being one of the most distressing aspects of the illness. A number of patients (P5, P7, P11, P21, P24, P25) remained unsettled by the lack of information available as to the *cause* of the condition. Although all patients demonstrated good adjustment to the illness, many still had occasional moments when they asked the common question "Why me?" P7 explained:

> It's a shame really, but, I don't know, I think what one would like to know [is] why it happens. Why's it gone wrong? I know it's something to do with these cells, the muscles they support, you know the jargon but 'Why me?' you know, 'What did I do?' I never smoked, I never drank, I didn't take drugs. You think, 'What went wrong?' You know? I take vitamins. I've taken vitamins for donkey's years, you know, so you think 'Well, what can you do?' It's been a reasonably healthy lifestyle, but if they could tell you why it happens, that would be interesting. If they knew that, they could probably find a cure then. You know, they'd be able to work on something. It would be nice to find out. (P7)

Knowing why *she* had the illness was important to P7. Rather than being interested in the "jargon" of what was happening, she wanted to know what had happened in her specific case and in the context of her own life. This was a personal issue, rather than a more general query. P7 wanted to make sense of why she had contracted MND in spite of the positive behaviours listed. There was an implication of injustice in the fact that she had not been a smoker, drinker or drug-taker and had taken vitamins and kept herself active, yet still developed the illness. Although she latterly bought the issue of a cure into the conversation, it seemed that being able to make sense of why she had the illness was a more pressing concern. It would clearly have been more than just "nice" to find out.

The feeling of injustice was shared by a number of patients, who felt defeated by the idea that the illness struck the good and undeserving. P11's illness seemed to have encouraged a fatalistic attitude towards life, he explained:

> Tough luck if you've got it. Thanks a lot [laughs]. It's changed my outlook on life really. [...] There just doesn't seem to be any justice – what more can a man do you know? [...] It just makes you think. [Long pause] [...] [wife's] brother got cancer and my sister died just before Christmas and that was cancer as well. You can't win, can you? (P11)

There was a sense of hopeless resignation in the idea that "you can't win". He later opened up further on this subject, revealing:

> I think, you know 'Why me? Is it my fault?' It wouldn't be so bad if it were my fault. You know, if it were cancer of the lung I could understand it then, it would be my fault, but this - This just came out of the blue. If I understood why, or if they could say that it was my fault, it wouldn't be so bad. [...] It's very rare, you know, this condition. That's what it said on that thing the doctor gave me; it only affects one or two people in every hundred thousand. So why did it pick on me? [Laughs] (P11)

That he described himself as being "picked on" indicated P11's belief that he was a victim. He even suggested that it being his "fault" would have been preferable to not knowing why he had contracted the illness; there had been no warning, no risk and no slow build up, the illness was perceived to have come "out of the blue" and without reason. A lack of understanding about the cause of the illness led to significant rumination and was a frustrating aspect of the experience.

Not every patient wanted to present themselves as explicitly asking the question 'Why me?' Indeed, P25 directly insisted that "...there's nothing in there that's saying, you know 'Why me?' You've just got to take it on the chin..." He seemed to equate this particular question with self-pity or even weakness, seeing it as a self-indulgent question posed by those who felt sorry for themselves. However, in a somewhat contradictory statement, he did reveal that even in the early stages of the illness: "I just wondered how I'd contracted it. And all the people I've spoke to, they wonder the same. Whether it's an inherited thing we don't know. [...] I suppose it's just one of those niggles in the back of your mind." P25 *did* question why he had the illness; though perhaps perceived negative connotations in explicitly posing the question "Why me?" It was interesting to note that P25 was genuinely concerned about the heredity of the illness and whether it was something that might be passed onto his son or grandson; yet he later placated these legitimate concerns by designating the illness as an act of God, random and unexplainable. This was perhaps a greater comfort than not knowing. Whilst hospitalised to have a RIG fitted, P25 shared a ward with a number of other patients and told the following story that revealed his feeling about the illness:

> Well, the one thing that upset me was that in the bed opposite me was a wino – self-inflicted. Next to him was a beer-drinker and next to him was scotch. So, out of the six people on that ward, three people were self-inflicted. So I chose the youngest one and I went and sat down and had a talk to him. I talked to him and I told him he doesn't want to go down that road, that poison, I said to him 'I'm here because God's put me here.' He didn't like it. ... It's right though. I was only trying to save the little bugger. You know, this MND is just through no fault of my own, but he was self-inflicted. He bought his misery on himself. (P25)

It was perhaps unnecessary for P25 to ask 'Why me?' if he had satisfied himself with the explanation that God had "put him" there. Collectively, P25's interview indicated that he was a man that had many questions about the illness, yet, perhaps through frustration at the answers not being available, found it necessary to seek spiritual answers to biological questions. Being able to explain the illness in relation to a wider spiritual 'plan' seemed to have made it easier for P25 to cope with the situation.

Most patients were not satisfied that they had all of the information they wanted and needed; indeed, there was clearly enduring frustration. P21 explained that at diagnosis, and even many months after, the issue of a cause was an actively contentious issue: "I still want to know. No one knows. [...] Drives me mad!" She felt that knowing why she developed the illness would allow her to "get on with it"' and would offer "peace of mind". This reflected the need for meaning within the situation and highlighted again the negative impact that uncertainty could have on patients. P21 suggested: "I want to make sense of it. It doesn't make sense to me. I ask 'Why?' a lot." There were many unanswered questions and many patients did at times find themselves asking 'Why?'. Although this was not a constant and direct source of distress, it was, as P25 himself acknowledged "one of those niggles in the back of your mind". It seemed that patients *wanted* their experiences to make sense; this was part of their psychological adjustment to the situation. Patients knew that there were no answers to the questions that they posed, but this did not negate the existence of these questions. A great deal of negative feeling stemmed from the fact that patients could not establish answers to questions about an illness that had literally become part of them; a strong sense of dissatisfaction and injustice endured.

Section 2: Attitudes and Coping Strategies

Contained within this section are themes that emerged as patients discussed the ways that they approached the illness on a day-to-day basis; this section is intended to give a general description of patients' attitudes towards the illness from a cognitive/emotional perspective. Table A6.2 provides a summary of all subsections, superordinate themes and subthemes contained within this section.

Table A6.2.
An overview of the subsections, superordinate themes and subthemes contained within 'The Experience of Diagnosis'.

Section	Subsection	Superordinate Theme	Subtheme
2. Attitudes & Coping Strategies		2.1 Degrees of Engagement	2.1.1 Out of My Hands 2.1.2 Getting On with It
		2.2 You Just Don't Know	2.2.1 Avoiding the Unknown 2.2.2 Hope in Uncertainty
		2.3 One Day at a Time	
		2.4 Fighting Spirit	2.4.1 Not Letting It Win 2.4.2 Keeping Going: The Physical Fight
		2.5 Positive & Negative	2.5.1 Staying Positive 2.5.2 Selecting/Controlling Responses

There are five superordinate themes in this section. The first superordinate theme, 'Degrees of Engagement', describes an avoidant attitude towards the illness, with the vast majority of patients demonstrating a

significant level of cognitive/emotional *dis*engagement that was clearly intended to circumvent potential distress. The second superordinate theme, 'You Just Don't Know', addresses the impact of the unpredictable illness trajectory and patients' focus on the uncertainty that existed in respect to the future. The third superordinate theme, 'One Day at a Time', reports patients' preference for coping on a day-by-day basis, adjusting to change *as* it occurred rather than looking ahead; this includes descriptions of the cognitive, emotional and practical benefits associated with this approach. The forth superordinate theme, 'Fighting Spirit' describes a more determined attitude, expressed via patients' insistence that they would not be 'beaten' by the illness. The fifth superordinate theme, 'Positive & Negative', explores the importance attached to being and staying positive and not succumbing to negative affect. For each superordinate theme there is a summary of the subthemes contained within.

