## Appendix C

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Ethics, Communication and Decision Making  

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INTRODUCTION

The ethical precepts pertinent to end-of-life decisions and management have been elaborated and disseminated widely, yet remain poorly understood by practitioners, as well as those affected by ALS (Carver et al., 2000). Decisions cannot be made in a vacuum, yet knowledge and understanding are imperfectly conveyed to patients and their caregivers. Health care providers accede to a lack of discussion with family and patients. A dance of denial can continue until faced with an issue of emergency care, often an urgent need for ventilation, among other life threatening situations. Based on the ALS CARE Database, 90% of those with ALS had their final wishes respected. However, this may only be representative of those clinicians (and patients) sufficiently committed to participate in a voluntary process (Bradley et al., 2000). The tools that clinicians could use to develop and practice good communication skills exist and could be applied in the field of ALS care to lay the groundwork for effective decision making and improved end-of-life management. Current guidelines provide a framework that, if applied systematically, could have profound impact on care. Improvement will come with the establishment of expectations of care, evaluation of the efficacy of the existing guidelines and the development of new tools to measure outcomes as well as those that can assist in the care itself.

Objectives

The over-riding objective of the project is to improve end-of-life care in ALS. Restated, the objective of improving care is to achieve a good death. As defined by the Institutes of Medicine, “[a] decent or good death is one that is free from avoidable distress and suffering for patients, families and caregivers, in general accord with patient and family wishes and is reasonably consistent with clinical, cultural and ethical standards.”

Methods

The ethical background to the decision-making process and the essential communication required to set the stage for good end-of-life care were reviewed. Literature reviews in the areas of ethics, communication and decision making were performed and pertinent citations, were extracted. The combined experience of the reviewers, and their interpretation and distillation of the literature from related fields, was used to select examples of communication strategies, decision making and to iterate the ethical basis where ethical issues arise.

WORKGROUP FINDINGS

Ethical Issues in End-of-Life Care in ALS

Ethical issues have emerged at the forefront of ALS-specific end-of-life care discussions in public and professional publications. The ethics of:

- disclosure of diagnosis and prognosis,
• decision making regarding institution and withdrawal of life-sustaining therapies, and
• physician-assisted suicide and euthanasia
are all pertinent to ALS care, and have been the focus of recent study. The latter two topics are immediately relevant to the mandate of the Promoting Excellence ALS Peer Workgroup.

Physician-assisted suicide (PAS) and ALS have been of particular prominence, with two editorials in the New England Journal of Medicine specifically addressing this issue in the last five years (Ganzini and Block, 2002; Rowland, 1998). The thrust of public discussion focusing on the rights of individuals with ALS to request assisted suicide and euthanasia is a major incentive to counter with initiatives to provide optimal care at the end of life. It is worth noting that even in the Dutch experience where both PAS and euthanasia have been condoned in practice, 80% of individuals with ALS did not choose either PAS or euthanasia – as reported in Veldinck (between 1994 and 1999) (Veldinck et al., 2002).

**Identification of Existing Resources**
A number of recent reviews summarize the prevailing North American ethical standards with respect to end-of-life decision making in general, with reference to ALS in particular (see Bernat, Section E). This area is not without debate (for example, see Loyal 2000) and controversy. Individual institutions have their own policies (whether health care facility or religious groups, for example) and for most mainstream organizations, the policies are consistent with the prevailing ethical standards. The legal status of Advanced Directives, patient autonomy in refusing or withdrawing life-sustaining therapies and proxy decision making regarding end-of-life care is also the subject of recent review. Information on a state-by-state basis is available on the Internet. Physician-assisted suicide is legal in only one state – Oregon – and euthanasia is illegal in the U.S.

**Defining the Gaps**
The major gaps are between the ethical theory and actual clinical practice (Carver et al., 2000). Missing from available studies are any which investigate the perception of the individuals making the decisions as to the validity of the ethical precepts in their particular circumstance. For example, patient seven in Bolsmjo (2001), although ventilator dependent, perceives having the ventilator withdrawn as euthanasia, as do physicians in his experience. Lacking are specific strategies for applying ethical standards in practice for patients and clinicians dealing with ALS that could be specifically endorsed by the applicable institutions and organizations.

**RECOMMENDATIONS TO THE FIELD**

**Practice Recommendations**
For individual patients the ethical ramifications of their decisions when opting for or against life-sustaining therapies must be identified and explored even as the need for the decisions is being broached. Triggers for end-of-life discussions and some of the ethical considerations each provokes are included in Table 1. Clinicians need to identify the policies and practices of their institutions, and of the major religious groups in their area, and be prepared to consult
with and refer patients to the appropriate contacts within these groups. Frequent interactions with an ethics committee will assist the ALS clinician.

Research Recommendations
Studies to better understand the barriers to applying the accepted ethical standards to clinical practice, and to test strategies to improve practice are needed. Better understanding of the perceptions of those making the decisions and the influence of these perceptions on decision making are needed. The factors that influence desire for assisted suicide and euthanasia, and the interventions that might reduce the perceived need for these acts, must be investigated.

Policy Recommendations
Health care institutions, whether hospitals, nursing homes or other, must have explicit policies to support end-of-life decision making. Together with institutional ethics boards and teams, consistency in application of the standards should be reviewed regularly by health care facilities. Medical organizations should supply avenues for individuals not associated with institutions a means to access ethical consultation and supportive ethical and legal counseling.
Communication Issues Affecting End-of-Life Care in ALS

Communication is the necessary process by which knowledge is shared. Both the manner and content of communication can affect the perceptions and responses of the communication partners. At key times as the disease progresses, there are responsibilities on the part of the health care providers to address issues that have a high impact on the terminal stages of ALS. Inadequately addressing the issues in a timely fashion can lead to misperceptions, default decision making and could increase suffering for all those affected by the illness. A major goal of communication between clinicians and those affected by ALS is the acquisition of insight into the illness both to achieve the best coping strategies and to ensure the best end-of-life planning for the individual with ALS. Insight, when applied to illness, means the patient’s awareness and personal significance of their illness and the need for treatment, which in turn is influenced by cultural, psychological and neurological factors (David, 1992).

The time to set up the communication strategy is at diagnosis. Establishing rapport with the patient and care partners cannot wait until critical issues arise. Appropriate and empowered decision making can only take place in the setting of good communication. Informed consent is the standard for implementing medical care; how and when the patient is informed strongly influences the decisions. Accurate information regarding the implications of the diagnosis must be made available, but introduced and discussed in a manner that fits with the needs and values of those affected. Ongoing conversations identify the psychosocial, spiritual and socioeconomic context in which an individual is working through the process and will enable appropriate decision making consistent with their values, and allow congruence of the care with these values.

WORKGROUP FINDINGS

Barriers to Communication:
- Clinicians’ lack of familiarity and subsequent avoidance of end-of-life discussions.
- Patient naïveté and avoidance of difficult topics. Denial of illness and/or its implications by patient, family and clinicians.
- Perceptions that any discussion of end-of-life care will be too time consuming, and therefore no discussion takes place.
- Perceptions that there is not adequate reimbursement related to the time communications will take.
- Fear of harm – discussion of the end of life is perceived as destroying hope. “What I don’t know won’t hurt me” is a prevailing attitude.
- Physical barriers – clinics and hospitals that are not set up for sit-down, relaxed and private discussions.
Appendix C  Ethics, Communication and Decision Making

- Prevailing medical attitudes conveyed in medical education – medicine is for curing diseases; the enemy is death – whereas the enemy is actually suffering.

- Separation of the medical and spiritual domains; medical education pays little heed to integrating psychosocial and spiritual inquiries with medical diagnostic queries.

- Physical state of the patient (particularly dysarthric patient).

Denial as a barrier to communication and decision making has been the subject of discussion and investigation in a number of medical and psychiatric conditions but not specifically ALS. Denial as a coping strategy can be positive, but at the extreme has been found to be a barrier to best management for prevention and treatment of diseases. Hope has been the subject of accounts of the experience of coping with ALS and hopelessness has been associated with, amongst other negative outcomes, higher interest in assisted suicide (Ganzini et al., 1998) including at the end of life (Ganzini et al., in press). Identifying coping strategies (and their correlates) that are positive as has been done in other chronic illnesses, as well as further investigation into the attributes of adaptation to ALS would enhance understanding of best approaches to end-of-life care. Approaching denial without destroying hope is one of the major challenges in effectively communicating about end-of-life choices in ALS.

Identification of Existing Resources
Strategies to improve communication skills of clinicians in sharing bad news include the CLASS and SPIKES approaches advocated by Buckman (Sections A and B). The EPEC program (referenced in Report Summary) is a workshop to improve teaching of end-of-life issues, although not specific to ALS. Sections D and F apply the basic principles to ALS and to neurological disorders specifically.

Defining the Gaps
- The major gap is that which lies between existing recommendations and best practice from the literature and actual practice.
- Studies specific to ALS regarding the impact and efficacy of the published communication strategies are lacking.
- There is a lack of research studies validating approaches to communication and decision making.
- There are no regulatory means to impart the responsibility of the appropriate communication strategies, and no measure of their impact in place.

RECOMMENDATIONS TO THE FIELD

Practice Recommendations
1. Practice parameters, derived from both evidence-based reviews and expert opinion, have been established for the treatment of ALS and include recommendations for communication and end-of-life decision making (Miller et al., Oliver, Borasio and Walsh). At different stages of ALS, key decision points are reached that require well-developed communication strategies to impart the choices available in an impartial
manner. The ethical basis of these choices must be understood by both the practitioner and the ALS patient and family.

2. Imparting the diagnosis together with its implications rarely can be accomplished in one clinical encounter, and the discussion is never completed in one visit. A dialogue should be initiated as soon as the evaluating physician views ALS as a potential diagnosis. Early discussions do not have to be specific, nor should they go beyond the expertise of the practitioner. However, orienting the patient and family to the possibility of a serious or disabling illness allows them to prepare to receive the more devastating details on a subsequent visit after confirmatory testing is complete. Definitive diagnostic disclosure should be planned to take place in keeping with the current standard of care (Buckman 1996, Doyle 1996, Ptacek 1996, Borasio, 1997). The content of the discussion will vary with the needs of the individuals. The use of blanket statements (e.g. death in 2 to 3 years) should be avoided. Tailoring the discussion to answer immediate concerns and summarizing with both immediate and long-term goals at the end of each visit will provide continuity and a sense of purpose that can counter feelings of hopelessness and anxiety.

