Decision-making for assisted ventilation in amyotrophic lateral sclerosis

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Abstract

Amyotrophic lateral sclerosis (ALS) is a progressive neurological disease that leads to respiratory compromise and eventually death within two to five years. Even though people with ALS must make many treatment decisions, none has such a significant impact on quality of life and survival as the one pertaining to assisted ventilation. A qualitative research study was undertaken to elicit factors that are pertinent to this decision-making process. Ten individual, semi-structured interviews were conducted with individuals with ALS. Six main themes emerged from the interviews. These are: meaning of the intervention, the importance of context, values, and fears in decision-making, the need for information, and adaptation/acceptance of the intervention. Based on these findings, it is argued that a pluralistic conception of autonomy as well as a shared decision-making model is better suited to give high priority to patient autonomy in this context. Some recommendations to improve clinical practice are proposed.

Abrégé

La sclérose latérale amyotrophique (SLA) est une maladie neurologique dégénérative qui entraîne des difficultés respiratoires et la mort dans un délais de deux à cinq ans. Tout au long de la maladie, les personnes atteintes doivent faire plusieurs choix d'interventions qui auront un impact sur la qualité et la durée de leur vie. Cependant, aucune décision n'a autant d'importance que celles concernant la ventilation assistée. Une recherche qualitative a été entreprise afin d'identifier les facteurs pertinents à la prise de décision dans ce contexte. Dix entrevues individuelles, semi-structurées ont été effectuées avec des personnes atteintes de SLA. Six thèmes principaux ont été identifiés. Ceux-ci sont : la signification de l'intervention, l'importance du contexte, des valeurs et des peurs, le besoin d'information et acceptation-adaptation. À la lumière des résultats, une définition pluralistique de l'autonomie ainsi qu'un modèle de décision partagé sont recommandés. Quelques recommandations sont suggérées afin d'améliorer l'approche clinique.

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Chapter I

Introduction

1.1 Amyotrophic Lateral Sclerosis

Amyotrophic lateral sclerosis (ALS) is a rapidly progressive neurological disease that causes degeneration of both upper and lower motor neurons leading to decreased motor control, paralysis, muscle atrophy, spasticity, speech loss, swallowing difficulties, respiratory compromise, and eventually death within two to five years. ALS affects men more than women, oftentimes in the prime of their lives. It is the most common degenerative disease in adulthood with a prevalence of around 6-8/100,000 (Brooks, 1996). Today, upon diagnosis, individuals are no longer told there is nothing that can be done for their condition. Medical advances including the use of various drugs, provision of early gastrostomy (Kasarskis, et al., 1999; Miller, et al., 1999), assisted ventilation (Lo Coco, et al., 2006; Butz, et al., 2003; Lyall, et al., 2001; Aboussouan, et al., 1997) and a multidisciplinary team approach (Traynor, et al., 2003) affect the course of the illness and enhance the person's ability to remain at home with a good quality of life.

Some cases of ALS are familial but the vast majority of cases seen in North America are sporadic cases. ALS can be divided into two categories depending on symptoms at onset. Individuals who suffer from the spinal form typically have symptom onset in their limbs whereas individuals who have the bulbar form first experience difficulty with speech and swallowing. The rate of progression of bulbar ALS tends to be more rapid than that of the spinal form.

Individuals with ALS typically have severe physical disability interfering with carrying out every day activities. Consequently, patients must rely on family members and home care health professionals to fulfill their basic daily needs, such as washing and feeding. They also require access to a variety of equipment, such as wheelchairs and communication devices. The presence of architectural barriers can severely limit their ability to function in every day activities. Thus, provision of such things as elevators and adapted vehicles is extremely important.

Even though recent studies (Wolley-Levine, et al., 2006; Ringholz, et al., 2005) suggest that individuals with ALS have a higher prevalence of cognitive/behavioral changes than initially thought, at present, the impact of theses changes on competency are unclear. Therefore, for the purpose of this thesis, ability to make treatment decisions is assumed. For it is also the case that, as stated by Borasio & Voltz; "The dichotomy of severe physical disability and unimpaired mental abilities up to the very end is almost unique to ALS" (1998, p.721).

1.2 Assisted ventilation and ALS

Individuals who suffer from ALS usually die from respiratory failure unless they are treated with long-term mechanical ventilation (LTMV) via a tracheostomy. LTMV remains the only means to considerably prolong the life of individuals with ALS (Borasio & Voltz, 1998). Yet, various studies report that LTMV through a tracheostomy remains a relatively rare occurrence with less than 5% of all individuals with ALS choosing LTMV (Gelinas, et al., 2000; Moss, et al., 1993).

Use of LTMV with individuals suffering from ALS raises issues that are specific to the disease. Because individuals with ALS retain their cognitive abilities, use of mechanical ventilation in this situation differs from use of ventilation to prolong the life of patients in a neuro-vegetative state. In addition, the level of disability of persons with ALS varies greatly at the time when invasive ventilation might be indicated.

Furthermore, compared to other neuro-muscular diseases and muscular dystrophy, the progression of symptoms with ALS is very rapid.

Despite medical advances in the care of individuals with ALS, the mainstay of treatment remains symptomatic management (Miller, et al., 1999). Non-invasive ventilation via a mask and a bi-level positive airway pressure (BIPAP) machine can palliate symptoms of respiratory failure. In 1999, the American Academy of Neurology (AAN) published the Practice Parameter: The care of the patient with ALS (an evidence-based review) (Miller, et al., 1999). In this document, the AAN addressed respiratory management and recommended that non-invasive ventilation be offered to ALS patients as an effective therapy for symptomatic management. They recommended BIPAP be offered to patients when they present with symptoms such as dyspnea, morning headache, daytime hypersomnolence or when they present with impaired pulmonary function as demonstrated by a forced vital capacity (FVC) <50%. Following the introduction of these practice guidelines, the use of BIPAP is common practice in ALS care (Lyall, et al., 2000).

Lo Coco, et al. (2006) have shown that patients who can tolerate using the BIPAP for 4 hours or more each day have a twelve-month increase in survival. In contrast, using Riluzole, the only medication approved in the treatment of ALS prolongs survival by an average of only three months (Gelinas, 2000). Nevertheless, many individuals with ALS are unable to tolerate using the BIPAP or simply refuse to try any means to palliate symptoms of respiratory distress.

There exists very little empirical data concerning how individuals with ALS go about making decisions concerning assisted ventilation to guide professionals who assist them in making treatment choices. The clinical consensus is largely based on years of clinical experience in caring for individuals with ALS (Mitsumoto, et al., 2005) and may make assumptions about factors that are important to patients and their families while making treatment choices. Furthermore, the paucity of empirical data may influence how and what information is provided in the clinic setting thereby introducing a bias in the recommendations and impacting patient decisions.

1.3 ALS care in Quebec

The ALS clinic of the Montreal Neurological Hospital (MNH) cares for close to 200 people with ALS. These individuals are mainly from the Montreal region but some come from other parts of the province of Quebec, from eastern Ontario, and the Atlantic Provinces. The patients followed at the MNH ALS clinic have access to a specialized multidisciplinary team to assist with symptom management and liaise with community agencies that provide services in the home. This is in contrast to other individuals with ALS who live in this province and are cared for mainly by a neurologist and home care agencies, such as the Centre Local de Services Communautaires (CLSC), without benefiting from a specialized multidisciplinary approach.

The MNH team, like other specialized ALS centers has integrated the use of BIPAP in its clinical practice and also offers LTMV to its patients. Careful assessment and monitoring of the patients' respiratory status is part of standard practice at the MNH. The professionals working on this team are often responsible for providing information and offering various interventions for respiratory symptom management to the patients. They work in collaboration with the Montreal Chest Institute (MCI) National Home Ventilation Program, which is responsible for the provision of services associated with use of assisted ventilation in the home (CUSM, 2001). The incidence of LTMV for patients followed at the MNH is roughly 3% as opposed to the 5% reported in the literature. Medicare in Quebec covers the actual cost of the BIPAP or the ventilator, the inpatient stay to initiate treatment, and some of the follow-up home visits through the home ventilation program.

1.4 Purpose of the study

As stated previously, there exists little empirical data concerning decision-making and assisted ventilation in ALS. Some studies provide insights into decision-making and assisted ventilation from the viewpoint of health care professionals. Moss et al. (1993) discovered that the percentage of ALS patients on home ventilation varied greatly amongst centers. Their study revealed that: "Physicians' personal attitudes toward home ventilation seemed to account for this difference" (Moss, et al., 1993, p.440).

In an attempt to explore patients' perspectives on the issue of assisted ventilation and examine the process and factors involved in their decision-making, Young et al. (1994) interviewed thirteen individuals followed in an ALS program in British Columbia and developed a list of factors that are important for decision-making and assisted ventilation. This Canadian study was carried out at a time when use of non-invasive ventilation was not yet common practice. Since then, the social views on chronic life threatening conditions have changed and management of ALS and the technology available to do so have evolved. Nevertheless, a review of the ALS CARE database with regards to BIPAP use in North America shows that only a small proportion of eligible patients are actually being offered the intervention (Bradley, et al., 2001). One possible explanation is clinicians' fear of prolonging survival in the face of severe disability (Bourke, et al., 2002). It would appear that physicians and health care professionals caring for people with ALS have continued to question the moral justification of using assisted ventilation with the aim of prolonging life. One cannot help but wonder whether use of assisted ventilation is still decided on an arbitrary basis instead of being based on sound morally justifiable criteria and whether the patients involved are making well informed and truly autonomous decisions about assisted ventilation.

Many studies have examined issues surrounding the use of LTMV and BIPAP. These discussions span the quality of life (QOL) imposed by the ventilator on patients and their families (Kaub-Wittemer, et al., 2003; Polkey, et al., 1999), issues of cost (Moss, et al., 1996), and patient's need for information and control in cases of request for discontinuation (Borasio & Voltz, 1998). Nevertheless, Heiman-Patterson & Miller (2006) suggest that we need to further explore factors affecting intolerance to non-invasive ventilation in patients including claustrophobia, secretions, mask discomfort, lack of caregiver, the enthusiasm of the clinicians, etc. as well as physician's personal views on assisted ventilation.

No study has attempted to widen the general understanding of how individuals make decisions about treatment options for management of symptoms of respiratory failure including the individuals that opt against BIPAP and LTMV since the introduction of the AAN's practice guidelines and the more common use of BIPAP in ALS. Furthermore, no study has been undertaken to elicit factors that are pertinent to the decision-making process strictly from the patients themselves in order to develop a patient-centered approach to decision-making about assisted ventilation even though:

> Respiratory care presents the greatest challenge for the ALS patient and the clinician. Deciding when to initiate non-invasive mechanical ventilation is critical because of the risk of either sudden death or ventilator dependence without proper advance planning. An early understanding of the patient's

> preferences will help ensure careful and timely planning (Miller, et al., 1999, p. 1317).

Thus, in this thesis, a qualitative research study involving individuals with ALS who opt for and against assisted ventilation is undertaken to further explore the decision-making process for assisted ventilation from the patient's perspective.

The purpose of this research is to understand the experience of persons with ALS followed at the MNH ALS clinic in making choices about treatment options to relieve respiratory failure. Through this study, it is hoped that the ALS team of the MNH and other treating teams will have a better understanding of the individuals' perception, understanding, thinking process, decisions, and experience of treatment options for respiratory failure. It is hoped that having a better understanding of the decision-making process will facilitate the dialogue between health care providers, patients, and their family promoting informed treatment decisions and maximizing the potential for patients to exercise autonomy.

1.5 Importance of the problem

Individuals with ALS are faced with treatment decisions throughout the course of their disease. At each new stage, they must decide how to proceed. They must decide whether they want to use Riluzole, whether they want to use a percutaneous gastrostomy to palliate symptoms of dysphagia, or a motorized wheelchair to compensate for their inability to walk. No other treatment choice has such a significant impact on quality of life and survival as the one pertaining to assisted ventilation. In addition, the use of assisted ventilation is so specialized that it raises specific issues.

Symptoms of respiratory failure do not occur solely at the end of life; a small number of ALS patients experience respiratory distress when other muscle groups are only minimally involved. Therefore, the level of disability of persons with ALS varies greatly at the time of ventilation and persons with ALS may need to make decisions about assisted ventilation at any point in the course of the disease making advanced planning difficult. Furthermore, because the presentation and progression of ALS is so variable, some individuals may suffer from symptoms of respiratory failure long before death. These symptoms are extremely distressing to patients and their families and severely affect quality of life. Consequently, patients must be equipped to make treatment decisions at opportune moments to enable potentially useful interventions, such as BIPAP and LTMV to positively impact the person's life.

Compared to the use of a BIPAP, which is non-invasive, often used on a "part-time" basis, and relatively easy for patients to start and stop using, the use of LTMV in particular necessitates more careful consideration. Because of the surgery required, initiation of invasive ventilation through a tracheostomy necessitates that others beyond the immediate treating team get involved. These professionals may further question the patient's decision and will need written consent prior to the procedure. What's more, even though discontinuation of life-sustaining treatment is permitted in Canada (Nancy B. v. Hôtel-Dieu de Québec et al., 1992), it is much more difficult psychologically to stop LTMV once it has been initiated than not to start it at all in the first place.

Nevertheless, establishment of BIPAP in ALS entails different concerns than its use with other pathologies, such as in the case of a respiratory condition where there is no concomitant physical disability. The initial BIPAP trial for an individual with ALS usually requires a brief hospital admission to monitor if it improves the person's breathing, comfort, and sleep and determine if the person can tolerate its use overnight. BIPAP use may relieve symptoms of respiratory failure but it does not stop the progression of the disease. Thus, as the person's condition declines, BIPAP use usually increases to daytime use as well. As a consequence, the specialized home ventilation team must frequently monitor the person's condition and ensure that the BIPAP is used optimally. In addition, initially, the person may be able to use the BIPAP independently but over time, help at home must be available to look after the machine and put the mask on and off once the person with ALS cannot do this alone. Thus, in the case of ALS, choosing to use assisted ventilation may be influenced by the availability of a caregiver, home services, and specialized personnel.

Family involvement in care and decision-making may impact how frequently patients are using assisted ventilation. In our Western culture, little is known about the role that families play in treatment decisions. However, Hirano (2005) argues that in Japan, the large portion of ALS patients who use invasive ventilation may result, in part, from the physician's obligation, as stipulated by their clinical guidelines, to discuss ventilation not only with the patient but the family as well. This obligation results from the Japanese culture's emphasis on family involvement in medical decision-making. In North America, such an obligation does not exist and our approach is centered mainly on respect for patient autonomy. In the case of ALS, both patients and families should be at the center of health care delivery. Oftentimes, they both live with the consequences of the various interventions since the patient must frequently rely on the family to act as caregiver. Since both patients' and their families' interests, values, preferences may impact autonomy, these factors must be of primary concern to health care workers during decision-making.

The principle of respect for patient autonomy is expressed through the doctrine of informed consent, which stipulates that for consent to be valid one must determine that:

1) the decision is made by a competent individual, 2) that the decision is informed, and 3) that it is voluntary. Because individuals with ALS typically retain their cognitive abilities, competence is rarely an issue in treatment decisions. Even so, an informed decision is one where the individual agrees or refuses treatment on the basis of an understanding of relevant information, gives permission for the action (or inaction) to be taken, and is not controlled by influences that would engineer the outcome in terms of what decision was made (Faden, et al., 1986). Little work has been done to elicit from patients themselves factors that are important to treatment decision-making about assisted ventilation and ALS. By and large, the patients' own views on what constitutes relevant information and what are the possible influential factors remain to be determined.

Patients are at a disadvantage in relation to health care professionals when it comes to decision-making. Illness, fear, uncertainty, the complexity of the medical jargon, and so on, all contribute to increased vulnerability of the patient and have been shown to affect patients' ability to understand information and make informed decisions. Discussions about end of life issues are particularly difficult for clinicians to initiate (Weissman, 2004; Von Gunten, et al., 2000). Patients, their families, and clinicians frequently collude to avoid mentioning death or dying even when the prognosis is poor and the patient's suffering is severe. Professionals may not want to be perceived as giving up, thereby eliminating the patient's hope (Quill, T. 2000) and the uncertainty of prognosis and life expectancy may also contribute to communication barriers. In addition, the clinician's role is at times fragmented and he/she may have very little knowledge of the patient's context and values making it even more difficult to provide relevant information for decision-making.

What information is provided and understood by patients and families is dependent, amongst other things, on the decision-making model used by health care professionals. To improve the care of individuals with ALS, some physicians have advocated the adoption of a palliative care approach (Mitsumoto, et al., 2005; Borasio, et al., 2001; Borasio, 2001). The palliative care philosophy adopts a more holistic approach to patient care and integrates patients' hope, values, goals, social context, as well as their medical condition in treatment decisions. It is imperative that key elements impacting on decision-making about assisted ventilation for individuals with ALS be explored and that patients' own views of their situation be examined.

This empirical study attempts to achieve the goal of gaining a better understanding of the perception, understanding, thinking process, decisions, and experience of treatment options for respiratory failure of individuals with ALS. It is hoped that having a better understanding of their experiences will allow recommendation of a medical decision-making model that facilitates the dialogue between health care providers, patients, and their family to ultimately improve the care as well as the decision-making process in the context of assisted ventilation while increasing the potential for more informed and autonomous decisions.

1.6 Outline of chapters

This thesis will be accomplished in several steps. Chapter II will consist of a literature review. Relevant literature on autonomy, informed consent, and medical decision-making models will be reviewed, as well as literature specific to ALS and decision-making about assisted ventilation. Chapter III will present the research project. The research design, sample, procedure, and analysis will be described. Chapter IV will present the results of the research study. The emergent themes from the interviews will be described in detail with the inclusion of sample quotations from the interviews to justify the analysis. In chapter V, I will discuss some of the emergent themes in more detail. I select those themes most relevant to the purpose for which the study was undertaken. Hence, the emergent themes that are discussed relate to the conception of autonomy to advocate for a pluralistic conception. This conception of autonomy better accounts for the many factors identified by the participants in relation to decision-making and assisted ventilation. The impact of the findings on a decision-making model will also be discussed to illustrate how a shared decision-making model is superior to other models found in the literature to promote autonomy in the context of ALS and assisted ventilation. Chapter VI will suggest recommendations for possible improvements in clinical practice and suggestions for future research.

Chapter II

Literature review

The importance of examining the decision-making process of individuals with ALS about assisted ventilation was introduced in chapter I. In this chapter, I will review the literature to explore the meaning of autonomy and autonomous decisions. The tenets of informed consent and some of the theoretical medical decision-making models will be reviewed. Whether patients can express preferences about the interventions they receive, choose to be involved or not in decision-making, and make autonomous decisions is influenced, amongst other things, by whether they are provided with appropriate and sufficient information during disclosure. Consequently, I will also review the ALS literature to explore the existing information pertinent to decision-making and assisted ventilation.

2.1 Notion of autonomy

Prior to developing the ALS practice parameters (Miller, et al., 1999), the AAN agreed on four guiding principles to improve the care and quality of life of people with ALS. The first principle addresses autonomy:

> High priority should be placed on patient self-determination or autonomy as an underlying assumption in the therapeutic relationship. Delivery of both information and care must take into consideration the cultural and psychological context of the patient and the family (p. 1311).

However, nowhere in the practice guidelines is the meaning of "autonomy" discussed or defined.

The term "autonomy" is used in every day discourse with different meanings. The terms "liberty", "self-rule", "sovereignty", "freedom of the will", "dignity", "integrity", "individuality", and "independence" can all be used to suggest autonomy. The specific meaning intended by the use of "autonomy" can vary depending on the perspective adopted. Liberal conceptions of autonomy tend towards an atomistic view of the individual while pluralistic conceptions accept the influence of a person's context, emotions, relationships, and society in defining who we are (Friedman, 1997; Sherwin, 1998; Donchin, 2001). A pluralistic conception may be more appropriate when discussing decision-making in ALS since the person with ALS is typically dependent on others for care. Furthermore, the certainty of imminent death resulting from the absence of a cure may trigger emotional responses that impact decision-making. Since the literature review on ALS and autonomy does not provide any empirical evidence to suggest the superiority of a pluralistic conception over a liberal conception of autonomy when dealing with ALS both conceptions will be reviewed.

2.11 Liberal conceptions of autonomy

A review of the literature on autonomy illustrates a wide disparity in the definitions of autonomy and what accounts for autonomous actions. Liberal theories have a commitment to the self that is free from contamination from other selves and undue social influences (Moody, 1989). From a liberal perspective, respect for autonomy means "respect and non-interference with others' goals, projects, and actions, chosen on their own conception of what is right" (Cummings Gauthier, 1993, p.24). Kant's conception of autonomy emphasizes the rational nature of the self, views the self as freely choosing one's end in life without regard to one's concrete circumstances, assuming that all rational beings will arrive at the same "answer" when faced with a moral question (Kant, 1988). His conception of autonomy does not center on pragmatic determinations of what accounts for autonomous actions but it serves as a philosophical foundation and, as such, has influenced modern definitions of autonomy

Also influencing modern definitions of autonomy is John Stuart Mill's notion of "liberty", which is the ability of the self to frame a life plan in accord with one's character, to do as one likes without interference from others so long as one does not harm them (Mill, 2003). Contrary to Kant's conception of autonomy, Mill's notion of liberty assumes the presence of alternatives. Consequently, in a specific situation, different individuals can use their rational faculties to choose among various options available the one that best fulfills their own well-being.