Degrees of Engagement (Superordinate Theme)

This first superordinate theme describes general attitudes towards the illness, with the vast majority of patients demonstrating a significant level of cognitive/emotional *dis*engagement intended to avoid distress and maintain wellbeing. This is expressed in attempts to purposefully limit the attention afforded to the illness wherever possible. This superordinate theme continues three subthemes: the first subtheme, 'Thinking & Not Thinking', describes the transient nature of patients' disengagement from the illness and their attempts to avoid rumination without entirely escaping the reality of the situation; the second subtheme, 'Out of My Hands', describes how disengagement endured as a product of patients' inability to exert an influence over the course of the illness; and the third subtheme, 'Getting on With It', explores patients' attempts to move forward and 'get on' with life rather than dwelling on the wider situation.

Thinking & Not Thinking (Subtheme)

It seemed that successful adaptation often relied on patients' ability to oscillate between involvement and avoidance in different areas of the illness; most patients' demonstrated variable levels of cognitive and emotional engagement over time. Patients lived with unavoidable physical reminders of change and so were not unaware of illness progression; yet most still made active attempts not to consciously dwell on their circumstance and seemed to avoid reflecting on the illness wherever possible. This resulted in patients' reporting that they were both thinking and not thinking about the illness. P22 explained: "I don't really even think about any of it. [...] I don't think about things unless I have to really. You can't not think about it, 'cause of this [gestures to own body], but you don't really <u>think</u> about it anyway..." Absolute denial was clearly not an option for patients with high levels of physical disability, yet P22 still preferred to limit the attention he gave to the illness. Most patients seemed to be capable of affording just *enough* attention to their condition; addressing physical needs and processing change without becoming overly reflective. P7 revealed:

> I'm not bothered, you know, it doesn't depress me. I just think, you know, I used to make cards, decorate goose eggs, but I can't do it as much anymore, you know, because I haven't got the dexterity and the feeling, you know? But other than that, I haven't got any problems. I don't think about it; I'm fine really. Well, when I say I don't think about it, I don't <u>dwell</u> on it. (P7)

Not *dwelling* on the illness was important; when patients spoke of thinking and not thinking about MND, they seemed to suggest that they accepted the illness but did not ruminate on the wider situation. This appeared essential in preventing patients from slipping into negative emotion states. The idea that P7 was "not bothered" by the illness appeared a surprising assertion, perhaps one best understood as part of this wider disengagement and avoidance, which served as a form of self-protection.

Most patients seemed to restrict (i.e. purposefully control) the cognitive resources allocated to the illness to some extent, which allowed them to avoid thinking about it without becoming entirely dislocated. Patients sought to maintain a degree of 'distance' until external triggers called upon them to re-engage. Catalysts for re-engagement typically included events such as hospital appointments and assessments; P4 explained: "I just think I don't need to think about it until they [doctors] tell me to." P5 concurred: "... don't really think about it unless I've got a test coming up. I just think 'Oh yes, put that one to the back of my mind'. And I don't think about it." It is important to note that most patients did re-engage when prompted and that the majority of patients only occupied a transient and/or selective disengagement from the illness; this differentiated it from a more enduring form of denial.

The terms 'avoidance' and 'denial' hold potentially negative connotations; reference is made to 'disengagement' to indicate that this type of 'not thinking' was often a positive strategy. However, it should be noted that much of the behaviour displayed by patients seemed to involve the *intentional* exclusion of illness-related thoughts and thinking, making avoidance an identifiable part of this process. Discussing her approach to the illness, P15 explained: "I think that the more you think about it, the more it will come up and pick on you, so I tend not to think about it." Thinking about the illness made it 'real' and increased the threat posed, so patients like P15 elected not to consciously attend to the illness unless absolutely necessary. This was a tendency away from the illness rather than a strict denial of it; patients like P15 would still engage with people and services when and where necessary. The notion that the illness was a threat likely to incur negative emotional responses was captured by P15's reference to the chance that it would "pick on" her. This underlined the strategic nature of disengagement as a means of protecting patients' psychological wellbeing.

On the periphery of the selective disengagement described by the majority of the sample, a minority of patients (P3, P17, P24) remained consistently engaged with the illness, and a similarly small number (P4, P14) seemingly sought not to engage with the illness at all. Patients who sought to remain engaged with the illness seemed to spend more time thinking about the illness than not thinking about it. They often actively sought out illness-related events and information on an almost daily basis; for example, P24 reported fervent information-seeking, explaining:

> I still look now [for information]. I mean there are so many forums out there and there are always people talking about it and, like, there's one that I go on everyday... I like to keep quite active and on top of that side of things. And the forums are a good way of doing that, you know, talking to other people on the forums. [...] I've tended to look things up in advance really, you know, so I know what to expect. (P24)

P24 presented as a patient who needed to retain control and to feel that he was "on top" of things so that he knew what to "expect"; he clearly thought about the illness a great deal and on a daily basis. P24's engagement with the illness was obvious from the content of his interview and his willingness to discuss all aspects of the condition; from the attitudes of strangers on public transport to his strong views on suicide and euthanasia, it seemed to be the illness that dominated his thoughts. In contrast, patients like P14 demonstrated more consistent avoidance and the act of being interviewed and placed in a situation where he was *required* to attend to the illness revealed the degree of disengagement that had come to dominate everyday life. He became tearful as he spoke about his current status, explaining: "It's okay, I just [long pause] talking about it, I don't even really think about it very much, but when you have to think about it, you realise what it is now; how it's changed now." It was clear that not thinking about the illness had protected P14 from anguish in the short-term, yet may only have delayed rather than negated distress entirely. The content of P14's interview was far more limited; he clearly felt uncomfortable having to engage on any level, often simply closing down questions by stating: "I don't want to talk about it. ...Can we move on? I don't want to talk about this anymore."

These active and avoidant outliers, though seeming to adopt antithetical approaches to the illness, also manipulated levels of engagement in order to maintain psychological wellbeing. It should also be noted that even those seeking to maintain an active involvement with the illness conceded that they often disengaged from negative information in order to maintain a positive outlook. The degree to which patients were thinking and not thinking about the illness varied. Although no patient could deny the illness completely, many clearly attempted to disengagement from it to some extent, with patients only differing in the balance that they struck between approaching and avoiding the illness at any given time.