End-of-life discussions are difficult due to lack of clinical role models or mentors for communication strategies, inadequate teaching and general avoidance of what is perceived as a problem area. Tools to improve communication exist (Buckman, EPEC) that have been validated in other diseases and may be modified to specific ALS situations. Misperceptions of the impact of bad news, especially information regarding life-threatening aspects of disease, lead health care providers to withhold or not initiate discussions. Misconceptions and misinformation conveyed in the early stage of the disease can hamper constructive approaches later on.

3. The process of communication includes identifying communication partners (patient, caregivers, also other clinicians, social agencies, volunteer organizations). It is an exchange, a dialogue, not a monologue. Clinicians, for example, impart information, provide experience and reassurance; patients and families share values and provide the context in which the issues facing the team will be proceeding. Family and social values have been shown to be the important determinants in advanced care planning, more than medical information.

Effective sharing of information is the responsibility of all practitioners, including the first practitioner to investigate or contemplate the diagnosis. It is important to establish a partnership with families, take a leadership role in end-of-life discussions, and communicate the decisions made by the patients and family to other physicians and other health care professionals who provide care for patients.

4. Disease Progression and Decision Points: The timing of discussion of the tools for functional adaptation, life-sustaining therapies (LSTs) and terminal process can be driven by the patient, but will also be determined by “physical imperatives” as the need to prevent injury, malnutrition or suffering arises. Identifying the parameters that are
driving the clinical adaptation (or not) include the evaluation of aspects of the patient and family’s outlook that fall outside of areas that we traditionally include in the clinical realm, particularly spirituality.

Key decision points are identified in Tables 1 and 2 and are prompted by patient and family concerns and/or the stage of the disease. All should be anticipated and identified as impending. The information must be imparted completely and impartially, difficult in some practice settings and for some practitioners. Identifying sources and resources other than the practitioner can help.

5. Communication Surrounding End-of-Life Issues: Specifically discussing the process of dying, dealing with myths and reassurance regarding the control of symptoms is essential.

Deal with requests, overt or covert, for assisted suicide, as an opportunity to provide as much detail as requested and additional reassurance about the relief of suffering at the end of life. Reinforcing the value of seeking a good death may obviate the need to refuse such requests. There is no obligation on the part of physicians to perform assisted suicide. We must explicitly discuss that assisted suicide must be distinguished from pain management and sedation at the end of life and also from the process of withdrawing medical interventions such as assisted ventilation.

Avoid using the term “hastening death;” use “assisted suicide.”

Specifically list the medical interventions that can be refused and the medical acts that relieve symptoms that are not suicide.

Research Recommendations

1. Develop overall disease assessment tool(s) for the patient with ALS that helps the physician identify specific areas of need for education and communication on managing their illness (e.g., spirituality, psychological, social). Investigate how physicians will know what and how they should be communicating to the patient.

2. Use available clinical practice data (retrospective) to get data on communication between the patient and the physician.

3. Develop prospective studies to determine the optimal way to communicate the diagnosis, illness progression, etc., with the ALS patient. Conduct studies to test the consensus-based recommendations in this report. One strategy would be to identify decision points during end-of-life care and investigate the impact of standardizing treatment decisions around these decision points.
**Policy Recommendations**

Improve physician education programming (medical school, residency, board certification) on the need for optimal communication on palliative care (for all illness, including ALS).

1. RRC (Residency Review Committee) & ACGME (Accreditation Council of Graduate Medical Education) require that “The resident must receive instruction in appropriate and compassionate methods of end-of-life palliative care, including pain relief and psychosocial support and counseling for patients and family members about these issues.” However, an examination of the end-of-life care content among fifty major medical textbooks, including the leading neurology texts, demonstrated that the neurology textbooks ranked in the lowest quartile of end-of-life care coverage (Rabow, 2000).

2. Support for general palliative care training for patient/nurse/physician is essential for all illnesses addressing end-of-life care. Recognize good communication and education as part of an optimal treatment strategy for ALS.

3. Include palliative care in hospital accreditation (from the nurse practitioners through the level of the physician care).

**End-of-Life Decision Making in ALS**

Both the Ethics and the Communication sections cover many of the issues relevant to end-of-life decision making. Ideally, the necessary preparation and discussion regarding end-of-life decisions would come well in advance of the period of the illness that constitutes the actual end of life. Failure to address wishes regarding ventilation, in particular, may result in emergency intubation in an acutely dyspenic patient or a failure to recognize respiratory failure that results in unplanned death. Table 1 outlines opportunities to initiate end-of-life discussions to establish the care path to be taken.

**WORKGROUP FINDINGS**

Factors that influence choice of life-sustaining therapies in ALS have been studied and include the role of religion and spiritual practices (Silverstein, 1991; McDonald, 1994). Presentation of the choices for life-sustaining therapies, and the potential for their implementation without specific prior discussion and consent, will reflect the values and practices of the clinician as much as the patient and family. The framing heuristic refers to the observation that the manner in which risk is presented influences the choice of action (Tversky and Kahneman, 1981). The issue of framing has been studied in subjects with no illness (ibid) as well as those facing decisions about medical interventions in, for example, cardiac diseases (Gurm, 2000). It has been studied in physicians’ decision making (McGettigan et al., 1999). It is clear from these studies that framing has a major impact on risk-taking decisions as well as choices regarding treatment. Framing has yet to be specifically studied in decision making in ALS.
Barriers to Decision Making:

- Denial,
- Knowledge deficits,
- Cognitive deficits or deficits of higher cortical function,
- Language and other communication barriers, and
- Mismatch between decision-maker’s perceptions and those of the care providers; expectation mismatch between individual and significant others.

Identification of Existing Resources

The use of advance directives represents an opportunity to discuss specifics of care and choices regarding life-sustaining therapies. A number of Web sites offer access to standardized Advance Directives that include state specific references. A recently published disease specific Advanced Directive is particularly useful (Benditt et al., 2001). The ALS Association and the Muscular Dystrophy Association have resources and information about life sustaining therapies including PEG, NIPPV and invasive ventilation.

Gaps

While there are studies about the use of Advance Directives in acutely ill hospitalized patients and in nursing homes, and there is a high utilization of Advance Directives in the ALS CARE database (Bradley, 2000), there are no studies on the specific utility of Advance Directives in determining the quality of end-of-life care in ALS. The preferences of patients and the public compared with actual utilization of life-sustaining measures (Alpert and Emanuel, 1998) suggests that this should be assessed in the ALS population and their families. The impact of the decisions on family remains to be evaluated, although the impact of ALS itself has been the subject of a retrospective survey (Turnbull and Martin, 2001).

The process of decision making and the influences of factors intrinsic to the patient as well as environmental and framing effects, are areas of major importance for development of prospective studies.

RECOMMENDATIONS TO THE FIELD

Practice Recommendations
Use the existing resources to standardize an approach to end-of-life decisions in ALS. Direct initiatives to the patient. Promote a patient bill of rights. Patients need to have access to what the standard of care is. This equates to education of patients and physicians so the two are in agreement and expectations are met. Courses at the American Academy of Neurology and other neurology meetings will be very helpful across the board. Translate practice parameter guidelines for patients and make guidelines available to patients.

Research Recommendations
2. Conduct follow-up studies on the impact of end-of-life decisions on the family.
3. Investigate the prevalence of the desire to end life and determine what factors or interventions should be enacted to obviate this desire.

Policy Recommendations
Institute end-of-life planning/Advance Directives as an institutional goal required for accreditation of hospitals and nursing homes. Assess adherence to the Advance Directives by the health care facilities as part of quality assurance required for re-accreditation.
Table 1: Talking points for “Triggers” for End-of Life Discussions:
(The talking points listed in each area are relevant to all discussion, but each trigger might bring forward immediate ethical issues, and highlight the communication and decision-making priorities.)

<table>
<thead>
<tr>
<th>Trigger</th>
<th>Ethics</th>
<th>Communication points</th>
<th>Decision-making points</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. Patient/family query</td>
<td>Requests to withhold information from patient; Cross-cultural differences in discussing death &amp; dying; Requests for assisted suicide, euthanasia; Patient autonomy of decisions about life-sustaining therapies (both use of and withdrawal of)</td>
<td>Identify the reason for the queries; Ascertain understanding of the nature of death, symptoms and strategies; Dispel myths about death in ALS</td>
<td>Introduce specific palliative care planning, identify maintenance of quality of life as major goal; review Advance Directives specifically for end-of-life scenarios pertinent to ALS at different stages</td>
</tr>
<tr>
<td>2. Distress or suffering</td>
<td>Appropriateness of inquiries regarding psychosocial and spiritual components of distress; Medication use (see pain)</td>
<td>Identify physical, psychosocial and spiritual components; Involve others as appropriate (pastoral care, etc.) and include in clinic correspondence</td>
<td>Formulate and implement short term plan, clearly identify lead clinician; Set timelines for progress, specify location (inpatient, outpatient, etc.) and level of interventions</td>
</tr>
<tr>
<td>3. Pain</td>
<td>Medication choice and dose directed at controlling symptom, not limited by possibility of hastening death</td>
<td>As for 2</td>
<td>As for 2</td>
</tr>
<tr>
<td>4. Dysphagia/PEG</td>
<td>Right to refuse nutrition and to stop PEG feedings; Evaluate cultural and religious practices for consistency with standard practices</td>
<td>Importance of concurrent respiratory assessment; Importance of nutrition for quality of life; Not all PEG patients are non par os (NPO); role of recreational eating</td>
<td>Set end-points for timing of PEG (e.g. % weight loss, duration of meals); If no decision re: PEG, discussion of risks of poor nutrition and aspiration</td>
</tr>
<tr>
<td>5. Dyspnea</td>
<td>Right to accept/refuse initiation of and to withdraw any type of respiratory support; Drug use for symptoms not to be limited by potential effects of respiratory drive</td>
<td>Explicit and detailed description of non-invasive and invasive ventilation, causes of death in ALS, process of dying with or without respiratory support; Need for communication strategies when speech lost; Drugs used for dyspnea, including end-points (symptom relief not sedation) both in absence of respiratory support and at its withdrawal</td>
<td>Set up guidelines for timing of respiratory interventions – “not wait and see;” Setting end-points (when to stop ventilation) based on explicit discussion of continual progression of ALS as respiratory support prevents death</td>
</tr>
<tr>
<td>6. 2 regions affected</td>
<td>Autonomy of decision making; Identifying denial, preserving hope</td>
<td>Progression to all regions; Clarify what is life-threatening progression; Review utility and implications of monitoring or respiratory and swallowing functions</td>
<td>Advance Directive review; Caregiver needs and limits; Timing of interventions</td>
</tr>
</tbody>
</table>
### Table 2: Communication Goals for Clinicians

<table>
<thead>
<tr>
<th>Stage</th>
<th>Ethical points</th>
<th>Communication points</th>
<th>Decision-making points</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pre-diagnosis</td>
<td>Addressing uncertainty</td>
<td>Orienting patient to potential of serious diagnosis</td>
<td>Referrals for symptom management without firm diagnosis</td>
</tr>
<tr>
<td>Diagnosis</td>
<td>When to introduce concepts related to the end of life; Cultural differences in discussing death and dying</td>
<td>Adopt Buckman’s SPIKEs protocol for initial and follow-up discussions</td>
<td>Introduce general concepts about decisions on life-sustaining interventions, need for Advance Directives and medical power of attorney</td>
</tr>
<tr>
<td>Follow-up visits</td>
<td>Dealing with denial without hampering hope</td>
<td>Explain the reasons for symptom monitoring including the rational for routine pulmonary function tests</td>
<td>More detailed discussions about decision making, including concrete examples; Reinforce completion of Advance Directives</td>
</tr>
</tbody>
</table>
REFERENCES


Gurm HS, Litaker DG. “Framing procedural risks to patients: Is 99% safe the same as a risk of 1 in 100?” *Acad Med, 75*: 840-842, 2000.

