

**Do Physicians of Amyotrophic Lateral Sclerosis (ALS) Patients Understand
Their Patients' Wishes Concerning End-of-Life Care?**

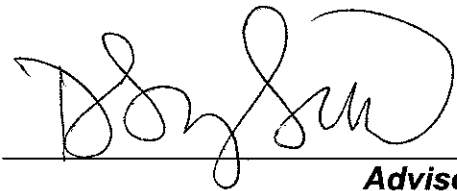
By

Aysha Akhtar, MD

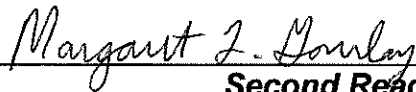
A Master's Paper submitted to the faculty of
The University of North Carolina at Chapel Hill
In partial fulfillment of the requirements for
The degree of Master of Public Health in
The Public Health Leadership Program.

Chapel Hill

2004



Advisor



Second Reader

6/4/04

Date

ABSTRACT

Background of Study Design

Physician-assisted suicide (PAS) and euthanasia have repeatedly emerged as issues of debate regarding end-of-life treatment for patients with intractable pain, terminal illnesses and /or other debilitating diseases. Amyotrophic lateral sclerosis (ALS) is a non-curable, debilitating neurological disease that causes progressive paralysis and eventual respiratory failure. Patients with ALS are in unique positions to contemplate end-of-life care issues due to the predictability of the course of the disease. While some have expressed an interest in pursuing PAS and/or euthanasia in order to control the circumstances of their deaths, it is not known how many have done so. In addition, little information is known about how much physicians of ALS patients understand their patients' interest in these options.

Methods

Physicians of ALS patients will be identified in neurology clinics throughout North Carolina. Mailed questionnaires will be sent to physicians over a 4-month period. They will be asked about their understanding of their ALS patients' wishes concerning end-of-life care including PAS and euthanasia. ALS patients, identified through these physicians, will be asked to complete questionnaires

pertaining to their wishes regarding end-of life care. These questionnaires will be completed through semi-structured interviews.

Results

To be completed

Conclusion

It is predicted that physicians of ALS patients will underestimate their patients' interest in PAS and euthanasia.

Research Question: Do physicians of Amyotrophic Lateral Sclerosis (ALS) patients know what their patients' wishes are concerning end-of-life care, particularly physician-assisted suicide (PAS) and euthanasia?

BACKGROUND

Natural History of ALS

Amyotrophic Lateral Sclerosis (ALS) is a debilitating and fatal neurological disease that affects approximately 1-2 per 100,000 individuals in the United States. It tends to affect males more than females at a rate of 2:1 and onset is usually after age 50. While the cause is unknown, approximately 5% of cases are familial (autosomal dominant), the remainder being sporadic. Many of the familial cases have been linked to the super-oxide dismutase gene on chromosome 21.

The disease is characterized by degeneration of upper and lower motor neurons. Initial manifestations in most cases include weakness of the hand muscles followed by progressive weakness of the remainder of the limbs, dysarthria (slurred speech) and dysphagia (difficulty swallowing). Cramping and fasciculations (twitches) of the muscles, particularly the forearm, upper arm, shoulder girdle and tongue appear along with muscle atrophy. Head

droop, a distinguishing feature of ALS occurs due to weakness of the thoracic and cervical muscles. Initially, loss of reflexes may occur (due to lower motor neuron disease), but is eventually replaced by hyperreflexia and spasticity as upper motor neurons become more affected.

As the disease advances, patients lose weight rapidly due to disuse of muscles and difficulty swallowing and thus inadequate caloric intake. All patients eventually become wheel chair bound. Activities of daily living become difficult and patients must rely on assistance from others. Pseudobulbar palsy, a term used to define inappropriate emotional outbursts due to disease of upper motor neurons, is common. Drooling becomes uncontrollable and aspiration of liquids, food and saliva pose a frequent threat. Paradoxically, bowel and bladder function are preserved. Sensory function and mental capacity are also preserved. Eventually, respiratory failure ensues and is the predominant cause of death unless ventilatory support is provided.

Time until death is, on average, three years after symptoms first appear. There is no cure and treatment consists of supportive care. Depression, hopelessness, anxiety, feelings of isolation and frequent sensations of choking and difficulty breathing typify the terminal phase.¹ Quality of life is greatly reduced.

End-of-Life Care for ALS Patients

Due to the predictability of the course of the disease, patients with ALS and their families find themselves in a unique position to contemplate issues concerning end-of-life care. Since the patients' intellects remain intact, they have the opportunity to make informed decisions regarding their care. Issues such as whether or not to have ventilatory support once respiration becomes impaired, to insert feeding tubes to maintain adequate nutrition and to receive palliative care for pain are all relevant. Not only are issues concerning the *maintenance* of life frequently addressed, but also issues concerning the *termination* of life. Specifically, ALS patients may find themselves exploring various methods of dying.