Drawing on both Kant and Mill, Faden, et al., (1986) introduced a pragmatic conception of autonomy that is suited to promoting autonomy during the informed consent process within a health care context. However, their definition centers on actions instead of the individual's characteristics. They argue that consent and refusal are actions and, consequently, a practical conception of autonomy grounding informed consent must focus on actions.

These authors advocate for a scaled autonomy where a threshold of "substantial" autonomy is necessary for an action to be considered autonomous. They identify three criteria to determine the substantiality of an action: intention, understanding, and voluntariness. On their view, an action can only be considered autonomous if it is intentional. Intentional actions are actions "willed in accordance with a plan, whether the act is wanted or not" (Faden, et al., 1986, p.243). Intentionality is an all or none phenomenon; acts are either intentional or they are not. However, understanding and voluntariness can be determined in degrees. An act can range from fully understood to fully ignorant in which case it would be considered non-autonomous. Similarly, the degree of voluntariness can range from the act being completely non-controlled to completely controlled eliminating the possibility of the act being considered an expression of autonomy. All three conditions must be fulfilled for autonomous actions.

Faden, et al. (1986) acknowledge that even though specific situations such as suffering from a fatal illness like ALS may reduce one's freedom, it cannot be assumed that the person is necessarily coerced by the situation. They argue that non-controlling influences are acceptable and do not necessarily interfere with an individual's abilities to exercise autonomy. So decisions made in such a context may remain voluntary and should be acknowledged as such. A pragmatic implication of the grading of voluntariness is the acceptance of some influence from the physician and others on the individual's decision so long as that influence does not control the outcome of the patient's decision. Nevertheless, even if more applicable to the medical context, this conception of autonomy is criticized by proponents of pluralistic conceptions (Sherwin, 1998; Friedman, 1997; Donchin, 1995). These authors argue that this conception of autonomy remains too focused on rational processes, mainly ignores that autonomy evolves over time, and is influenced by emotions, personal and social relations.

Gerald Dworkin (1988) has developed a different liberal theory of autonomy where autonomy is linked to the **capacity** of the individual to reflect upon his/her motivational structures and to affect change in that structure. Dworkin's conception of autonomy acknowledges that persons have the capacity to make their own decisions and others should not interfere with this decision-making capacity. In addition, this conception of autonomy implies that persons can make rational decisions that are compatible with whom they are. Accordingly, patients can best judge what decisions match their specific life plan and what is in their best interest. Discussion to explore the individual's preferences, values, and beliefs should therefore be encouraged to determine what is the patient's best interest instead of adopting an approach that assumes that physicians know best.

One objection to Dworkin's individualistic conception of autonomy is its tendency to promote non-involvement from professionals who may want to avoid being accused of interference with patients' right to self-determination. Non-interference may actually prevent communication between patient and physician. The emphasis on self-sufficiency and independence of the individual may also create situations where patients are presented with treatment options without consideration for possible impacts of those options on others. For instance, as Anne Donchin (2001) argues, in the medical context it is essential to recognize that the patient's decision often impacts the care that the family must provide, which may have disruptive effects on the projects and life plans of caregivers. Therefore, a different definition of autonomy may be needed.

Even though liberal, individualistic conceptions of autonomy are typically referred to for grounding of the doctrine of informed consent, some scholars such as Donchin (1995) believe that a pluralistic model of autonomy is better suited for today's health care context. She argues that the traditional liberal conceptions of autonomy are better suited to the marketplace, impersonal relations whereas the nature of our public health care system is more suited to a model of relational interdependence. In fact, today's aging population, the high incidence of chronic diseases that create dependence on family and community resources, the multiculturalism of our populations, and the public nature of our health care system are all factors that contribute to the necessity of adopting a pluralistic view of autonomy (Donchin, 1995).

When discussing decision-making in ALS, a pluralistic conception of autonomy that acknowledges people's context, interactions, and influences on decision-making may better promote autonomy. This may contribute to the high priority placed on patient autonomy in the AAN guidelines. However, the theoretical literature to date does not provide empirical evidence to suggest the superiority of a pluralistic conception of autonomy in the context of decision-making and ALS.

2.12 Pluralistic conceptions of autonomy

A pluralistic conception of autonomy acknowledges different perspectives, including feminist perspectives of autonomy. As opposed to liberal conceptions of autonomy, most feminist theories endorse including context as a central element in moral reasoning and the emphasis on strictly individualistic, rational processes in decision-making is absent (Friedman, 1997; Donchin, 1995). Such a view more clearly acknowledges that one's wants, values, and desires affect autonomy and, as such, autonomy evolves over time as one acts consistently and frequently according to the same values.

As opposed to the liberal models of autonomy that emphasize mainly the rationality of decisions, from a feminist perspective, emotions, such as love and affection, do not altogether eclipse the self; instead, they enrich the conception of the authentic self (Meyers, 1987). Decisions can be based on emotions and still qualify as autonomous, as long as they are reflected upon and remain in accordance with one's deep values, wants and desires.

Feminists acknowledge that persons do not function in a vacuum. Relationships with others help to shape, influence, and develop individuals. In their conception of autonomy, some authors (Friedman, 2003; Friedman, 1989; Meyers, 1987) emphasize the influences of personal interactions, while others (Sherwin, 1998) place as much emphasis on relations with societal agencies. Importantly, feminist authors recognize that human relations are unequal; hence, their particular conception of autonomy not only aims at defining autonomy but also at ending traditional patterns of oppression (Donchin, 1995; Sherwin, 1992; Friedman, 1989; Meyers, 1987).

From a feminist perspective, autonomy involves an individual dimension and social components. Autonomy is socially rooted because no individual can be completely separated from social influences. Friedman (1997) integrates the notions of interdependence and solidarity in her account of autonomy by emphasizing that humans are social beings who are connected and dependent on one another for material and emotional support, for the development of our capacities, and for the sources of meaning in our lives. The social nature of human beings contributes to individuals building their skills, confidence, and very own identities. Pluralistic societies promote autonomy by providing a wide range of alternatives, values, and perspectives on which the person can self-reflect and exercise the range of abilities related to autonomy. Consequently, Friedman (1997) argues that relationships such as familial, social, or personal relationships like friendship are necessary for the realization of autonomy however she does not emphasize the influence of societal relationships.

In contrast, Susan Sherwin (1998) includes all influential human relations in her relational approach to autonomy. She accepts the influence of both personal relations and public relations. Personal relations may provoke influences from our role as mother, spouse, or friend whereas public relations influence autonomous decisions from our culture, our religious affiliation, our social class, and our education. From her feminist perspective, these influences cannot result solely from tradition; like emotional influences, they must have been reflected upon, integrated into the self, and correspond to deep, important aspects of how we define ourselves.

Accepting a variety of relational influences in a pluralistic conception of autonomy maximizes the potential to exercise autonomy during decision-making. In the case of ALS, a degenerative disease that results in severe functional dependence, it is usually necessary for family and society to provide long-term medical support. For example, the CLSC, a publicly-funded agency, may provide personal care services such as bathing. In addition, CLSC personnel and the patient's family may be involved in typically medical interventions such as suctioning of secretions. Consequently, disclosure for informed consent may require providing information on availability of community services in light of limited resources, or on the family's burden of care associated with a specific intervention. Further, in accord with a pluralistic conception of autonomy that acknowledges the potential impact of health care decisions on others who depend on the patient for care or financial security, discussions about treatment options may need to encompass the interventions' impacts on the children or the income of the family.

Accepting that one's personal relations impact autonomy not only affects disclosure, it also promotes involvement of significant others in the decision-making process. It promotes professionals' awareness of the possible influences others may have on the patient's decisions bearing in mind that these influences may not necessarily constitute coercion or duress if integrated in the person's conception of self. Consequently, a decision that may have been considered non-autonomous with a liberal conception may be deemed autonomous if examined with a pluralistic conception, increasing the potential for patients to exercise autonomy.

2.2 Meaning and elements of informed consent

Regardless of the philosophical conception of autonomy adopted, respect for autonomy is central to our Canadian culture as evidenced by Canadian law. For example, it is expressed through our Canadian Charter of Rights and Freedoms (Charter) S.7 where the right to liberty and security encompass autonomy (Downie, 2004). Furthermore, the right to security included in S.7 also refers to basic human dignity, the inviolability of the person, and the right to bodily integrity, that is, the right to decide what shall be done with one's body. Similarly, Article 10 of the Civil Code of Quebec also expresses the importance of the principle of autonomy, inviolability and integrity of persons:

> Every person is inviolable and is entitled to the integrity of his person.
>
> Except in cases provided by law, no one may interfere with his person without his free and informed consent.

Consequently, a medical intervention such as use of assisted ventilation demands *a priori* informed consent. Failure to do so may constitute battery and justifies the patient's right to refuse treatment even though this may result in the person's death. The principle of respect for autonomy is therefore central to the doctrine of informed consent and the patient's right to be involved in his/her own treatment decision-making.

While the legal doctrine of informed consent expresses the principle of respect for autonomy and specifies which criteria must be fulfilled for informed consent to be considered legally valid, as Benjamin Freedman (1975) argues, informed consent in medicine is more than a legal requirement; it is a substantial requirement of morality. From both legal and moral perspectives, it is justified in the patient's right to self-determination and inviolability, and the professionals' duty to not interfere and promote autonomy. In the medical context, informed consent introduces the dilemma of how much to tell the patient to allow a carefully assessed, informed decision as opposed to how much information to withhold to avoid alarming the patient. This quandary can be interpreted in terms of the sometimes competing principles of respect for autonomy and beneficence. While health care professionals often struggle with this quandary, in North America, promoting autonomy has taken precedence over beneficence in many aspects of medicine including during the informed consent process (Brody, 1986). With the advent of more standardized and international practice guidelines in medicine, this emphasis on autonomy has also become integrated into the European culture and has led to profound changes in clinical practice (Pasetti, 2000). Even the Japanese culture, which up to recently tended to adopt a paternalistic approach to decision-making, is struggling now to integrate the notion of informed consent and respect for autonomy into clinical care (Narita, et al., 2006).

In assessing whether a decision constitutes an informed consent one must determine that: 1) the decision is made by a competent individual, 2) that the decision is informed, and 3) that it is voluntary (Faden, et al., 1986). Recent research findings (Wolley-Levine, et al., 2006; Ringholz, et al., 2005) suggest that individuals with ALS have a higher prevalence of cognitive/behavioral changes than initially thought. Yet, at present, the impacts of theses changes on competency are unclear. Thus, for the most part, persons with ALS can understand the nature, purpose, cause, and consequences of their decisions and in the present discussion, competency of the person is assumed. The next step in appraising consent is then to assess whether the decision is truly informed.

2.21 Disclosure

> The "informed" aspect of an informed decision has two components: a) there has been accurate and adequate disclosure of information and b) the individual understands the information that has been disclosed (Downie, 2004, p.73).

Of these two, disclosure is the element of informed consent most widely discussed in the literature. According to the law, seeking informed consent requires the professionals to share relevant information with the patient, but what is to count as "sufficient relevant information" is not always clear. The courts have used three standards to determine if disclosure is adequate: 1) the professional standard, 2) the objective or reasonable person standard, and 3) the subjective standard (Katz, 1984).

The "professional standard" of disclosure requires disclosure of information the medical community as a whole deems relevant to make a rational decision about an intervention. Assessment of a situation according to the professional standard applies to other fields of medicine where medical skills or knowledge is necessary to establish a consensus and determine the best medical approach. For example, a medical consensus based on specialized knowledge determines whether an oral or an intravenous agent is best to prevent recurrence of breast cancer in a specific case. In the context of disclosure of information, the consensus to determine what information is relevant to allow an informed decision can be determined without specialized medical knowledge. Thus, use of the professional standard to assess adequacy of disclosure does not ensure adequate disclosure. On the contrary, its use undermines autonomy by leaving the assessment of how much information is relevant to the medical professionals who, as a group, may not be committed to patients making autonomous decisions. Therefore, to promote autonomy during informed consent, adequacy of disclosure should not be evaluated according to the professional standard.

The "objective" or "reasonable person" standard of disclosure is objective in that it is not patient or professional specific. Rather, what information is required to make an informed decision in a particular situation is determined according to what information a typical, reasonable person would need as opposed to what is agreed upon by the medical community. Even though at first glance the reasonable person standard of disclosure is an improvement over the professional standard, it still undermines the right of an individual to decide for him/herself. The reasonable person standard assumes a universal response from all individuals confronted with a similar medical situation and disregards differences of opinions between two patients, a patient and a physician, or even between two physicians (Katz, 1984). Furthermore, this standard may tempt physicians to introduce their own subjectivity into the disclosure process by letting physicians believe that what information they, as "reasonable persons", would require is necessarily the information that any reasonable person would need.

The "subjective standard" of disclosure seeks to determine what information should be disclosed according to the specific individual's needs and circumstances. It requires professionals to clarify the specific patient's context, values, and preferences. Use of the subjective standard to determine adequacy of disclosure encourages physicians to attempt to determine what information this specific person requires to make an informed decision. Usually, this would involve discussion with the patient, exploration of the patient's values and beliefs, and confirmation that informational needs have been satisfied. Use of the subjective standard of disclosure is the best for promoting autonomy since it makes use of patients' specific contexts, circumstances, values, and beliefs, acknowledges that these factors influence informational needs, and corresponds with a pluralistic conception of autonomy. Yet, it has been criticized because it puts an unfair burden on physicians to guess the needs of a particular individual. Even so, it is the standard used in the Quebec legal system to determine adequacy of disclosure as opposed to most jurisdictions in North America that still use the reasonable person standard of disclosure (Downie, et al., 2002).

2.22 Understanding

The requirement for understanding of the information disclosed implies that a valid, informed consent requires more than adequate disclosure of information on the part of the physician. The patient must understand the nature of the disease and the suggested treatment, as well as the foreseeable consequences of performing or not the suggested intervention (Faden, et al., 1986). Understanding demands comprehension and appreciation of all the information of significance to the person's decision (called "material information" in law). What constitutes material information for one person will vary according to one's values, beliefs, needs, interests, etc. (Faden, et al., 1986). Even though the law provides some guidance with regard to disclosure of material information, it is less specific with regard to the requirement for understanding. For example:

> In Reibl v. Hughes, the Supreme Court of Canada hinted at understanding as a requirement. However, it did so in passing, in the context of language comprehension, and without any clear statement of the nature and scope of the understanding required (Downie, 2004, p.72).

Various means such as use of analogies, layman terminology, written material, and videos can be used to facilitate understanding of specialized medical information and its impact on a specific person's context. Patients' understanding can be verified by asking them to explain in their own words what they understood of the information disclosed and the impacts of the information in their specific context, and ultimately clarifying points that remain nebulous. Even though the law is vague with regard to the requirement of understanding, professionals have a duty to promote understanding of information as stipulated by various codes of conducts such as the Canadian Medical Association Code of Ethics (2004)

2.23 Voluntariness

As just discussed, for consent to be informed, the competent individual must agree or refuse treatment on the basis of an understanding of relevant information that was disclosed. In addition, consent must not be controlled by influences that would engineer the outcome and it must involve the giving of permission (Faden, et al., 1986). The presence of coercion, force, fraud, duress, deceit, constraint, etc. eliminates the possibility of a valid consent (Downie, et al., 2002). Pressure arising from the patient's interactions with others might influence decisions. Similarly, a person's context, situation, presence of disease may also influence consent. Nevertheless, the impact of such factors does not automatically render the consent involuntary. For example, the negative impact on the family of a long-standing chronic condition may influence a patient's decision to stop all medical interventions. Such influences do not necessarily make the decision involuntary as long as the decision is grounded in the person's longstanding value of caring and priorizing the family's welfare. Consequently, in the case of ALS and decision-making about assisted ventilation, relations, context, and impact of the disease on the person's life are amongst the many issues that should be examined during the consent process.

In light of the requirements for informed consent, it is evident that the nature of the physician-patient relationship and the approach to decision-making influence a person's ability to exercise autonomy and provide informed consent for an intervention. Thus, I will now review the various medical decision-making models to clarify whether one is more likely to maximize the patient's potential to exercise autonomy when making decisions about assisted ventilation.

2.3 Medical decision-making models

In today's medical context, with its tendency towards impersonal and highly technical care, relying solely on a process of informed consent does not ensure or promote autonomy. Merely fulfilling the elements of informed consent ensures compliance with the legal requirements in Quebec. It does not meet the moral requirements and ensure that the decision is in line with one's life plan and truly reflects the desires, values, and beliefs of the individual. In addition to the requirements of informed consent, several authors endorse models of informed decision-making that rely on ongoing communication between all the parties involved to promote informed decisions (Charles, et al., 1997; Pres. Com., 1988; Katz, 1984).

Research on ALS and use of a ventilator or BIPAP has shown that, amongst other things, physicians' own views and attitudes towards ventilation impact the decision whether to use assisted ventilation (Lechtzin, 2004; Moss, et al., 1993). In some cases, the availability of assisted ventilation for ALS is simply not discussed or invasive ventilation is initiated without consent from the patient even though there was ample time to discuss the issue (Hirano, et al., 2005; Moss, et al., 1993). Consequently, use of assisted ventilation may be based on factors unrelated to the patient's desires, beliefs, or values, precluding them from the opportunity to exercise autonomy and make informed choices. This is similar to research done on use of invasive ventilation and muscular dystrophy (Gibson, 2001). A review of the literature reveals that physician and patient involvement in medical decision-making vary depending on the decision-making model adopted. Thus, in the next section, I will discuss three prominent models discussed in the literature; the paternalistic model, the informed model, and the shared decision-making model.

2.31 Paternalistic model

The underlying assumption of the paternalistic model of decision-making is that the physician knows best what is in the patient's interest. In this model, the physician selects and provides information to encourage the patient to agree to the treatment proposed. Furthermore, patient involvement in decision-making is limited. The emphasis of this model is on the patient's "good" or "well being", not on choice or autonomy (Emanuel & Emanuel, 1992).

The paternalistic approach to medicine is threatened by the requirement of disclosure. The paternalistic view holds that sharing information with patients may make them more anxious, cause fears, and increase the likelihood of foolish and irrational decisions (Katz, 1984). Consequently, physicians may avoid giving information to patients, thus fulfilling the requirements of beneficence but simultaneously neglecting to respect patient autonomy. A paternalistic approach ignores the findings of a study performed by Silverstein, et al. (1991), which revealed that individuals with ALS want a better understanding of the disease, more information about breathing problems they experience, and use of ventilators. Seventy percent of the respondents in that study agreed with the statement: "I prefer to participate in decisions about my own medical care" while the remainder preferred to leave decisions about care and treatment to their physician.

The notion that doctors can determine what is in the best interest of their patients is challenged if physicians attempt to understand their patients' values, beliefs, and other personal factors. Even though a paternalistic model of decision-making may be appropriate under specific circumstances, such as in emergency medicine, routinely adopting such a view with ALS does not promote decisions that are autonomous.

Through a paternalistic model of decision-making, initiation or foregoing of assisted ventilation risks being based on the professional's subjective, and possibly erroneous, assessment of the patient's quality of life. To allow persons the opportunity to make decisions that are in accordance with who they are, patient preferences and preferred approach to decision-making should be explored. Following these discussions, it is possible that the patient actually prefers the physician to make treatment decisions and perceives this to be in his/her best interest. Nonetheless, these discussions provide the patient with the opportunity to decide and exercise autonomy. Thus, a decision-making model that promotes patient involvement and open communication between patient and physician may be more suitable to ALS and decision-making for assisted ventilation.

2.32 Informed model

At the other end of the spectrum of decision-making models is the informed model (Charles, et al, 1997) or the independent choice model (Quill & Brody, 1996) where professionals provide medical information to patients who have the sole responsibility for decision-making. This model assumes that autonomy is equivalent to control over decision-making and that information is empowering the patient by increasing their knowledge of the possible risks and benefits of various treatment options therefore promoting decisions that reflect the patient's preferences and the medical knowledge available (Charles, et al., 1997).

In this model, the role of the professional is restricted strictly to that of information provider. Professionals may avoid sharing some of their clinical experience to prevent their own values from affecting the patient's decision (Charles, 1997). Withholding this type of information may actually encourage poorly informed decisions. For example, the physician may hesitate to disclose to a patient that invasive ventilation requires so much care that, from his clinical experience, should the patient choose this intervention, the family will most likely be unable to provide the necessary care resulting in the patient living in an institutions. Withholding this important piece of information may create a poorly informed decision that ignores a patient's wish to remain at home.

With the informed model, professionals also withhold their opinion and recommendation and refrain from being involved in the decision-making *per se.* Thus, with the informed model, the patient may feel extremely vulnerable, abandoned, or poorly supported if left to make important treatment decisions alone (Gray, 1990) with no possibility of delegating decision-making onto the professionals. The model of shared medical decision-making where the patient and the professional interact and share their views at each stage of the decision-making process may be better suited to promote a pluralistic conception of autonomy in decision-making and assisted ventilation.