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Section A: CLASS: Communicating With Patients

Context

Show the patient that you are there to listen. The location should be quiet and private. Care provider should be seated. When the care provider and the patient are at the same level, the locus of control is somewhere between them and the patient feels he or she has the care provider's attention.

Listen

You don't have to sit with the patient endlessly. In most medical interviews, physicians interrupt their patients after they've been speaking about 19 seconds. If allowed to run on, 90% of the time patients finish speaking in less than two minutes.

Acknowledge

According to Robert Buckman, MD, "The secret of empathetic response is to state what is covertly obvious." Acknowledge and respect the reactions the patient expresses. Remember – it may not be a "minor" or "routine" event to the patient. Your acknowledgement is of the patient's perception/reaction to the information – not of your opinion about the seriousness of the situation.

Strategy

Encourage patient input into the treatment strategy or plan of care. This will increase the likelihood of adherence to the plan.

Summary:

Close the conversation by creating a clear verbal contract. It may be as basic as, "Ok, you'll do _______; I'll do _______ and we'll re-evaluate _________."

Section B: SPIKES: Breaking Bad News

Setting:

Setting should foster communication by providing for privacy and comfort. Body language should be culturally appropriate and may include facing the patient, eliminating distractions and making eye contact.

Perception:

Learn what the individual already knows so you can break the bad news in a way he or she will understand. Listen to the way the individual describes the situation to identify level of comprehension and degree of denial.

Invitation:

Ask the individual if he or she is willing to accept information and leave the door open for the individual to request information at a later time. He or she may not understand serious and complicated information unless prepared to receive them.

Knowledge:

Deliver bad news at the individual's level of understanding and using the same vocabulary he or she does. You can then begin educating by giving important information in small chunks and checking periodically for understanding.

Empathy:

Acknowledge and respect the reactions and feelings the individual expresses.

Summary:

Summarize the important points discussed, ask if there are any other questions or issues to discuss, and set the time and place for the next meeting.

Palliative care in neurology
An overview

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Palliative care in neurology is an evolving discipline of interest among neurologists who care for chronically ill, critically ill, and dying patients with neurological diseases. Increasing appreciation for the importance of palliative care for patients with neurological disorders is emphasized in the American Academy of Neurology (AAN) Ethics and Humanities Committee 1996 Position Statement: "It is imperative that neurologists understand and learn to apply the principles of palliative care as...many patients with neurological disease die after long illnesses during which the neurologist acts as the principle or consulting physician."[1] In an editorial accompanying this position statement, Bernat et al.[3] identified data on the physiology of death and of mature knowledgeable physicians with expertise in symptom control and palliative care as some of the critical barriers to the delivery of competent end-of-life care for patients with neurological disease.

The AAN Ethics and Humanities Committee has published a series of position statements to highlight the ethical responsibilities of neurologists to their dying patients. These position statements constitute a set of ethical guidelines for neurologists in the management of patients with advanced incurable disease (Display Box 1).

<table>
<thead>
<tr>
<th>AAN Position Statements Related to End-of-Life Care</th>
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What is Palliative Care and Palliative Medicine?

For purposes of discussion, adopted here is the World Health Organization (WHO) definition of palliative care. Numerous groups have developed variations of this definition and, recently, *Last Acts®* published a document entitled “Precepts of Palliative Care” (Display Box 2).

Five Principles of Palliative Care

1. Palliative care respects the goals, likes and choices of the dying person.
   - It respects a patient's needs and wants as well as those of the patient's family and other loved ones.
   - It finds out from the patient who he or she wants to help plan and give care.
   - It helps the patient understand his or her illness and what to expect in the future.
   - It helps the patient figure out what is important.
   - It tries to meet the patient's likes and dislikes: where to get health care, where to live, and the kinds of services wanted.
   - It helps the patient work together with his or her health care provider and health plan to solve problems.

2. Palliative care looks after the medical, emotional, social, and spiritual needs of the dying person.
   - It knows that dying is an important time for the patient and family.
   - It offers ways for the patient to be comfortable and ease pain and other physical discomfort.
   - It helps the patient and family make needed changes if the illness gets worse.
   - It makes sure the patient is not alone.
   - It understands there may be difficulties, fears and painful feelings.
   - It gives the patient the chance to say and do what matters most.
   - It helps the patient look back on life and make peace, even giving the patient a chance to grow.
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3. Palliative care supports the needs of the family members.
   It understands that families and loved ones need help.
   It offers support services to family caregivers, such as time off for rest, and advice and support by telephone.
   It knows that caregiving may put some family members at risk of getting sick themselves and plans for their special needs.
   It finds ways for family members to cope with the costs of caregiving, like loss of income, and other expenses. It helps family and loved ones as they grieve.

4. Palliative care helps gain access to needed health care providers and appropriate care settings.
   It uses many kinds of trained health care providers – doctors, nurses, pharmacists, clergy, social workers, and personal-care givers.
   It makes sure, if necessary, someone is in charge of seeing that the patient's needs are met.
   It helps the patient use hospitals, home care, hospice and other services, if needed.
   It tailors options to the needs of the patient and family.

5. Palliative care builds ways to provide excellent care at the end of life.
   It helps health care providers learn about the best ways to care for dying people. It gives them the education and support they need.
   It works to make sure there are good policies and laws in place.
   It seeks funding by private health insurers, health plans and government agencies.

Palliative care is the term used throughout this article to broadly define the approach to the patient with incurable disease, with both hospice and supportive care services included within this concept of palliative care.

As defined by the World Health Organization, palliative care is the active total care of patients whose disease is not responsive to curative treatment. Control of pain, of other symptoms, and of psychologic, social and spiritual problems is paramount. The goal of palliative care is the achievement of the best possible quality of life for patients and their families. Many aspects of palliative care are also applicable early in the course of the illness in conjunction with disease modifying treatment.

Palliative care includes the following:

- Affirms life and regards dying as a normal process;
- Neither hastens nor postpones death;
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- Provides relief from pain and other distressing symptoms;
- Integrates the psychological and spiritual aspects of patient care;
- Offers a support system to help patients live as actively as possible until death; and
- Offers a support system to help the family cope during the patient's illness and in their own bereavement. [14]

Palliative medicine is the formal term used to describe the medical specialty that is the study and management of patients with active, progressive, far-advanced disease for whom the prognosis is limited and the focus of care is quality of life. [10] At present, several countries including Britain, Sweden, Norway, Australia and Canada have adopted palliative medicine as a specialty and have designated University Chairs in Palliative Medicine.

Recent data suggest that the current U.S. health care system provides inadequate palliative care for the seriously ill and dying. This was perhaps best documented in the 1995 Study to Understand Prognoses and Preferences for Outcomes and Risk of Treatment (SUPPORT) study involving over 9,000 patients in five academic medical centers. [15] This Robert Wood Johnson Foundation supported study demonstrated serious inadequacies in the care of the dying. Fifty percent of dying patients experienced moderate to severe pain in the last three days of life; physician-patient communication was poor; few patients had Advance Directives, and if they did, the documents were often not followed; family needs were minimally addressed; and the cost of care was high. The SUPPORT study confirmed the abundant anecdotal data claiming inadequacy of palliative care in the United States.

In 1997, the Institute of Medicine (IOM) of the National Academy of Sciences published its commissioned report entitled "Approaching Death." [11] This IOM report noted serious inadequacies in the current provision of palliative care in the United States and included seven recommendations for care at the end of life (Display Box 3).

**Recommendations from the Institute of Medicine's "Approaching Death"**

1. People with advanced, potentially fatal illnesses, and those close to them should be able to expect and receive reliable, skillful and supportive care.

2. Physicians, nurses, social workers and other health care professionals must commit themselves to improving care for dying patients and using existing knowledge effectively to prevent and relieve pain and other symptoms.

3. Because many problems in health care stem from system problems, policy-makers, consumer groups, and purchasers of health care services should work with health care practitioners, organizations and researchers to:
   
   a. Strengthen methods for measuring the quality of life and other outcomes of care for dying patients and those close to them.

   b. Develop better tools and strategies for improving the quality of care and holding health care organizations accountable for care at the end of life.

   c. Revise mechanisms for financing health care so that physicians encourage rather than impede good end-of-life care, and sustain rather than frustrate coordinated systems of excellent care.
d. Reform drug prescription laws, burdensome regulations, and state medical board policies and practices that impede the effective use of opioids to relieve pain and suffering.

4. Educators and other health care professionals should initiate changes in undergraduate, graduate, and continuing education programs to ensure that practitioners have relevant attitudes, knowledge and skills to care well for dying patients.

5. Palliative care should become, if not a medical specialty, at least a defined area of expertise, education and research.

6. The nation's research establishment should define and implement priorities for strengthening the knowledge base for end-of-life care.

7. A continuing public discussion is essential to develop a better understanding of the modern experience of dying, the options available to patients and families, and the obligations of communities to those approaching death.

The report clearly identified profound gaps in scientific knowledge about care at the end of life that needed serious attention from biomedical, social science and health services researchers. It pointed out significant organizational, economic, legal and educational impediments to good care. Particularly germane to neurologists, the report indicated health care professionals' lack of education and knowledge about end-of-life care as one of the major barriers to providing care for patients and families at the end of life.