Controversies Surrounding Physician-Assisted Suicide and Euthanasia

When surveyed, many ALS patients have expressed interest in assisted suicide and/or euthanasia as possible options.² Physician-assisted suicide (PAS) is defined as assistance to a patient's termination of life by a physician, usually by prescribing lethal doses of narcotics that the patient then self-administers. Euthanasia refers to the active administration of lethal medications directly by the physician in cases where the patient is unable to do so herself due to paralysis or inability to swallow. Today, medical advances are allowing prolongation of lives that would normally have succumbed to many diseases. Because artificially induced life prolongation is increasingly more available through parenteral nutrition, ventilatory support

and cardiac support, questions pertaining to withdrawal of artificial support become relevant.

To many, PAS and euthanasia are natural extensions to the discussions of withdrawal of care. Indeed, ethicists argue that PAS/euthanasia are, in essence, the same as withdrawal of care since the end goal of all three methods is the same, namely to facilitate death, regardless of the means.³ They further argue that to allow withdrawal of care, but to deny PAS and euthanasia to those who also wish to facilitate their deaths is a form of discrimination. In other words, those patients whose lives are not dependent on artificial support are the “unlucky” ones who are denied the option of terminating their lives painlessly even though they may suffer equally or more than those living with life support. Even more unfortunate, according to this argument, are those incapable of self-administering lethal doses of painkillers due to paralysis, for example, and thus must rely on euthanasia to end their suffering in a desirable manner, a practice that is considered even more controversial by many than PAS.

Opponents of PAS and euthanasia fear that if these practices are legalized, then the potential for abuse of these practices becomes much higher. One such “slippery slope” argument is based on the concern that PAS and euthanasia will be used too readily before other options have been exhausted.⁴ For example, depression in patients might not be recognized or

adequately treated before termination of life is considered. Patients suffering from unbearable pain might opt for ending their lives without having had appropriate pain management. Alternatively, there is a fear of discrimination against the poor or members of minority groups. Opponents ask if these groups will be coerced into PAS or euthanasia as alternatives to more expensive treatments. ⁵ Thus, they ask, will end-of-life decisions be tainted by subtle or not so subtle economic coercion? Not only could the medical system be tainted, but also so could the patients' families and the patients themselves. Patients might be influenced by family members or by their own concerns of being a "burden" to others and as a result, too readily opt for ending their lives. Opponents fear that there are too many "slippery slopes" that could arise if PAS and euthanasia are allowed. They argue that we cannot adequately ensure that these practices would be used appropriately and without abuse, and thus should not be legalized.

In the United States today, while withdrawal of life-support at the patient's request is legal and recognized as a Constitutional right nationwide (a liberty to autonomy protected by the Fourteenth Amendment), PAS is only legal in the state of Oregon and only under certain conditions. Euthanasia is illegal throughout the US. This is not the case in many parts of the world, however. In the Netherlands, for example, both PAS and euthanasia are accepted practices for alleviating suffering in the terminally ill.

Although PAS and euthanasia are currently illegal in most of the US, both the legalities and ethics of these practices continue to be hotly debated. Ethicists, legal and medical experts, patient advocacy groups and politicians are bringing these issues to the forefront of public discourse. The ethical dilemmas concerning PAS and euthanasia will not magically disappear until we have adequately addressed all facets of these dilemmas. As we attempt to evaluate and clarify these issues, it becomes vital that any further decisions made be based on appropriate and comprehensive information. It is important that we understand all viewpoints on these topics.

Understanding the Patient's Wishes

Perhaps most importantly, it is vital that we understand the wishes of the patients themselves. Cancer patients, those with chronic pain and/or debilitating disease and the terminally ill are potentially the most affected by any further decisions that either allow or forbid these practices. We need to understand what they want. Do they wish to have the options of PAS and euthanasia? If so, are we failing as medical practitioners by not providing a much-desired service? Are we being remiss in our obligations to appropriately treat suffering?

ALS patients have a unique role in this debate due to a combination of several factors. Their disease is presently non-curable and terminal, their quality of lives are often markedly reduced and their mental capacities remain

preserved, allowing them to continue to make informed decisions regarding their health care through the final phases of their disease. ALS patients have expressed many common concerns and fears about the dying process itself.⁶ Fears about choking to death or dying painfully through respiratory failure are frequently cited. In addition, since most patients are paralyzed by the terminal phase, the issue of euthanasia becomes particularly relevant since they are unable to voluntarily terminate their lives on their own. Thus, their views on these issues and our understanding of their views are central to this debate.

This proposed research study will further our understanding of the views of ALS patients concerning these issues. Furthermore, it will explore whether or not the physicians of ALS patients understand their patients views. The hypothesis of the proposed study is that physicians of ALS patients are underestimating their patients' desires for euthanasia or PAS. The ultimate goal of this study is to determine if the US medical community (with the exception of Oregon) is appropriately addressing end-of-life concerns and if we are failing to provide services that are commonly desired by ALS patients.

The following section summarizes prior research that has attempted to understand the views of ALS patients regarding these issues. The strengths and weaknesses of these studies are explored. In addition, gaps in the

literature where more research is needed is discussed as well as further rational for this proposed study.