Out of My Hands (Subtheme)

As evidenced by patients' immediate responses to diagnosis, a significant determinant of disengagement was the fact that there were no treatments available. The sense of detachment that many patients felt as a result of this fact appeared to endure and patients frequently directed interviews back to the fact that there was 'nothing' they or anyone could do to change the situation (P3, P5, P7, P10, P11, P14, P18, P20, P22, P25). Some patients intended this statement as a show of negative emotional response to the powerlessness inflicted, whilst others used it as a more positive affirmation of acceptance and moving forward. In all instances this statement justified or explained disengagement. P7 seemed to capture the mood of many patients in her descriptions of the illness experience when she explained:

> Not much you can do. I mean, my idea is that there's not much I can do about it, 'cause as they say, there's no cure for it, so it's just waiting for God really, isn't it? I mean, yes, I take on board what they're saying and you must be sensible... I'm [pause] well, what can you do? It's out of my hands. (P7)

A situation appraised as being "out of my hands" affords a natural sense of dislocation; however, it seemed that P7 wanted to show her understanding and recognition rather than to express hopelessness. Insisting that there was 'nothing' she could do to change her circumstance appeared to be a pragmatic reflection that indicated acceptance. P7 notably labelled the fact that there was not much she could do as "my idea",

intimating that this may have been a considered appraisal and perhaps even a purposeful way of looking at the situation to incur the least possible negative feeling. P7, like many others, understood that she could listen to the doctors and be "sensible" in her approach, yet ultimately could not *do* anything about the illness. It was recognising the limited potential to exert influence that seemed to encourage many patients to maintain disengagement. It seemed that patients protected themselves from feelings of helplessness and/or hopelessness by 'stepping back' from the illness.

Although physical changes naturally sparked negative emotional responses, most patients appeared determined not to dwell on these transitions; the fact that there was 'nothing' that patients could do actually seemed to encourage some patients to move forward. Stressing their inability to influence the speed and direction of changes was one way of rationalising a need to disconnect from them, thus defusing/diluting emotional responses. P25 acknowledged that his failing speech had made things "difficult" in many situations, yet when asked how he *felt* about these changes, he explained: "Doesn't bother me; there's nothing I can do about it, so I can't sit down and say 'Oh well...'..." It seemed that there were specific instances in which patients used the fact that there was nothing they could do to avoid recognising negative cognitive and emotional responses to change.

Many patients seemingly *evaded* questions about emotional aspects of the illness experience by giving answers that only stressed the limits of their control and involvement. The idea that there was nothing that anybody could do to change the situation seemed to be used to deflect attention from patients' affective responses to it. This was also evident as P11 discussed respiratory change; he explained:

> It doesn't make no odds whether my breathing goes or not – well, it's going to go isn't it, so...
> **How do you feel about that?** Nothing I can do. Can't change that. [...] Nothing I can do with the MND, is there? It's gonna happen no matter what. (P11)

This response captured the sense of fatalism that many patients identified in the situation; it was understood that there were aspects of the illness that rendered patients powerless and vulnerable; disengagement was part of accepting this position and protecting oneself from it. When asked how he *felt* about change, P11 only made reference to the inevitability of the situation and not to his emotional response at all. In suggesting that it made "no odds" whether his breathing declined, P11 seemed to imply that his emotional responses to changes were negated by their inevitability; P11 was aware that changes would occur, yet his answer gave no indication of any emotional connection. He appeared either unwilling or unable to identify and express this component of the experience or perhaps felt that his emotional response was self-evident in the facts themselves.

It should be noted that a strategy of selective disengagement did not mean that negative feelings were absent; it appeared more probable that patients had simply elected to focus on the fact that there was nothing they could do as a *response* to distress. It seemed most likely that patients were aware of negative affect – or the potential for it – and so sought disengaging strategies to ameliorate these reactions. Patients made frequent references to the fact that there was 'nothing' they could do, which seemed to have become a reassuring

mantra cited to create a comforting distance from change. This was also a way to deflect questions about emotionally sensitive subjects, as a number of patients skilfully demonstrated.

Getting On with It (Subtheme)

Where patients sought to disengage from the illness, they often consciously sought to re-engage with life. A number of patients (P3, P4, P11, P12, P15, P16, P19, P20, P24, P25) described themselves as 'getting on with it'; this idiomatic phrase was used, in varying ways, to demonstrate that the illness was not impeding patients and was, as far as possible, being put aside to enable them to move forward. This sentiment revealed patients' positive ideals of *living* with MND rather than dying from it; however, it also appeared as a further reflection of disengagement and avoidance. The illness was acknowledged where it needed to be, yet in most instances patients were happier to 'get on' in a way that excluded the illness in a deliberate way.

For many patients 'getting on with it' displayed purposeful attempts to shift the focus of attention away from the illness and back to more positive experiences. These patients (P3, P5, P7, P12, P16, P17, P18, P24, P25) successfully learnt how to integrate important aspects of an 'old' life into a 'new' life with MND, complete with its challenges and rewards. P11 explained "I've just been getting on with it, just living with it..." and it seemed that most patients concurred with that sentiment. P16 was determined that the illness would not prevent him from enjoying the aspects of life that he found meaningful; he was proud of his "positive attitude" and endearingly explained:

> The thing with life, you see, is I do not like stopping it and doing what I enjoy. [Wife] has got a sister you see, and she's 83 and she just sits in a chair and never goes out the house. I just couldn't do that. I get bored if I'm stuck in the house. Like you, you're here talking to me and you're keeping me interested, you know? I hate to be left on my own in the house or in the hospice. **Yeah?** I mean, the disease, I wish I never had it but I just wanna get on with my life as anyways. (P16)

For P16, being able to "get on" was desirable because it implied a focus on the positive (i.e. life) rather than the negative (i.e. illness). Engagement with people and places kept him occupied and interested. Patients who were "getting on with it" were not dwelling on the illness and wanted to communicate this fact.

Interestingly, P25 inquired about the wellbeing of Stephen Hawking during his interview, as it had recently been reported that he had been hospitalised. He concluded: "I don't know whether he's better or not. I expect <u>he</u> must be getting on with it too." This gave the impression that this was what *all* patients did, perhaps regardless of choice. P25 expressed a belief that Professor Hawking was "not really living", which may have indirectly revealed his own view that one could be getting on with simply being alive and dealing with the illness, rather than actually thriving and living a full life. For some patients (P15, P21, P26, P22) an expression of 'getting on with it' did seem to be a less positive descriptor. These patients appeared to apply the term to show that they were not succumbing to psychological distress despite the size of the challenges they faced. These patients had higher levels of physical disability and might be said to have been 'getting by' as much as 'getting on'. P15 explained:

> I haven't had any really bad episodes where I'm screaming in agony for a while now. Still have the odd fall over, but that's normal, that's how we got to know something was wrong, me falling over... I can't sit down for too long and I have to get up and I'm really bad screaming. ...never do anything by half. Go the whole hog. But, I just get on with it. ...that's life isn't it? (P15)

P15's remarkable acceptance echoed descriptions of MND patients as 'stoical' in the face of extreme adversity. Her description of events and her responses to them felt somehow incongruent, which seemed to reflect the way that cognitive appraisals could appear dislocated from emotional responses in patients' narratives. It seemed that patients like P15 were not stopping to think about challenging situations, they were just aiming to get through them. It is likely that disengaging from the illness was helpful in this sense; patients were just getting on with what they had no choice but to get on with.