Neurologists care for many patients who die, because cerebrovascular disease is the third leading cause of death in the United States, following heart disease and cancer. Over 40% to 50% of patients with advanced cancer have neurologic complications, and pain is the most common presenting symptom requiring neurologic evaluation. The increasing prevalence of dementia in the aging population and changes in the trajectory of dying for patients with multiple sclerosis, amyotrophic lateral sclerosis (ALS), Huntington's disease, Parkinson's disease, stroke, and AIDS emphasize the growing need for neurologists to be competent in symptom control and end-of-life care.

Recent advances in the management of chronic pain and the development of reliable valid data assessment methods and evidence-based guidelines on the use of opioid, non-opioid, and analgesics in somatic and neuropathic pain states, provide the opportunity for neurologists to undertake sequential trials of analgesic treatments tailored to individual patients’ types of pain. In addition, advances in symptom control research provide treatment protocols for managing patients with nausea and vomiting, delirium, cachexia and anorexia, and dyspnea. Improved methods to assess mood disorders, such as anxiety and depression in the medically ill patient, coupled with clinical trials demonstrating antidepressant efficacy, now allow for therapeutic trials to reduce patients' psychological distress. Lastly, the use of Advance Directives and ethical guidelines on withholding and withdrawing care, the role of nutrition, hydration and sedation in the imminently dying are available to help neurologists provide high-quality symptom control to improve the quality of living for their dying patients.
**Epidemiology and Ethnography of Death**

In the year 2000 in the United States, approximately 2.5 million Americans died. This is slightly less than 1% of the population. The major causes of death have shifted during this century. Death from chronic illness now accounts for approximately 70% of all deaths annually and, as noted, heart disease, cancer, emphysema, stroke and dementia are the leading causes. The last century saw the life expectancy of American women rise to 79 and men to almost 75. Significant racial differences in life expectancies exist, with the average African-American woman expected to live 74 years and the average African-American man 65.4 years. In the next 10 years, this aging population will triple, placing the increasing burden on the current health care system that inadequately cares for a population with increasing disability. In 2003, one in five persons, or 20% of the population, will be over the age of 65 in the United States.

With this changing trajectory of death, the last century has witnessed the over-medicalization of death. The majority of Americans now die in hospitals (up to 70%), with approximately 15% dying in hospice care, and 10% to 15% dying in nursing homes. There is, however, regional variation in where patients die. For example, only 31% of deaths occur in hospitals in Oregon. In contrast, in New York, 65% of patients are hospitalized during the last days of life. The SUPPORT study demonstrated that it was the number of hospital beds in the region, not the autonomous choice of patients, that determined whether patients died in hospitals.

Significant health care resources are expended to care for the dying. Thirty percent of Medicare benefits is spent caring for patients in the last six months of life. Increasing concern that over treatment or inappropriate treatments are driving these escalating costs has led to a broad public discussion about the financing of end-of-life care.

Numerous studies have attempted to understand the barriers to providing appropriate, humane and compassionate care at the end of life. Institutional-, patient-, and physician-related barriers have been identified, ranging from a restrictive Medicare hospice benefit that prevents patients with neurologic disease from entering hospice care to lack of physician knowledge and skills in palliative care.

The current Medicare hospice benefit is a capitated reimbursement program that provides home care, prescription drugs, and an interdisciplinary care team to patients with a prognosis of six months to live or less. However, difficulty in predicting prognoses in patients with chronic neurologic disease has prevented dying patients with neurologic disease from fully accessing and using hospice care. Studies demonstrate that even for doctors who work regularly with dying patients, prognostic estimates are incorrect 50% of the time. Often, patients with dementia, ALS, and brain tumors do not appear discernibly different in the last few days before death than they did several weeks before. Because of the difficulty in identifying who is dying, there is increased advocacy for a change in the hospice benefit to a "Medicaring" benefit. This benefit would be based on a patient's level of function and severity of illness – not on their prognosis.

Cancer patients represented more than 60% of the 700,000 patients who received hospice care in the United States in the year 2000. In fact, only 15% to 20% of Medicare eligible patients access hospice care as they are dying. Focusing on the current barriers to access and the provision of hospice care for patients with neurologic disease is one of the policy issues that neurologists should be aware of in advocating for access of such care for their patients.
As identified in the IOM report, neurologists, like other medical specialists, lack training and knowledge in palliative care. In a survey of neurologists, funded by the American Academy of Neurology in 1996, a sizable gap was identified between established legal, medical and ethical guidelines for the care of seriously ill patients with neurologic disease and the beliefs and practices of many neurologists surveyed. For example, although most neurologists supported a patient's right to refuse life-sustaining treatment, many believed they were killing their patients by supporting such refusals. Thirty-seven percent of neurologists surveyed thought it was illegal to administer analgesics in doses that risk respiratory depression among terminal ALS patients, and 40% believed they should obtain legal counsel when considering stopping life-sustaining treatment. In addition, 44% thought they would participate in physician-assisted suicide and 28% in voluntary euthanasia if these practices were legalized. The lack of knowledge of the law, and confusion, disagreement, or both concerning medical and ethical guidelines suggests a need for education in palliative care and end-of-life decision making.

In a survey of practicing neurologists and residency program directors about pain medicine education, 89% of practicing neurologists stated that more pain medicine education is needed for resident training, and 91% agreed this education was needed for practicing neurologists. Only 5% of neurology residencies include a pain clinic rotation. Only 29% of residency program directors report having a neurology pain specialist on the faculty, and they rated the importance of a pain subspecialty seventh out of eight subspecialties; in this survey, practicing neurologists report that 25% of their new patients present with pain as a major complaint. Seventy-four percent of practicing neurologists feel adequately trained to diagnose headache and 67% to treat headache conditions.

A recent survey of medical textbooks rated general neurology texts as having minimal to no content on topics indexed as palliative care, symptom control, ethics and care of the dying. In contrast, family medicine and geriatrics covered these topics in detail.

Although the Residency Review Committee (RRC) for Neurology has clearly stated that residents should receive adequate training in "appropriate and compassionate methods of end-of-life palliative care, including adequate pain relief and psychosocial support such as counseling for patients and family members about these issues," such training is not included in most programs. To help United States neurology residency programs meet this accreditation standard, the Graduate Education Subcommittee of the AAN recently unanimously endorsed a proposal by Drs. David Weissman and Alan Carver to integrate end-of-life care into neurology residency training. During the 2001-2002 academic year, 30 neurology residency programs will send representatives to a seminar on palliative care training in neurology and learn how to enhance the teaching of delivery of end-of-life care in their respective programs and institutions.

In 1999, the AAN, along with 17 other subspecialty medical and surgical organizations, including the Joint Commission on Accreditation of Healthcare Organizations, agreed to a set of core principles and a consensus statement emphasizing the importance of training neurology residents and practicing neurologists in end-of-life care (Display Box 4).
Core Principles for End-of-Life Care

Clinical policy of care at the end of life and the professional practice it guides should:

1. Respect the dignity of both patient and caregivers.
2. Be sensitive to and respectful of the patient's and family's wishes.
3. Use the most appropriate measures that are consistent with patient choices.
4. Encompass alleviation of pain and other physical symptoms.
5. Assess and manage psychologic, social and spiritual/religious problems.
6. Offer continuity (the patient should be able to continue to be cared for, if so desired, by his/her primary care and specialist providers).
7. Provide access to any therapy that may realistically be expected to improve the patient's quality of life, including alternative or nontraditional treatments.
8. Provide access to palliative and hospice care.
10. Respect the physician's professional responsibility to discontinue some treatments when appropriate, with consideration for patient and family preferences.
11. Promote clinical and evidence-based research on providing care at the end of life.

The AAN has also expanded courses in palliative care, pain management and ethics at its annual meeting and has developed a clinical ethics rotation for its residents. There is a strong impetus to include palliative care curricula at the undergraduate level. Billings and Block have outlined the principles that should guide these undergraduate palliative care programs (Display Box 5).

Basic Principles for Enhancing Undergraduate Medical Education in Palliative Care

1. The care of the dying and their families is a core professional task of physicians. Medical schools have a responsibility to prepare students to provide skilled, compassionate end-of-life care.

2. The following important content areas related to end-of-life care must be appropriately addressed in undergraduate medical education:
   a. Communication
b. Pain and symptom management

c. Provision of home and hospice care

d. Patient wishes

e. Ethical issues

f. Interdisciplinary teamwork

g. Professionals' personal stress

h. Personal awareness of attitudes, feelings and expectations regarding death and loss

3. Medical education should encourage students to develop positive feelings about dying patients and their families and about the role of the physician in terminal care.

4. Enhanced teaching about death, dying and bereavement should occur throughout medical education.

5. Educational content and process should be tailored to students' developmental stage.

6. The best learning grows out of direct experiences with patients and families, particularly when students have an opportunity to follow patients longitudinally and develop a sense of intimacy and manageable personal responsibility for the suffering.

7. Teaching and learning about death, dying and bereavement should emphasize humanistic attitudes.

8. Teaching should address communication skills.

9. Students need to see physicians offering excellent medical care to the dying and their families and finding meaning in their work.

10. Medical education should foster respect for patients' personal values and an appreciation of cultural and spiritual diversity in approaching death and dying.

11. The teaching process itself should mirror the values physicians aspire to in working with patients.

12. A comprehensive integrated understanding of and approach to death, dying, and bereavement is enhanced when students are exposed to the perspectives of multiple disciplines working together.

13. Faculty should be taught how to teach end-of-life care, including how to be mentors and model ideal behaviors and skills.

14. Student competence in managing prototypical clinical settings related to death, dying and bereavement should be evaluated.

15. Educational programs should be evaluated using state-of-the-art methods.
Summary
There is a consensus that education in palliative care needs to be integrated at the undergraduate, graduate, and continuing medical education levels, and new initiatives and model programs are underway in medical schools, residency, fellowship, and faculty development programs. For example, the Veterans Administration Healthcare System supports a broad faculty development program in end-of-life care and funds fellowships in palliative care. Also, to further develop palliative care expertise and practice by neurologists, a Promoting Excellence ALS Peer Workgroup is creating guidelines and practical assessment tools for neurologists who care for patients with ALS and other neuromuscular diseases. An International Working Group in Palliative Care in Neurology has formed and is developing a core curriculum for neurologists. New texts such as "Palliative Care in Amyotrophic Lateral Sclerosis" and "Hospice Care for Patients with Advanced Progressive Dementia" serve as model monographs to educate health care professionals, and specifically neurologists, in a comprehensive approach for patients with these progressive, incurable neurologic diseases.