REVIEW OF MEDICAL LITERATURE

A Medline electronic search was conducted of studies published between 1970 and February 2004 using the search terms “amyotrophic lateral sclerosis AND assisted suicide”, “amyotrophic lateral sclerosis AND euthanasia”, “ALS AND euthanasia” and “ALS AND assisted suicide”. Limits included English language and human subjects. From the 130 total articles displayed, only those that were either original research studies or reviews of studies were chosen. Additional searches included the key words “amyotrophic lateral sclerosis AND end of life” or “ALS AND end of life”. Related articles on Medline were reviewed for the following two articles from the earlier searches:

Silverstein MD, Stocking CB, Antel JP, Beckwith J, Roos RP, Siegler M. Amyotrophic lateral sclerosis and life sustaining therapy: patient’s desires for information, participation in decision making, and life-sustaining therapy. *Mayo Clin Proc.* 1991;66(9): 906-13.

Schneiderman LJ, Kaplan RM, Rosenberg E, Teetzel H. Do physicians’ own preferences for life-sustaining treatment influence their perceptions of

patients' preferences? A second look. *Camb Q Healthc Ethics*. 1997; 6(2): 131-7.

All articles identified by the search were screened by the author. Only articles which were either available on-line or were available in the UNC Health Sciences Library were selected. A total of nine articles were assessed as relevant and were included in this review.

STUDIES OF GENERAL EXPERIENCE WITH PAS/EUTHANASIA

After PAS became legalized in Oregon, Ganzini et al, surveyed 2649 physicians eligible to prescribe lethal medications in Oregon.⁷ Physicians were asked for information on all terminally ill patients who had requested lethal medication for assisted-suicide. The physicians covered a wide range of disciplines, including neurology, internal medicine, family practice, and gynecology.

Between the years 1997 and 1999, 165 patients had requested assistance with suicide. The majority of these patients were male (52%), Caucasian (97%), had completed high school (95%) and had medical insurance (98%). The average age was 65. Most patients (93%) were deemed mentally competent by their physicians and most suffered from cancer (67%). According to the physicians, the most common reasons for requesting PAS were loss of independence (57%), poor quality of life (55%), pain (43%) and desire to

control circumstances of dying (53%). Reasons such as perceived financial burden on others or lack of social support were given for only 11% and 6% respectively.

In total, 18% of patients actually received prescriptions for PAS and 10% ultimately died via PAS. There were many reasons for not honoring the patients' requests including the presence of depression.

Sullivan et al, studied PAS in the second year after it became legal in Oregon.⁸ Through interviews with physicians and family members of decedents of PAS and through death certificates, they found 33 patients who had requested prescriptions for lethal medications. Out of these 33, 26 actually died via PAS. The median age of these patients was 71. Cancer was the most frequent diagnosis (63%) followed by ALS (15%). As with the previous study, most patients had at least high school education (92%), all had some form of insurance and most were male and Caucasian (96% and 59% respectively). According to the physicians and family members interviewed, the predominant reasons for wanting PAS were concerns of loss of autonomy, inability to participate in joyful activities, physical suffering and loss of control of bodily activities. One patient was concerned about medical costs and eight were concerned about being burdens on others.

Limitations to both of these studies include using surrogates rather than interviewing the patients directly. Since these interviews were conducted after the patients committed PAS, there was also potential for recall bias. However, since PAS is mandated as reportable in Oregon, all cases of PAS could be analyzed.

The results of both studies suggest that even though legal in Oregon, PAS is requested by a small minority of patients. Of those who do request it, the majority are terminally-ill Caucasians with at least high school education and with some form of medical insurance. Among their top reasons for requesting PAS are loss of independence and desire for control over their lives. Perhaps most importantly, lack of financial or social support was rarely cited as reasons for their requests.

STUDIES ASSESSING ALS PATIENTS DIRECTLY

Rabkin et al, surveyed a total of 56 ALS patients on their willingness to consider assisted suicide and on other quality of life issues.⁹ The patients in this study were consecutively selected from ALS patients being seen at the Lou Gehrig Muscular Dystrophy Association (MDA)/ALS Center of the New York Presbyterian Hospital. Semi-structured interviews were conducted. The patients also completed the Beck Depression Inventory and other quality-of-life surveys. Among the ALS patients, 34% stated that they would consider, under certain circumstances, asking for a prescription of medicine for the

purpose of ending their lives. Fifty percent said they would not and 8% were undecided. The authors found that those patients who were willing to consider assisted suicide did not differ significantly in degree of suffering, distress or depression from those who would not consider such an action. Euthanasia was not addressed in this study.

There are several limitations to this study. First, the patients were selected from a tertiary and specialty clinic and thus may have had more severe symptoms of disease or were more knowledgeable of their disease. This selection bias may have led to an overestimate of the frequency of willingness to consider PAS. Second, the majority of the patients (80%) were Caucasian and thus not necessarily representative of all ALS patients. Third, the small number of subjects limits the strength of this study. Finally, no information was given regarding characteristics of patients who refused to participate in the study. This information is necessary to evaluate for potential selection bias.

There were several strengths of this study that includes the use of validated instruments for surveys, such as the Beck Depression Inventory (BDI). The authors also included spirometry measures of FVC to evaluate degree of impairment in addition to using levels of function questionnaires. Also the patients in this study, other than being predominately white, represented a

wide range of characteristics with regards to education level, religious affiliation, and sex and employment status.