2.33 Shared model

Charles, et al. (1997) describe the shared decision-making model where patients have the opportunity to exercise autonomy and make informed treatment decisions. In this model, autonomy in the decision-making process is understood as an opportunity for self-growth. They identify three necessary components to shared decision-making. First, shared decision-making assumes at least two participants, usually a professional and a patient. In addition, they acknowledge the possible involvement of others, such as family members, in the decision-making process. This is essential in the case of a serious illness like ALS because of the potential impact of the illness and its treatment on the family as well as their involvement in supporting and caring for the patient throughout the illness.

Secondly, in the shared decision-making model, both parties must take steps to share information during the decision-making process. This model acknowledges that while medical professionals have more knowledge on the medical aspect of treatment, patients have a better understanding of how interventions specifically affect their context. Shared decision-making involves patient and physician participation at each stage of the decision-making process, that is, during information exchange, deliberation, and decision (Charles, et al., 1999).

The third requirement of the shared decision-making model pertains to the decision *per se*; it must be accepted by the parties involved. However, this does not mean that every one must be convinced that the decision is the best for the patient. Through mutual acceptance, all parties share the responsibility of the decision (Charles, et al., 1997).

The use of a shared decision-making model requires a commitment to substantial collaboration among all parties involved. This model acknowledges that people's emotions, beliefs, values, motives, context, etc. evolve over time, over the course of an illness, or a life span, which is consistent with the pluralistic conception of autonomy discussed in section 2.12. Its emphasis on ongoing communication can contribute to improved disclosure and understanding during the informed consent process. Since sharing of information entails sharing the clinical experiences as well as scientific evidence with the patient (Charles, et al., 1999), with the shared decision-making model, physicians are expected to share clinical experience, scientific evidence, and recommendation with patients making disclosure more complete. Ongoing communication also allows exploration of the factors that influence decisions potentially improving the voluntariness of decisions. In addition to the improvements the shared decision-making model brings to the process of informed consent, it is more conducive to psychological and emotional support making this model the most suitable to the context of decision-making for assisted ventilation and ALS, one of the most stressful milestones in the course of this disease.

2.4 Assisted ventilation in ALS

As discussed previously, patients require information that is material to the decision in order to make autonomous, informed choices. However, the question remains: What constitutes appropriate and significant information for individuals with ALS making decisions about assisted ventilation? Young et al., (1994) attempted to answer this question and found that the factors cited most frequently by individuals with ALS as influencing decision-making about assisted ventilation were: QOL, severity of disability, availability of ventilation by nasal mask, and admission to a long-term care facility. This study was carried out before BIPAP use became common practice for respiratory symptom management following the publication of the AAN Practice Parameter (Miller et al., 1999).

Many other studies contribute part of the answer to what constitutes pertinent information for decision-making and assisted ventilation in ALS. These studies have mainly explored the expected benefits and burdens of invasive and non-invasive ventilation and will be reviewed in the next two sections.

2.41 Invasive ventilation

Invasive ventilation refers to the continuous use of a ventilator via a tracheostomy typically resulting in the patient loosing the ability to eat, communicate verbally, and increasing the burden of care. The aim of initiating invasive ventilation in ALS is prolongation of life. This intervention does not change the course of the disease and, over time, patients nonetheless become progressively more debilitated. In addition, invasive ventilation carries increased risks of respiratory infections and other complications.

Some studies have illustrated that ALS patients can survive many years by using invasive ventilation (Hayashi & Oppenheimer, 2003; Kaub-Wittemer, & et al., 2003). In these cases, death usually results from cardiac complications or infection. Use of invasive ventilation can increase patient comfort by eliminating the sensation of shortness of breath, improving alertness through better ventilation, and increasing general physical endurance, which may contribute to QOL.

While LTMV entails the considerable benefits of prolonging and possibly maintaining QOL, it may pose significant burdens and important financial consequences for the family. Improvement of patient QOL is addressed in a study performed by Kaub-Wittemer, et al. (2003), which revealed that ALS patients ventilated via a tracheotomy showed a good overall QOL. However, almost one-third of their caregivers felt that their own QOL was lower than that of the fully ventilated patient they were caring for. This is similar to the results obtained by Gelinas, et al. (1998) and Moss, et al. (1996) who found that ALS caregivers have a lower QOL than the general population.

Another study of ALS patients on home ventilation in Illinois conducted by Moss, et al. (1993) quantified some of the burdens associated with ventilator use; "families provided 9 hours of care daily and nurses and aides, 15; there was a mean of 3.5 persons per patient involved in daily care" (p.439). The mean yearly cost of home ventilation was $153 252 and most of the expenses related to nursing care. These figures illustrated only the direct care costs. However, as suggested by Polkey, et al. (1999): "When assessing these costs, it is necessary to consider both financial costs such as missed employment opportunities as well as the enormous emotional burden inherent in the domiciliary ventilation of a severely disabled relative" (p.369).

Patients' preferences and actual treatment choices for use of invasive ventilation was investigated in a study by Albert, et al. (1999). Through the use of a questionnaire, they examined the impact of patient mental health, attachment to life, and severity of ALS on patient preferences and choices. They found that patients with shorter disease duration and strong attachment to life were more likely to favor use of invasive ventilation. Depression and hopelessness were unrelated to patient preferences. This group also points out that other factors, such as, physician recommendations, changes in medical care, patient's judgments about QOL, and insurance coverage may impact the decisions, but these factors were not investigated in their study.

A German study by Kaub-Wittemer, et al. (2003), indicated that only 33% of patients ventilated via a tracheostomy had been informed in advance about the impeding respiratory failure. Moreover, 81% of these individuals underwent tracheostomy without prior informed consent. These results are similar to those of Moss et al. (1996) who reported that most persons with ALS who underwent invasive ventilation in the United States were not well prepared to reach a decision. Thus, even though the literature on ALS and invasive ventilation provides some information that may be pertinent to decision-making, it would appear that patients still lack information to make informed decisions about LTMV. Nevertheless, these studies provide some insights into factors that may be pertinent for decisions about assisted ventilation and can be used to initiate sharing of information such as described in the shared decision-making model.

2.42 Non-invasive ventilation

Non-invasive ventilation via a mask and BIPAP machine can palliate symptoms of respiratory failure. Typically, the ALS patient starts using the BIPAP only at night to improve the quality of sleep and improve daytime endurance. As the disease progresses and the respiratory function declines, BIPAP use progresses to daytime as well. The person with ALS usually needs help to use the BIPAP (e.g., to maintain the machine, put the mask on and off, etc.) because of the physical limitations caused by the disease. Towards the end of the illness, if invasive ventilation is not initiated, the patient may need to use the BIPAP almost continuously to prevent or minimize symptoms of respiratory failure such as shortness of breath and decreased alertness. These impacts of BIPAP use on the patient are confirmed both in clinical practice and in the literature.

A study by Butz, et al. (2003) has shown that use of BIPAP at night significantly improves sleep quality, daytime sleepiness, and physical fatigue. In addition, research has shown that use of BIPAP can slow the decline in forced vital capacity and dramatically increase survival of individuals with ALS (Lo Coco, et al., 2006). It was also shown that respiratory muscle weakness has a greater impact on QOL of the patient than overall ALS severity (Mustfa, et al., 2006). Thus, early, aggressive use of BIPAP is advocated (Heiman-Patterson & Miller, 2006; Butz, et al., 2003; Bourque, et al., 2003).

The use of non-invasive ventilation and its impact on patients and families has been extensively studied. Some studies reported on its positive impact on patients' QOL (Mustfa, et al., 2006; Kaub-Wittemer, et al., 2003; Bourke, et al., 2002; Lyall, et al., 2001). These same studies highlight that the impact of BIPAP use on caregivers' QOL, burden, and stress is minimal when compared to that of invasive ventilation. The main negative impact of BIPAP use was found to be increased anxiety score in caregiver at the time of initiation of BIPAP, however this was not sustained over time (Mustfa, et al., 2006).

Gender of the patient was shown to significantly impact whether BIPAP and invasive ventilation are used (Lechtzin, et al., 2004; Kaub-Wittemer, et al., 2003). In those studies, male patients were significantly more likely to use assisted ventilation. Since tolerance to BIPAP is not influenced by gender (Gruis, et al. 2005), it has been suggested that a possible reason for this disparity is that women are generally more willing caregivers (Kaub-Wittemer, et al., 2003). This finding highlights the importance of a dedicated caregiver for persons to be able to use assisted ventilation.

Similar to invasive ventilation, aspects of cost seem to be related to BIPAP use. A study performed by Lechtzin, et al. (2004) at Johns Hopkins University identified that a higher income level was associated with BIPAP use. However, this may not be the case in Quebec since Medicare covers the actual cost of the equipment.

The information provided in the studies discussed in section 2.41 and 2.42 constitute some of the information that can be shared with the patient and family to promote informed consent. Sharing strictly this information is insufficient to promote decisions that are in accordance with a specific patient's values, beliefs, desires, and context. Other pertinent information that may not be found in the literature, such as availability of adapted equipment or home care services for ventilated patients in a specific region also need to be discussed. Only the use of a shared decision-making model requires that clinicians share the scientific evidence, as well as their personal clinical experience, opinion, and recommendation to better inform patients. By recognizing the pertinence of a wide range of information, the shared decision-making model maximizes the potential for patients to exercise autonomy as defined from a pluralistic perspective.

2.5 Summary of literature review

This chapter has highlighted the fact that even though the AAN has agreed to give high priority to the principle of patient autonomy and self-determination, and that they agree that patients and families need information in a timely fashion, they do not define the meaning of autonomy or the pertinent information needed to make autonomous and informed decisions. Liberal and pluralistic conceptions of autonomy were reviewed and it was suggested that, in the context of ALS and assisted ventilation, autonomy is best viewed in a pluralistic fashion encompassing the person's context, relations, values, emotions, and beliefs. Furthermore, the doctrine of informed consent was reviewed to illustrate that an informed decision requires more than mere disclosure of facts and is much more than just giving one's authorization. Various medical decision-making models were reviewed to illustrate that the process of decision-making goes beyond the required standards of informed consent. It was suggested that the shared decision-making model is better suited to the context of ALS and assisted ventilation. A review of the current scientific evidence available for this context was provided to illustrate what may constitute material information for informed decisions. Even though one other study (Young, et al. 1994) provides an initial examination of the factors pertinent to decisions about assisted ventilation from the patients' viewpoint, it was carried out in a different context and, thus, its findings may not be applicable to the patients followed at the MNH clinic. The next chapter will present the empirical research undertaken as part of this thesis to explore, from the point of view of the ALS patients followed at the MNH, the information that is required to make informed choices and actual aspects of the decision-making process about assisted ventilation.

Chapter III

Research design and analysis

3.1 Method and rationale

As discussed in chapter II, most research on ALS and assisted ventilation has focused on matters such as, aspects of care, costs, and patient and caregiver quality of life. Few studies have attempted to widen the general understanding of how individuals with ALS make decisions about treatment options for management of symptoms of respiratory failure including the individuals that opt against BIPAP and LTMV. Some studies (Kaub-Wittener, et al., 2003; Lyall, et al., 2001; Albert, et al., 1999; Moss, et al., 1996; Young, et al., 1994; Silverstein, et al., 1991) have used a combination of quantitative and qualitative approaches, developing surveys and closed-ended questionnaires to study ALS and assisted ventilation. However, these methods may make assumptions about the factors that are important to patients when they consider various interventions to relieve respiratory distress.

Quantitative research is not well suited to gain a detailed understanding about what people are actually thinking when making treatment decisions for end of life care (Morse & Field, 1995; Brock, 1994). Qualitative research, however, has the potential to reach into a research topic where other methods cannot reach (Pope & Mays, 1995). By asking open-ended questions, qualitative research uses the participants' own words to describe a specific experience. Because understanding the decision-making experience with respect to ALS and assisted ventilation is precisely what is sought, for this research a qualitative methodology is favored.

Within the scope of qualitative methodologies, a phenomenological approach was chosen as "it is considered to be a highly appropriate approach to researching human experience" (Wimpenny & Gass, 2000, p.1486). Phenomenology itself is the study of the essence of the "lived experience" of humans. It asks: What is it like to have a certain experience? It aims at describing, analyzing, and understanding an experience from the participants' perspective. Phenomenology is distinct in that it assumes that there is an essence to shared experience and that there exists basic elements in the phenomenon under study (Patton, 2002; Morse & Field, 1995).

Through phenomenology, the experience of individuals with ALS who consider various ways to manage respiratory symptoms was explored. How these people live through the process of deciding for an intervention, how they describe the respiratory symptoms, what they think of the available management strategies, who they discuss the topic with, are all issues that were studied to understand the basic elements involved in the individual's decision-making process. Individual, semi-structured, in depth interviews allowed exploration of the different values, beliefs, emotions, understanding, and consequences of the choices made by the individual pertaining to management of respiratory failure. This approach facilitated understanding of the elements that these persons have in common when exploring treatment options to manage respiratory symptoms while remaining attentive to the unique features of every individual case.

In using a phenomenological approach, it is an asset to have an understanding of the phenomenon that is being explored. As an occupational therapist, I have worked with the ALS population for over ten years. My knowledge and experience in dealing with individuals who have ALS was useful in formulating the research and developing interview questions and probes so that rich data could be gathered from the participants.

Individuals with ALS frequently experience difficulties talking because of the disease process. Oftentimes, their speech is dysarthric and difficult to comprehend. They frequently complement their use of speech to communicate by using various technological communication devices. Because of my work in the MNH ALS clinic, I was already familiar with their particular voices and communication devices. Furthermore, I could anticipate impediments to communication and compensate by providing the interview guide in advance, asking some yes/no questions to clarify answers, and repeating participants' answers for their validation to ensure my understanding of their speech. During the interviews, participants were asked to discuss sensitive topics, such as their fears, their experience of respiratory distress, and their eventual death. My previous involvement with them in the ALS clinic facilitated conducting sensitive interviews even when participants cried and became highly emotional.

3.2 Study sample and recruitment

A maximum variation sampling strategy, which aims at getting a range of subjects who live a variety of experiences, was used. Individuals were therefore approached for participation in the study regardless of age, gender, ALS type, speed of progression, or choice of strategy to manage respiratory symptoms. Language spoken was limited to English and French because the investigator has no knowledge of other languages and the use of an interpreter would have made it difficult to truly reflect each person's experience in his/her own words.

The study sample targeted individuals with whom discussion pertaining to treatment choice for respiratory failure was documented and whose forced vital capacity (FVC) was below 60%. This was to ensure that the potential participants had already considered the various strategies available to manage respiratory symptoms prior to the study and were able to reflect on the experience of breathing difficulties and considering interventions for symptom control.

Individuals with ALS followed at the MNH ALS clinic were recruited to participate in the research. This group was chosen for convenience and feasibility, as resources did not permit a multi-centred study. The respiratory therapist or the clinical nurse specialist of the treating team identified potential participants for the research. On a subsequent clinic visit, the clinic secretary gave these individuals an index card (Appendix A) describing the study and inviting their participation. If interested in learning more information about the study, they were asked to write down their name and telephone number on the card and to return it to the secretary. The principal investigator (PI) then contacted these persons by phone and described the research study and its purpose in detail, the advantages and disadvantages of participating, and the means used to ensure confidentiality. Potential participants were encouraged to ask questions and discuss the research study with their family and friends before agreeing to participate. In some cases, the consent form (Appendix B) was sent to the individual by e-mail prior to meeting the participants in person. If they agreed to participate, an appointment was set up.

On the day of the interview, the PI asked the individual with ALS if they preferred that their caregiver participate in the interview (often times to facilitate communication and expression of their thoughts). This is consistent with clinical practice where caregivers often ask questions for and assist the individual with ALS during interviews and interventions. The consent form was re-examined with the participant and their caregiver. The interview process, the need to record the interview, and the importance of participants' involvement later on for confirmation of the emergent themes were reviewed and all their questions were answered. Only one signature was requested on the consent form; because of physical disability in all but one case where the caregiver participated, the caregiver signed the consent form for the person with ALS.

Most participants received the interview guide (Appendix C) by e-mail prior to the interview to allow them to prepare in advance. Some persons who experienced communication difficulties because of the ALS drafted written preliminary answers to minimize fatigue during the interview. These preliminary answers were further explored during the interview.

3.3 Procedure

Semi-structured, in-depth interviews were carried out. The interviews lasted approximately one hour with a range of 45 minutes to two hours and were performed in the patient's home to minimize fatigue.

An interview guide with open-ended questions was used to gain a detailed understanding of the individual's experiences with treatment strategies for management of respiratory failure. The interview guide provided topics of relevance to the decision-making process about assisted ventilation that the interviewer was free to probe and clarify. Questions evolved over time as ongoing analysis of the material was performed and emergent themes were identified. This is consistent with the methodology that recognizes "discovery during inquiry" (Patton, 2002).

Demographic information was also gathered during the interview regarding age, marital status, community services received, date of diagnosis, date of approximate onset of respiratory symptoms, and type of ALS.

Fieldnotes were kept throughout the study to document participants' reactions and behavior. The fieldnotes also documented the PI's reactions, thoughts, feelings, and opinions throughout the study. The fieldnotes were used to promote reflexivity, sensitivity, and criticality of the interviewer to improve the quality of the research as it proceeded (Mays & Pope, 2006; Whittemore, et al., 2001).

3.4 Data analysis, credibility, and rigor

The interviews were tape recorded to allow precise transcription and use of the individual's own words to reflect the person's experience as directly as possible during data analysis. The PI listened to all the interviews prior to the transcription to familiarize herself with the material and the mode of communication used by the participant. The PI then transcribed verbatim the taped interviews within 2 weeks of the interview, listened again to each interview, and verified the transcription to ensure that it was precise and truly documented the participants' own words.

As recommended by Pope & Mays (1995), the stages of data collection and analysis were intertwined, going backwards and forwards between the interview transcripts and the emergent themes thereby understanding and analyzing the data throughout the period of data collection. This also allowed identification of areas that needed to be developed further in subsequent interviews. Each interview transcript was read and re-read to gain a general impression of the material. Each transcript was then examined for significant statements related to the decision-making process for management of respiratory failure. As data collection and analysis continued, categories of significant statements were grouped together and broad categories or related themes were identified for all interviews. This process is called "within case analysis" and "across case analysis" by Ayres, et al. (2003).

Validation of the emergent themes was sought from the participants to promote rigor of the analysis of the results (Barbour, 2001; Mays & Pope, 1995). Confirmation from the participants was originally planned once all interviews were completed. However, after four interviews were performed, the emergent themes and justifying quotes that emerged so far were forwarded to the participants for validation of the analysis to avoid misinterpretation of the phenomenon. This was done also to ensure participants' feedback in light of the rapid deterioration of their medical condition. This provided an opportunity for reflection, questioning, and new interpretations. The analysis continued throughout data collection and themes were progressively refined. Involvement of the participants for confirmation of the emergent themes and analysis was repeated after completion of all ten interviews to confirm the common elements that emerged from all the interviews. In cases where the person with ALS had passed away, confirmation of the emergent themes was sought from the caregiver who had participated in the interview.

Validity and rigor were addressed through frequent verifications of the PI's interpretation of the data through formal and informal discussions with researchers experienced in qualitative research and with my thesis supervisor. Once all ten interviews were completed, the emergent themes were also presented for feedback to colleagues on the MNH ALS team and other teams specialized in ALS who attended the Allied Health Forum of the 17th International ALS/MND Symposium where this research was presented. Credibility, defined as an accurate interpretation of the meaning of the data (Whittemore, et al., 2001), was promoted by my intensive involvement with the participants.

3.5 Ethical and moral implications

Prior to enrolment of participants, approval from the MNH Research Ethics Board (REB) was requested and granted in August 2005 (Appendix D). This process and all other requirements of the *Tri-Council Policy Statement: Ethical Conduct for Research Involving Humans* (TCPS) (2002) were followed.

To avoid coercion, the clinic secretary initially alerted potential participants about the study. Potential participants were also given ample time and opportunity to discuss with friends and family their possible involvement in the study prior to meeting with the PI about the study.

Free and informed consent was obtained from all participants prior to the interview. Participants were informed that they had the right to withdraw from the study at any time or to refuse to answer any of the questions without affecting the care to which they were entitled at the MNH.

There were no known risks to participating in this study. However, to reduce possible anguish in discussing this potentially sensitive issue, participants were only approached for participation after it was ascertained that they had already considered assisted ventilation in the clinical setting. Furthermore, to maximize the participants' comfort, some of the participants received the consent form and the interview questions prior to the interview to allow them ample time to consider participation, reflect, and prepare their answers. My years of experience in dealing with these people in the clinical setting and my training as an occupational therapist contributed to facilitate communication and the conduct of the interviews in a sensitive manner.