Despite these efforts, nothing would have a greater impact on providing palliative care to patients with neurologic disease now than instituting current knowledge. Within this issues, articles provide disease-specific palliative care discussions about patients with brain tumors, multiple sclerosis, ALS, stroke, dementia and Huntington's disease written by experienced clinical neurologists whose programs emphasize the importance of a comprehensive palliative care approach. Also included are articles specifically addressing palliative care domains common to all patients who are seriously and incurably ill, including symptom assessment and control, psychiatric issues, ethical and legal concerns, and the specific need to address the uniqueness of caring for the dying. As is evidenced by the unavoidable repetition in such a text, patients requiring palliative care share common symptoms and can benefit from common management approaches. It is this very observation that makes us enthusiastic about our goal that palliative care education can have a broad impact on the care of patients with neurologic disease over the course of their illness. The expanding role of neurologists in therapeutics, the enormous strides in the ability to improve patients' quality of life through symptom control, and the management of psychosocial distress provide the opportunity to better care for the chronically ill and to enhance the quality of living for the dying.

References


8. Department of Veterans Affairs: VA Faculty Leadership Project for Improved Care at the End of Life. Compendium of Abstracts and Curricula. June, 2000


Amyotrophic lateral sclerosis (ALS) (i.e., motor neuron disease in the United Kingdom and Lou Gehrig disease, or ALS, in the United States) is the most common degenerative disorder of the motoneuronal system occurring in adults. The estimated incidence of ALS is 1.5 to 2/100,000/year (the corresponding figures are 3 to 5/100,000/year for multiple sclerosis and 0.4/100,000/year for myasthenia gravis), a figure which appears to be increasing. The upper limit for prevalence is approximately 6 to 8/100,000, which means that 20,000 US citizens and 25,000 Western Europeans suffer from ALS at any given time. Although rare cases may begin before the age of 20 years, most cases begin after the age of 40 years, the mean age at onset is approximately 58 years.

The clinical picture is characterized by fasciculations and slowly progressing paresis of voluntary muscles, coupled with hyperreflexia and spasticity caused by concomitant involvement of upper and lower motor neurons. Bulbar onset with slurred speech (dysarthria) or difficulty in swallowing (dysphagia) occurs in 20% to 30% of all cases, particularly in older women. Extraocular movements and sphincter continence usually are spared, and sensation is normal. Although subtle neuropsychologic deficits can be detected on careful examination, frank dementia is a rare occurrence. The main symptoms of ALS are shown in Table 1.

<table>
<thead>
<tr>
<th>Direct</th>
<th>Indirect</th>
</tr>
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<tbody>
<tr>
<td>Weakness and atrophy</td>
<td>Psychological disturbances</td>
</tr>
<tr>
<td>Fasciculations and muscle cramps</td>
<td>Sleep disturbances</td>
</tr>
<tr>
<td>Spasticity</td>
<td>Constipation</td>
</tr>
<tr>
<td>Dysarthria</td>
<td>Drooling</td>
</tr>
<tr>
<td>Dysphagia</td>
<td>Thick mucous secretions</td>
</tr>
<tr>
<td>Dyspnea</td>
<td>Symptoms of chronic hypoventilation</td>
</tr>
<tr>
<td>Pathological laughter/tearfulness</td>
<td>Pain</td>
</tr>
</tbody>
</table>

Table 1. Symptoms of ALS
It should be noted that some of the indirect symptoms, such as psychologic problems, thick mucus secretions, or symptoms of chronic hypoventilation, may at certain stages have the greatest impact on the patients' quality of life.

The rate of disease progression in patients with ALS remarkably is variable. It is not uncommon to see patients with long phases of slow progression, but genuine remissions exceedingly are rare. Sudden worsening or relapses, such as in multiple sclerosis, usually do not occur in patients with ALS. Average disease duration is three to four years; 10% of patients survive more than 10 years, and single cases may run over several decades.

Diagnostic measures and causative treatments for ALS have been reviewed recently. The available disease-specific treatment options for ALS are still unsatisfactory. Therapeutic nihilism, however, is not justified because a large array of palliative measures is available to enhance the quality of life of patients and their families. It is important to stress that palliative care is neither restricted to pure symptomatic treatment nor to the terminal phase of the disease; it is a multidisciplinary approach that starts with how the diagnosis is communicated to the patient and continues to bereavement counseling for the family.

A first attempt at establishing evidence-based guidelines for clinical management of ALS recently has led to the publication of the Practice Parameters by the American Academy of Neurology. Many questions in this field, however, have not yet been addressed in proper randomized trials, and standards of palliative treatment in ALS are still largely based on expert consensus. This article covers the most important aspects of palliative care in patients with ALS using an evidence-based approach whenever possible.

Who is Involved in Palliative Care?

Palliative care in patients with ALS involves not only physicians but also a large number of different professionals (Table 2) and family members, for whom caring for the patient often becomes a full-time job.

<table>
<thead>
<tr>
<th>Table 2. Palliative care in ALS: Who is involved?</th>
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<tbody>
<tr>
<td>Chaplain</td>
</tr>
<tr>
<td>Counselor</td>
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<tr>
<td>Dietitian</td>
</tr>
<tr>
<td>Hospice worker</td>
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<tr>
<td>Lay associations</td>
</tr>
<tr>
<td>Nurse</td>
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<tr>
<td>Occupational therapist</td>
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</table>

The physician's role is to coordinate the efforts and discuss the appropriate time for each intervention with the patient and family. In the United Kingdom, at least 75% of the inpatient palliative care/hospice units are involved in the care of patients with ALS. This figure is lower (between 25% and 50%) for the rest of Europe (no U.S. data are available so far). Early and close cooperation between the neurologist and the local palliative care/hospice team can be of invaluable help for the patients and family. Ideally, all members of the palliative team should be trained and receive ongoing support and education in this area of expertise.
**Breaking the News**

Palliative care begins by how the diagnosis is communicated to the patient. If the communication between physician and patient is inadequate, the patient will be left uncertain about the diagnosis, with a vague sense that he/she has some serious and probably incurable disease without any reasonable hope for the future. Often, such a situation leads to a pilgrimage from one doctor to another and from one hospital to another, until a physician establishes a good therapeutic relationship with the patient based on mutual openness. Now, many patients gather information concerning their diagnoses from the Internet, which can sometimes lead to their knowing more about their disease than their physician.

Communicating the news of an ALS diagnosis is not a routine procedure. Although the physician often feels a sense of frustration in communicating an incurable diagnosis, this should not lead to withholding information from the patient or, worse still, providing information only to relatives and giving the patient "reassuring" statements. Conversely, it is an undeniable right of the patient to not be informed about the condition if so wished. Accordingly, it is the patient who should dictate the pace and depth of information flow, while the doctor retains the difficult task of responding appropriately to the patient's desires.

Once the diagnosis has been confirmed, the patient should be told that he/she has a progressive disease of the motor nerves. The name of the disease should be mentioned and explained to avoid confusion (e.g., with multiple sclerosis). Positive aspects of the disease (e.g., no pain, no disturbances in sensation, mentation or continence) should be emphasized. It is important to point out to the patient and family ongoing trials of new therapeutic agents and the existence of promising new drugs being studied and to encourage participation in clinical trials, which is often beneficial.

For subsequent clinical visits, the physician should remember that patients often recall a select portion of the information that has been discussed; therefore, care should be taken at each visit to find out how much the patient understands and move on from there. As the great existentialist philosopher Kierkegaard wrote in 1849, "If you really want to help somebody, first you must find out where he is. This is the secret of caring. If you cannot do that, it is only an illusion if you think you can help another human being. Helping somebody implies your understanding more than he does, but first of all you must understand what he understands."

The relentless progression of the disease is a burden for patients and caregivers alike. Indeed, the psychic burden of caregivers sometimes exceeds that of the patient. It is therefore mandatory to involve the patient's family in every step of palliative care beginning with disclosure of the diagnosis. If the patient and family express the wish for a second opinion, this should be encouraged, and appropriate tertiary referral centers should be offered.

Many patients with ALS (at least 54% according to a recent survey) turn to alternative treatments because of dissatisfaction with the effectiveness of available drugs. Often, this is not discussed with the physician because of fear of "condemnation." It is therefore advisable to approach this topic proactively when discussing the diagnosis, offering to discuss whatever therapeutic options the patient might wish to explore. This enables the physician to protect the patient from treatments that might entail serious financial or medical risks, while preserving hope and maintaining trust in the patient-physician relationship.
At the onset of symptoms of dyspnea, chronic nocturnal hypoventilation (Table 3), or when the vital capacity drops below 50%, the patient should be offered information about the terminal phase of the disease, because most patients at this point fear they will "choke to death."

**Table 3. Medications for fasciculations and cramps***

<table>
<thead>
<tr>
<th>Medicine</th>
<th>Dosage</th>
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<tbody>
<tr>
<td>Quinine sulphate</td>
<td>200 mg bid</td>
</tr>
<tr>
<td>Carbamazepine</td>
<td>200 mg bid</td>
</tr>
<tr>
<td>Phenytoin</td>
<td>100 mg qd-tid</td>
</tr>
<tr>
<td>Magnesium</td>
<td>5 mmol qd-tid</td>
</tr>
<tr>
<td>Vitamin E</td>
<td>400 IE bid</td>
</tr>
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</table>

*In all medication tables, the usual range of adult daily dosages is indicated; some patients may require higher dosages, e.g., of antispastic medications.*

Describing the mechanism of terminal hypercapnic coma and the resulting peaceful death during sleep will produce relief from this fear in most patients. Patients and relatives must be informed that the array of medication available in the terminal phase, if correctly applied, is sufficient to prevent "chooking to death" in every patient. This information needs to be reiterated in subsequent visits.

Concurrently, the patient should be asked whether intubation and ventilation are desired in the event of a terminal respiratory insufficiency. Patients who have been informed about the possible subsequent clinical course and who may end up in a "locked-in" syndrome or in an intensive care unit, will usually will deny permission to use such a procedure. This denial must be documented in writing by the physician and should be incorporated into an advance directive. The consequences of such a decision must be discussed with the patient, family, and primary physician (e.g., concerning the use of opioids in the terminal phase). It is important to revise the Advance Directive at six-month intervals, because ALS patients' preferences for life-sustaining treatments change over this period of time.

**Symptoms Directly Caused by ALS**

**Weakness**

Progressive weakness is the major symptom of ALS. Initially, patients find their stamina undergoes greater than usual day-to-day fluctuations, and they need to be reassured that this is a normal phenomenon and does not herald an increase in pace of disease progression. Active and passive physiotherapy can help in the prevention of muscle contractures and joint stiffness. The maximum exercise load may vary greatly from one day to another; as a rule, patients should never exercise to the point of exhaustion.