Ganzini et al, surveyed both ALS patients and their caregivers in Washington and Oregon.¹⁰ Subjects were selected from patients in the ALS clinic at Oregon Health Sciences University in Portland, Oregon, or had either participated in or expressed prior interest in participating in research studies. In total, 100 patients and 91 family caregivers agreed to participate. Fifty-six percent of patients surveyed were willing to consider assisted-suicide (via prescriptions to end their lives). These patients were more likely to be men, were less religious, more educated and had lower quality of life scores than those who would not consider assisted death. In addition, even though there were no significant differences in depression scores between these two groups, the former group did collectively have higher scores of hopelessness. Euthanasia was not addressed.

Among the caregivers, most of whom were spouses, 62% would support the patient's decisions to end their lives via PAS. There was agreement on this decision between caregivers and patients 73% of the time. The caregivers who supported PAS were less religious than those who did not. Overall, the authors found no significant differences between patients with an interest in PAS and those without in education level, extent of social support, degree of

disability or presence or absence of suffering, depression or perceptions of being a burden to the caregivers.

Strengths of this study include direct responses by ALS patients rather than by surrogates. Selection criteria were adequately described. Also, standardized instruments for evaluating depression, social support and hopelessness were used. Limitations of this study include a selection bias since all patients were selected from a tertiary clinic or had shown or expressed interest in research participation. It is possible that these patients, in general, are more educated about their disease and may be more willing to discuss or consider assisted death. No information was provided on the characteristics of subjects who refused to participate which may influence the outcome. The majority of patients were highly educated Caucasian males, which limits the generalizability of these results. Finally, the authors used education level as a marker for socioeconomic status, which may not be accurate.

These studies support many of the results from the studies in the previous section. Again, it appears that the majority of patients who would consider PAS are Caucasian males with less religious affiliation and are better educated, on average, than those patients who would not consider PAS. Also, depression and perception of being burdens to others were not significantly

associated with interest in PAS. Hopelessness, however, was positively correlated with interest in PAS.

STUDIES ASSESING VIEWS AND OF ALS PATIENTS VIA SURROGATES

In the Netherlands, PAS and euthanasia, while not legal, are not punishable. Physicians of 279 patients with ALS who had died between 1994 and 1998 participated in a study by completing questionnaires about the end of life decisions of their patients.¹¹ Two hundred and three physicians participated; the remainder who refused (26%) predominately cited lack of time as their reason for refusal. The patients were identified through referrals to two national ALS centers.

According to the responding physicians, 17% of the patients died by euthanasia and 3 % by PAS. The patients who died by euthanasia differed from those who died by PAS in the level of functioning of their arms. Patients who had a physician-assisted death (either by suicide or euthanasia) did not differ significantly from those who died by other means in education level, income, and disease-related or care-related characteristics. There were also no significant differences in the severity of pain, despair or fear. Interestingly, physician-assisted death was negatively associated with feelings of anxiety before death. Reasons for the decreased anxiety in these patients were not pursued.

There are obvious limitations to this study. Perhaps the most significant is the fact that physicians, rather than the patients themselves, completed the surveys. There is great potential for erroneous responses, particularly to those questions evaluating patients' emotions. Thus, information bias, as well as recall bias, could be a factor. A second limitation, as the authors discuss, is the potential for selection bias since the patients were identified through university clinics. Also, this study may only pertain to Dutch patients for whom income status may not play a large role in their decisions on dying since almost all Dutch are insured. In addition, greater public acceptance of physician-assisted death in the Netherlands may mean that these results cannot be generalized to patients in the US.

Ganzini and colleagues conducted a second study in this area.¹² In this study, 50 family caregivers of decedent ALS patients in Oregon were surveyed. Selection criteria included prior enrollment of patients in either the Portland Veterans Affairs Medical Clinic or the Oregon Health Sciences University muscular clinic or participation in the previously discussed study.¹⁰ Caregivers were asked a number of questions pertaining to the last month of the patients' lives.

According to the caregivers, 32% of patients had discussed wanting PAS in the last month. Only one patient, however, actually died via PAS. Patients who had expressed interest in PAS were more likely to have insomnia, to feel

as a burden on their families, and to have more discomfort or pain than those who did not explicitly express a desire for PAS. Of the caregivers who had participated in the previous study, almost half of the patients who had expressed an interest in PAS early on maintained that desire, according to the caregivers.

This study was a follow-up of the prior study by Ganzini et al.¹⁰ Thus, the authors were able to ascertain how many of the patients who had previously expressed an interest in PAS actually died by that method. Another strength is the use of validated survey instruments.

However, since this study relied on caregiver reporting, the accuracy of their answers is unknown. There is also the potential of recall bias. Selection bias may play a role due to the same reasons discussed in the previous study.

There is no information on the characteristics of those who did not participate in the study and the small sample size limits the strength of this study.

Finally, a lack of overt expression of desire for PAS does not imply that the patients did not privately wish for this. There may be many reasons for not discussing this desire with caregivers, including fear of disagreement, of alienation or of distressing their caregivers.

Unlike the previous studies discussed thus far, the Netherlands study revealed no significant difference in education level between those patients who died

via PAS and those who did not.¹¹ In addition, the study by Ganzini and colleagues does suggest that perceptions of being burdens on caregivers is associated with desire for PAS.¹² Both of these results contradict the results of the prior studies in regards to these factors. It is unclear why this is so, however, the discrepancies could be due to differences in culture (between the US and the Netherlands) and in using surrogates to obtain answers.