Finally, erring towards the more avoidant edge of the disengagement described, two patients (P14, P4) seemed to view 'getting on with it' as a strategy more akin to denial; they were trying to 'get on' with a life *without* MND. Lower levels of physical and functional disability meant that this strategy was more readily available to these patients. Significantly slurred speech was the only notable sign of illness for P4; he was extremely frustrated by the fact that medical appointments and assessments interfered with his attempts to 'get on with it'. He explained:

> Now I know what I got, just let me get on with it and deal with it. I want to carry on. **Yeah?** I don't even want to think about it [becomes upset] [...] I just want to get on with it; just like I was before. **Yeah.** I'm sick of it. ...**there's been a lot of change?** A lot of change. A lot of hospitals, doctors, MND. I just wanna get on with it. I want it to just – I'm sick of hearing about it. Hearing about it's worse than the MND [laughs exasperated]. All the time. I wanna forget about it. (P4)

For P4, 'getting on with it' seemed to be an alternative to actually confronting the reality of the situation; he wanted to escape his illness. He was visibly upset as he explained his attempts to go back to life as it was *before* his diagnosis, being staunchly against any change to his routine. The idea of things being "normal" was very important to P4, who stressed on a number of occasions that "I just want to be treated normal" The idea of being treated "normal" implied being treated like somebody who did not have an illness; this reflected his desire to exclude all traces of MND from his life so that he did not have to acknowledge the implications of his diagnosis. P4 had returned from a holiday just prior to his interview and reported:

> She [doctor] said 'If there's anything we can do to help you, let us know'. **Right?** And I said 'Send me back to Spain' [laughs]. **[Laughs] Send you back to Spain; that would do the trick?** Oh it was good in Spain; no worries or nothing. I just walked round in the sun. No doctors, no MND, just the sunshine. ...it was good. Over here though, I come back to reality; hospital and tests and breathing and all this. And people can't even understand me. [...] I'm more normal over there. (P4)

P4 wanted to be left alone to 'get on with it' so that he could be "normal" again and so that he could indulge his denial. Tellingly, he suggested that in Spain there had been "no MND", which was a psychological construct and a product of his disengagement. He would not be able to live without acknowledging the illness forever, yet whilst he *could* sustain an illusory MND-free life, he wished to live it. This was a more absolute disengagement. All patients had defined a level of engagement/disengagement that they found comfortable and reassuring. These differing approaches were all focused on avoiding psychological distress and promoting wellbeing. What was clear was that most patients had elected to disengage from the illness on some level.

You Just Don't Know (Superordinate Theme)

The second superordinate theme describes the way that many patients seemed to focus on uncertainty. One would be correct in asserting that a diagnosis of MND implies an assured and irrefutable fate: physical degeneration and hastened death. However, within the constrictive parameters of this future, patients were bombarded by *un*certainties; how fast the illness would progress, which parts of the body might be affected and in what order, how and when the end would come – all were ambiguous and contentious elements of a future that seemed far from assured. Even though patients might be expected to have struggled with the certain threat to life that MND inflicted, many focused predominantly on the *un*certainty that lay ahead. P13 explained:

> ...she [doctor] said to me that everyone is different, you know, where MND hits and all that, so I guess you could say, you just don't know. I mean, you don't know when it's going to hit you where and if it will hit you there at all. [...] She did say that it doesn't affect everyone in the same way – so I thought 'Okay, it may well do or it may not'. (P13)

Many patients seemed to build their coping around the fact that the future was 'unknowable'. Generally, illness progression did not offer patients any further assurances; thinking back to diagnosis, P21 reflected: "I didn't know what to expect..." before adding amusedly "I still don't! [Laughs]" Impressions of the future remained precarious and even patients in the latter stages of the illness were confronted by uncertainties. Often patients seemed to strategically use uncertainty to reduce or avoid potential distress.

This superordinate theme contains two subthemes; the first subtheme, 'Avoiding the Unknown', describes how patients used uncertainty to justify avoiding questions about the future, which they seemed to find comforting; the second subtheme, 'Hope in Uncertainty' explores how an uncertain future allowed patients to invest hope in the prospect of positive outcomes.

Avoiding the Unknown (Subtheme)

Uncertainty justified a degree of avoidance and many patients (P4, P7, P10, P11, P13, P14, P16, P18, P19, P20, P22, P25) actively *deflected* questions about the future on the grounds that "...you don't know how things are going to go..." (P13). For some patients, an unpredictable illness trajectory was presented as a legitimate reason not to contemplate – even hypothetically – the negative changes that potentially lay ahead. Indeed, this

was often considered a 'pointless' endeavour. These patients insisted "...you just don't know, so what's the point of going into it?"(P7) and "...I don't know what's down the line. ...It's almost like there's no point thinking about it now." (P18) Most patients presented themselves as *unable* to talk about the future; this reluctance was not restricted to discussions of physical changes and illness progression, it often included an unwillingness to answer questions about how they might *feel* should certain situations arise, perhaps part of a wider reluctance to address emotional responses to the illness. When P16 was asked about his breathing and how he might feel if it transpired that there had been a change in his respiratory status, he was defensive in answering questions, stating: "I don't know anything in three months, six months, twelve months time, do I? And with you coming, you know, and the tests, we'll just wait and see." Although P16 had been asked about his *feelings*, rather than being asked what might happen, he was still reluctant to delve into the uncertain future to answer the question.

This strict "wait and see" policy was common; facilitated by the unpredictable illness trajectory and seemingly motivated by a desire to avoid distress. In some cases it seemed that patients were not sure whether they had the cognitive, emotional and even physical resources to cope with looking ahead if there were no guarantees. P22 simply seemed to lack any incentive at all, asserting: "It's better to wait and see how things go. ...because you never know. You can't say what might happen around the corner. I can't be bothered really knowing about that yet. **Yeah.** So I'll wait and see." The idea that P22 could not be "bothered" suggested that looking ahead could be demanding, involving effort, investment and, potentially, distress. Motivation to look to the future was naturally low given that all patients knew that physical degeneration of some form would dominate the view. It seemed "better" – preferable – to wait until things happened; that is, to wait until the future became the present and shifted into certainty. This allowed patients to deal with what was actually happening rather than what might happen.

Patients who were generally more avoidant of the illness used uncertainty to justify their more comprehensive disengagement; they did not *want* to think about such a foreboding future and uncertainty provided grounds not to. P4 was prone to using this strategy more than most, explaining his avoidance as a direct consequence of uncertainty. When asked about the future, he explained:

> I don't want to <u>think</u> about that before I <u>have</u> to. [...] I know my mouth - my voice - is gone to the MND, but I don't know if it will get to my legs or my arms. You know, I just don't know. So I don't know how I'll be, so [I] don't think about it. (P4)

It seemed comforting to suggest that particularly negative aspects of the illness might not happen or at the very least were a distant prospect. Not addressing these difficult issues appeared to enable patients to avoid distress in the short-term, as patients seemed to use uncertainty as a shield against a feared future.

Hope in Uncertainty (Subtheme)

Although one might assume that it would be challenging to maintain hope in an illness like MND, most patients did seem to sustain some form of positive expectation. Few patients claimed to hold realistic hopes of a cure, yet a small minority did intimate that they had not entirely abandoned this ambition; most patients

focused hopes on maintaining the physical and functional status quo and living for as long as possible. In this domain, uncertainty created the potential for positive expectations and aspirations to thrive; an unpredictable illness trajectory allowed patients to invest in specific hopes about the path that the illness might or might not take and this was often a comfort.

Prior to diagnosis, uncertainty had appeared as an unbounded threat, which increased anxiety, yet the uncertainty that stemmed from the unpredictable illness trajectory seemed confined and understood. This uncertainty existed within the clearly defined parameters of a diagnosis that had already demarcated a negative extreme. As P18 suggested, "...they've already told me the worst thing, there's not a lot else they can throw at you." From a point of things already being the "worst" they could be, uncertainty seemed to lose some of its ability to evoke fear and could instead be reframed as the possibility that positive alternatives (e.g. prolonged survival) might emerge. Given that diagnosis and the accompanying prognosis had already presented patients with a worst-case scenario, it seemed that many patients concurred to some degree with P11's reasoning that "...you gotta hope for the best, haven't you?"