To protect confidentiality, a coding system was used to link a transcript to a specific individual. This coding document, the consent forms, and the interview tapes are kept in a locked filing cabinet at the MNH. Data from the interview transcripts are kept on a protected computer system at the MNH. All audiotapes and transcriptions will be kept for five years and then destroyed. The specific content of the interviews were not discussed with colleagues from the clinic. The person's care at the MNH ALS clinic was not affected by their participation or refusal to participate in the research. Use of citations, quotations, and case descriptions is necessary in qualitative research to support the emergent themes and ensure authenticity. However, this method makes it difficult to ensure anonymity with such a small number of individuals with ALS followed at the MNH. Participants were aware of this factor.

This chapter has specified the method, study sample, procedure, and analysis used in the research. The following chapter will present the results, including the emergent themes and selected quotations from participants' interviews to justify the findings.

Chapter IV

Results and Emergent Themes

In this chapter, I will describe the results of the study. In doing so, I report the profile of the study participants and the themes that emerged from the ten interviews. Each theme will be described in detail and quotations from some of the interviews will be used to substantiate the emergent themes. Deviant cases will also be discussed to illustrate individual differences where applicable.

4.1 Profile of the participants

A total of ten semi-structured interviews were carried out with nine individuals followed at the MNH ALS clinic. As noted in section 3.2, the study sample targeted individuals with whom discussion pertaining to treatment choice for respiratory failure was documented in the chart and whose FVC was below 60%. The sample included six men and three women. Table 1 illustrates the characteristics of the nine participants interviewed.

One woman was interviewed twice; once while she was using BIPAP intermittently and once again after she had initiated invasive ventilation through a tracheostomy. Six individuals were using BIPAP: some at night only, however one subject used the BIPAP all the time except for brief moments when eating. Two persons were using invasive ventilation through a tracheostomy, and two participants were not using any form of assisted ventilation at the time of the interview.

Persons with ALS were asked if they wished a family member to be present during the interview. In total, six family members who also act as main caregivers participated in the interview, sometimes clarifying the person with ALS' answers, sometimes giving their own interpretation of the experience. One family caregiver declined to stay for the interview and one was not present at home at the time of the interview. Seven of the individuals with ALS were still capable of communicating verbally. One person was using a communication device to supplement verbal communication. Four interviews were performed with the use of an assistive communication device. Only two individuals had no speech impairments at all.

Table 1: Characteristics of the participants

#	SEX	AGE	MARITAL STATUS	LANGUAGE	COMMU-NICATION	EDUCATION	ALS TYPE	MONTHS SINCE DX	MONTHS SINCE RESP. FAILURE	INTERVENTION USED
1	M	72	M	E	VERBAL/ DYNAWRITE	UNIVERSITY	SPINAL	18	6	BIPAP
2	F	43	M	F	DYNAWRITE/ E-MAIL	UNIVERSITY	BULBAR	30	11	BIPAP/ TRACH+VENTILATOR
3	F	45	D	F	VERBAL	UNKNOWN	SPINAL	65	30	BIPAP
4	M	47	M	F	VERBAL	UNIVERSITY	SPINAL	18	10	BIPAP
5	M	69	M	F	VERBAL	HIGH SCHOOL	SPINAL	66	52	BIPAP
6	M	54	M	E	VERBAL	UNIVERSITY	BULBAR	16	2	NONE
7	M	46	D	E	DYNAVOX/ SIGNAL	UNIVERSITY	SPINAL	60	>36	TRACH+ VENTILATOR
8	M	36	D	E	VERBAL	COLLEGE	BULBAR	19	2	NONE
9	F	66	W	F	VERBAL	COLLEGE	SPINAL	132	12	ON/OFF BIPAP

All the participants lived at home at the time of the interview and all but one main caregiver consisted of a family member. All the participants received home care services from their CLSC and four were also paying for extra private home care services. The

National Home Ventilation Program of the MCI was responsible for the services pertaining to assisted ventilation of the persons using assisted ventilation. One participant started using assisted ventilation prior to the establishment of the MCI home ventilation program and was initially followed by a private company specialized in the rental of respiratory equipment. Table 2 illustrates the participants' caregiver and home care service profile.

Table2: Home care service profile of the participants

PARTICIPANT	MAIN CARE-GIVER	CLSC SERVICES *	PRIVATE SERVICES	MCI INVOLVED **
1	SPOUSE	YES	YES	YES
2	SPOUSE/ CHILDREN	YES/ 39 HRS PER WEEK	NO	YES
3	SISTER	YES/30 HRS PER WEEK	NO	YES
4	SPOUSE	YES/14 HRS PER WEEK	NO	YES
5	SPOUSE	YES/19 HRS PER WEEK	YES	YES
6	SPOUSE	YES/14HRS PER WEEK	YES	NO
7	PRIVATE/ SPOUSE	YES/15 HRS PER WEEK	YES	YES
8	COMMUNITY SERVICES	YES/28HRS PER WEEK	NO	NO
9	DAUGHTER	YES/25 HRS PER WEEK	NO	YES

*CLSC: Centre local de services communautaires

**MCI: Montreal Chest Institute

4.2 Emergent themes

Six main themes pertinent to decision-making about assisted ventilation from the patient's perspective emerged from the interviews. These are: meaning of the intervention; the importance of context, values, and fears in decision-making; the need for information for decision-making; and adaptation/acceptance of the intervention. Feedback on analysis and emergent themes was received by five of the nine participants, all of whom confirmed the emergent themes.

4.21 Meaning of the intervention

Participants expressed a clear distinction between what the BIPAP and invasive ventilation represented to them. The BIPAP was considered as a means to relieve symptoms of respiratory failure, whereas all the participants who discussed use of invasive ventilation clearly expressed that it represented a choice between life and death.

a. Meaning of the BIPAP

Eight individuals interviewed discussed what the BIPAP represented to them. The one person who did not discuss the meaning of the BIPAP had been using invasive ventilation for over three years and, since communication was extremely tedious for this person, the interview focused mainly on his decision-making pertaining to the use of the ventilator.

Statements from all participants reflected on the fact that ALS involves continuous adaptation to a changing condition and that assisted ventilation is perceived as one more adaptation.

> *...c'est un appareil de plus qui débarque, et à la fois pour faciliter mais à la fois pour concrétiser que ça devient de plus en plus lourd... C'est une machine de plus, encore des tubes, c'est un appareil de plus, là, O.K. ?* (spouse participant # 2)

Also, the participants expressed that the BIPAP represented a means to palliate symptoms of respiratory failure.

> *...c'est comme une suce pour moi! Bien, c'est parce que c'est inconfortable d'avoir de la difficulté à respirer... Puis quand j'ai ça, bien, c'est le confort total... Ça fait que c'est pour ça que j'appelle ça ma suce... moi je pense que j'appelle ça ma suce parce que justement, ça enlève cet inconfort là, puis comme on s'habitue à porter un équipement, bien je le sens même plus sur moi puis je respire bien, je force pas Ça fait que, tout est là, en tout cas pour moi c'est du confort de porter ça.* (participant # 4)
>
> *...I see it [BIPAP] as a way of helping me, like my chair helped me to move around, this will help me to... [breathe better?] Yes.* (participant # 8)

This was true even for the one individual who used BIPAP 24 hours a day except for brief moments to eat. He clearly expressed that the BIPAP was the machine that allowed him to go on living:

> *...Maintenant, si je ne le portais pas le BIPAP, je crèverais...J'étoufferais... Cet appareil là, [suction machine] puis le BIPAP, c'est pour rallonger la vie seulement...* (participant # 5)

b. Meaning of the ventilator

Invasive ventilation was discussed with seven of the nine individuals interviewed. Two participants, including the person who used BIPAP nearly continuously denied knowing anything about the use of tracheostomies and ventilators. All the individuals who discussed the use of invasive ventilation clearly expressed that it represented a choice between life and death.

> *...[using a ventilator] That's a big decision... If no tracheostomy, might die sooner.* (participant # 1)

> *... Pour l'instant j'en vois pas beaucoup d'avantages à part que de respirer! Bien à part que de prolonger ta vie, j'imagine là! En quelque part le but de ça [ventilateur] c'est ça hein!* (participant # 4)

Four participants expressed misconceptions surrounding the use of tracheostomies and ventilators. For example:

> *...si là, je suis plus capable d'avaler parce que là ça passe dans le conduit des voies respiratoires, bien là, il y a un problème, là! Majeur! Là ça veut dire, il faut bloquer l'accès aux voies respiratoires pour éviter que j'avale, soit des sécrétions ou de la nourriture. Donc, c'est une trachéo qu'il faut pour pallier à ça.* (participant # 4)

In addition, some of the participants expressed that using a ventilator brought up visions of people in a neuro-vegetative state, on the verge of dying, restricted to a bed, unable to move around and be involved with others.

> *... Moi je m'attendais à renconter quelqu'un dans un lit, blanc comme un drap avec aucune expression dans la face! Tu sais vraiment quelqu'un sur le bord de la mort!* (spouse participant # 7)

In half of the interviews, the importance of being in control of one's condition was emphasized. The fear of developing a dependency towards the machine and the fear of triggering a more rapid deterioration in one's condition was discussed in relation to assisted ventilation.

> *...le ventilateur, dont [elle] parlait, c'est un appareil, un respirateur artificiel. Ça, ça respire pour elle, ça l'oblige de continuer à respirer, à moins d'un arrêt cardiaque là, si elle arrive en arrêt respiratoire, la machine elle pompe pour elle...* (spouse participant # 2)

> *...Parce que, pour moi si je commence ça, je ne peux pas respirer sans machine après. Mais pourtant c'était pas ça parce que je pensais que si je suis habituée, que je ne peux pas respirer sans le BIPAP...* (participant # 3)

The invasiveness of the intervention was another component of meaning of assisted ventilation that was discussed by the participants. In contrast, for many people interviewed, the BIPAP offered the advantage of being non-invasive, having no risks, and being easy to introduce or discontinue.

> *...BIPAP machine is not invasive. It's easy to put on... it's the best you can have for breathing.* (participant # 1)

4.22 Context affects decision-making

In some cases, the meaning the individuals attached to the intervention affected decision-making about assisted ventilation. The context of the individual also impacted decisions. Three aspects of the individual's context were repeatedly reported by participants. These are the functional status and need for purpose of the individual, the need for support, and the financial implications associated with assisted ventilation.

a. Functional status and need for purpose

All the participants reflected on the importance of their functional status and the need to feel that they have a purpose in life. For example, when questioned about his motivation to go on living, one individual who has been using invasive ventilation for three years wrote:

> *...The world still needs me.* (participant # 7)

The participants emphasized that assisted ventilation had functional impacts on their every day life. How assisted ventilation limited the person's functional status in relation to, for example, being able to move around, be with people, live at home, talk, or eat, were discussed by everyone interviewed.

> *...[With BIPAP] you're restricted...* (participant # 1)
>
> *... Quand téléphone, il faut que j'enlève pour répondre, je ne peux pas manger. A part ça, c'est correct. Je ne peux pas répondre au téléphone parce qu'il n'y a personne qui comprend.* (participant # 3)

Four participants stressed that trying to anticipate how disabled they would be at the time of the intervention was very problematic and impacted their choices of interventions.

> *...Là je ne suis pas encore totalement paralysé ça fait que, j'ai encore une qualité de vie mais c'est sûr qu 'à un moment donné, j'imagine alité tout le temps puis regarder la télévision, il me semble que c'est pas une vie ça!* (participant # 4)
>
> *...'let's leave the trach as a possibility'. We're not saying 'no' to it but I have the feeling that I don't care for that situation I would probably be paralysed, and unable to talk, and unable to eat. I would only be able to see and think. I would not wish this life.* (participant # 6)

b. Need for support

All the participants identified that the need for support in a wide variety of domains impacted decision-making about whether to use assisted ventilation. The availability of adaptive equipment, accessible housing, family and community services to facilitate care of the person with ALS were aspects of support that subjects identified as pertinent to using assisted ventilation. For example, one lady used the BIPAP until her husband passed away. After his death, she was unable to continue using the equipment because her context and the support available had changed. She had tried to hire home help but to no avail.

> *Bien, j'en cherche [de l'aide]. J'ai le livre ici qu'on m'a envoyé là. Il y a des noms dans ça, des madames qui veulent travailler. Quand j'ai appelé, il y a deux semaines, tout le monde était pris!* (participant # 9)

Even though the participants expressed that BIPAP use is relatively simple, it still entails care and support.

> *...Ça prend des soins, là, comme là j'en ai un dans le nez, ce qu'ils appellent un nasal, là... Un masque nasal, et puis là bien évidemment, euh... ça prend de l'humidité, puis il faut mettre un lubrifiant pour éviter que les narines s'assèchent, là... Ça fait que, ça prend des soins, régulièrement, pour éviter d'avoir des problèmes...* (participant # 4)

The family members of the two individuals using invasive ventilation also emphasized the enormous amount of time, energy, and skills required to manage the care of the person with ALS.

> *...Il est venu avec certaines infirmières, à trois c'était pas trop! Certaines quand elles ne viennent pas souvent, elles ne sont pas habituées. Parce que mine de rien c'est complexe! Beaucoup de petits ajustments! Juste pour qu'il soit capable de bien écrire comme ça, c'est beaucoup, beaucoup d'efforts!* (spouse participant # 7)

When the support needed for assisted ventilation affected the place of residence, this too became significant in the decision-making process.

> *...With a tracheostomy, I'm stranded to a nursing home.* (participant # 1)
>
> *... That [being at home] would influence the decision!* (participant # 6)

c. Financial implications

Six participants discussed the financial implications associated with using assisted ventilation.

> *...The cost of being at home means hiring 24 hour help, over $500/day...The other option is a nursing home. The other option costs $4000/month.* (participant # 1)

However, some participants stated that they did not consider the financial consequences of their choices when deciding whether to use assisted ventilation. The people with ALS expressed more concern about the financial implications of their choices then most family members did.

> *...We experience a lot of fear, a lot of shock but it has nothing to do with the financial aspect!* (spouse participant # 6)

4.23 Values affect decision-making

The third theme that emerged from the interviews was that participants' values affected their decisions about assisted ventilation. Communication, relationships, autonomy, life, quality of life, and spirituality were some of the pertinent values discussed by the participants in relation to assisted ventilation.

a. Communication

Four participants emphasized how important the ability to communicate was on their decision-making. Communication was identified as the most important factor especially with regards to deciding whether to use invasive ventilation.

> *...as long as I can properly communicate with my voice, my eyes or a machine or whatever, I want to have a respirator... But as soon as I can no longer communicate, that's it! I don't want anything else to be done.* (participant # 8)

The two participants who used a ventilator discussed the impact invasive ventilation has on communication. For them, the ability to communicate was identified as the decisive factor to request discontinuation of the ventilator.

> *...[La communication] C'est critique au point que demande-[lui] quel va être le déclencheur réel pour dire "on arrête tout", c'est quand elle ne sera plus capable de communiquer! C'est pas respiratoire là! Quand au niveau de ses habilités de communication, clignements des yeux, quand on ne sera plus capable d'entretenir une communication soutenue, réelle, c'est entendu avec elle qu'on.... Ça va être le déclencheur!* (spouse participant # 2)

Communication with others is so important that the spouse of one person using a ventilator discussed how if she could grant her spouse one wish, it would be for him to regain the ability to talk.

> *...si j'avais un miracle à faire pour [mon conjoint] ce serait de lui redonner la parole! Ce serait pas la ventilation!* (spouse participant # 7)

b. Relationships

Related to the ability to communicate and the social nature of human beings is the capacity to interact with each other. All the participants discussed the impact that the significant people in their life had on their decisions. These relationships impact decisions in three ways. First, participants frequently discussed the treatment options available to them with the significant people in their life. They asked for the opinion of others, such as family, friends, and health care professionals, to assist with treatment decision.

> *...il faut une personne qui nous aide à accepter le BIPAP... Oui, que quelqu'un... que t'ais un "bond", qui t'explique comme il faut aussi... Ça, ça aide beaucoup.*

> *Comme [l'ergothérapeute] du CLSC, elle m'a poussée, mais d'une bonne façon.* (participant # 3)

Secondly, many participants expressed that their decisions were influenced by a desire to protect their loved ones. In many cases, both the individual with ALS and the caregiver are concerned with each other's welfare. Participants expressed being concerned with not overworking their caregivers, not inflicting suffering on their loved ones, and allowing them to go on living a normal life.

> *...J'ai pensé surtout à mes enfants, les protéger.* (participant # 2)
>
> *...Bien moi ma crainte c'est sûr que d'avoir une trachéo., ça on parle, je vous dirais, de la connaissance que j'en ai présentement, ça me fait peur pour le maintien à domicile!...Bien, c'est parce qu'un moment donné, c'est bien beau d'en demander aux aidants naturels, comme mon épouse mais là, tu sais, il y a une limite là! Si moi ça me prend des soins continus de jour et de nuit, un moment donné, tu peux plus aider quelqu'un sans affecter la santé de la personne qui s'occupe de toi. Mais moi, c'est ça qui me fait peur!* (participant # 4)

Four family members also identified the importance of protecting the person with ALS from hurtful information, bad news, or just everyday worries. They express a desire that decisions represent what the individual truly wants and not what others expect.

> *...je veux m'assurer qu'elle ne porte pas le fardeau de dire "je ne veux pas imposer ça à mon conjoint, aux enfants"* (spouse participant # 2)
>
> *...I just want to be sure that everything is put on the table, that we have an idea of what, fully, of what we are going through, that [my husband] is going through, and what us, as his family, as his wife and children are going through! I want the children*

> *and me to be prepared and to support him whichever decision he would like. Either way! And I feel the need for that!* (spouse participant # 6)

Thirdly, the pleasure and sense of purpose that results from the relationships participants have affected decisions. Seven of the nine individuals with ALS expressed that relationships with others motivated their decisions about assisted ventilation, as evidenced by the individual using invasive ventilation who wrote:

> *...The world still needs me.* (participant # 7)

Whereas another person used the following terms to discuss the impact his relationships have on his treatment decisions and his desire to live.

> *...Oui pour moi il y a deux affaires dans la vie qui sont importantes... Quand t'es entouré d'amour là, ça c'est le premier facteur! Ça là c'est la première affaire qui fait que t'as toujours le désir de vivre.* (participant # 4)

Even although many participants discussed the importance of relationships, at times, the value of relationships conflicted with that of autonomy.

c. Autonomy

All the participants discussed the value of autonomy. People with ALS and family members alike emphasized the importance of autonomy and self-determination in decisions about assisted ventilation. Persons with ALS reported valuing their decisional autonomy, wanting to be involved and respected in the decision-making process, and wanting to have the final say in what interventions they will try.

> *...on a toujours le choix de se le faire faire ou pas.* (participant # 4)

Eight people interviewed clearly expressed that they felt the decision to use assisted ventilation was their own decision even though they had considered the opinion of others.

> *...[What factors would come into play for you to decide to use a respirator?] If my doctor or whoever tells me I need one, and most of all, if I feel I need one, so one on one, that's all.* (participant # 8)

However, one person who used the BIPAP nearly continuously discussed how he got the impression that the disease itself had taken away his ability to exercise autonomy and how he felt he had no control over his decision to use assisted ventilation.

> *...Je suis obligé. Si je l'ai pas [BIPAP], je suis pas capable. Non. Je sais pas combien de temps je 'toughe' mais c'est pas longtemps certain!* (participant # 5)

This individual also discussed how he perceived that he had no choice in using assisted ventilation after the physician suggested it.

> *...Tu sais, quand le [neurologue du MNH] dit quelque chose, par habitude, t'as pas le choix!* (participant # 5)

The values of autonomy, self-determination, and control are also reflected in the meaning attributed to the BIPAP and the ventilator: the former is perceived as an assist for breathing whereas the later is felt to take over the individual's breathing.

> *...quand la décision a été prise [to have invasive vent.], les médecins croyaient, nous avaient informés qu'elle serait capable d'être indépendante du respirateur, en partie. Donc, elle ne serait pas branchée en permanence. C'est sur ce principe là que [ma conjointe] avait pris sa décision.* (spouse participant # 2)

Family members consistently expressed that they endorse the value of autonomy and right to self-determination of the person with ALS.

> *...je lui ai dit que si on avait rien d'écrit, c'est certain qu'on prendrait quand même des décisions pareil, on resterait pas là à rien faire, puis on prendrait très probablement des décisions comme on pense qu'elle les aurait prises, là... Mais, surtout pas en situation*

> *de crise, dans le sens que, pausée, reposée, elle aurait voulu faire quoi ?* (spouse participant # 2)

Consequently, if family members have to act as surrogate decision-makers, they need some guidance to make decisions that are in accordance with the person's wishes.

> *...I have come to realize that this condition might kill me and my wife has also expressed the fear of having to make some difficult decisions. She'd prefer that I write them down so that there is no pressure on her to make my decisions for me.* (participant # 6)

However, in two cases, the conflict between the value of autonomy and the need of the family for some guidance from the person with ALS gave rise to tensions within the family.

> *...If it was just for me and him, then it's fine! But it's not that simple! That's why I want to make sure... And I know that the three of us have been pushing [him] a lot, and a lot, and a lot into approaching the living will, in approaching the feeding tube, with this and that, and with the trach...* (spouse participant # 6)
>
> *...il y a deux choses qui est source de... et de frustration, c'est pour moi d'accepter que [ma conjointe] fasse les choses à sa façon et c'est probablement différent de ce que moi j'aurais fait à sa place.* (spouse participant # 2)

Both individuals who used invasive ventilation discussed how this decision and the ability to determine the moment of one's death is an expression of autonomy.