STUDIES ASSESING PHYSICIANS' KNOWLEDGE OF PATIENTS' WISHES

In this literature search, no studies directly comparing ALS patients' preferences on end-of-life care and their physicians' understanding of these preferences were found. Studies involving other patients are discussed below.

Schneiderman et al, identified 36 patients who had advanced directives with life-threatening illnesses from specialty clinics in San Diego.¹³ Physicians of 22 of these patients participated in the study while 8 refused due to lack of knowledge of their patients. Thus in all, 22 patients and their corresponding physicians (16 in all) were interviewed about end-of-life decisions. Patients were asked to give their preferences on end-of-life care for 4 different scenarios and their physicians were asked to predict what their patients' responses would be. Overall, there was very poor correlation between patients' wishes and their physicians' predictions. For example, while 93% of

the patients wished for CPR in cases of cardiac arrest, only 61% of their physicians accurately predicted this. In all, physicians' predictions more closely matched what they would prefer for their own care rather than what their patients truly desired.

Limitations to this study include the very small sample size. Also, since all patients had advanced directives, it is possible that discussions between physicians and their patients may have led to greater understanding of patients' wishes than would occur with the general patient population. Despite this, a significant portion of physicians initially approached (50%) refused to participate in this study; citing insufficient knowledge of their patients as their reason for refusal. Thus, there is a clear bias for patient and physician selection, which may have resulted in data skewed toward positive correlation between patient and physician responses. This study appears to support a lack of physician understanding of patients' wishes.

A second study surveyed 28 physicians of 35 patients with advanced AIDS or cancer.¹⁴ Thus there were a total of 35 patient-physician pairs. Again, physicians were asked to predict their patients' choices on various end-of-life scenarios. In all, there was 63% perfect agreement between patient wishes and their physicians' predictions. In general, the correlation was highest in situations involving very invasive treatments such as CPR and mechanical ventilation and lowest in situations involving less invasive treatments such as

use of antibiotics or pain medication. As with the previous study, physicians' predictions more closely matched their preferences for their own care rather than that for their patients.

Limitations include a small sample size and selection of patients from a tertiary clinic (and thus limited generalizability). Also, no detailed information regarding the patients' and physicians' demographics was included. Thus vital information such as years of practice of physicians and race, sex and socioeconomic status of patients were not provided, all of which could influence the results.

Coppola et al, conducted a similar study as above involving 41 physicians (24 primary care and 17 hospital-based) and 82 elderly outpatients.¹⁵ As with the previous studies, their goal was to assess the accuracy of physicians' predictions of patients' preferences for treatments in several different life-threatening scenarios with and without advance directives. These physicians were allowed to review advance directives on approximately half of the patients before completing the questionnaire. Consistent with other similar studies, physicians did not, as a group, accurately predict their patients' preferences. Overall, predictions were accurate 66% of the time for primary care physicians and 64% of the time for hospital-based physicians. The accuracy of physicians' predictions with their patients' advance directives improved by 20% overall to a total of 70%.

Limitations in this study include a small sample size and limited demographic range of both physicians and patients. For example, physicians and patients were predominantly Caucasian and either Protestant or Catholic. Subjects were also selected from a limited area in Ohio. A selection bias may have been present if those who chose to participate in this study were already interested in the subjects of end-of-life care and advance directives. Thus accuracy of predictions may be higher in this study than would be with the general patient and physician population. Unfortunately, the authors did not provide information on the number of potential subjects who refused to participate or their reasons for refusals.

From these above studies, it appears that physicians as a whole have minimal understanding of their patients' preferences regarding end-of-life care and treatments. However, the number of these studies is small. Also, none of these studies specifically assessed ALS patients or views on euthanasia and/or PAS. This is an area in obvious need of investigation.

METHODS

Study Population

The data gathered for this proposed study will be obtained primarily through cross-sectional surveys. Both ALS patients and physicians of ALS patients will be surveyed. To meet the criteria for eligibility in the study, physicians must be licensed neurologists practicing in the state of NC and actively

caring for at least one ALS patient. Criteria for subject eligibility for ALS patients will include the following:

- Patients must be adults with either documented, confirmed or suspected diagnosis of ALS
- Must be currently receiving care from a neurologist in North Carolina
- Must have had documented ALS for at least 1 year
- Must be aware of their diagnosis
- Must be mentally competent
- Must have no other major untreated mental or physical co-morbid disorders
- Must be documented citizens of the US
- Must be able to give either verbal or written consent
- Must live in North Carolina
- Must speak English
- Must have a home telephone

Neurologists throughout the state will be contacted via phone for verbal consent to participate in surveys. ALS patients will be identified through the participating neurologists.

Initially, the patient survey will be piloted by 3-5 ALS patients who meet the above criteria and who will be identified in the waiting rooms of the Duke and UNC neurology clinics. Permission will be obtained for participation in 30- 45-minute semi-structured interviews. Those who consent will be

presented with a sample survey. Their answers to the survey questions will be sought. In addition, they will be asked for any recommendations regarding the format and content of the survey. The goal would be to include questions that the patients themselves deem relevant and omit those that they don't. After these initial interviews, the survey will be further refined and finalized. The patients who participate in this initial series of interviews will not be eligible for participation in the final survey.