As the disease progresses, patients require additional devices to maintain mobility (from a cane to an ankle-foot-orthosis to a wheelchair) and independence in daily activities (e.g., special eating utensils, higher toilet seats, or bath tub lifts). It is important to discuss the need for these devices ahead of time, so the patient and family will have time to adjust mentally to the new degree of disability and be ready to accept help when it becomes necessary. Home evaluation by an occupational therapist can help to pinpoint the exact needs of the patient.
Acetylcholinesterase inhibitors may lead to a short-term improvement in muscle strength, especially early in the disease process. This effect is more pronounced in bulbar patients; however, it is not seen in all patients and may only last for days to a few weeks. Acetylcholinesterase inhibitors do not alter the course of the disease, and there is no rationale for long-term therapy with pyridostigmine in patients with ALS.

**Muscle Fasciculations, Cramps and Spasticity**

Fasciculations are often the first symptom of the disease. They arise through degeneration of the intramuscular motor axons and can lead to painful muscle cramps. Spasticity of the extremities, caused by degeneration of the upper motor neurons, can sometimes be clinically severe. These symptoms can be relieved effectively by appropriate medications (Tables 4 and 5).

<table>
<thead>
<tr>
<th>Table 4. Medications for spasticity</th>
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<tbody>
<tr>
<td>Baclofen</td>
</tr>
<tr>
<td>Tizanidine</td>
</tr>
<tr>
<td>Memantine</td>
</tr>
<tr>
<td>Tetrazepam</td>
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<table>
<thead>
<tr>
<th>Table 5. Symptoms of chronic respiratory insufficiency</th>
</tr>
</thead>
<tbody>
<tr>
<td>Daytime fatigue and sleepiness, concentration problems</td>
</tr>
<tr>
<td>Difficulty falling asleep, disturbed sleep, nightmares</td>
</tr>
<tr>
<td>Morning headache</td>
</tr>
<tr>
<td>Nervousness, tremor, increased sweating, tachycardia</td>
</tr>
<tr>
<td>Depression, anxiety</td>
</tr>
<tr>
<td>Tachypnea, dyspnea, phonation difficulties</td>
</tr>
<tr>
<td>Visible efforts of auxiliary respiratory muscles</td>
</tr>
<tr>
<td>Reduced appetite, weight loss, recurrent gastritis</td>
</tr>
<tr>
<td>Recurrent or chronic upper respiratory tract infections</td>
</tr>
<tr>
<td>Cyanosis, edema</td>
</tr>
<tr>
<td>Vision disturbances, dizziness, syncope</td>
</tr>
<tr>
<td>Diffuse pain in head, neck, and extremities</td>
</tr>
</tbody>
</table>

To effectively use antispasticity drugs, the patient has to titrate the dosage against the subjective clinical effect, because a moderate degree of spasticity is usually better for mobility than a fully flaccid paresis. Dantrolene should not generally be used as a first-line medication because it enhances weakness; however, a case of extreme spasticity in the terminal phase that could only be relieved by high doses of IV dantrolene was seen. If severe spasticity is a problem early in the disease, intrathecal baclofen delivery should be considered.

**Dysarthria**

One of the most feared consequences of ALS is losing the ability to communicate because of progressive dysarthria. Speech therapy training is especially helpful in cases with slow progression. When speech becomes unintelligible, electronic devices may be employed as
communication aids. A simple alphabet chart, however, can also be effective. Modern computer technology offers several options that enable even patients with almost total paresis of voluntary muscles to communicate and view the Internet (e.g., through myoelectrically controlled switches). Given the great variety of options, decisions about communication devices should be discussed on an individual basis.

**Dysphagia**

Dysphagia in ALS results from disturbed motility of the tongue, pharynx and esophagus. It can lead to choking and aspiration, especially with fluids and crunchy/chewy foods like popcorn or pumpernickel bread. The first step is a change in diet consistency. Food should be easy to chew and rich in calories. Recipe books for patients with ALS are available from patient associations such as the Muscular Dystrophy Association (MDA) or The ALS Association (ALSA) in the United States or the Deutsche Gesellschaft für Muskekranke (DGM) in Germany. Swallowing techniques, such as supraglottic swallowing, can be taught by specialized speech therapists or physiotherapists and can reduce the risk of aspiration. If, despite these actions, the caloric intake is still insufficient and the patient continues to lose weight (>10% of normal body weight before diagnosis) and oral food intake becomes intolerable because of frequent choking, a percutaneous endoscopic gastrostomy (PEG) should be discussed. The placement of a PEG is a simple procedure that can be performed using local anesthesia; however, if a PEG is postponed until the patient is in respiratory distress, the procedure becomes dangerous because of the possible insurgence of basal atelectasis through pressure of the air-inflated stomach against the weakened diaphragm. The patient and family should be encouraged to make an early decision regarding PEG placement (to minimize this risk before forced vital capacity [FVC] falls below 50% of predicted). Whether an early PEG placement may result in an increased life span has not yet been demonstrated convincingly; however, as with all other palliative measures, the primary goal is improvement of the quality of life, rather than prolonging life. It is important to remember that a PEG does not prevent aspiration pneumonia, which is especially frequent if overfeeding by PEG occurs.

**Dyspnea**

Respiratory insufficiency is the most feared symptom of ALS. Patients often react to the first dyspneic bouts with strong anxiety. In all stages of respiratory insufficiency, it is important first to break the vicious cycle of dyspnea-anxiety-dyspnea. The calm, reassuring presence of relatives, trunk elevation, and chest physiotherapy may provide relief. In cases with a pronounced panic component, lorazepam given sublingually (0.5-1 mg) is helpful. The subjective feeling of shortness of breath is reduced by the administration of morphine (2.5 to 5 mg by mouth or 1 to 2 mg IV/subcutaneous every four hours). Titration of the morphine dose against the clinical effect almost never leads to a life-threatening depression of respiratory drive. Treatments for chronic hypoventilation and terminal dyspnea are outlined below.

**Pathologic Laughing and Crying (Pseudobulbar Affect)**

A major symptom of ALS, which must be differentiated from a depressed mood state, is the insurgence of uncontrollable bouts of laughter or tearfulness, which is also referred to as the pseudobulbar affect and occurs in up to 50% of patients with ALS. It is not a mood disorder but an abnormal display of affect, which can be disturbing for the patient in social situations. Because this symptom is seldom disclosed, physicians should inquire about it and point out that it responds well to medication. The drug of first choice is amitriptyline, but positive effects also have been reported with fluvoxamine, dopamine and lithium (Table 6).
Table 6. Medications for pathologic laughing and crying

<table>
<thead>
<tr>
<th>Medication</th>
<th>Dosage</th>
</tr>
</thead>
<tbody>
<tr>
<td>Amitriptyline</td>
<td>10-150 mg</td>
</tr>
<tr>
<td>Fluvoxamine</td>
<td>100-200 mg</td>
</tr>
<tr>
<td>Lithium carbonate</td>
<td>400-800 mg</td>
</tr>
<tr>
<td>Levodopa</td>
<td>500-600 mg</td>
</tr>
</tbody>
</table>

Symptoms Indirectly Caused by ALS

Psychologic Problems

Most, if not all, patients with ALS undergo a phase of reactive depression after diagnosis. Counseling is of paramount importance at this stage. The reported prevalence of depression in ALS varies depending on the assessment method. Although full-fledged major depression, according to Diagnostic and Statistical Manual of Mental Disorders, 4th edition (DSM-IV) criteria is infrequent (around 10%), self-reported depressive symptoms have been described in 44% to 75% of patients. Clinically significant depression should be treated at all disease stages, particularly because the psychologic status of the patients strongly correlates with survival. The most widely used drug is amitriptyline (starting with 25 mg/d and slowly increasing to 100-150 mg/d, as tolerated), which may exert favorable effects on other symptoms such as drooling, pseudobulbar affect and sleep disturbance. If side effects such as dry mouth or constipation are a problem, selective serotonin reuptake inhibitors such as sertraline or paroxetine may be employed. Anxiety disorders are less common but may arise in conjunction with dyspneic bouts that can lead to panic attacks and should be treated with short-acting benzodiazepines as outlined earlier. Importantly, the concordance of depression and distress levels between patients and caregivers is high, reinforcing that attention to the mental health of the caregiver also may alleviate the patient's distress.

Symptoms of Chronic Hypoventilation

With progressing respiratory insufficiency, symptoms of chronic nocturnal hypoventilation (see Table 5) may develop. These symptoms can severely hamper the patient's quality of life. Noninvasive intermittent ventilation (NIV) by through mask is an efficient and cost-effective means of alleviating these symptoms, which may prolong the patient's life span considerably. As outlined before, this treatment should be discussed with the patient and family at the onset of symptoms of chronic hypoventilation. They should be informed about the temporary nature of the measure, which is primarily directed toward improving quality of life rather than prolonging it (as opposed to tracheostomy). The problem with mechanical ventilation usually is not related to cost or technical difficulties, but to the increasing care needs of the ventilated patients. A slow progression, good communication skills, mild bulbar involvement and, above all, a motivated patient and a supportive family environment argue in favor of the initiation of NIV. It is important to reassure the patient that whenever the decision to stop NIV is made, all necessary care and appropriate medications will be available to prevent death by choking. Collaboration with hospice can be helpful in such cases.

If the patient refuses NIV, intermittent oxygen application may be tried; however, oxygen is inferior to NIV because it may only be administered during the day when the patient is awake because of the danger of respiratory depression in chronically hypercapnic patients receiving oxygen during sleep.
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A full, 24-hour mechanical ventilation by tracheostomy is rarely chosen by fully informed patients, although single instances of patients with good quality of lives over a period of 10 years and longer have been reported. Conversely, unwanted intubation by the emergency physician because of terminal respiratory failure is not infrequent when patient and family are poorly informed about the disease. Such patients may survive for years in intensive care units and progress to a total "locked-in" syndrome where any possibility of communication with the outside world is rendered impossible by a complete tetraplegia including extraocular muscles. The understandable wish for discontinuation of life support that arises in such cases is fraught with medical, ethical and legal problems. One important aim of a good patient-physician relationship in ALS must be to prevent such a situation through early discussion and the completion of Advance Directives, including agreement upon conditions of withdrawal of ventilatory support when communication is no longer possible.