> *...[deciding to stop to live, again, is going to be an expression of your control, if at one point it comes to that ?]* [1] *(signals yes with his eyes)*(participant # 7)

---

[1] Interviewer's question was a summing up for confirmation of what participant # 7 had communicated.

However, one person discussed how the emphasis on autonomy also brought extreme discomfort as she felt she was expected to decide when she wanted the ventilator turned off therefore deciding on the moment of her death.

> *...C'est pas l'étape d'après qui l'inquiète, c'est à quel déclencheur elle va dire "O.K, aujourd'hui je fais un appel téléphonique et on passe à d'autre chose, on enclanche la fin de la course". Ça c'est un sujet qui est pratiquement impossible à aborder.* (spouse participant # 2)

Even though she felt well informed prior to agreeing to use the ventilator, she had not anticipated this difficulty before making her decision.

> *...[Qu'est-ce qu'il aurait fallu que tu saches?] Que ça allait me faire du bien mais que la décision de mourir allait être difficile!* (participant # 2)

d. Life

Many participants brought up the importance of life and the will to live even in the face of severe physical disability. All the individuals with ALS discussed how they were not ready to die.

> *...J'aimerais ça continuer à vivre encore un petit peu, là. Il me semble que 66 ans, j'suis pas prête à partir tout de suite, même si je ne fais pas une aussi belle vie que j'ai déjà faite, ça fait rien, j'ai pas l'intention de partir, ça ne me tente pas! Faire le grand voyage, là...* (participant # 9)

Two individuals discussed how their hope for a cure triggered a strong will to go on living.

> *...Hoping that they'll come up with a cure. Maybe if you stick around. Because there's no known cause, there's no cure.* (spouse participant # 1)

For some of the participants, the desire to go on living affected their decisions.

> *...Any living person's will to survive is primordial and outweighs many other concerns or reasons.* (participant # 1)

Even the two individuals who were using invasive ventilation discussed the importance of life and expressed a desire to go on living in spite of their severe functional limitations.

> *... I love life.* (participant # 7)
>
> *... Je ne peux me résoudre à mourir. Je suis bien dans cette vie-ci.* (participant # 2)

Assisted suicide and euthanasia were discussed by two people to emphasize that they thought these were not acceptable options.

> *...Suicide, ce n'est pas une option.* (participant # 2)

However, three participants brought up discontinuation of treatment in the form of stopping assisted ventilation. This was acceptable to these people in cases where communication became impossible.

> *...If I'm on a respirator [and cannot communicate anymore], I think I would want to be unplugged... That's what I wanted and I wrote that all on my document.* (participant # 8)

e. Quality of life

The value of life was clearly an important factor affecting decision-making. In addition, all participants talked about aspects of QOL that affected decisions about assisted ventilation. Aspects of QOL that were discussed by participants included the ability to communicate, eat, move around, live at home, and be surrounded by loved ones.

> *...Oui pour moi il y a deux affaires dans la vie qui sont importantes... Quand t'es entouré d'amour là, ça c'est le premier facteur! Ça là c'est la première affaire qui fait*

> *que t'as toujours le désir de vivre. Puis aussi dans mon cas à moi, personnel là, comme on disait manger et être encore capable d'utiliser un ordinateur...* (participant # 4)

> *... C'est qu'ici on reste quand même dans un milieu familial. Je veux dire, il peut écouter la musique qu'il veut, au son qu'il veut, il peut écouter la télé, il peut faire ce qu'il veut, il a ses soins quand il le veut, il n'y a personne qui lui impose.* (spouse participant # 7)

These aspects of QOL were considered when people with ALS faced decisions about assisted ventilation.

> *...We are at the point where I think 'let's leave the trach as a possibility'. We're not saying no to it but I have the feeling that I don't care for that situation I would probably be paralysed, and unable to talk, and unable to eat. I would only be able to see and think. I would not wish this life. I would not be able to do anything else but why not see the children grow older.* (participant # 6)

Some people talked about the importance of where one lived and how this could be affected by using assisted ventilation. As a result, this was also taken into consideration when they made their decisions.

> *...With a tracheostomy, I'm stranded to a nursing home.* (participant # 1)

Three participants discussed the impact of assisted ventilation on mobility and took this aspect into account when considering assisted ventilation.

> *...[What changes within your quality of life if you have a Peg and a trach?] To be able to move around, mobility...* (participant # 1)

f. Spirituality

Two participants talked about how spirituality influenced their decisions. For one person who chose to use invasive ventilation, using technology to go on living was an expression of his spiritual values.

> *...[l'aspect que tu as la possibilité de contrôler ces choses là [la technologie] et qui tu l'exerces dans la limite de tes capacités, ça , ça va avec l'aspect de la spiritualité que tu vis?] (signals yes with his eyes)*[2] (participant # 7)

One person discussed being preoccupied by the moral dilemma involved if one chose to use invasive ventilation. For this person and his family, having the ability to control the circumstances of one's death was problematic.

> *...[How do you perceive the tracheostomy?] It is a moral question for me, because when I have the trach, I think that I would want to decide the time of my death...because I can live for a long time with it. Where I die, where I do it, is too easy unless I can choose my death...* (participant # 6)

4.24 Fears affect decision-making

Participants' fears affected decisions about assisted ventilation. The main fears that were discussed were the fear of being a burden, discomfort, the process of death, respiratory failure, and the fear of disease progression and uncertainty of outcome.

a. Fear of being a burden

Five individuals discussed their fear of being a burden for their loved ones and again wanting to protect them. As a result, they wanted to make decisions to relieve them of some of the responsibilities associated with being the main caregiver.

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[2] Interviewer's question was a summing up for confirmation of what participant # 7 had communicated.

> *...I told everyone I wanted to find the right place before I became too much of a problem.* (participant # 8)
>
> *...I'm afraid to be a burden to my family... I don't necessarily want them taking care of me when I am in that state, in that terminal phase. I figured I would be much more of a burden for them and I know now it is difficult for them because it's a lot of work to take care of me now.* (participant # 6)

Two people expressed worries that if their caregivers burned out they would be left in a vulnerable position.

> *...J'ai aussi peur que les personnes autour de moi s'épuisent.... Parce qu'ils ne pourront pas s'occuper de moi et je devrai mourir.* (participant # 2)

b. Fear of discomfort

Six participants discussed the fear of discomfort. Discomfort includes both physical and psychological discomfort that one may experience during the illness.

> *...Ça peut être au niveau financier, ça peut être physique, tu sais...ça peut, supposons qu'il y a une période ou c'est vraiment difficile à respirer ou des fois c'est d'autres fonctions du corps qui fonctionnent pas bien, il dit "regarde, là, j'suis tanné...j'suis tanné, je passe mon temps à souffrir, qu'est-ce que ça donne ?" Dans ces moments là, c'est plus difficile.* (spouse participant # 7)

The fear of discomfort was a preoccupation for both the people with ALS and their family.

> *...I'm afraid... The fear that I'm afraid the most is that... I'm afraid to see [my husband] trapped within his body without being able to do anything but witness with pain, witness life with pain...* (spouse participant # 6)

The amount of discomfort experienced influenced the decisions about assisted ventilation.

...Quand je disais que le fait de manquer d'air ça peut créer de l'anxiété puis de l'angoisse, puis d'avoir ça, [BIPAP] bien, c'est sécurisant, là... (participant # 4)

...My physical comfort because my mind is no problem but, psychologically I will support it but, physically, I don't like to have any pain! If pain comes, if pain comes with ALS, I don't want to be alive and suffer and I will let the natural course of my disease! (participant # 6)

c. Fear of the process of death

All the people with ALS discussed the imminence of death. They expressed fear related to not knowing when they will die and how death from ALS usually proceeds.

...le sujet délicat à aborder avec [ma conjointe], parce qu'elle a pas voulu, elle le sait, je suis persuadé qu'elle le sait qu'est-ce que ça implique, ces dernières journées là, ces dernières heures là, avec BIPAP ou non, comment ça se passe, puis comment c'est compliqué, puis comment c'est paniquant ou pas paniquant selon les cas... (spouse participant # 2)

The family members also expressed preoccupation with the eventual death of their loved one. They talked about their apprehension and their concern about their own ability to support the person with ALS through the process of dying.

...Si je vais être à la hauteur! Qu'est-ce que je fais? Oui parce que tu sais pas comment tu vas réagir... Ah, oui, oui! Oui. Parce que je ne sais pas si lui a de la misère à respirer, je peux avoir le sang froid et faire tout ce que j'ai à faire ou bien peut-être que je vais paniquer et je vais dire... je vais tourner en rond. (spouse participant # 5)

d. Fear of respiratory failure

The fear of respiratory distress, chocking, and running out of air was very significant and affected the decision-making of those interviewed.

> *...Tu respires dans ça [BIPAP], puis, j'ai peur de perdre mon souffle, j'ai peur d'étouffer.* (participant # 9)

In many cases, the fear of respiratory distress was more significant than the fear of death itself. One individual who was using a ventilator expressed that she lived with the constant fear of a mechanical breakdown, power failure, and so on.

> *...Mes peurs égals à j'ai peur que mon tube s'arrache et que personne ne le replace. J'ai peur de manquer d'électricité et de mourir de suffocation.* (participant # 2)

The fear of respiratory distress was shared both by those with ALS and the family members who themselves were afraid to witness such distress.

> *...Qu'il étouffe, qu'il manque d'air puis que je ne sois pas capable de lui installer correctement!...Moi j'avais peur aussi aux sécrétions, j'avais peur à étouffement, vomissements pris dans un masque.* (spouse participant # 4)

e. Fear of disease progression and uncertainty of outcome

Eight participants talked about the unpredictability of ALS and how this impacted their decisions.

> *...with ALS there's no ruler... To measure where you're at. So, sometimes, I'll feel pain or cramps or numbness on my ... and I'm wondering is it normal, is it temporary, so that's the main problem, but as far as therapy or whatever, if I present a problem, there's usually a way to find a solution. So that's the only problem. It's not being able to know what's normal or what's next.* (participant # 8)

The only participant who did not discuss this aspect in decision-making is the person who used the BIPAP nearly continuously.

Many participants talked about being uncertain of the benefits of the various interventions, questioning whether assisted ventilation really helped.

> *...« Bien oui, monsieur ça peut prolonger votre vie de six mois ». Mais la réalité des fois c'est Ha! Tu peux vivre deux ans, je ne sais pas! Tu sais les gens qui ont le cancer puis qu'ils disent : « Monsieur vous avez le cancer et vous en avez pour trois mois ». Ça c'est déjà vu qu'il y a du monde qui ont vécu. C'est sûr qu'il n'y a personne qui le sait. Ça avec c'est un coup de dés! Ils vont peut-être me dire : « Ça va être bon pour trois mois ». Puis la réalité va être différente. Ça fait que...* (participant # 4)
>
> *...On ne sait pas dans le cas à [ma conjointe], cinq, six mois, un an de plus combien ça vaut par rapport aux avantages qu'on en retire.* (spouse participant # 2)

They also discussed how they were unable to ascertain the benefits of an intervention before actually trying it.

> *...Au début, c'était difficile à s'habituer parce que là je mettais ça la nuit. La première nuit, je me souviens à l'hôpital, je l'ai porté quatre heures peut-être, de dix heures à aller jusqu'à deux heures le matin là j'ai dit « enlève-moi ça cette affaire là, ça a pas de bon sens » je ne dormais pas, ça m'empêchait de dormir, puis ça me soufflait dans la face, puis j'aimais pas ça le feeling de ça! Puis la deuxième nuit, je l'ai mis six heures. J'ai « toughé », j'ai dit « je vais le tougher ». La troisième nuit, ici après, « j'veux une bonne nuit, je veux dormir pas de masque ». Là je me suis rendu compte la différence de sommeil que j'avais! C'est là que j'ai réalisé : « non, il va falloir le porter! »* (participant # 4)

4.25 Need for information

The participants' need for information is another theme that emerged from the interviews. All the participants interviewed discussed their need for various types of information and emphasized the need for adequate timing of information. However, as opposed to the previous themes where people with ALS and the family members' opinions concorded, their informational needs differ. The importance of continuously providing information along the disease continuum was also discussed.

a. Types of information

The seven participants who used assisted ventilation discussed the importance of being provided with adequate technical information about the various interventions.

> *...Comment l'utiliser, quand est-ce qu'il faut l'utiliser, comment faire pour l'humidificateur. Comment il faut laver, comment il faut nettoyer, quand est-ce qu'il faut changer le filtre, tout ça.* (participant # 3)

Participants also expressed that they needed the information to be specifically related to respiratory management in ALS.

> *...the technical aspects and also at that point, we weren't too sure about ALS and choking and digesting and different aspects of just every day living... of eating and drinking, of the liquids going down the wrong way. Now he came and explained to us that people don't really choke.* (spouse participant # 1)
>
> *...Ils ne donnent pas juste l'aspect technique. Ils donnent aussi l'aspect de la maladie. Donc, ils donnent l'information aussi sur les problèmes pulmonaires qu'on peut vivre. Style, un moment donné si je m'étouffe, s'il y a de la salive ou de la nourriture tout simplement qui se loge au niveau des poumons, bien ils donnent l'information à savoir c'est quoi qui peut se passer, puis qu'est-ce qu'il y a à faire, puis il y a de l'information qu'ils nous donnent là-dessus!* (participant # 4)

Even if they felt well informed, some participants had a poor understanding of how respiratory symptoms and use of assisted ventilation could impact their daily life.

> *[Est-ce que, si je comprends bien, ta perception c'est que si la personne a une trachéostomie, puis un ventilateur ou un respirateur artificiel, elle est branchée et elle ne peut pas se déplacer ?][3] Oui.* (participant # 2)
>
> *Puis je me suis aperçue que j'étais mieux ventilée. Alors que je ne pouvais tenir ma tête avant la trachéotomie, après l'intervention je pouvais la tenir. Je n'avais pas réalisé à quel point ça allait m'aider. Ça été une décision prise sans en connaître toutes les conséquences. Je suis contente de l'avoir prise cette décision. Je remarque que je me sens beaucoup mieux.* (participant # 2)
>
> *[The impact it can have on the family, the impact it can have on the people who live with the person with ALS should be discussed more is what you're saying?][4] (signals yes with his eyes and then typing) Give the information on high probability to be alone.* (participant # 7)

In addition, evidence of some misconceptions about the technical aspects of assisted ventilation were revealed in three of the interviews.

> *...je lui ai parlé de l'idée de la trachéo sur laquelle on pourrait brancher le BIPAP. Le BIPAP se branche pas directement sur la trachéo. sauf que la trachéo elle a l'avantage que si tu respires uniquement par la trachéo directement, tu by-pass si tu veux certaines voies respiratoires mais le BIPAP ne peut pas se brancher là-dessus !* (spouse participant # 2)
>
> *...The BIPAP is a different model. There's different masks. There's also different types of air. There's normal air, or mixed with oxygen or sometimes only oxygen and*

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[3] Interviewer's question was a summing up for confirmation of what participant # 2 had communicated.

[4] Interviewer's question was a summing up for confirmation of what participant # 7 had communicated.

> *also the tracheostomy depending on the severity or the choice of the patient whether they want something on their face or in the throat, it's less in the way of the face.* (participant # 8)

The need to discuss how death from respiratory failure can proceed in individuals with ALS was talked about by three participants.

> *...Bien des cas de SLA, de ce que j'ai vu, de ce que j'ai lu, que un soir ils se couchent puis le lendemain matin il ne se sont pas réveillés tout simplement. Ça, c'est très délicat...c'est fréquent dans plusieurs cas, là. Ça, ça fait parti des choses qu'on veut valider, justement. Tu sais, rassurer ça, est-ce que forcément tout ce monde là rentre en détresse respiratoire tout le temps ? Si c'est le cas de tout le monde, c'est important de le savoir puis si c'est le cas d'une minorité, c'est important de le savoir aussi.* (spouse participant # 2)

> *[Il n'y a personne qui parle de ça, hein ? De comment ça va finir ou qu'est-ce qui va se passer. Il n'y a personne qui parle de ça.]*[5] *Non, non, non. [Devrions-nous ?] Je ne sais pas. Peut-être. Je ne sais pas. M'informer peut-être plus, mais... Pour elle, oui. Pour pas que tu paniques.* (participant # 5)

Both individuals who used invasive ventilation expressed the need to be informed about discontinuation of ventilation prior to actually initiating the intervention.

> *Le jour où [elle] a accepté d'aller sur un respirateur artificiel, elle s'est imposée la possibilité d'avoir à prendre une décision, q'elle décide d'arrêter ça d'elle même. Et ça, je pense qu'elle n'avait pas réalisé à ce moment là...* (spouse participant # 2)

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[5] Interviewer's question was a summing up for confirmation of what participant # 5 had communicated.

> *C'est une personne [infirmiere du MNH] à qui je n'aurais pas de crainte de communiquer avec. Je me sens super à l'aise avec elle. Je pense qu'à un moment donné, on voit qu'on se dirige vers ça [arreter la ventilation], je prendrai rendez-vous avec elle..* (spouse participant # 7)

The participants did not identify a specific method that they preferred to provide information however some participants wished that the information be provided verbally by individuals who knew them well and whom they perceived to be ALS specialists.

> *...you have to develop a personal relationship with the patient first before you can talk about it. I suggest that you ask the person one question at the beginning. Would you like me to tell you a few things about ALS, for example, if you want me to tell me about breathing, talking, as opposed to giving me a booklet of 45 pages of all kinds of information, to give a broad talk with a person and tell them personally, to help to think a little bit of things you can possibly expect.* (participant # 6)

b. Importance of timing

The importance of timing of information was highlighted in seven interviews. There was no consensus amongst participants regarding when was the best time to provide information. Nonetheless, these individuals agreed that timing affected how they processed information and, in turn, could impact the decisions they made.

> *C'est toujours bien d'avoir l'information, le plus d'information possible, le plus tôt possible... On s'entend là-dessus, sauf qu'il y a un moment donné ou des fois tu veux pas en entendre parler, tu es peut-être pas prêt à en parler tant qu'on pense. Moi, ce sujet-là [la mort] je l'ai abordé avec [ma conjointe] déjà, quelques fois, depuis deux ans et demi qu'on a le diagnostic mais c'était toujours, "on en reparlera" ou "éventuellement on a du temps en masse devant nous" alors, d'aller chercher*

l'information, d'être confronté, alors là, ça compte pour vrai, là, c'est vrai faut être attentif en classe parce que le test arrive, on étudie plus de la même façon puis l'information est plus compilée du tout de la même façon... (spouse participant # 2)

Il faut la [information] donner tranquillement, pas vite parce que le gars qui l'a reçoit.... Les personnes qui ont la maladie sont tellement bouleversées, ça prend un bout de temps...pour tout absorber ça. Après, quand ils sont calmés, là ils lui disent. Parce que plus la maladie avance, ils ont besoin de... Alors c'est là qu'ils préparent les gens. (participant # 5)

c. Differences in informational needs between persons with ALS and family members

All but one individual with ALS interviewed talked about not wanting to have information before it was needed to make decisions since the information tended to discourage them.

...It's a two-sided sword. Knowing too much can be devastating. It's like when you're pregnant, you want to know the sex of the baby and everything but you don't really want to know. (participant # 1)

...je veux pas penser maintenant... Parce que ça va être des choses pires que ça!... Oui, je veux prendre jour à jour. (participant # 3)

...So in that sense, when I need, when I'm ready. Meaning also, that when the condition of my illness arise, brings me to a certain point, then I'm ready! But if my body, my physical condition is not there yet, don't talk to me about it! (participant # 6)

The one deviant case involved a person who did not have a family caregiver. He discussed how he sought information about interventions before he experienced the symptoms that warranted the intervention.

> *...As soon as I see that whatever problem has a negative effect on me, physically or psychologically or emotionally whatever, then I know it's time to find proper help I need for a specific problem. Usually I know before anyone else notices so it's already in progress... when I don't know I want to know, I believe knowledge is the best thing in the world. I'm a sponge!* (participant # 8)

For their part, family members who were present during the interviews highlighted the need for information about ALS, symptoms, and management strategies at the onset of the disease. They felt that information was a means for them to feel empowered and in control as opposed to patients who were ambivalent about the effects information had on them.

> *...On veut savoir! Donnez-nous de l'information puis on est capable de comprendre l'impact. Tu sais c'est de l'inquiétude et c'est de l'insécurité. On ne sait pas où on va puis d'aller chercher un peu partout, regardes, c'est pas bon là! Tu sais n'importe qui dit n'importe quoi là! Ça fait que moi c'est ce que j'aurais aimé! Donnez-nous de l'information! Dites-nous c'est quoi la maladie! C'est quoi la maladie dans toutes les sphères là! Toutes, toutes! L'alimentation, la respiration allez-y! Quitte à ne pas comprendre tout maintenant puis à revenir vous poser des questions. Tout se dit, tout se dit! On peut pas comprendre, mais on veut savoir, tu sais, t'as toujours des questions puis t'as pas de réponses! Puis c'est qui qui les donne!... Moi, j'aurais aimé le savoir maintenant! Tu sais quand tu rentres dans un inconnu, de savoir où tu vas c'est tellement sécurisant là!* (spouse participant # 4)

> *...I think from my perspective and the family and the main caregiver, I felt that I wanted, I needed, I would emphasize on the word NEED, to know as much as I could, everything about the disease!* (spouse participant # 6)

d. Continuing need for contact and information

The need for information about assisted ventilation varied across the disease continuum. However, because ALS is a degenerative condition, all the participants who used assisted ventilation brought up the need for regular contact and sustained information. For individuals with ALS and their family, the services provided by the home care ventilation program were appreciated and provided a sense of security and empowerment.