After the surveys are finalized, eligible candidates will be phoned and asked to consent to participate in the study and appointments will be made for survey completion through personal interviews. These interviews will be conducted either at the patients' homes or in health clinics, at their discretion.

Thus 2 different questionnaires will be developed, one for the physicians and the other for patients, both based on results of the interviews with patients. Since this is a pilot study, the goal will be to have surveys completed by at least 15 physicians and up to 15 patients. As previously stated, patients will be surveyed in person for mainly two reasons; 1) to allow participation of patients with physical limitations that may prevent their completing the questionnaire and 2) to eliminate any confusion or misconceptions regarding the questions. Physician subjects, however, will be surveyed through mailed questionnaires, the rationale being that they have more limited time for participation and thus written questionnaires sent through the mail will lead

to greater participation. In addition, physical limitations of the physicians will be of minimal concern.

Prior to initiation of this study, approval will be obtained by the Human Subjects Committee of the University of North Carolina, Chapel Hill School of Public Health.

SURVEYS

The initial draft survey presented for evaluation by ALS subjects will contain various questions pertaining to end-of-life issues. Patients will be asked for their opinions on various aspects of end-of-life care including the options of physician-assisted suicide and euthanasia. They will be adapted from other surveys asking similar questions. The goal is to develop a survey that closely matches those of other studies so that comparisons of results can be more easily made between this and other studies.

General questions asking patient and physician demographics, such as race, religion and income will be included to evaluate for any trends based on these characteristics. Race, marital status and education level have been shown to differ between those who would consider PAS and those who would not in several studies.^{8,10} However, other studies have revealed no significant differences.^{7,11} This study will measure these same characteristics in order to assess if these characteristics do influence patient decisions. In addition,

similar characteristics of physician subjects will be assessed to evaluate for influencing factors on physicians' assumptions of patient preferences.

Religiosity is negatively associated with interest in PAS/euthanasia in two of the studies reviewed.^{10, 11} The author of this study will measure religiosity by a question similar to that used in one reviewed study.¹⁰

Since studies assessing depression, pain and discomfort as possible indicators of interest in PAS or euthanasia have given mixed results, the author of this study will further explore these issues by asking questions based on those used in these studies.^{10, 11, 12} Questions concerning present and future treatments have been developed for this study in order to assess for consistency in patients' treatment desires. The purpose of these questions is to gauge the overall attitudes of patients to various end-of-life issues. Their responses will be compared to those by the physicians in order to assess the degree of agreement between what the patients' attitudes are to these treatments and what the physicians believe their patients' attitudes are. The influence of advanced directives on physician understanding of their patients' preferences has only been evaluated by one study and thus will be further evaluated in this study using similar questions.¹⁵ In addition, questions concerning physician-patient interaction have been developed for this study in order to assess influencing factors on physician understanding of patients.

All questions will be closed-ended and answers will either be yes/no or in a Likert scale format. The Likert scale is traditionally a 5-point ordinal scale in which subjects are asked to express degree of agreement or disagreement on a 5-point scale. Each degree of agreement is given a numerical value from 1-5. This is used predominately to measure attitudes and perceptions and is commonly used for surveys such as those in this study. For the purpose of this study, a 5-point scale will be used. For example, to the question “how likely would you consider having a tracheotomy placed if your breathing became labored?” answer options would be 5): very likely, 4): somewhat likely, 3): neutral, 2): somewhat unlikely or 1): very unlikely . In addition, patients will be asked to complete a mini-depression scale for evaluation for potential depression, which could affect their end-of-life care decisions.

The physician surveys will contain the exact questions as in the patient surveys, except that the questions will be directed for the physicians to contemplate how they believe their ALS patients would answer these questions. For example, the above question would be written as “How likely do you think your ALS patient would consider having a tracheotomy placed if his/her breathing became labored?” Answer options would again be 5): very likely, 4): somewhat likely, 3): neutral, 2): somewhat unlikely or 1): very unlikely. Depression scales, however, will not be included in these surveys.

Questionnaires

Patient Questionnaire-Sample Questions

Category	No. of Items	Sample Questions
Patient characteristics	5-8	What is your marital status?
Patient religious affiliation	1	Are you affiliated with any religious organization?
Depression scale	5	Have you had feelings of worthlessness in the past 2 weeks?
Pain/discomfort scale	5	Are you experiencing pain? How frequently? How severe?
Advanced directives	2	Do you have any advance directive?
Present treatment	5	Do you currently have or have had in the past a tracheotomy? A feeding tube?
Future treatments	10	How likely would you consider having the following treatments if they were deemed necessary for life?
PAS/euthanasia	5	If it were legal, how likely would you consider having help by a physician to end your life either now or in the future?
Physician/patient Interaction	5	Has your primary physician discussed end-of-life care issues with you?
Wrap-up	1-2	After having completed this survey, do you think any of your prior views/wishes may have changed?