When P7 was asked how she felt about the future, she suggested: "Oh, just not looking forward to it, thinking 'Oh god', that's it really. Having to be fed and different tubes and machines for breathing... Oh my god. But it might not come to that, you never know." Appending her statement with "you never know" seemed to have been an attempt to placate P7's own concerns; it served to justify the hope that these things might not happen, even if it was likely that they would. This final statement appeared to have been hastily added when the content of her answer started to overwhelm her. A number of patients used the unpredictable illness trajectory to console and reassure; referring back to this uncertainty when they felt themselves starting to worry about what the future might hold. P7 continued:

> I know I'll deteriorate, but in saying that, in the magazine that you get from the MNDA there are people who have various – some people have seven years and there's a chap in the magazine from Australia and he's had twenty-six years and they've give him three, you know, you just don't know... (P7)

Again, where you "just don't know" there was hope and P7 directly countenanced the acknowledgement that she would experience physical decline with hopeful examples of others who had lived for long periods regardless of poor prognosis. Favourable accounts of the illness added credibility to hopes, affording them legitimacy, and other patients (P3, P11, P25) also used this type of information to bolster hopes of survival. Evidence of positive illness experiences may have persuaded these patients that they were not merely indulging in 'wishful thinking' when they hoped for the optimistic outcomes, that an uncertain illness trajectory encouraged.

Hope seemed to *evolve* in MND, often facilitated by some form of uncertainty. Prior to diagnosis, P19 had hoped that her illness would prove treatable, yet even when she was informally told that MND was likely to be the diagnosis, hope endured. She recalled:

> Yes, I was told that it was MND but they couldn't write that officially on my records yet, but it was that any way. [Long pause] But it being unofficial sort of thing, well, made me feel a bit suspicious about it all really [laughs], a bit hopeful, like maybe it isn't if they can't say for sure, but then [doctor+] in the April was able to make it all official and it's just gone along that path since then. ...Yeah, I think you always hold out that bit of hope until they tell you it is actually MND - officially. (P19)

Even post diagnosis, P19 explained that hope remained where uncertainty existed; she suggested that there was something innately "human" about the need to keep positive expectation alive:

> ...I was [laughs] I was hoping it was a mistake. But as it's turned out it's not. I suppose that's a little bit of human nature coming out, isn't it? You just hope it's wrong and you're going to be okay. But it was just difficult to accept it at first and I really, well, I *wanted* them to be wrong. But maybe I knew in my heart of hearts that they weren't. (P19)

Hope was a robust concept that could be maintained in the face of rational acceptance to the contrary; although on many levels P19 "knew" that her diagnosis was not wrong, this did not extinguish all hope, which is perhaps a necessary component of coping when confronted with one's own mortality. It might be suggested that this type of hope protected patients. P19 suggested, there was a "need" to believe that she was "going to be okay" that is perhaps fundamental to psychological wellbeing. Acceptance for P19 – as for P25, P16 and P15 – only came when physical symptoms progressed and forcibly removed any doubt that the illness *was* MND. P19 seemed aware of the seemingly paradoxical state of 'uncertainty within certainty' and used this complex situation to channel her hopes towards specific aspects of illness progression. She explained:

> So, well, we'll all just have to see what the future brings. ...let's just hope that it won't all go downhill too fast; you know, that it doesn't happen quickly. Especially with the breathing, let's hope I'm still doing alright when I next see you. You just don't know, but you can keep your fingers crossed for that much [laughs]. (P19)

This seemed to be a different form and quality of hope, a more modest aspiration tinged with a slightly more defeatist air, yet it represented "hope" nonetheless. Like many patients, P19's hopes seemed to have become less ambitious; P19 focused more on negative events not happening, rather than positive events occurring, yet in many patients this type of aspiration seemed sufficient. The unpredictable illness trajectory provided space for patients to prepare for the worst *and* hope for the best, even if it seemed that expectations might shift over time.

'One Day at a Time' (Superordinate Theme)

The third superordinate theme stands alone without subthemes. This theme addresses patients' preference towards coping on a 'day-by-day' basis, adjusting to change *as* it occurred rather than looking ahead. This limiting technique has clear links to the disengagement of the first superordinate theme and the avoidance expressed in the second superordinate theme, with the notion of uncertainty again being important.

The unpredictable illness trajectory meant that often patients could not look to the future to prepare for *specific* changes or events, only the general direction of change. It was seemingly to cope with this ambiguity that most (P4, P5, P7, P10, P12, P15, P16, P18, P19, P20, P21, P25, P26) preferred to operate, both cognitively and emotionally, within an increasingly limited temporal sphere, only living in the tangible and assured present. Many patients had clearly estimated a safe radius of time within which they felt they could exist and plan and were, as P12 described, "...just getting on with it, one day at a time." It was considered favourable to just "...take each day as it comes..." (P12). This approach seemed to lessen the psychological impact of the illness *and* break down the practical challenges that it presented; it reduced the magnitude of uncertainty and closed down the illness parameters that needed defining.

It seemed that most patients had decided that attending to everything that might potentially happen over the course of the illness would be emotionally overwhelming; thinking about the future was a prime cause of distress and the 'one day at a time' approach was a form of protection. Patients knew that the future held further physical decline and this was sufficient to deter most from probing further. P7 explained:

> I think I would be a nervous wreck if I knew half the things out there. Far better to deal with what you know, what you can see, what's happening to you really. ...you think 'Oh God!' you know? I'd just rather wait and see. [...] Just take it and deal with it as it comes. That's what I thought, just deal with it. That's it really. (P7)

P7's ideas about the future and the threat that it posed are revealed in her suggestion that pre-empting changes would leave her a "nervous wreck"; just dealing with what was happening seemed to reduce this threat and make the illness itself appear more manageable. This seemed to help patients to maintain psychological wellbeing. It seemed that many patients conceptualised coping resources as finite and sought to focus precious efforts on problems that were of immediate concern, rather than wasting them on hypothetical problems that may or may not actually emerge. P10 explained in good humour: "I think I've got enough going on for me to worry about without having to think about every possible problem in the road ahead." With so much "going on", patients did not feel it would be constructive to look to the future where more challenges awaited them; many, like P10, were already facing significant physical challenges in the present.

It seemed that patients wanted to be able to engage with life and enjoy the present as much as possible and felt that pre-empting change by looking ahead would cause the negative events of the future to pollute the present. This idea was expressed by P19, who explained: "I think anything is easier when you don't know what's coming, rather than knowing what's coming and waiting for it to happen." It seemed that it was both knowing about change and then anticipating that change that patients wanted to avoid. Looking ahead was seen to be inviting an imposing future into patients' lives earlier than necessary and most patients preferred to do only what needed doing and only to think about what really needed thinking about. They did not deny the illness absolutely; they merely disengaged from the future by trying to only consciously acknowledge the present. This strategy directly appeased anxiety, as P4 explained:

> I don't want to know about what's going to happen - in the future. ...[I just want to] deal with it now; like my voice has gone worse, so I've gone more on the computer and I email and things and [mimes texting on his phone]. I deal with it. [...] I don't want to know anything. It isn't worth it worrying about that so I just think if I don't know, I can't worry and I can't get [unclear] and stressed, 'cause it stresses me out. **It stresses you out to think about it?** [Nods head] So I don't [laughs]. (P4)

For P4, the future was a somewhat traumatic prospect; he felt he could cope better, both practically and psychologically, by living in the moment and avoiding what was yet to come. P4 was coping successfully with the illness in its current state – which involved small, manageable changes – yet the future seemed to be something more abstract and uncontrollable. This made it sinister and 'stressful'.