**Sleep Disturbances**

These are usually secondary to other causes, the most common of which are:

- Psychological disturbances, anxiety, depression, nightmares
- Inability to change position during sleep because of weakness
- Fasciculations and muscle cramps
- Dysphagia with aspiration of saliva
- Respiratory insufficiency with hypoxia and dyspnea

The underlying reasons for sleep disturbances should be evaluated carefully and treated. Nocturnal oximetry in the home is an inexpensive and quantitative measure of oxygen desaturation during sleep. Patients with symptoms of excessive daytime sleepiness or disruptive sleep patterns at night should be evaluated with nocturnal oximetry or, if unavailable, polysomnography. A trial of NIV may be initiated when desaturation of less than 90% partial pressure of oxygen ($P_{O_2}$) occurs for more than one minute cumulative total, and many patients will report symptomatic improvement. Sedatives should be administered sparingly, although higher doses may be used if the patient is using NIV (Table 7).

<table>
<thead>
<tr>
<th>Table 7. Sedatives for sleep disturbances</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Dosage Noote</strong></td>
</tr>
<tr>
<td>Chloral hydrate</td>
</tr>
<tr>
<td>Diphenhydramine</td>
</tr>
<tr>
<td>Flurazepam (beware of respiratory depression)</td>
</tr>
</tbody>
</table>

**Drooling**

Drooling is a frequent complaint in ALS and is caused by a combination of facial muscle weakness and pseudohypersalivation stemming from reduced swallowing ability. Medications reducing salivary output are helpful (Table 8); the drug of choice is glycopyrrolate.

<table>
<thead>
<tr>
<th>Table 8. Medications for drooling</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Dosage Noote</strong></td>
</tr>
<tr>
<td>Glycopyrrolate</td>
</tr>
<tr>
<td>Amitriptyline</td>
</tr>
</tbody>
</table>
Salivary gland irradiation and transtympanic neurectomy have been partially successful in anecdotal reports \(^{36}^{68}\) but have not yet been evaluated in controlled studies. Preliminary encouraging results have been reported with botulinum toxin in ALS\(^{28}\) and await confirmation in larger series.

**Thick Mucous Secretions**

This is one of the most difficult symptoms to treat in ALS. Late-stage ALS patients often suffer greatly because of thick mucous secretions blocking the upper airways that result from a combination of diminished fluid intake and reduced coughing pressure. N-acetylcysteine is helpful only in a minority of cases because it requires large amounts of fluid intake and only dilutes the secretions, resulting in a higher secretion volume that does not ameliorate the problem. Suction may become necessary but usually is not fully effective unless performed by a tracheostomy. Manually assisted coughing techniques and mechanical insufflation-exsufflation (In-Exsufflator, JH Emerson Co, Cambridge, Massachusetts) can assist in extracting excess mucus from the airway.\(^6^{7}\) Intermittent positive vibration devices are specialized inhalators that deliver a pressurized, intermittent flow of nebulized saline with or without expectorants. They are used for 10 to 15 minutes at a time and can assist in the clearing of pulmonary and bronchial secretions.\(^{25}\) Physical therapy with vibration massage also may be helpful, especially in the initial stages.

**Constipation**

Although the autonomic fibers innervating the intestine are not affected overtly by the disease, lack of exercise can promote the development of constipation in patients with ALS. The first step is dietary (e.g., adding foods with high fiber content such as "power pudding," an equal measure of prunes, prune juice, bran, and apple sauce to the diet). Care should be taken to ensure adequate fluid intake because dysphagia-induced dehydration may worsen constipation. The next step is a review of current medication, because muscle relaxants, sedatives and anticholinergics reduce bowel mobility. Mild laxative therapy should be initiated prophylactically in bedridden patients and in those receiving opioids. If bowel pain arises, an ileus should be suspected and appropriate tests performed.

**Pain**

Although ALS itself usually does not involve sensory fibers at the clinical level, musculoskeletal pain often arises in the later stages of the disease as a result of stress on bones and joints that have lost their protective muscular ensheathment because of atrophy. In addition, muscle contractures and joint stiffness (e.g., frozen shoulder) may be painful. These symptoms are usually best treated with nonsteroidal anti-inflammatory drugs (NSAIDs) and physiotherapy. Another cause of pain in patients with ALS is skin pressure pain caused by immobility. The patient may be unaware of how severe the pain has become and talk only of "discomfort."\(^{61}\) Special attention must be given to the nursing care, which requires frequent changes in the

<table>
<thead>
<tr>
<th>Transdermal byoscine patches</th>
<th>1-2 patches</th>
</tr>
</thead>
<tbody>
<tr>
<td>Atropine/benztropine</td>
<td>0.25-0.75 mg/1-2 mg</td>
</tr>
<tr>
<td>Clonidine</td>
<td>0.15-0.3 mg</td>
</tr>
</tbody>
</table>
patient's position at night and during the day. If NSAIDs are insufficient, opioid medication should be started according to the World Health Organization (WHO) ladder, with appropriate antiemetic medication (e.g., chlorpromazine) and laxatives.

**Other Indirect Symptoms**

**Gastroesophageal Reflux Disease**

Gastroesophageal reflux disease (GERD) may occur in patients with ALS because of diaphragmatic weakness involving the lower esophageal sphincter. Particular care is needed when starting a patient on PEG because of possible overfeeding that may lead to GERD and even aspiration. Treatment includes peristaltic agents (e.g., metoclopramide) and antacids.

**Dependent Edema of the Hands and Feet**

Dependent edema of the hands and feet occurs in weak limbs because of reduced muscle pump activity. Limb elevation, physiotherapy and compression hose are helpful. If pain develops or swelling persists despite prolonged elevation, a deep venous thrombosis should be ruled out.

**Urinary Urgency and Frequency**

Urinary urgency and frequency in the absence of urinary tract infections can be caused by spasticity of the bladder and responds favorably to oxybutynin (2.5-5 mg one to three times a day).

**Jaw Quivering or Clenching**

Jaw quivering or clenching may develop in patients with pseudobulbar involvement in response to noxious stimuli such as cold, anxiety or pain and may be relieved by benzodiazepines (e.g., lorazepam sl or clonazepam).

**Laryngospasm**

Laryngospasm (a sudden reflexory closure of the vocal chords) can cause panic because of the sensation of choking. Several types of stimuli (e.g., emotions, strong flavors or smells, cold air, fluid aspiration, sinus drainage or gastroesophageal reflux) may provoke this symptom, which usually resolves spontaneously within a few seconds. Repeated swallowing while breathing through the nose can accelerate resolution. Patients also benefit from reassurance and education about this distressing symptom. $H_1$ or $H_2$ blocking agents (antihistamines or antacids) also may be helpful in selected patients.

**Nasal Congestion**

Nasal congestion in bulbar patients with a weakening of the nasopharyngeal muscles can be helped by elevating the nasal bridge with nasal tape and applying topical decongestants.

**Quality of Life in ALS**

The dilemma of quality of life (QOL) in patients with ALS begins with its definition. The definition followed here is that of professor Ciaran O'Boyle, of Dublin, who said "Quality of life is whatever the patient says it is." Professor O'Boyle developed a QOL measure based on patient-generated cues, the Schedule for the Evaluation of Individual Quality of Life-Direct Weighting (SEIQOL-DW). In a randomized study, this scale was judged by patients with ALS as being more valid (i.e., reflecting more accurately what they perceive as their QOL) and less emotionally distressing than either the Sickness Impact Profile (SIP) or the Short-Form-36 (SF-36), two standard QOL questionnaires used widely in research and clinical trials. The QOL
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domain most often mentioned by the patients was "family" (100%), and health-related issues were perceived as being relevant to QOL in about half of the cases. Correspondingly, a recent study indicates that QOL in patients with ALS depends on factors other than strength and physical function.[16]

In a separate study involving ventilated ALS patients,[37] there was no difference in QOL as measured by standard questionnaires between patients on tracheostomy ventilation versus NIV. Additional questions, however, showed a significant difference in QOL of the primary caregivers (mostly the spouses), with a high burden falling on the caregivers of tracheostomy ventilation patients. Almost one third of tracheostomy ventilation caregivers (versus 3% of the NIV caregivers) rated their own QOL as being lower than that of the severely disabled ALS patient they were caring for.

These data should prompt a reconsideration of the way QOL is defined and assessed in ALS palliative care. The role of health status may have been overestimated so far, and the role of the family appears preeminent.

Psychosocial Care

In palliative care, as defined by the WHO, "the control of psychological, social, and spiritual problems is paramount."[80] Psychosocial care is often the most important cornerstone of palliative care in ALS and cannot be administered by physicians alone. A multi-disciplinary team approach is necessary, as has been exemplified by several excellent models of care around the world.[34] Palliative care in patients with ALS involves a number of different professions (see Table 2). Close collaboration between the team members is essential. At any one time, a different member of the team may be the most important person for the ALS patient and the family. Patients associations, such as The ALS Association or the Muscular Dystrophy Association in the United States, the Motor Neuron Disease Association in the United Kingdom, or the German Society for Neuromuscular Diseases, may provide invaluable help and assistance to patients and families and should be involved in patient care from the beginning (a worldwide directory of ALS associations can be found at www.alsmndalliance.org). Referral to a tertiary care center with an interdisciplinary team may ease the burden on the single practicing neurologist and also may be a means of providing hope (a list of ALS centers worldwide can be found at www.wfnals.org).

According to published data, a large proportion of patients with ALS show an interest in physician-assisted suicide,[24] which is often because of the fear of becoming a burden on their families or feeling isolated or disempowered in their relationships with the health care team; however, suicidal actions relatively are rare in ALS.[52] The psychosocial care of family members is as important as that of the patient.[27][30] A particularly sensitive problem are the needs and fears of the patient's children and how to help patients in their roles as parents. An in-depth discussion of psychosocial care in ALS would go beyond the scope of this article; a comprehensive review on this topic recently has been published.[22]

Spiritual Care and Bereavement

As with any terminal illness, spiritual care is an important but often overlooked part of palliative care. The word spiritual has several implications and is difficult to define. Sykes[76] defines it as "the need to find within present existence a sense of meaning," which may or may not involve a religious framework. A recent study indicates that spirituality or religiousness may affect the use
of PEG and NIV in ALS and may be a source of comfort to the patients. Cases of patients whose spiritual practice greatly enhanced their ability to cope with ALS have been reported.