Physician Questionnaire- Sample Questions

Category	No. of Items	Sample Questions
Physician characteristics	5	How long have you been practicing?
Physician religious affiliation	2	Are you affiliated with any religious organization?
Patient types	5-10	What are the majority of diseases that you treat?
ALS patients	5-10	How many ALS patients do you treat? What are their major disabilities, if any?
ALS patients and depression	5-10	Are any of your ALS patients clinically depressed? Are any being treated for depression?
ALS patients and pain	5	How much pain or discomfort do you think the majority of your ALS patients are in?
Advanced directives	1-2	What proportion of your ALS patients has advanced directives?
Present treatments	5	What proportion of your ALS patients has or have had tracheotomies?
Future treatments	5-10	How likely do you think your ALS patients would consider having the following treatments if they were deemed necessary for life?

Physician/patient interaction	5-10	Do you discuss end-of-life issues or advanced directives with any of your patients? If so, how often?
Physician's views on PAS/euthanasia	5	How likely do you think that PAS/euthanasia might be appropriate in certain cases?
Physician's views on practicing PAS/euthanasia	5	If legalized, how likely do you think you would offer these services for your patients?
Physician's understandings of ALS patients' wishes regarding PAS/euthanasia	5-10	How likely do you think your ALS patients would consider PAS? Euthanasia?
Wrap-up	1-2	After having completed this survey, do you think any of your prior views may have changed?

Statistical Analysis

Descriptive statistics will be tabulated for the study participants. Chi-square tests will be used to determine the significance of differences between patients and physician responses for all categorical variables. Differences in continuous variables will be calculated using the Mann-Whitney U Test. All tests will be two-sided and a p value of .05 or less will be considered statistically significant.

RESULTS

Table- Patient Characteristics

Age Range (mean)	
Female number (%) Male number (%)	
Race –Caucasian no. (%) African-American no. (%) Hispanic no. (%) Other no. (%)	
Number of years since ALS diagnosed range (mean)	
Are you married? Yes (%) No (%)	
Do you have any children? Yes (%) No (%)	
Do you have any religious affiliation? Yes (%) No (%) If yes, which type? -Protestant -Catholic -Judaism -Islam -other	
Education level <HS no. (mean) =HS no. (mean) >HS no. (mean)	
Advanced medical directives? Yes? no. (mean) No? no. (mean)	
Pain Score range (mean)	
Depression score range (mean)	
Insurance? Yes no. (%) No no. (%)	

Overall Quality of life score range (mean)	
---	--

Table- Physician Characteristics

Age range (mean)	
Sex -Female no. (%) -Male no. (%)	
Race Caucasian no. (%) African-American no. (%) Hispanic no (%) Asian no. (%) Other no. (%)	
Religious affiliation? Yes no. (%) No no. (%)	
Practice type University-affiliated no. (%) Private clinic no. (%) Practice setting Rural no. (%) Urban no. (%)	

Patient Preferences (in blue ink) and Physicians' Assumptions (in red ink)

Type of Treatment	Very likely no. (%)	Somewhat likely no. (%)	Not at all likely no. (%)	Unsure no. (%)
Ventilation				
Nutrition/hydration				
Resuscitation				
Pain medication				
Would consider suicide				
Would consider suicide with physician help				
Would consider euthanasia by physician				

DISCUSSION

Based on previous studies, it is likely that the results of this study would indicate a significant gap between the wishes of ALS patients concerning end-of-life care and the assumptions made by their physicians. It is also likely that ALS patients would be more interested in pursuing PAS and/or euthanasia as options than their physicians would predict. There are many possible explanations for this. First, as revealed in the literature search earlier, physicians assumptions about their patients' desires might be influenced more by their own wishes rather than what their patients would want. Second, physicians, on average, spend too little time discussing such issues for many reasons including already-busy schedules, their own discomfort in broaching such topics with their patients and the belief that their patients would be offended or upset by these discussions.¹⁵ Third, physicians may perceive offering PAS and euthanasia as failures on their part to combat their patients' disease or symptoms.¹⁷ Fourth, fear of discussing these polemic topics under current legal constraints may impede understanding of their patients' wishes.¹⁸

If physicians are underestimating their patients' interest in PAS and/or euthanasia, then ALS patients might be denied a desired service as long as PAS and euthanasia remain illegal. As previously stated, ALS patients have the potential to suffer immensely physically and emotionally. If a large portion of this population does indeed wish for the options for PAS and

euthanasia as a means to control the circumstances of their deaths, then the medical community needs to further explore all the issues concerning such practices.

If the main hypothesis of this study is supported, then these results would be consistent with those of similar studies reviewed regarding several factors. For example, it is likely that the majority of patients who express interest in PAS or euthanasia will be Caucasian, educated, males with little or no religious affiliation. As previous studies revealed, interest in PAS or euthanasia does not imply that these acts will actually be committed. Reasons for this include unexpected death and improvement in treatments for depression and/or pain. Also, it is possible that patients who received lethal prescriptions found comfort in knowing that assisted death was an option even if never used. It is important to note, however, that this study will not simply reiterate the results of previous studies, but will go a step further by assessing how much physicians understand their patients.