> *... Ils viennent à chaque trois mois, ils vérifient la machine, ils vérifient l'oxygène, qui vient avec, compression trois mois. Comme je me sens plus en sécurité, que tout va bien.* (participant # 3)
>
> *...Dans le fond ils sont là aussi pour répondre à nos inquiétudes, à tout l'aspect... Parce que ça change. Là je suis dépendent d'un équipement, ça fait que ça change un peu le mode de vie là! Puis ils sont là pour répondre à toutes nos questions, des inquiétudes qu'on a par rapport à ça.* (participant # 4)

All but one participant felt they were well informed throughout the decision-making process. The deviant case was a person who was unable to consistently use the BIPAP. She emphasized how she felt she had been poorly informed and supported throughout the process.

> *...Ils viennent, ils te laissent la machine, ils te montrent comment ça fonctionne mais quand tu es toute seule. Elle ne s'est jamais vraiment bien, bien occupée de moi.* (participant # 9)

One person reflected on the improvement of the assisted ventilation services provided to her since the instigation of the home ventilation program of the MCI.

> *...Avant je pensais « Est-ce que c'est correct, est-ce que je fais bien? » Comme j'étais pas sûre. Maintenant avec l'hôpital je suis très à l'aise. C'est un bon service qu'ils ont commencé! Qu'avant il faut appeler la compagnie, puis la compagnie ne fait que travail, c'est pas comme l'hôpital.* (participant # 3)

This was confirmed by six of the seven participants using assisted ventilation who expressed a high appreciation of the services provided by the home care ventilation program.

> *...Eux s'occupent que les équipements soient adéquats à la maison, qu'on ait toute la formation et l'information nécessaire pour l'utilisation des équipements. Ils s'assurent que les équipements soient bien entretenus, donc il y a tout un côté euh... information et formation qui est donné aux aidants naturels... Ils sont efficaces, ils sont dynamiques, ils ont des connaissances, ils sont rassurants. Ils sont disponibles : tu les appelles, ils sont là. Ils veulent t'informer.* (spouse participant # 4)

4.26 Adaptation/acceptance of the intervention

As a result of repeated functional losses and acceptance of one's changing condition with ALS, ongoing adaptation to various interventions in order to palliate symptoms of the disease is necessary. All the participants interviewed who used assisted ventilation discussed the process of acceptance of assisted ventilation. The interviews highlighted that the acceptance process was intervention specific. Initially, subjects needed to objectify that they required ventilatory support. Once they had tried an intervention, they sought to confirm the benefits of the intervention. Acceptance of assisted ventilation was a lengthy process that involved gradual familiarization with the equipment. It followed a similar pattern as the one used to accept other assistive devices, and was influenced by past experiences.

a. Acceptance is intervention specific

Five of the seven participants who used assisted ventilation reflected on the fact that accepting to use assisted ventilation was a lengthy process. In all but one case, the decision to use the BIPAP followed some kind of a crisis situation.

> *... [before BIPAP] you cannot sleep, you think demons come out in the night, whatever unlikely or unthinkable occurs to you in the night when you can't sleep or even move your limbs make you feel you are in a grave or a casket. You are completely at the mercy of others, a virtual prisoner in your own body.* (participant # 1)
>
> *...[before BIPAP] elle étouffe, elle se cherche de l'air, on a beau succionner, ça succionne rien, puis là ça rentre dans une phase où là elle panique, elle s'énerve, elle a besoin de plus d'air, elle cherche son air, elle s'étouffe.* (spouse participant # 2)

At the time of the interview, some participants had not yet considered using a tracheostomy and a respirator, however those who had, preferred that invasive ventilation be planned ahead to avoid intubation in a crisis situation.

> *...Par ce que l'idée de la trachéo., on peut l'avoir en situation d'urgence, on souhaite pas ça. On pourrait, d'après ce qu'on sait... la scheduler d'avance, prévoir qu'il y aura une intervention chirurgicale et qu'il y aura une trachéo qui va être posée. Et ça, c'est l'option que ma conjointe a retenue.* (spouse participant # 2)
>
> *...I basically wrote a document for my final decisions and as long as I can properly communicate with my voice, my eyes or a machine or whatever, I want to have a respirator.* (participant # 8)

b. Objectify need for and benefit of assisted ventilation

All the participants discussed the deterioration in their FVC (which was measured regularly in the ALS clinic) to objectify and confirm the need for assisted ventilation.

> *...Quand on a vu que ça descendait, donc, de 130 à 73, là ils nous ont parlé "il va falloir penser à un BIPAP" fait qu'on a abouti avec un BIPAP ici.* (spouse participant # 2)

> *...Il m'a dit que j'avais perdu... quand j'étais debout c'était correct, mais quand j'étais couché, j'avais déjà perdu 15%....de forces... alors il m'a prescrit, le, comment ça s'appelle...Le BIPAP.* (participant # 5)

Participants interviewed also talked about their need to experience the benefits of assisted ventilation in order to decide to continue using it.

> *...Mais là ce bloc là de deux heures où elle l'a porté [BIPAP], on s'est entêté à ce qu'elle le garde. Ce qui fait qu'elle a commencé à acquérir certains bénéfices de ce deux heures là, pour s'apercevoir finalement que ça lui avait fait du bien ! Ça fait qu'elle l'a reporté vendredi toute la soirée et le samedi presque toute la journée!* (spouse participant # 2)

c. Normalization and familiarization with the intervention

All seven individuals who used assisted ventilation described that a process of normalization and familiarization with the equipment was necessary before it was used consistently. Acceptance took time and was facilitated if the person could gradually come to grips with the idea before being confronted by the equipment itself.

> *...les premiers six mois du BIPAP ici, on s'en est pas servi du tout, ou presque pas. Il est resté là, 'parké' dans une boîte, puis après ça mis sur une table de chevet puis... On*

> *s'en est servi réellement du BIPAP là, ça fait quelques semaines à peine...* (spouse participant # 2)

> *... J'ai pensé que j'avais besoin de l'habitude. J'ai acheté la machine juste pour une sécurité mais j'ai resté six mois sans lui toucher. Après six mois, j'ai accepté avant, après j'ai commencé tranquillement.* (participant # 3)

Even the use of invasive ventilation was normalized.

> *...Tu sais, il y a des gens qui se font transplanter un coeur parce qu'ils sont plus capables, parce que le coeur fonctionne plus, j'ai dit : "toi, il te fallait un petit trou au travers de la gorge pour respirer". Il dit que c'est ça que lui avait vu aussi. Pourquoi est-ce que moi je ne pourrais pas avoir de respirateur alors que certains se font transplanter des organes ? C'est juste... une intervention chirurgicale qui permet de continuer, qui permet de laisser une personne vivre.* (spouse participant # 7)

The need to gradually get used to the equipment and to become more confident with its use was discussed by those with ALS and their caregivers.

> *...Il n'y a plus de gêne à enlever le tube et insérer la canule. C'est plus le stress que c'était les premiers jours.* (spouse participant # 2)

d. Influence of past experiences

Four participants compared acceptance of assisted ventilation to acceptance of other assistive devices. The acceptance pattern was similar in that it often took time to get used to and accept a new piece of equipment.

> *...Il y a eu bien d'autres situations aussi là, tu sais, d'apprendre à... d'accepter d'avoir un fauteuil dans la maison, ça a pris une certaine étape, là... Le fauteuil, il a été parké dans la maison pendant plusieurs semaines avant qu'elle embarque dessus, là, tu sais. Le BIPAP ça a été pareil...* (spouse participant # 2)

Many participants with ALS and their caregivers reported that past experiences with illness and death influenced the decisions about assisted ventilation.

> *...I am reluctant to go into a hospital because I had a very bad experience with my Crohn's operation and I almost died.* (participant # 6)
>
> *...Je donne souvent l'analogie avec [ma conjointe]...elle est allée accoucher le pied sur le "break"! Dans le sens qu'elle n'a pas été deux semaines en retard pour rien! Approche évitement! ...dans le cas à [ma conjointe maintenant], on fait la même analogie! Comme on ne sait pas ce qui s'en vient, on ne sait pas comment douloureux que c'est ou non* (spouse participant # 2)

4.3 Summary

This chapter provided a profile of the study participants and described the six main themes that emerged from the interviews. These themes are: the meaning of the intervention; the importance of context, values, and fears in decision-making; the need for information for decision-making; and adaptation/acceptance of the intervention. In many cases, sub-themes were noted as well. While there was much similarity among experiences and opinions of participants with regards to, for example, the importance of others, context, and fears in decision-making, some variations in areas such as, support by the healthcare team were also noted. Collectively, these themes illustrate information pertinent to decision-making about assisted ventilation from the perspective of those interviewed, that is, persons with ALS followed at the MNH ALS clinic. In the next chapter, I will discuss some of the themes in relation to the existing literature on ALS. My analysis will support and reinforce the importance of a pluralistic conception of autonomy. In addition, I will advocate for a shared decision-making model in order to improve the decision-making process of people with ALS with regard to assisted ventilation giving high priority to autonomy and self-determination as stipulated in the AAN practice guidelines (Miller, et al., 1999).

Chapter V

Discussion

The themes described in chapter IV represent the common elements that emerged from the interviews with individuals with ALS who choose to try the BIPAP, those who do not, and those who use LTMV via tracheostomy. These themes capture the essence of the decision-making process of the ALS population seen at the MNH clinic and define, from these patients' perspective, the pertinent information for decision-making about assisted ventilation. Similar elements discussed in chapter II, in the literature review on ALS and assisted ventilation, support the credibility of the research findings (Mustfa, et al., 2006; Kaub-Wittemer, et al., 2003; Bourke, et al., 2002; Lyall, et al., 2001; Albert, et al., 1999; Young, et al., 1994; Moss et al., 1996; Moss, et al., 1993). Some of the themes that emerged from this study are also found in the literature on decision-making and use of assisted ventilation for other conditions (Gibson, 2001; Dales, et al., 1999). In this chapter, I will describe and discuss the high priority given to autonomy in decision-making by participants of this study. In doing so, I am lead to advocate for a pluralistic conception of autonomy when addressing the use of assisted ventilation for persons with ALS. I argue also that a shared decision-making model is better suited to the context of decision-making for assisted ventilation and ALS in order to give high priority to patient autonomy and self-determination as stipulated in the AAN practice guidelines (Miller, et al., 1999). By taking this approach, I forego in this thesis discussion of some of the themes that emerged from this study. I hope to return to them in the future.

5.1 Importance of autonomy

The empirical evidence provided by this research supports, from the patients' and family members' perspectives, the underlying assumption of the AAN (Miller, et al.,

1999) that high priority should be placed on patient autonomy and self-determination in the therapeutic relationship. The value of autonomy and its impact on decision-making about assisted ventilation was discussed in Chapter IV, section 4.23c. Individuals studied here expressed the importance of autonomy and discussed how they value the ability to make their own treatment decisions. This is similar to Young, et al. (1994) who found that their participants also believed that they should make the final decision about the use of assisted ventilation. In the present study, however, the importance of autonomy is reflected in other areas of the moral deliberations and is not related strictly to the decision *per se.*

The importance of patient autonomy is illustrated, in part, by the meaning attributed to the intervention, which appears to impact patient autonomy. The BIPAP was clearly perceived by the study participants as a tool to palliate symptoms, whereas the ventilator is an "artificial" means that allows one to go on living. From the perspective of the individuals interviewed, the artificial nature of the ventilator contributes significantly to their reluctance to agree to its use. Many participants emphasized the importance of not being dependent on a machine for breathing and remaining in control: "*…on sait très bien que le BIPAP respire pas pour elle mais ça peut devenir une béquille, avec laquelle elle doit avoir tout le temps, ou à peu près, pour dormir, puis pour tout faire, là… On s'entend que c'est pas un respirateur artificiel….*" (spouse participant # 2).

The relation between functional independence and autonomy is not restricted to the ability to breathe. Many participants discussed how their present and anticipated physical status impacted their decision-making about assisted ventilation. This is similar to the finding of Young, et al. (1994) where the severity of disability was identified as the second most important factor influencing decisions about assisted ventilation. In the present study, some of the participants expressed that since ALS brings about physical dependence, involvement in decision-making is one of the few areas where one continues to exercise control, self-determination, and autonomy. Consequently, the value attached to decisional autonomy in ALS may be due in part to the patient's lack of ability to exercise autonomy in many other areas of daily living.

The interviews highlighted that, at times, the high value attributed to autonomy can give rise to tensions and discomfort. For example, autonomy can give rise to moral tensions when the individual with ALS feels pressure from the family to make decisions, is unable to do so, and, simultaneously, wants to relieve them from the burden of decision-making. This is illustrated by the following quotation from participant # 6: "*I have come to realize that this condition might kill me and my wife has also expressed the fear of having to make some difficult decisions. She'd prefer that I write them down so that there is no pressure on her to make my decisions for me*". In this case, respect for patient autonomy may interfere with autonomy of the family if they feel that a decision must be made or is inappropriate.

Another example of moral tension resulting from the high value attributed to autonomy in decision-making is highlighted in section 4.23c by participant # 2 (a participant who uses LTMV) when she verbalizes wanting to go on living while having to consider the circumstances for discontinuation of invasive ventilation. In this case, within the same individual, the value of autonomy conflicts with the value of life. Thus, even although professionals, individuals with ALS, and their family members all agree on the high value of autonomy in decision-making, it is important to note that moral tensions that may be detrimental to the patient can arise.

In spite of conflicting values, all but one participant in this study felt that the decision about assisted ventilation reflected who they truly are. In contrast, subject # 5, the man who used the BIPAP constantly except for brief moments when eating, expressed that he felt he had no choice in the decision to use assisted ventilation. He felt that the physician's approach to decision-making took away his decisional autonomy. This patient's approach to decision-making, the physician's attitude, and the decision-making model used may all have influenced this patient's decision. These influences may have been significant to the point of engineering the outcome of the decision. Thus, one may question whether the decision to use the BIPAP was grounded in the person's longstanding values and whether the voluntariness of the consent was sufficiently substantial to make it an autonomous decision as defined in Faden's scaled conception of autonomy (1986).

This participant also expressed being unaware that LTMV could be used as an alternative to constant use of BIPAP. For one to be able to exercise autonomy in decision-making, one must first be aware that a decision is needed and, subsequently, be provided with information on interventions, alternatives, and anything else necessary for one to make an informed choice. One may question whether, in this case, LTMV as an alternative to constant BIPAP use was ever discussed with the patient. Sharing of information and possibly its understanding may have been insufficient to allow this participant to exercise autonomy in the context of decision-making about invasive ventilation. This case illustrates that the decision-making model used, as well as the attributes of a specific individual can have a definite impact on ability to exercise autonomy in treatment decisions.

The responses of individuals who opted for and against the use of assisted ventilation in this study contribute to a confirmation that individuals with ALS do arrive at different conclusions when presented with the need to make decisions about assisted ventilation. Furthermore, the data indicates that the decisions may not necessarily result strictly from rational reasoning. The reasoning may involve much more than deciding that one cannot tolerate using the BIPAP because of secretions or bulbar involvement as suggested by some studies on use of BIPAP (Lo Coco, et al. 2006; Gruis, et al., 2005). Notably, the decision-making process revealed by this study draws heavily on moral reasoning that takes into account the person's context, relations, values, beliefs, and emotions. Consequently, a conception of autonomy that assumes only rational deliberations and a single conclusion to moral reasoning is il-suited for people with ALS making decisions about assisted ventilation.

5.2 Factors influencing decision-making

The context of ALS is complex: the preservation of cognitive function, the typical rapid physical deterioration and high level of disability, the absence of a cure, and the possibility of using artificial methods to prolong life all contribute to the complexity of decision-making in ALS. The interview data in Chapter IV confirms the pertinence and possible influence of the scientific evidence with regard to decision-making discussed in section 2.4, particularly with respect to the risks and benefits of the interventions. Some of the studies also address aspects of the patient's context, QOL, and relationships that may influence autonomy and decisions (Mustfa, et al., 2006; Kaub-Wittemer, et al., 2003; Bourke, et al., 2002; Lyall, et al., 2001; Young, et al., 1994). Similarly, many participants of this study also discussed these aspects in relation to decision-making. In addition, they highlighted the significance of communication, emotions, and availability of services and adapted equipment with regard to autonomy and decision-making. The next sections will discuss these factors that influence autonomy of persons with ALS making decisions about assisted ventilation in order to, subsequently, define the conception of autonomy that is best suited to this context.

5.21 Patient context

The participants in this study confirm the significance of patient context as described in section 2.4 in relation to decision-making. They confirm the influence of factors, such as, disease type, rate of progression, and functional status on their ability to make autonomous decisions. Participants also confirm the influence of aspects such as knowing which symptoms are due to respiratory failure, how they are relieved by the intervention, and for how long. These factors make up a specific individual's medical context.

In addition, participants talked about the impact of their specific personal context. One's perception of the self or self-identity was brought up by many people interviewed and affects decisions about assisted ventilation. For example, for participant # 7, an educated man who, prior to the illness, had always been in control of his decisions, held a high profile position, was financially secure, and fiercely independent, using LTMV was an expression of his ability to continue to exercise control and was consistent with how he defined himself.

Participant # 2 also discussed how her definition of the self influenced her decision to use BIPAP and LTMV. She included her role as a mother, spouse, and nurse with a Master's degree in her self-identity. In the interviews, she and her spouse frequently discussed how these roles define the self and impacted deliberations about assisted ventilation. The importance of her role as a mother is illustrated by the following quotation: *"Je veux que mes enfants puissent finir leur année scolaire avant que je décède"* (participant # 2).

In addition to one's medical context and self-identity, the study highlights that the presence of a dedicated caregiver also constitutes one's context and, as such, may influence one's decision-making about assisted ventilation. For example, participant # 9 describes how her husband's death impacted her ability and willingness to use the BIPAP: *"Je l'ai déjà mis [BIPAP], quand mon mari était ici, lui, il s'installait dans le fauteuil, il me mettait toutes mes affaires là... [Depuis que votre conjoint est décédé vous ne l'avez pas utilisé du tout?] Pas une fois!"* (participant # 9). The impact of the availability of a dedicated caregiver is even more significant in the case of LTMV as illustrated by the following quotation from the spouse of participant # 7 who uses LTMV: *"Il y est venu un bout de temps, là, je regardais, là, je faisais mon horaire en fonction de l'horaire des infirmières. Je veux dire, quand "elle" est là, il faut que je sois là!"*

How rarely participants talked about the impact that one's financial context may have on decisions about assisted ventilation is a surprising finding of the study. Yet, the impact of assisted ventilation on one's financial situation is unmistakable as illustrated by the following citation: *"With a tracheostomy, I'm stranded to a nursing home... The cost of being at home means hiring 24 hour help, over 500$/day...The other option is a nursing home. The other option costs $4000/month."* (participant # 1). Thus, it is difficult to understand this study's finding that one's financial context is not an influential factor on one's ability to exercise autonomy and make decisions about assisted ventilation. This may result partly from some participants' perception that assisted ventilation does not entail anymore assistance and, hence, anymore financial resources than that already required in light of their severe physical disability. This finding may relate also to the fact that, in Quebec, the cost of the equipment is covered by the public health care system.

Since professionals generally recognize the importance of the patient's personal and medical context, I suggest it continue to be explored during decision-making in relation to assisted ventilation. Factors, such as, expected benefits of intervention on symptom management, physical and technical support required, burden of care, and financial impacts, influence decisions. Yet, they do not preclude autonomous decisions since they do not necessarily constitute coercive, persuasive, or manipulative forces that would control the decision *per se* and void it of autonomy Consequently, professionals should disclose to patients the scientific evidence presented in section 2.4 to acknowledge these influences on autonomy and decision-making. Nevertheless, considering strictly the person's medical and personal context is insufficient to ensure autonomous and informed decisions.

5.22 Quality of life

Another influence on decision-making identified by the participants of this study is the value of QOL. During the interviews, participants discussed different aspects that make up quality of life. These include: physical function, and the ability to eat and move around, which are similar components of good QOL discussed in other studies (Mustfa, et al., 2006; Kaub-Wittener, et al., 2003). However, as stated by Professor O'Boyle who contributed to the development of a QOL assessment tool (Hickey, et al., 1996), the meaning of "good" QOL is whatever the person defines it to be.