Focusing on what was currently happening or appeared likely to happen in the immediate future allowed patients to find a high degree of certainty *within* the uncertainty that existed. This meant that they did not waste practical resources, such as time and energy. Patients were far more likely to adjust to physical and functional change *as* it occurred rather than planning ahead. P7 continued:

> I don't know a lot about it but I just know that it's terminal but you don't know when. It depends on your deterioration and that's about it really. I don't think you can really pre-empt it, you know, what's going to happen. You think 'Okay, well, I'll just wait and see'. So that's been my attitude. I'll just keep going. [...] I'm quite positive you see and so I just try and find ways around doing things, take it easy, you know, don't do this and - I just really thought, well, adjust, basically, adjust. (P7)

Most patients wanted to *adjust* to change rather than prepare for it. By dealing in increments of change rather than absolute losses, patients could avoid seeing these smaller changes as part of a more significant transformation. This also made 'adjusting' to change easier, as it became a process of continual minor amendments rather than intimidating step-changes. It seemed that patients did not want to contemplate adjustments that might be made in the future nor dwell on how much change had already occurred; they remained resolutely in the present.

A further impetus for living in 'one day at a time' related to issues of control; this approach was often seen as the most effective means of retaining mastery over the illness. This seemed to be inherent in P7's feeling that if she knew too much she would be a "nervous wreck", which depicted a lack of confidence and control. This was an issue of great importance to P25, who explained:

> I mean I don't have any problems about asking [about the illness] and nobody has any problems telling me either. But it's when I want it; in my own time and on my terms. ...that's a great comfort. That puts me in control, which - in this illness you must try and retain that control or it will run away from you. I decided early on that I would be sensible about this and I would approach it in this way, just step-by-step and day-by-day. (P25)

Approaching the illness this way seemed to deconstruct it into less daunting portions; this allowed patients to confront MND in 'steps', rather than trying to control all elements at once. In an illness that threatened self-efficacy on so many levels, most patients sought to actively maintain control wherever they could. This approach seemed to have been encouraged by the nature of MND itself, with its progressively degenerative course occasionally seeming overwhelming; P19 compared the illness to a "steam train" suggesting:

> You know, takes a bit to get going but then once it picks up pace, you're pretty hard pressed to stop the damn thing [laughs]. I don't know, I suppose [exhales] sometimes it seems a bit out of your control. (P19)

Breaking down the illness into single days of MND seemed to reduce the size of the challenge and to protect a sense of mastery over the illness. P25 was also aware of the threat that his illness might "run away" from him if he did not approach it strategically, this suggested that an underlying anxiety was being moderated by this coping strategy. Indeed, P25 had purposefully "decided" to adopt this approach "early on" to be "sensible" about how he coped with change; this suggested that this means of coping was the result of a rational and pragmatic appraisal of the problem.

One may see this 'one day at a time' approach as an example of selective disengagement; patients wanted to shut out thoughts of the future, yet would engage with the present and with problems that needed addressing. Most patients were adjusting well to the changes that the illness incurred, however, unless change was tangible and seen to encroach on everyday function. Many patients were happier not to attend to it or to 'pre-empt' its progression. It is often pondered how MND patients can avoid succumbing to significant psychological distress when they are faced with such a devastating future; in this sample, living 'one day at a time' was a significant coping strategy. This approach enabled patients to feel that they did not have to face MND in its entirety, they only needed to address the challenges that MND posed at that time and on that given day. These challenges – the practical day-to-day challenges, such as negotiating the stairs or finding alternative modes of communication – were smaller and more manageable. The full ramifications of the illness as a whole appeared 'boxed off' and hidden away; these were not to be avoided forever but did seem to be evaded in the short-term.

Fighting Spirit (Superordinate Theme)

The third superordinate theme describes a more determined attitude not to be 'beaten' by the illness. Most patients accepted the consequences of physical degeneration and adjusted well to lifestyle changes. However, this did not mean that all patients submitted passively to the illness; in reality, many revealed rather determined attitudes. A number of patients understood that their influence within the physical domain was limited, yet adopted the view that there was 'no harm' in trying to maximise the likelihood of positive outcomes. These patients developed a mentality aimed at winning a psychological battle and, tellingly, six patients (P3, P15, P16, P17, P25, P26) explicitly positioned themselves as 'fighting' the illness. In most cases this was little more than an attitude or gesture, yet it often seemed to play a significant role in maintaining patients' wellbeing over time.

This superordinate theme contains two subthemes: the first, 'Not Letting It Win' describes a determination not to surrender to the illness, with patients differentiating effects of the illness on the physical body from those on their spirit and sense of self. The second, subtheme, 'Keeping Going' outlines attempts to physically fight illness progression by exercising and keeping muscles in use; patients conceded that this was probably futile, yet felt comforted by these positive actions.

Not Letting It Win (Subtheme)

In some patients, combative imagery dominated discussions and defined coping strategies; although all patients appeared to have accepted the illness, this did not preclude them from wanting, as P26 explained, to "fight it" for as long as they could. P15 presented her experiences as a dialogue between herself and the disease, using this approach to vent frustrations and confront negative aspects of change. Describing her reaction to illness progression, she recalled:

> I said 'You're not getting the better of me, you can go and run'.... when the voice started going and the breathing started picking on me 'Ah, fishcakes! You've caught up with me already – it's only been five minutes' and you sort of like, resign to the fact. But, I ain't giving in and it ain't gonna win. So, until something else kicks in 'You're still not gonna win' [directed at own body]. (P15)

P15 conceptualised the illness as an opponent and adversary rather than merely a presence or process; personifying it in this way allowed her to communicate her feeling towards it. By directing the sentiment that the MND was "not gonna win" at her own body, she demonstrated the complexity of the 'fight' in which many patients engaged; MND was an enemy within. There was a clear sense that the illness was gradually encroaching on the body and the physical self was slowly being eroded; most often, it was only in the psychological domain that patients were *able* to fight back. In many cases this was expressed where patients determinedly continued to perform a function after muscle weakness began to make the process more challenging; this reassured patients that they were not "giving in". By framing each area of physical decline as an individual fight against the illness, patients were able to keep fighting in the face of illness progression; this also meant that they could address smaller increments of change rather than trying to fight an overwhelming tide of physical adjustment.

Being able to verbalise her determination and to channel this directly at the illness seemed a powerful coping strategy for P15; she defiantly continued:

> I say to the illness 'No you don't, you're not getting me'. [...] ...I won't let it get me. I say 'Motor neurones? Fuck off!' [Laughs] I'm gonna fight it all the bloody way. ... I won't let it beat me. It may have got me, but there was nothing I could do about that. It can have a go at me body but it's not getting me; I won't let it. This disease can fuck right off [laughs]. (P15)

The strength of P15's feeling was evident in the language used and, although she laughed intermittently as if to diffuse the tension, there seemed to be a genuine anger and frustration motivating her confrontational

approach. Increasingly, the physical body was portrayed as separate from the 'true' self; MND could attack the body, but P15 felt that the she ("me") could not be eroded in the same way. This was part of the illness that she could control and the reason that the MND would never truly "get" her. For all that patients could *not* change; their sense of self was often something that they maintained mastery over. Putting effort into the 'fight' was an attitude rather than an action; however, it enabled patients to feel that there was still something that they could do, creating an active rather that passive role within the illness experience for those who wanted it. In many cases it seemed as if the simple act of being able to state that one was 'fighting' – even if this was an empty or ineffective gesture – had a significantly positive psychological impact.