Of course, questions regarding the ethical and practical issues of legalizing PAS/euthanasia will remain after this study. The concerns of abuse potential would need to be further addressed. However, we may find that rather than leading to abuse of patients, legalized PAS and euthanasia causes patients to receive better palliative care with improved treatment of pain and depression than is currently occurring. This may be due to increased awareness among

physicians of palliative treatments if they must comply with procedures mandating that all these alternatives be exhausted before PAS and/or euthanasia can be enacted.³

Limitations to this study will include a small and selective sample size. Both physician and patient subjects would be selected only from North Carolina and thus are not necessarily representative of both general populations. There may also be a selection bias. Both subject populations may be comprised of those who already have an interest in the subjects of PAS/euthanasia and end-of-life care. They may be more partial to legalizing PAS and euthanasia than the general populations. On the other hand, even with prior interest in these topics, both physicians and patients interviewed may tend to underreport any interest in legalization. There are many possible reasons for this. Physicians may fear being “labeled” and may have concerns that, despite the anonymity of the study, their responses may be revealed to colleagues. Patients may be reluctant to express true feelings for fear of upsetting family members, especially if family members are present during interviews. Both physicians and patients may be reluctant to openly consider treatments that are currently illegal in most of the US.

One of the major strengths of this study is that information is directly obtained from the ALS patients themselves, rather than via surrogates. This leads to less recall bias. In addition, this study will directly compare patients’

wishes and physicians' assumptions. While some of this has been done, as reviewed earlier, none of these studies involved ALS patients specifically. The results of this pilot study can be used to guide further and much needed research in this area.

REFERENCES

1. Ganzini L, Johnston WS, Silveira MJ. The final month of life with ALS. *Neurology*. 2002; 59: 428-431
2. Carver AC, Vickrey BG, Bernat JL, Keran C, Ringel SP, Foley KM. End-of-life care: A survey of US neurologists' attitudes, behavior, and knowledge. *Neurology*. 1999; 53: 284-293
3. Steinbock B, Arras JD, London AJ. *Ethical Issues in Modern Medicine*. "The philosophers' brief". New York, NY: McGraw-Hill Companies, Inc; 2003, pages 386-394
4. Steinbock B, Arras JD, London AJ. *Ethical Issues in Modern Medicine*. "Assisted suicide: The philosophers' brief". New York, NY : McGraw-Hill Companies, Inc; 2003, pages 382-385
5. Steinbock B, Arras JD, London AJ. *Ethical Issues in Modern Medicine*. "Physician-assisted suicide: A tragic choice". New York, NY: McGraw-Hill Companies, Inc; 2003. pages 394-400
6. Silverstein MD, Stocking CB, Antel JP. Amyotrophic lateral sclerosis and life-sustaining therapy: Patients' desires for information, participation in decision making, and life-sustaining therapy. *Mayo Clin Proc*. 1991; 66: 906-913
7. Ganzini L, Nelson HD, Schmidt TA, Kraemer DF, Delorit MA, Lee MA. Physicians' experiences with the Oregon death with dignity act. *NEJM*. 2000; 342: 557-563
8. Sullivan AD, Hedberg K, Fleming DW. Legalized physician-assisted suicide in Oregon- the second year. *NEJM*. 2000; 342: 598-604
9. Rabkin JG, Wagner GJ, Del Bene M. Resilience and distress among amyotrophic lateral sclerosis patients and caregivers. *Psychosomatic Medicine*. 2000; 62: 271-279
10. Ganzini L, Johnston WS, McFarland BH, Tolle SW, Lee MA. Attitudes of patients with amyotrophic lateral sclerosis and their caregivers toward assisted suicide. *NEJM*. 1998; 339: 967-73
11. Veldink JH, Wokke JHJ, Van Der Wal G, Vianney de Jong JMB, Van Den Berg LH. Euthanasia and physician-assisted suicide among patients with amyotrophic lateral sclerosis in the Netherlands. *NEJM*. 2002; 346: 1638-44
12. Ganzini L, Silveira MJ, Johnston WS. Predictors and correlates of interest in assisted suicide in the final month of life among ALS patients in Oregon and Washington. *J Pain Symptom Management*. 2002; 24: 312-317.
13. Schneiderman LJ, Kaplan RM, Pearlman RA, Teetzel H. Do physicians' own preferences for life-sustaining treatment influence their perceptions of patients' preferences? *J Clin Ethics*. 1993; spring: 28-32
14. Schneiderman LJ, Kaplan RM, Rosenberg E, Teetzel H. . Do physicians' own preferences for life-sustaining treatment influence their perceptions of patients' preferences? A second look. *Cambridge Quarterly Healthcare Ethics*. 1997; 6: 131-137

15. Coppola KM, Ditto PH, Danks JH, Smucker WD. Accuracy of primary care and hospital-based physicians' predictions of elderly outpatients' treatment preferences with and without advance directives. *Arch Int Med.* 2001; 161: 431-440
16. Layson RT, Adelman HM, Wallach PM, Pfeifer MP, Johnston S, McNutt RA. Discussions about the use of life-sustaining treatments: A literature review of physicians' and patients' attitudes and practices. *J Clin Ethics.* 1994; 5: 195-203
17. Emanuel EJ, Fairclough D, Clarridge BC, Blum D, Bruera E, Penley WC, Schnipper LE, Mayer RJ. Attitudes and practices of US oncologists regarding euthanasia and physician-assisted suicide. *Ann Intern Med.* 2000; 133: 527-532
18. Back AL, Starks H, Hsu C, Gordon JR, Bharucha A, Pearlman RA. Clinician-patient interactions about requests for physician-assisted suicide. *Arch Intern Med.* 2002; 162: 1257-1265.