Many participants in this study discussed that where one lives impacts QOL and decisions about assisted ventilation. This is similar to the findings of Young et al. (1994) who found that the possibility of being admitted to a long-term care facility impacted negatively the decision to use assisted ventilation. For many individuals interviewed in this study, use of invasive ventilation was automatically associated with placement in a long-term care facility. For some, the importance attributed to being at home was related to the ability to do as one pleases and to control one's care, schedule, choice of activities, etc. Yet for many participants, the impact of remaining in the home setting on QOL pertained mainly to its impact on relationships and being surrounded by loved ones.

The finding that relationships impact QOL is confirmed by a study done by Borasio (2001) where individuals with ALS were surveyed in an attempt to define QOL. One hundred percent of the individuals surveyed in that study identified "family" as one domain defining good QOL. Not only is it identified as one domain impacting QOL; it is the most important aspect for 75% of the people interviewed. Borasio (2001) goes on to question whether many ALS studies that use standardized QOL measures focus excessively on physical function and, as such, are found to be distressing and not valid by the patients themselves. Thus, the scientific evidence found in the ALS literature about QOL may be biased. Sharing strictly the scientific evidence about QOL and assisted ventilation with patients and caregivers may be insufficient to promote informed decisions and may overlook essential components of good QOL from the patient's perspective. Thus, exploration of the patient's own definition and experience of QOL is an essential element to an informed decision since QOL is a subjective notion, varies over time, and is most likely not accurately estimated by others.

5.23 Personal relations

Individuals with ALS interviewed in this study acknowledge that their decisions about assisted ventilation are influenced, amongst other things, by their context and definition of QOL. As discussed previously, the impact of relationships in defining QOL is significant, however, the participants of this study do not restrict the influence of relationships to its impact on QOL. In his biography about a former professor confronting his own experiences with ALS, perhaps Mitch Albon in Tuesdays with Morrie best describes the importance of relationships:

> The fact is, there is no foundation, no secure ground upon which people may stand today if it isn`t the family… If you don`t have the support and love and caring and concern that you get from a family, you don`t have much at all. Love is so supremely important. As our great poet Auden said: "Love each other or perish". (1997, p.91)

The reports of the individuals interviewed make it clear that there are various ways that relationships impact on decision-making about assisted ventilation. For some, significant others were involved in gathering information and deliberating for decision-making. For others, the needs of their loved ones as well as the impact each intervention might have on the family was taken into account.

One of the impacts of the interventions on the family discussed by participants was increased burden of care. The need for intensive involvement of others for use of assisted ventilation and its associated increase in burden of care was already discussed in studies reviewed in section 2.4 (Mustfa, et al., 2006; Kaub-Wittemer, et al., 2003; Bourke, et al., 2002; Lyall, et al., 2001; Moss, et al., 1993) . The fear of being a burden is not specific to the MNH population. A study by Hirano (2005) also found that ALS patients using invasive ventilation are concerned with being a burden on their family. Thus, sharing the information on burden of care associated with use of assisted ventilation with the patient and family members contributes to promoting autonomy and informed decisions by acknowledging that this factor impacts decisions. Furthermore, these discussions allow the patient and the family to make a more accurate risk/benefit assessment of the intervention and its global impact and to plan accordingly.

In addition to the help others can provide, the impact of treatment decisions on family may be a serious, critically important component of the patient's definition of self. Participant # 2 and 9 addressed the importance of their personal relations in defining who they were. Both discussed their role as mothers in defining their values and how this affects their decision-making. When questioned about the reasons for deciding to use LTMV, participant # 2 responds: *"J'ai pensé surtout à mes enfants, les protéger".* Similarly, participant # 9 is motivated to keep trying to use the BIPAP in order to go on living and be able to buy a new house for her daughter and grandson. The significance of personal relations in defining the self and one's values is not restricted to parental relations. Participant # 4 also highlights the importance of protecting his spouse from the negative impacts that may result from his treatment decisions.

The importance of relationships with respect to autonomy is not restricted to the context of ALS. It is also recognized in the context of other serious illnesses. For example, the importance of relationships on autonomy is found in the The Body Silent written by Robert Murphy (1990) in which he tells the story of a man (Murphy himself) who progressively becomes quadriplegic as a result of a spinal tumour. His reflections draw on his anthropological research of quadriplegia and also recognize the significance of relationships in defining the self. This text gives credibility to the finding that personal relationships impact autonomy and decision-making in a much more complex fashion than strictly from a QOL perspective. Thus, professionals who only address the family in relation to burden of care or QOL during decision-making risk ignoring some factors very important to their patients' sense of self and how decision-making should take place within their own context.

5.24 Communication

The present study highlights another factor that influences decisions about assisted ventilation: the ability to communicate. The ability to communicate is closely related to the value of relationships and the ability to interact with others and, as such, it also affects whether one is willing to use assisted ventilation. Hirano's study (2005) on the needs of ALS patients who use LTMV also confirms the significance of communication and its impact on decision-making about assisted-ventilation. Yet, the importance of communication is not restricted to individuals who use invasive ventilation. A study by Hecht, et al. (2002) where persons with ALS identified the worst aspect of the disease as the "reduction of speech" also substantiates the importance of communication for individuals with ALS in general, similar to the findings of the present study. Based on their research, Hecht et al. suggest that losing one's ability to communicate may lead to greater emotional suffering, hence, it is more significant than loosing one's mobility.

The importance of communication in decision-making highlights the need to inform patients and caregivers of the technological means available to them to palliate loss of speech. Providing this information may help to reduce the fears associated with not being able to predict one's functional status and ability to communicate once the need for assisted ventilation arises as discussed in section 4.24e. The use of advanced directives may also reassure the person with ALS (or their family) that even when they can no longer communicate, their wishes can be known and respected. Participant # 8 used this strategy: *"If I'm on a respirator [and cannot communicate anymore], I think I would* *want to be unplugged... That's what I wanted and I wrote that all on my document".* The impact of communication ability on some people's willingness to continue using assisted ventilation may also support discussing the circumstances for discontinuation of ventilation before it is actually initiated.

In addition to patient context, QOL, and personal relations, the value of communication is another factor that influences autonomy and decision-making about assisted ventilation. Consequently, to promote patient autonomy in decision-making, professionals would benefit from better recognizing its importance and discussing with their patients information on communication devices and their availability even if the importance of communication in relation to decision-making about assisted ventilation is not well documented in the literature. Even though communication may not, at first glance, be considered a pertinent value to be explored in this context, it is identified as such by the participants of this study.

5.25 Emotions

Participants in this study talked about the impact that various emotions may have on their decisions about assisted ventilation. As discussed in section 5.23, emotions such as love are part of the reason why relationships have such a significant impact on one's definition of the self and decision-making. Fear is another emotion that was repeatedly discussed as affecting decisions about assisted ventilation. As noted in section 4.24, participants' fears were varied and included such things as fear of being restricted to bed with a ventilator, being locked into one's body, choking to death, or even for one person who used LTMV, the constant fear of a mechanical breakdown or power failure. These findings are not unique to the present study and have been documented in other studies on ALS (Borasio, 2001; Hirano, 2005).

This study identified one fear that was shared by all the participants: the fear of death and how it proceeds. For individuals with ALS, the absence of a cure and the certainty of death are omnipresent. From the moment of diagnosis, individuals with ALS and their family live with the anticipation of death. Some participants expressed that every time there was a noticeable change in their condition, they would wonder if the moment of death had arrived. The data from the interviews highlights that the inability to predict how the disease typically progresses to result in death generates concerns for patients and family members alike and impacts decisions: "*…est-ce que forcément tout ce monde là rentre en détresse respiratoire tout le temps? Si c'est le cas de tout le monde, c'est important de le savoir puis si c'est le cas d'une minorité, c'est important de le savoir aussi…*" (spouse participant # 2). Patients and families expressed that the fear of death related mainly to fear of discomfort, chocking, running out of air, and suffocating at the moment of death. Over time, as ALS progresses, these fears impact one's values and beliefs so significantly that they contribute to the definition of the self and the decisions one makes.

Even though most of the scenarios feared by participants are not supported by clinical experience and scientific evidence-particularly since "the terminal phase in ALS is peaceful" (Borasio, 2001, p.S33)-they relate to events that are possible in ALS. The lack of empirical data pertaining to the impact of fears on decision-making about assisted ventilation may contribute to professionals' current avoidance of discussions about death. In fact, patients, their families, and clinicians frequently collude to avoid mentioning death or the fears raised by this topic. Yet this research suggests that they should acknowledge and explore the impact that fears have on decisions. If not, they risk ignoring emotions and beliefs that influence decisions, thus preventing patients from making autonomous decisions. More importantly, by exploring patients' and families' fears, professionals can better provide psychological and emotional support.

In addition to fears that are founded on events that can possibly occur in ALS, a few participants discussed misconceptions they held about assisted ventilation. For example, some of them compared the use of invasive ventilation in ALS with that of people in a neuro-vegetative state. These participants discussed how they feared that use of a ventilator would bring them to a state similar to that of individuals in a neuro-vegetative state. Use of invasive ventilation with ALS does not imply a neuro-vegetative state, thus this fear is based on false information or false beliefs. Decisions made under the influence of such fear are misinformed and cannot be considered autonomous. Addressing patients' fears, therefore, allows clarification of possible false beliefs and misconceptions and provision of accurate information increasing the potential for informed decisions that are an expression of autonomy.

Since the fear of death in general and, more specifically, the fear of death from respiratory failure are omnipresent and impact decisions about assisted ventilation, professionals must provide information on the intervention itself as well as its alternative, namely no intervention and death. The typical events that result in death in ALS may be discussed to relieve some of the concerns associated with death from respiratory failure. For instance patients and families should be informed that, typically, ALS patients do not choke to death and that death is peaceful. The use of medications to relieve feelings of shortness of breath and discomfort in addition to assisted ventilation or as alternatives can be discussed as well. The possibility of the patient dying in the hospital or in a palliative care unit if the prospect of dying at home is too distressing can also be explored. More importantly, patients and family members can be reassured that support and care will be provided regardless of the treatment decision.

5.26 Societal relations

Participants of this study discussed how societal influences were also included in the moral deliberations for decisions about assisted ventilation. Many people interviewed talked about how the availability of services from agencies such as the CLSC or respite care affected decision-making. For example, the spouse of participant # 2 expressed the importance of respite care in the following terms: "*..l'option du répit, pour nous, va devenir importante parce que... si on a pas, à un moment donné, un "break" pour se ressourcer un peu... C'est pas chaque soins, chaque journée qui devient difficile. C'est la chronicité de tout ça!... Si on est pas capables à un moment donné de décrocher...* ".

The data from this study illustrates that the impact of societal relations on decisions is not limited to one's relations with health care agencies. The availability of homes that are accessible to wheelchairs, the provision of financial assistance to allow home modifications, the supply of subsidized wheelchairs, communication devices, or computerized environmental controls were identified by many as significant influences on decision-making about assisted ventilation. The influence of these factors on decisions may result, in part, from the important impact these aspects have on QOL as supported by a recent study done by Foley, et al. (2007) where participants confirmed that services and support received contribute to well being and coping with the disease.

One's access to such services and equipment is dependent, among other things, on society's views on disability and chronic life threatening diseases, and on societal decisions for allocation of resources, which may vary from sector to sector. At first glance, the influence of whether one has access to an adapted wheelchair to carry the equipment for assisted ventilation may not be considered a significant component of autonomy. However, in some cases, its impact is so significant as to determine the outcome of the decision about whether to pursue or forgo assisted ventilation. Thus, to promote autonomy and informed treatment decisions, professionals must acknowledge the influence that patients' relations with societal agencies and availability of publicly-funded equipment have on decisions. To do so, they may need to involve the various agencies involved in patient care, such as the CLSC or the rehabilitation center that is responsible for provision of various adaptive equipments, in the deliberations about assisted ventilation.

In the next section, I will argue that even although a liberal conception of autonomy, such as Faden's definition described in section 2.11, may acknowledge some of the factors identified by the participants in this study as being influential to decisions about assisted ventilation, this is better accomplished by adopting a pluralistic conception of autonomy.

5.3 Superiority of a pluralistic conception of autonomy

The interview data from this study confirms the importance of autonomy in decision-making about assisted ventilation. This is consistent with the AAN's guidelines, which states that: "High priority should be placed on patient self-determination or autonomy as an underlying assumption in the therapeutic relationship" (Miller, et al., 1999, p.1311). The AAN does not define its use of the term "autonomy". The factors that impact decisions identified in this study contribute to clarifying the definition of autonomy in the context of ALS and assisted ventilation. The interview data illustrates that patient's medical and personal context, including their self-identity, QOL, personal relations, and ability to communicate influence decisions about assisted ventilation. The influence of these factors is compatible with a liberal conception of autonomy (see section 2.11).

In addition, participants emphasized that these factors evolve over time. They expressed that it is extremely difficult for them to anticipate how they will perceive and feel about their situation before actually living the experience of respiratory failure. "*C'est pour ça que ça c'est un petit peu difficile à prévoir! Parce que là à brûle pour point comme ça, moi je peux dire : « Non, moi j'en veux pas! » Quand t'es rendu au pied du mur, ça peut être bien différent! Ça fait que ça c'est pas quelque chose qui tu peux jouer d'avance là!*" (participant # 4). Thus, they stressed the importance of timing of information in relation to decision-making since the assessment of their situation as well as their decisions may change over time. A liberal conception of autonomy accounts for the notion that one's wants, values, desires, and definition of the self evolve over time as one lives through various experiences.

A liberal conception of autonomy may be adequate in many decision-making contexts however, its emphasis on primarily rational processes in decision-making is poorly supported by the findings of this study. In this study, participants discussed the impact that emotions such as love and fear have on decisions. These influences more readily suggest a pluralistic conception of autonomy, which recognizes that the influence of emotions does not necessarily lead to irrational decisions and, as such, to non-autonomous decisions. With a pluralistic conception of autonomy, decisions that may be out of character, unusual, or opposite to what the medical evidence suggests may be influenced by deep-rooted fears or emotions that are truly a part of someone's identity. This is especially important in the context of ALS since, over time, as the disease progresses, these emotions impact one's values and beliefs so significantly that they contribute to the definition of the self and the decisions one makes. Thus, adopting a pluralistic conception of autonomy is especially well suited to acknowledging the influence of various emotions on autonomy and their impact on decisions about assisted ventilation.

By adopting a liberal conception of autonomy, professionals risk focusing solely on personal relations and their impact on QOL or aspects of burden of care while ignoring more subtle and complex ways in which relationships impact decisions. A pluralistic conception of autonomy, on the other hand, acknowledges patients' desire for involvement of significant others in decision-making. In contrast to a liberal conception, it also recognizes that individuals identify with the interests of their significant others (something also noted by those interviewed in this study). Thus, when patients act in accordance with these interests, whether that involves the process of coming to a decision or the actual decision made, they also act in their own interest and, as such, they exercise autonomy.

Participants in the study also discussed the significant impact societal relations have on autonomy and decision-making in the context of ALS and assisted ventilation. As discussed in section 2.12, the influence of societal relations on autonomy is better addressed by pluralistic conceptions of autonomy. By adopting a liberal conception of autonomy, professionals risk ignoring this very significant influence on their patients' autonomy and decisions. In contrast, adopting a pluralistic conception of autonomy encourages professionals to explore the influence of availability of services in the home, respite, or adapted equipment in relation to decision-making and assisted ventilation.

Only by adopting a pluralistic conception of autonomy does one acknowledge the wide range of influences identified in the present study, giving high priority to autonomy and self-determination as stipulated by the AAN and maximizing the potential for ALS patients to exercise autonomy. A pluralistic conception of autonomy not only acknowledges the factors that are congruent with a liberal conception of autonomy, it also accounts for other influences on decision-making that are identified in the present study. Moreover, since it recognizes a wider range of influences on autonomy and decisions, a pluralistic conception of autonomy promotes more in depth exploration of patient's and family's concerns and encourages sharing of information that is not restricted mainly to the scientific evidence.

In addition to information sharing being influenced by the conception of autonomy adopted, it is impacted, among other things, by a patient's approach to decision-making and the physician's attitude. These factors are influenced by the medical decision-making model adopted. Thus, the next section will address themes from the study that substantiate that a shared decision-making model is better suited to promoting a pluralistic conception of autonomy in the context of ALS and assisted ventilation.

5.4 Impact of findings on decision-making models

In section 5.1, I confirmed the importance of autonomy for individuals with ALS making decisions about assisted ventilation. In most cases of decision-making and assisted ventilation, the use of a paternalistic approach is unjustified since it prevents individuals from using any form of autonomy, which is contrary to the desires expressed by the participants in this study.

The emphasis of the paternalist decision-making model is not on autonomy; its emphasis is on patient "good". It assumes that professionals know best what is in the interest of the patient. Disclosure of information is limited to professionals sharing the information that encourages patients to agree to the treatment proposed. Even though professionals may have a better understanding of the medical context, the study data illustrates that one's personal context, QOL, values, relations, and emotions affect decision-making. The impact of an intervention on one's personal context is best determined by the individual with ALS provided he/she has a correct understanding of the intervention itself. Partial information sharing typical of the paternalist model may contribute to limited understanding of the information since patients are unable to fully explore the impacts an intervention may have on their specific context. Furthermore, its use precludes individuals from sharing and discussing what matters most to them and the factors they would want decisions to take into account.

The people interviewed discussed at length their desire for information. Even though in this study there are differences between patients' and family members' informational needs, there exists a clear consensus that information is empowering and essential to decision-making. Therefore, one may be tempted to adopt an informed model of decision-making. However, many findings from this study suggest that an informed model may be il-suited to the context of ALS and assisted ventilation.

According to the participants, the information needed for decision-making spans the technical information about the intervention, the symptoms of respiratory failure, and the impact of symptoms and interventions on every day life. Participants also talked at length about their preoccupations with their imminent death from respiratory failure as illustrated by the spouse of participant # 2: *"...[la ventilation assistée} c'est un facteur de stress et de fatigue, d'inquiétude constante! Et à toutes les fois que je laisse un message pour un des enfants, ils sont toujours en mode "tu m'appelles-tu pour me dire qu'il faut qu'on aille à l'hôpital là?"* Participants expressed that they need more information on sensitive issues such as death resulting from respiratory failure and discontinuation of invasive ventilation.

The ALS literature is scarce on these issues. Professionals may need to rely more on their clinical experience in caring for people with ALS in order to share and discuss these matters with their ALS patients. Thus, professionals withholding their experience or opinion, as stipulated in the informed model, may create a situation where patient and family feel abandoned at crucial points of time in the course of the disease. In practice, use of the informed model may actually provoke poorly informed decisions since the person with ALS is missing crucial information (e.g. to address the fears that were highlighted in section 4.24).

5.41 Superiority of a shared decision-making model

The superiority of the shared decision-making model in the context of ALS and decision-making about assisted ventilation is supported by findings from this study. As the data from the interviews points out, many individuals are involved in the decision-making process. Many participants commented on the involvement of family and friends in the decision-making process as discussed previously in section 5.23. Furthermore, participants described how at the MNH ALS clinic, many of the professionals such as, the neurologist, pneumologist, nurse, and respiratory therapist are involved in the decisions about assisted ventilation. As well, the individuals with ALS interviewed rely on many other agencies, such as the CLSC or the MCI for provision of services. Consequently, many participants involved these professionals as well in their deliberations about assisted ventilation. This is consistent with a shared decision-making model that acknowledges the possible involvement of more than two individuals in decision-making.

The context of decision-making about assisted ventilation in the population interviewed is complex. At various stages of the decision-making process (i.e., during information exchange, deliberation, and decision), the different participants take on different roles. Family members and professionals may provide information, act as “sounding boards”, or be involved with the patient in actually reaching a decision. For example, the caregiver of participant #2 discussed his involvement in his wife’s decision-making as: “*...moi, mon rôle comme conjoint là dedans, c'est de valider avec elle "est-ce que t'as vraiment toutes les informations, est-ce que tu sais vraiment tout ce qu'il nous faut ?"* A shared decision-making model allows the various participants to take on different roles at the different stages of decision-making.

A shared decision-making model is also supported by the participants’ emphasis on timing of information. As discussed in section 4.25b and 4.25c, both the individuals with ALS and their families want information to make informed decisions, however, they are ambivalent about when this information should be provided. They describe a process of information exchange, deliberation, and decision that is continuous, evolves over time, over the course of the disease. Many participants stressed the importance of having continuous support and information as their condition declines and their context changes. They also expressed their appreciation of the expertise and the services provided by the various agencies. Most of the participants expressed feeling supported throughout the experience of assisted ventilation no matter what decisions were made. This is consistent with a shared decision-making model where all the parties involved have a commitment to sharing and assessing various sources of information, sharing the responsibility, and accepting the treatment decision made.

Throughout the interviews, participants discussed their longstanding involvement with the various professionals engaged in their care. They described that a trusting relationship evolved over time. Thus, some participants came to rely on the professionals' assessment of their best interest to support their decisions about assisted ventilation. For example, participant # 3 talked about how she was grateful that one of the professionals from the CLSC had encouraged her to persevere in using the BIPAP even though she herself doubted its usefulness. She also described how her acceptance of assisted ventilation resembled her acceptance of other assistive devices, a process that required time and patience. Thus, through the long-standing therapeutic relationships that evolve over the course of ALS, professionals may be in a better position to estimate patients' capacity and pattern of adaptation to assist and support them to make informed treatment choices that ultimately match longer term goals or patterns of acceptance and adaptation. Thus, the medical context described by the participants in this study is similar to the context of chronic diseases described by Cathy Charles (1997) where she argues for the superiority of a shared decision-making model.