The dichotomy between the 'self' and the physical body seemed to be important in enabling patients to disengage from the illness and retain control. Maintaining the sense of self seemed to be a significant motivation in patients' 'fighting spirit'. P25 reported:

> ...psychologically [long pause] I have not got a downer on this thing at all. It's not that I don't give a monkey's but I do my best to hold my head up and not let it beat me... ...it isn't going to get my mind, my spirit, you know? ... I'm just gonna hold my head high for as long as I can. (P25)

P25 was determined not to let the illness "get" his mind and spirit, which showed how the fight to protect the emotional and psychological became a significant part of the illness experience. If there was nothing that patients could do to stop the physical changes, they could at least seek to protect these aspects of self. Low mood and having a "downer" on the illness also seemed to be equated with defeat; for P25, "letting" the illness have that impact was tantamount to letting it "beat" you. As such, maintaining psychological wellbeing and displaying a positive attitude were pitched as the fights that patients *could* win and this was a means of asserting defiance. P25's approach appeared to have been carefully crafted; he acknowledged that a positive attitude was not necessarily a natural reaction and that it was not that he did not care, yet he asserted "I do my best to hold my head up." This indicated that he consciously invested efforts in not succumbing to distress. Holding one's head up is the determined act of someone who might reasonably have been subdued by negative circumstance; it is a show of courage in challenging times and suggests bravery and determination.

Keeping Going: The Physical Fight (Subtheme)

Only a small number of patients (P7, P15, P16, P17, P25) explicitly extended their fight to the physical realm, doing so by trying to increase physical activity. This was a more literal fight against the illness. P17 revealed that he targeted potential muscle loss at the gym, explaining that he was:

> Not getting weaker, lifting more weight. Don't want to lose the muscles, keep using them and building up. **Right, so you're going to the gym now to try and keep the muscle there.** [Nods head] Important to fight it. Hopefully slow it down. (P17)

Similarly, P7 revealed:

> I'm going to these motorcise beds. **Oh right?** I do them because I feel that that's my muscles working without me having to do it, you know? If you go the gym you're knackered, you know what I mean? **Yes.** And you don't need to, you go for half an hour and all these machines are working all your muscles. So, I try that, you know? Because if you don't it's like, it atrophies doesn't it, if your muscles don't work. So I'm giving them something to do, keeping them all going, you know? And that's the kind of thing – it's all you can do. So there you go. So I'm looking after myself [laughs]. (P7)

Patients wanted to do *something* and any attempts to slow the illness were preferable to no attempts – this was *despite* patients acceptance of the fact that there was 'nothing' they could do to influence the rate of physical degeneration. In this way, this physical strategy was actually initiated to placate psychological unease at the situation. Patients often played down hopes and ambitions, seemingly to avoid disappointment; however, a small number seemed to feel that there was 'nothing to lose' by physically trying to slow the rate of decline. P25 confirmed that these physical efforts were actually a component of a wider psychological strategy, explaining: "...it's winning that mental battle as well as the physical one. Part of the going to the gym and working out and fighting the MND is tied up with psychological wellbeing for me." Being able to declare that they were 'fighting' the illness in one way or another meant that patients were not passive victims of circumstance; it created the illusion of power and control and patients seemed to need this psychological boost.

Positive & Negative (Superordinate Theme)

This superordinate theme explores the 'affective valence' of attitudes towards the illness. It contains two subthemes: the first subtheme, 'Staying Positive', describes the importance that patients attributed to a 'positive' attitude; the second subtheme, 'Selecting/Controlling Responses', reports the way that a number of patients tried to actively avoid being 'negative', even in the face of significant challenges. These were considered to be two related but separate responses. Patients were not simply trying to be positive so that they could avoid negativity; a positive attitude was seen as desirable in its own right. However, negativity was seen as a threat to the positive attitude that patients aspired to *and* an undesirable outcome. There appeared to be a careful process of selection and control between positive/negative responses to the illness.

Staying Positive (Subtheme)

The importance of being and staying 'positive' consistently emerged in patient narratives and positivity was an ideal that a number of patients suggested they aspired to (P3, P5, P7, P15, P16, P18, P19, P21, P25, P26). When first learning about the illness, P18 suggested that for all of the negative information she received, she always "...tried to put a positive on it..." Looking for positives and being positive seemed to be ways of protecting patients from distress, yet positivity also appeared to be a desirable state in its own right. Even with limited communicative ability P26 stressed that one must "stay positive", whilst P21 directly stated "Stay positive or the MND will win. I won't let it win. [Laughs]..." Such statements showed how patients constructed the idea of a psychological victory over the illness through cognitive and emotional channels. P25 felt that being positive was "vital" in MND and was keen to stress "You won't find many more as positive as

me, I can assure you." He clearly saw his positive attitude as a laudable quality and this was felt to give him an advantage within the context of this illness.

Two patients (P3, P16) also felt that being positive had influenced the physical course of the illness. Having lived with the illness for many years with relatively moderate physical impairment, P3 attributed his impressive physical status to his positive attitude:

> I think that's why I'm doing so well now, because at first they didn't think I would be like I am now, they thought I'd be a lot worse. I think being positive has kept me going and made the illness not as bad. [...] ...there's so many people that, like, are in a worse position than what I was after two years. I'm going into my seventh year now, so I think keeping positive has helped. (P3)

P16 expressed a similar view, explaining:

> I wish I didn't have it but it don't seem to bother me because I think that I'm very positive. [...] I think 'Thank God I've been positive, so it hasn't affected me that bad.' [...] I'm doing okay cause I'm positive see. That's the key to it. (P16)

It appeared that attributing moderate physical symptoms to a psychological approach allowed these patients to feel that they maintained influence and control over the situation. Patients felt that winning the psychological battle over MND had allowed them to advance in the physical fight. This seemed to have been comforting; however, it is not clear how effective this type of coping strategy would be over time and as physical degeneration spread in spite of their positivity. It seemed as if patients viewed positivity as both a means to an end *and* an end within itself; many simply saw a positive attitude as being an inherently good thing to have and being positive was often a calculated strategy.

Selecting/Controlling Responses (Subtheme)

A number of patients spoke of the importance of not 'letting' themselves succumb to negative affect (P7, P15, P20, P21, P22, P25). Curiously, this sometimes presented psychological states as choices. P21 revealed: "I never let the MND get me down. It can be scary but I stay very positive." There was a sense of incongruence between the subjective experience and the overt psychological response; P21 suggested that she stayed "very positive" even during the "scary" times and never "let" the illness get her down. This might reasonably be taken to suggest that negative emotions were being stifled or even repressed. Patients seemed to try and 'overwrite' negative emotions with a positive facade because being positive was itself desirable.