The model of shared decision-making emphasises ongoing communication, which is better suited to the context of ALS and decisions about assisted ventilation, and fulfills the needs expressed by the participants of this study. Its use allows for better exploration of patients' and families' concerns and preoccupations thus improving the quality of care provided to these individuals. However, adopting such a model on a consistent basis at the MNH ALS clinic has implications for clinical practice. These will be briefly discussed in the last chapter of this thesis.

Chapter VI

Recommendations for clinical practice

Many participants expressed their gratitude for my undertaking this project, trying to better understand their lived experience, and ultimately attempting to improve the services provided to individuals with ALS. Thus, I will make some recommendations pertaining to the clinical practice in an attempt to facilitate the decision-making process of these individuals and improve the care provided to them at the MNH ALS clinic.

6.1 Involvement of multiple parties in decision-making

In light of the research findings, decision-making about assisted ventilation should involve many individuals. Participants in this study have highlighted the importance of family involvement in decision-making. Thus, unless requested otherwise by the patient, I suggest that the patient and family be viewed as a unit to be involved in decision-making.

The various professionals of the multidisciplinary teams involved in specific patient care are also part of the decision-making process as discussed in this study. Their collective involvement appears preferable to solely calling upon the experience of the physician to address the various concerns highlighted by the participants. For example, while the physician may better address the medical benefits and alternatives of treatment, the impact of assisted ventilation on mobility may be better addressed by the occupational therapist, exploration of the spiritual meaning of the intervention by the pastoral worker, and the impact on communication by the speech-language pathologist. Thus, I suggest that a multidisciplinary approach be maintained with regard to decision-making and assisted ventilation.

A multidisciplinary approach has an increased potential to be uncoordinated and undirected leading to more anguish for patients and families. To minimize this possibility, some of the discussions could take place with multiple disciplines present, being careful not to overwhelm the patient and family while addressing their concerns with regards to the day-to-day impacts of the intervention. In addition, I suggest the use of a respiratory protocol, advanced directives, and a checklist of topics to be covered when discussing assisted ventilation to promote a coordinated approach to decision-making and assisted ventilation.

6.2 Respiratory protocol

As discussed in section 4.21b, some of the individuals interviewed appeared to be unaware of the need to make decisions about LTMV or lacked information to make truly informed treatment decisions. Others, clearly had misconceptions about assisted ventilation. Recall, for example, one comment with regard to one's mobility: "*La tracheo. je ne sais pas si... Je n'ai pas plus de détails que ça là. Je ne sais pas si on est encore plus limité dans les déplacements? J'imagine que, oui*" (participant # 4). These misconceptions may result in part from patients and families getting their information from various sources such as, television, internet, ALS support groups, and so on, and making incorrect analogies to their specific situation.

In addition, these misconceptions may be explained by the team's attitude in specific situations or the fact that, as the disease progresses, some individuals may be less informed as a result of being seen less frequently at the MNH ALS clinic and being cared for mainly by homecare teams. Thus, in such a context, professionals from other agencies, such as the MCI or the CLSC may need to take on the responsibility of sharing information and assisting with treatment decisions. In an attempt to improve communication, increase sharing of information, clarify misconceptions, and promote a standardized approach across the various teams involved in patient care, I would suggest the use of a common respiratory protocol.

Some aspects of a respiratory protocol are already integrated into practice in relation to use of BIPAP at the MNH ALS clinic. For example, as stipulated in the AAN Practice Guidelines (Miller, et al., 1999), FVC is systematically measured in clinic and patients are offered BIPAP when they present with symptoms such as, dyspnea, morning headache, daytime hypersomnolence or when they present with impaired pulmonary functions as demonstrated by a FVC <50%. Unless requested otherwise by the patient, this practice guides professionals in timing concrete discussions about the need for non-invasive ventilation. Since, as discussed in section 1.5, professionals often find decisions about end of life issues difficult to initiate, it would be beneficial to integrate specific guidelines pertinent to LTMV as well, to minimize the impact of the professionals' personal attitude on timing of information and decision-making. For instance, it may be agreed that use of LTMV is discussed with all patients who refuse BIPAP and those who have progressed to daytime use of BIPAP.

Use of a respiratory protocol could possibly prevent the initiation of assisted ventilation that is based on an arbitrary basis (eg., on the professionals attitudes towards the interventions more than on the patients' preferences). In addition, using such a protocol could minimize the professionals' subjective contribution to decision-making and standardize the timing for sharing of information about assisted ventilation.

As illustrated by the data from the interviews, there is no consensus with regards to what is the best time to provide information for decisions about assisted ventilation. Early on in the disease, patients should be encouraged to explore how they prefer to proceed to get information and make decisions about potential interventions. The respiratory protocol could serve to document the patient's preferences for information and who they wish to be included in the process of decision-making. As suggested by participant # 6: "*I suggest that you ask the person one question at the beginning. Would you like me to tell you a few things about ALS, for example, if you want me to tell me about breathing, talking, as opposed to giving me a booklet of 45 pages of all kinds of information, to give a broad talk with a person and tell them personally, to help to think a little bit of things you can possibly expect.*" Thus, in order to meet the patient's expectations, at the initial stages of the disease, professionals may discuss assisted ventilation only in general terms and, at the onset of respiratory symptoms, in more concrete terms.

Many individuals with ALS want their family involved in decision-making. However, as discussed in section 4.25c, patients and family members have distinct informational needs. For example, upon diagnosis, the individuals with ALS interviewed preferred not knowing too much about the disease: *"I admit it that I was familiar a little bit with the definition of the disease but I felt maybe formerly that I wanted to keep myself ignorant of the disease"* (participant # 6). On the contrary, the family members expressed that they preferred getting all the pertinent information on the typical course of ALS : "*Puis moi ce que je trouve déplorable, quand ils nous ont dit la sclérose, donnez-nous l'information maintenant...c'est quoi qui vous attend, les voix respiratoires, l'orthophonie....On veut savoir! Donnez-nous de l'information puis on est capable de comprendre l'impact...*" (spouse participant # 4). As a consequence, at times, each may need to be seen separately to better meet their respective needs.

I suggest that upon diagnosis family members be provided with general information on various aspects of the disease such as, equipments available to palliate physical disabilities, home modifications, dysphagia, and respiratory failure. This may be achieved through the use of information sessions designed specifically for family members. Later on in the course of the disease or at the onset of respiratory symptoms, while the various professionals of the multidisciplinary team meet with the patient to discuss assisted ventilation in more concrete terms, the family members can be present as well to support the person with ALS and ensure that both parties have a similar understanding of the information.

As discussed in section 4.26b, patients feel ill equipped to make decisions about assisted ventilation until they actually experience symptoms of respiratory failure and try the intervention. In light of the degenerative nature of ALS affecting decisions over time, decisions made must be revisited periodically to ensure that they still represent the person's wishes. The frequency at which decisions are revisited may vary depending on the rate of progression of the disease. For example, every 3 months may be suitable for someone with a rapid progression, however it may be distressing for the patient and the family to review decisions this frequently when the illness progresses more slowly or when the patient feels that she/he has already made her/his final decision. Thus, the respiratory protocol may include a grid, established in conjunction with the patient, to remind professionals about the need to review decisions at set intervals.

6.3 Advanced directives

Use of advanced directives may also promote autonomous decisions. However, similar to informed consent which requires much more than a mere signature, advanced directives (especially those that contain specific instructions) are only as good as the exchange of information upon which decisions were made. In some cases, use of advanced directives (even if only to name a mandator for decision-making) contributes to sharing of information between patients, families, and professionals. In other cases, use of advanced directives is an opportunity to clarify general principles for decision-making. Thus, use of these documents may promote autonomous decisions by encouraging participation of all parties implicated in the three stages of shared decision-making and contributing to a consistent approach that reflects the patient's wishes and preferences.

In addition to improved consistency of approach, advanced directives may also contribute to improved communication among the multiple professionals and agencies involved in the care of the individuals followed at the MNH. Therefore, I suggest that once completed, these documents be routinely included in the patient's medical file as well as shared with the various teams involved in patient care.

6.4 Topics to be discussed for decisions about assisted ventilation

The information provided to patients in clinic for decision-making about assisted ventilation currently focuses mainly on the justification of the need, the potential medical benefit or expected outcome of the intervention, the medical and technical requirements of assisted ventilation, and its impact on quality of life and burden of care. Professionals must continue to share up-to-date information to facilitate decision-making. However, as highlighted in this study, current practice is insufficient to fulfill patients' and caregivers' needs for information and neglects many areas, especially with regard to the impact of assisted ventilation on daily activities, patients' fears, and death.

The data highlighted the fact that MNH ALS clinic patients have many misconceptions about the impact of assisted ventilation on daily living. These misconceptions span the use of assisted ventilation and its impact on mobility, secretion management, ability to eat, home care, and, most importantly, communication. Thus, I would suggest further involvement of the various professionals in, for example, clarifying misconceptions about the need to remain in bed with invasive ventilation or sharing specialized information pertaining to the numerous technological devices available to maintain communication as the disease progresses. Each professional should be encouraged to get involved in sharing information to promote decisions that are based on accurate information.

Thorough exploration of patients' fears and open discussion about death should be undertaken to facilitate coping with the disease as well as decision-making about assisted ventilation. Sensitive and individualized discussions about assisted ventilation, its alternatives (namely no intervention and progression of respiratory failure), provision of medications for comfort, hospitalization if necessary, etc. should be undertaken with patients and caregivers. Open discussion on these sensitive topics can contribute to providing information that participants identified as pertinent to their decision-making. End of life discussions can also provide professionals with a context to verify patients' understanding of the situation and information, and to explore the voluntariness of decisions. More importantly, addressing death and respiratory distress can be an opportunity for professionals to follow up on misconceptions identified previously, provide support, reassurance, and comfort to the patient and the family.

Withdrawal of assisted ventilation also constitutes an alternative intervention and, as such, should be discussed at initiation of treatment. Merely stating that withdrawal of ventilation is legally permitted in Canada is insufficient, since, as discussed previously, fear of the unknown impacts treatment decisions. Thus, the specific circumstances and means for withdrawal of assisted ventilation should also be explored during the initial discussions.

6.5 Limitations of the study

A limitation of this study may arise from my previous involvement with the participants. Even although precautions were taken to distinguish my role as a researcher from that of an occupational therapist, my clinical role on the ALS team may have influenced some aspects of the findings. For example, participants being aware of my role as a clinician on the team may have discouraged them from highlighting negative aspects of their experience with the ALS team and may have contributed to the mainly positive description of the involvement of professionals in treatment decision-making. However, that some participants expressed dissatisfaction with the care they received and the role played by professionals leads me to believe that any negative effect of my involvement on the team on the accuracy of the results was minimal and that my dual role as occupational therapist and researcher in fact facilitated the research.

The greatest challenge during the research was communicating with the subjects. Because of the speech difficulties and to facilitate the communication of persons with ALS, it was sometimes impossible to use strictly open-ended questions. In addition, at times, I had to re-formulate the participant's comments so that a yes/no reply was sufficient to communicate the person's thoughts. This is contrary to a pure qualitative methodology and may have limited the participants' ability to fully express their experience and influenced some of the findings. Nonetheless, I believe that my familiarity with these people and the way they speak contributed considerably to the ability to do this research.

In light of the small sample size and the qualitative nature of this study, the findings cannot be generalized to the ALS population as a whole. This is consistent with a qualitative methodology that aims at understanding a phenomenon rather than generalizing new knowledge. Nevertheless, many of this study's findings on factors that affect decision-making about assisted ventilation replicate findings from a previous study done by Young et al. (1994) suggesting that the findings may be transferable to other similar settings.

6.6 Contribution to knowledge and areas for future research

This study reveals various factors about decision-making and assisted ventilation that are pertinent to individuals who suffer from ALS and their families. The themes that emerged confirmed that when making decisions about assisted ventilation, patients consider much more than just the scientific evidence that is available in the literature. Relationships, functional status, emotions, and other factors greatly influence decision-making. This study highlighted specifically the importance of communication and how, in many cases, ability to communicate is the most important factor in deciding whether one wants to use assisted ventilation.

This study identifies that people with ALS live with the constant fear of death and anticipate respiratory distress. In some cases, these fears are overwhelming and contribute to the definition of the self. As such, in the context of ALS and assisted ventilation, fears impact decisions about assisted ventilation without necessarily leading to irrational and non-autonomous decisions.

The study also highlights the importance of societal influences, such as, availability of adapted equipment, accessible housing, and home care services on decision-making about assisted ventilation. These considerations were significant to the participants' deliberations and, even though they are not typically included in liberal definitions of autonomy, in the context of ALS, societal influences contribute to autonomous decisions.

To better take these factors into account, healthcare professionals should adopt a pluralistic conception of autonomy in the context of ALS and assisted ventilation. A pluralistic conception of autonomy acknowledges that autonomous decisions do not result solely from rational processes and are influenced by one's context, emotions, personal relations, and societal relations. This finding clarifies the meaning that should be attributed to the word "autonomy" in the AAN's guidelines that suggest giving high priority to patient autonomy in the therapeutic relationship.

This study also highlights the need for a shared decision-making model to maximize the patient's potential to exercise autonomy from a pluralistic perspective. This model requires ongoing communication focusing on the factors that impact decisions and encourages participation of patients, families, and other health care professionals in the various stages of decision-making as discussed by the participants of this study.

In an effort to integrate these findings into clinical practice, I suggest the implementation of a respiratory protocol that could be communicated and shared across the various service providers, such as, the MNH ALS clinic, MCI home ventilation program, and CLSC home services. In addition, use of advanced directives is recommended to encourage an individualized approach to decision-making and assisted ventilation that respects patients' wishes and preferences. Several topics need to be better explored during deliberations for decision-making about assisted ventilation, including death and discontinuation of ventilation. These recommendations apply strictly to the patients followed at the MNH ALS clinic and are provisional pending further assessment of their effectiveness in improving the care and meeting the needs of patients.

This study highlights the need for future research concerning the numerous factors that may affect decision-making of individuals with ALS about assisted ventilation. Future research is needed to explore whether these factors are consistently being addressed by the health care professionals involved with the MNH patients during decision-making about assisted ventilation contributing to informed and autonomous treatment decisions. In addition, more research is needed to determine if the themes identified here are representative of all the people with ALS in the province of Quebec or if they are more representative of patients followed by a multidisciplinary clinic, such as the one at the MNH. This would allow subsequent studies to determine whether use of a multidisciplinary team improves exploration of the factors that are pertinent to decision-making and assisted ventilation.

This study is an initial examination of the perspective of individuals with ALS who consider the use of assisted ventilation. In addition to patient context, QOL, and personal relations, which have already been addressed in the ALS literature in the context of assisted ventilation, this study highlights the importance of communication, fears, and societal relations. Only by thorough exploration of all these factors can professionals caring for individuals with ALS hope to facilitate their decision-making process and provide care that promotes truly autonomous decisions.

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APPENDIX A

RECRUITMENT FORM

The Amyotrophic Lateral Sclerosis (ALS) clinic at the Montreal Neurological Hospital is looking for individuals who would be interested to participate in a research project to look at how people decide which treatment to try when they have difficulties with their breathing. This study involves a one hour interview in your home. If you are interested in finding out more about the project please write down your name and telephone number on this form and return it to the secretary. Josée Lemoignan will call you with further information.

La clinique de sclérose latérale amyotrophique de l'Hôpital Neurologique de Montréal recherche des personnes qui seraient interessées à participer à un projet de recherche pour explorer comment les patients atteints de SLA décident quel moyen utiliser quand ils ont de la difficulté à respirer. Cette étude implique une entrevue d'environ une heure qui aura lieu chez vous. Si vous êtes interessé à en savoir plus sur l'étude, s'il vous plaît, inscrivez vos noms et numéro de téléphone sur cette feuille et ramenez-la à la secrétaire. Josée Lemoignan vous contactera sous peu.

Name/Nom : ____________________________ Tel. : _________________________

APPENDIX B

CONSENT FORM

Montreal Neurological Hospital

HOW DO INDIVIDUALS WITH AMYOTROPHIC LATERAL SCLEROSIS MAKE DECISIONS ABOUT ASSISTED VENTILATION?

Reason for the study: The goal of this study is to understand how people with ALS who develop increased difficulty breathing decide which treatment (if any) they choose to try. It is hoped that by understanding your experience as a person with ALS who has breathing difficulty and how you feel about the treatments that are used to help, the Montreal Neurological Hospital ALS team and other teams that care for people with ALS will be better able to help other patients who are in a similar situation.

Description of the study: I (researcher Josée Lemoignan) will interview you in your home, asking questions about your experience of breathing difficulties and how you decided about which treatment to use. The interview should last about one hour. I will record our discussion with a tape recorder so that I do not forget what you say and so that I can report your experience in your own words as precisely as possible. After I have interviewed 10 people, I will combine everyone's answers and look for what is similar and different in what people with ALS told me. Once I have a summary of these similar and different points of view, I will mail or e-mail it to you to get your feedback on whether my summary of what everyone said reflects what you told me during my interview with you.

As part of the study, I will also ask you some questions such as your age, marital status, the date of the diagnosis, the type of ALS you have, the approximate date that your breathing became more difficult, and details about your choices of intervention to manage your breathing difficulties.

Once finished, I will present the results of the study to the MNH ALS team and I will publish an article in a journal to share the knowledge with other treating teams. The study results will also be part of my Master's thesis on *Informed Consent for Assisted Ventilation in ALS*

<u>Confidentiality:</u> I will use a code to distinguish each interview. The document to link the code to you, the interview tape and the consent form which will have your name on them will be kept locked in a filling cabinet at the hospital for five years and only I will have access to it. After that, they will be destroyed. I will not identify you by name in any report or publication of this research. If I want to quote from you to justify or explain a general topic and there is a risk that someone might recognize you because of what is said in the quote, I will change details to hide your identity or I will ask for your specific permission before using the quote. The use of e-mail may jeopardize confidentiality. Therefore, it will be possible to use the postal system to send you the summary of the themes that arise from the interviews should you prefer this.

<u>Advantages and disadvantages of the study:</u> This study offers you no specific advantage and does not compensate you financially for your time. However, I hope that you will benefit from thinking about your experience and from contributing to increasing the knowledge of the professionals caring for people with ALS who will then be better able to assist other patients who have to decide about treatment for breathing difficulties. The only potential disadvantage of the study is that someone might recognize you from what is being quoted. However, if there is such a risk, your permission to use your quotes will be requested.

Withdrawal from the study: If you choose to participate in this research and later change your mind, you can stop participating at any time. You may also skip any questions that you do not want to answer. Doing so will not affect the care you are entitled to receive at the MNH.

Contact information: Should you have any questions regarding your rights as a research subject in this project, you should contact the MNH Patient's Committee (a group established to protect the rights of patients and research subjects, Room 354, tel. 514/398-5358)._ The subject should be made aware that the Research Ethics Board or Quality Assurance Officers duly authorized by it may access study data.

If you have any questions about the research or a research-related problem, you may contact Josée Lemoignan at (514) 398-4640.

RESEARCH PARTICIPATION CONSENT

I, ___________________________ , have read this consent form. The present study has been explained to me by Josée Lemoignan and my questions have been answered to my satisfaction. I understand the procedures, advantages and disadvantages of the study. I freely and voluntarily agree to participate in this study.

______________________________	________________________	______
Name of research participant	Signature	Date
______________________________	________________________	______
Name of researcher	Signature	Date

APPENDIX C

INTERVIEW GUIDE

- I know that you have given some thought to whether or not you want to use a BIPAP or a ventilator if breathing becomes too difficult. How do you experience the decline in your respiratory functions? Or: What is it like for you to have/experience some reduction in your respiratory functions (in your ability to breathe)?

Probes-If patient is using a BIPAP- How does it feel to use the BIPAP?

- Tell me about all the options that were presented to you by the team to help with your breathing?
- Tell me what you know about the BIPAP?
- Tell me what you know about the ventilator?
- How do you feel about people using a BIPAP or a ventilator?
- How is using a ventilator different from using a BIPAP?
- What is the meaning of these interventions for you, in your life?
- Tell me about your thoughts when you considered the decision to use mechanical ventilation?

Probes-Whom did you discuss this with?

When did you make the decision?

How did you make the decision?

- Tell me about the factors that influenced your decision?

Probes-What do you think of the QOL of people who use a BIPAP? Who use a ventilator?

What do you think of the caregiver's QOL?

What are the financial implications?

- What are the values or beliefs that influenced your choice?
- Why did you decide to use or not the interventions proposed to you?
- What do you think are the consequences of your decision about your breathing?
- How was the topic of mechanical ventilation discussed with you by the ALS team?

Probes- When did this happen?

What was said?

- How would you have liked your treating team to address this topic?

Probes- Whom would you want to discuss this with?

When would you like this discussion to take place?

What information would you like?

- Is there anything else you would like to tell me?