Much is made of MND patients' appearing stoical in the face of adversity and this certainly emerged from data. P21 described herself as "...stuck between a rock and a hard place." before adding "Never let it bother me though..." Again the idea that patients exerted some form of control over these reactions was apparent; indeed, there were multiple references to the idea that there was choice or assertion in this regard. P3 explained: "I don't <u>let</u> myself feel sorry or down." and likewise, P15 stressed:

> I'm not letting it get me down – see this [points to face] I'm still smiling [laughs]. And I'll keep smiling, you wait and see. Positive – mental – attitude; that's what I got. [...] Without a positive attitude I don't think I'd be this buoyant. I'd be more down in the dumps and depressed if I let myself, so I don't let myself be. I won't bloody get like that. (P15)

Although this attitude allowed patients to maintain an outward display of psychological wellbeing, it did at times feel forced. Patients appeared aware that, objectively, the situation could – and maybe *should* - incur distress and some seemed to maintain a positive approach to avoid that distress rather than being positive for its own sake or as a natural response. Patients could not do anything about the illness; however, they could make the choice of whether to live with MND in a positive way or live with it and also be low in mood or angry about the situation. A number of patients seemed to have *elected* to remain positive as a means of opposing the illness and this seemed to be a further strand of avoidance.

Indeed, many patients stressed how important it was that they did not fall into a negative mindset; in one of the only intelligible sentences that P23 was able to communicate he revealed that he often reminded himself: "There's always someone worse off than you." This seemed remarkable coming from a patient with such a high level of physical disability, yet it seemed important to patients that they did not allow themselves to wallow in negativity. P7 also described herself as "positive" in her approach to the illness, stressing "...there's no point worrying about it." Indeed, the idea that there was "no point" being upset about physical change was used to explain a lack of negative response and appeared to be part of the reason why patients would not "let" themselves succumb to it; P15 suggested: "...that's life isn't it? No point in crying over that – what a bloody waste of tears." These patients suggested that since there was nothing they could do to change the situation there was little point in becoming upset about it. Often patients emphasised the fact that negative emotional reactions would not objectively change anything. P15 continued:

> I think 'Yeah this is quite bad isn't it? But will you make it better by sulking? No. What's the point? I get by okay. So I won't let myself be down. I will keep smiling and keep laughing right until the very end. [...] If I feel myself getting down about it I say 'Don't you dare!' [Laughs]. (P15)

Objectively, P15 recognised that the situation was indeed "quite bad", yet there was a clear sense that her cognitive strategy was keeping her emotional states in check; she appeared to have *decided* not to take the route of negative reactions. She acknowledged that there were occasions where she felt herself "getting down" and had to act to restore her "positive" approach; patients appeared apprehensive about letting themselves indulge in low mood, appearing to fear where that path would lead them. P25 explained: "You'll not change anything by being angry and bitter and feeling sorry for yourself..." Taking a negative path was perceived as potentially harmful and certainly unhelpful. Patients were not immune to distress; however, they did seem capable of purposefully manoeuvring away from negative affect - be that by acceptance, avoidance, denial, repression or any other mechanism that they could use. If MND was going to win the physical battle, patients would not let it win the psychological fight too. They could retain some sense of mastery, control and power by determining how they responded to the illness itself. In this way, the MND did not absolutely "win".

Appendix 7: Research Dissemination

Conference Presentations

Piggin, L.H., Thornton, E.W., Angus, R.M., Chakrabarti, B., & Young, C.A. (2008); Non-Invasive Ventilation (NIV) in Motor Neurone Disease: A Qualitative Exploration of Patient & Carer experiences. Poster Presentation, Dementias & Neurodegenerative Diseases Research Network (DeNDRoN) Annual Conference, Newcastle, UK.

Piggin, L.H., Thornton, E.W., Angus, R.M., Chakrabarti, B., & Young, C.A. (2009); Non-Invasive Ventilation in Motor Neurone Disease: Qualitative Accounts of Initiation and Impact; Oral Presentation, British Thoracic Society Winter Meeting, London: UK.

Piggin, L.H., Thornton, E.W., Angus, R.M., Chakrabarti, B., & Young, C.A. (2009). Non-Invasive Ventilation in Motor Neurone Disease: The Utility of the Epworth Sleepiness Scale as an Outcome Measure; Poster Presentation, British Thoracic Society, Winter Meeting, London: UK.

Piggin, L.H., Thornton, E.W., Angus, R.M., Chakrabarti, B., & Young, C.A. (2009). Patient and Carer Experiences of Non-Invasive Ventilation in Motor Neurone Disease: A Qualitative Exploration; Oral presentation, British Psychological Society, Division of Health Psychology Annual Conference. Birmingham, UK.

Piggin, L.H., Thornton, E.W., Young, C.A., Chakrabarti, B., & Angus, R.M. (2009). Patient and Carer Experiences of Non-Invasive Ventilation in MND: A Qualitative Exploration; Poster Presentation, European Respiratory Society, Annual Conference, Vienna: Austria.

Piggin, L.H., Thornton, E.W., Young, C.A., Chakrabarti, B., & Angus, R.M. (2009). The limitations of quantitative measures: assessing approaches to research into NIV use in MND; Poster Presentation, European Respiratory Society, Annual Conference, Vienna: Austria.

H. Ando, L. H. Piggin, E. W. Thornton, B. Chakrabarti, R. M. Angus, C. A. Young (2010); "For the rest of your days..." an Interpretive Phenomenological Analysis of Motor Neurone Disease Patients who declined or did not tolerate Non-Invasive Ventilation. Oral Presentation, European Respiratory Society Annual Congress, Vienna: Austria.

Ando, H., Piggin, L.H., Thornton, E.W., Angus, R.M., Chakrabarti, B., & Young, C.A. (2009). Health Benefits of Non-Invasive Ventilation in MND: longitudinal study design examining the psychosocial and physical impact on people with MND and their carers. Poster Presentation, Dementias & Neurodegenerative Diseases. Research Network (DeNDRoN) Annual Conference. Southampton, UK.

Ando, H., Piggin, L.H., Thornton, E.W., Angus, R.M., Chakrabarti, B., & Young, C.A. (20010).; The lived experience of patients with Motor Neurone Disease who declined or did not tolerate Non-Invasive Ventilation. Poster Presentation, Motor Neurone Disease Association, Annual Conference.

Academic Papers

Piggin, L.H., Thornton, E.W., Angus, R.M., Chakrabarti, B., & Young, C.A. (xxxx). The Experience of non-invasive ventilation in motor neurone disease: a qualitative exploration of psychological responses to treatment. (Under Review: Psychology & Health).

Piggin, L.H., Thornton, E.W., Angus, R.M., Chakrabarti, B., & Young, C.A. (2012). Non-invasive ventilation in motor neurone disease: a qualitative study of patients' responses to respiratory masks. (In Press: Palliative Medicine).

Grant Reports

This thesis forms part of a wider research project based at the Walton Centre for Neurology and Neurosurgery NHS Trust (Liverpool). It was funded by a grant from the Motor Neurone Disease Association UK (www.mndassociation.org). The full study is entitled "Health Benefits of Non-Invasive Ventilation in Motor Neurone Disease: The Psychosocial and Physical Impact on People with MND and their Carers". Requests for further information from external grant reports should be made to Motor Neurone Disease Association, PO Box 246, Northampton, NN1 2PR or enquiries@mndaassociation.org citing the grant reference: Young/Mar07/6026.

Other Sources of Information

Information about the study is also available from the UK Clinical Research Network (UKCRN), where it is registered in the study portfolio database (http://public.ukcrn.org.uk/search). The UKCRN reference is: 4025.