Spiritual care is not limited to patients but should encompass the whole family as a means of preventing problems during bereavement. First data indicate that bereavement in relatives after an ALS patient's death may be particularly severe and prolonged. Conversely, strategies to reduce the impact of the loss of a loved one through appropriate counseling are available. It is important to acknowledge that the process of bereavement in ALS actually starts immediately after the diagnosis is communicated, in the form of anticipatory grief, and that callous delivery of the diagnosis may affect the psychologic adjustment to bereavement. A simple structured interview to assess the patient's spiritual needs has been recently developed. Further research into spiritual care and bereavement in ALS is needed to improve the quality of interventions during this pivotal segment of palliative care.

Terminal Phase

A retrospective survey of 171 patients with ALS showed that around 90% of the patients died peacefully, mostly in their sleep, and none of the patients choked to death. If patients with ALS are not artificially ventilated, the death process usually begins with the patients slipping from sleep into coma because of increasing hypercapnia. In this phase of the disease, efforts are directed solely at maintaining patient comfort. If restlessness or signs of dyspnea develop, morphine should be administered, beginning with 2.5 to 5 mg by mouth, subcutaneously, or IV every 4 hours (if necessary in combination with chlorpromazine as an antiemetic). Because morphine is not an anxiolytic drug, if anxiety is present it should be treated with lorazepam sublingually (beginning with 1 to 2.5 mg) or midazolam by mouth or subcutaneously (beginning with 1 to 2 mg). The dosages of morphine and anxiolytics should be increased until satisfactory symptom control is achieved. The potential of these drugs to induce respiratory depression (which is usually overestimated) is irrelevant in the terminal phase according to the doctrine of double effect. Administration of oxygen may be helpful psychologically but is usually of minor importance.

Most patients with ALS wish to die at home. This often can best be achieved through enrollment of the patient in a hospice program, which can be of invaluable help to patients and families. It is advisable for the physician to initiate contact with the hospice institution, where available, well in advance of the terminal phase. If death at home is not possible, inpatient hospice or palliative care units should be considered. Hospice teams also can assist during the relatives' bereavement period after the patient's death.

Summary

Patients with ALS witness their progressing debilitation with a clear mind. This situation is regarded as a nightmare by most neurologists; however, the intact mentation offers patients with ALS the possibility to develop coping mechanisms that can lead to a surprisingly serene acceptance of the disease. Indeed, most professionals dealing with patients with ALS and their families would agree that they are often exceptionally pleasant and warm people. This observation, although unexplained, is so striking that it has been the subject of presentations at scientific meetings. As physicians, it is a privilege to work with patients with ALS and to witness the formidable amount of inner strength that often develops in the wake of seemingly unbearable adversity. Patients with ALS and their families usually wish to be actively involved
in the decision processes regarding symptomatic treatment. It is the physician's responsibility to establish a working relationship with the patient and family that may enable their full participation in all aspects of palliative care.

Hopefully, new research developments will lead to drugs that can promote a significant prolongation of life in patients with ALS. This will increase the prevalence of the disease and is likely to be paralleled by a strong demand for optimal palliative treatment. Accordingly, QOL measures have become a standard feature of therapeutic trials in patients with ALS.¹⁶ Physicians, therefore, need to struggle to enlarge the evidence base for palliative interventions in this disease. Especially in patients with ALS, it is not enough to add years to life but also to learn how to add life to the remaining years.

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Section E

Ethical and Legal Issues in Palliative Care

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The contemporary practice of palliative care in neurology raises important ethical issues that deserve thoughtful consideration. A comprehensive analysis of these issues requires a discussion of moral theory and ethical practice beyond the scope of this article, but interested readers can consult a monograph on this subject. The annually updated monograph by Alan Meisel offers the most authoritative compendium on legal issues in the care of dying patients. This article outlines the ethical principles underlying palliative medicine and discusses several areas of current ethical controversy. The ethical discussions are highlighted with a brief summary of the relevant legal issues.

Ethical Principles

Patients Have a Right to Refuse Life-Sustaining Treatment

A defining characteristic of 21st century Western medicine is the recognition that patients have a right to refuse all tests or treatments recommended by physicians, including life-sustaining treatment, even if they die as a consequence. The ethical and legal rights to refuse life-sustaining treatment is a component of the ethical and legal doctrines of informed consent and informed refusal. The right to consent or refuse derives from the ethical and legal concepts of respect for self-determination and autonomy and the right to be left alone. People are not permitted to touch other people without their consent. Physicians are allowed to help patients only to the extent that patients permit them to. Physicians can make strong treatment recommendations, but ultimately, only patients can decide whether to accept or refuse these recommendations.

The doctrine of informed consent and refusal has three elements, all of which must be met for validity: adequate information must be conveyed to the patient, the patient must be able to decide, and the patient must have freedom from coercion. Physicians must provide patients with information adequate for a rational decision. Adequate information is information a reasonable person needs to know to make the decision in question. Reasonable people need to understand their options, the general risks and benefits of each option, the physician's treatment recommendation, and the reasons for the recommendation.
Patients must be able to consent or refuse. This capacity entails the ability to comprehend the available choices and their risks and benefits, to think rationally, and to express a treatment preference. Competence, in a clinical and nonlegal sense, is used by many physicians to refer to the capacity to make medical decisions. (For the purposes of this article, this is the definition followed.) In the absence of capacity, patients do not forfeit the right to consent or refuse. Rather, the right is transferred to a surrogate decision-maker who consents or refuses on behalf of the patient. Patients must make their consent or refusal freely without coercion from physicians, family, friends or agencies.

The evaluation of competency to consent or refuse medical decisions usually can be made at the bedside by neurologists based on a neurologic examination. Although a formal legal determination of competency can be made only by a court of law, this step rarely is necessary in practice. If neurologists remain uncertain whether a patient has the capacity to consent or refuse therapy because of a fluctuating mental status, intermittent confusion or depression, psychiatric consultation may help to clarify the patient's competence to consent or refuse.

In practice, the physician and patient unite to create a decision-making dyad in a process known as shared decision making. In shared decision making, the physician supplies the necessary technical information about treatments, side effects, beneficial effects, outcomes, and prognoses, and then makes a treatment recommendation. The physician attempts to separate fact from opinion in the patient's mind. The patient brings his or her personal system of values and treatment preferences and uses it as a basis to consider each option. Together they identify a mutually agreeable treatment plan and carry it out.

When a patient refuses life-sustaining treatment, the physician should make certain the refusal is valid before respecting it. The physician can investigate to find any evidence of reversible depression in which treatment might change the patient's refusal, and then treat the depression to improve the patient's psychologic state. The physician can investigate whether the patient has shown evidence of irrational thinking and try to help the patient think rationally. The physician can improve the patient's physical and communicative capacities to fully enable him/her to participate in decisions. The physician can test if the refusal remains consistent over time and is not merely an impulsive reaction to a particular medical, personal or social event and can weigh the extent of the family's support as a measure of the rationality of the refusal. The physician can support the patient and family as a social unit. Once a physician has concluded that the patient's refusal is valid, however, he/she must respect it by discontinuing life-sustaining treatment.

During the discontinuation of life-sustaining treatment, physicians have a duty to provide proper palliative care to minimize suffering. As noted by other contributors to this volume, proper palliative care in this setting includes the appropriate use of opiates, benzodiazepines, barbiturates and other agents. There is evidence that rapidly dialing down ventilator settings is preferable to summary extubation or prolonged terminal weans in ventilator-dependent patients refusing life-sustaining treatment.

**Withholding and Withdrawing Treatment are Ethically and Legally Identical**

Many physicians wrongly believe there is an important ethical or legal distinction between withholding life-sustaining treatment that has not been started and discontinuing life-sustaining treatment that has already begun. The psychologic basis of this incorrect belief is the intuitive sense of causation. Some physicians conceptualize that if a patient dies without life-sustaining
treatment, the patient died of natural causes, and the withholding of life-sustaining treatment was not directly causative; however, if the patient was dependent on life-sustaining treatment and died as soon as the physician withdrew the life-sustaining treatment, the act of withdrawing life-sustaining treatment was causative in the patient's death. Therefore, these physicians may be willing to withhold life-sustaining treatment, but not to withdraw it, because they wish to avoid causing the patient's death.

Although this psychologic explanation is helpful in understanding a physician's feelings and motivations, it is irrelevant in addressing the physician's duty once a patient validly refuses life-sustaining treatment. Once validly refused, life-sustaining treatment not started must be withheld and life-sustaining treatment already begun must be withdrawn. There are no important ethical or legal distinctions between the two actions. Patients (or surrogates acting on their behalf) who consent to life-sustaining treatment and later change their minds should not be required to continue receiving life-sustaining treatment because of the physician's psychologic difficulty in discontinuing it. The law makes no distinction between withholding and withdrawing life-sustaining treatment once a patient has refused it.³⁸

Some neurologists fear liability for criminal homicide if they withdraw life-sustaining treatment and the patient dies.¹⁸ This fear is grossly unrealistic. A number of high court rulings, beginning with Quinlan and ending with Cruzan, have made clear that physicians who discontinue life-sustaining treatment that validly has been refused by a patient or a legally authorized surrogate incur no liability for criminal homicide; however, not discontinuing life-sustaining treatment once it has been validly refused creates a potential liability for negligence.

**Surrogates are Authorized to Refuse Life-Sustaining Treatment on Behalf of Incompetent Patients**

A patient who lacks a decision-making capacity requires a surrogate decision-maker. Surrogates can be appointed formally through legal mechanisms such as health care proxy or durable power of attorney statutes operational in most states.⁸² For the incapacitated patient without a legally authorized surrogate, the united family can be an acceptable surrogate even though they lack legal authority. If the family remains intractably divided, however, it may be advisable to ask a judge to formally appoint a guardian or legal surrogate. In some states, legally authorized surrogates are appointed automatically in the absence of a previously named surrogate from a legally designated list of close relatives.⁶⁵

Physicians should explain to surrogates that how they reach their decision on behalf of patients should follow established ethical and legal standards. Surrogates first should try to follow the patients' previously expressed wishes, when known. When unknown, surrogates should use the standard of substituted judgment and attempt to reproduce the decision the patient would have made under the circumstances, using the patient's system of values and preferences. When the patient's values and preferences are unknown, the surrogate should attempt to balance the benefits and burdens of the patient to a particular course of therapy and determine what course is in the patient's best interest.¹⁵

Surrogates need counsel and emotional support from physicians as they execute their important and difficult roles. Physicians continually need to emphasize that they will work with the surrogate using shared decision making to try to do the right thing, namely to treat the patient exactly as the patient would have wished to be treated. Physician communication, explanations,
and treatment recommendations should be conducted with surrogates to the same extent as with patients. Explaining to surrogates that their roles merely are to communicate the patient's preference helps alleviate the guilt felt by many surrogates about their decisions to refuse life-sustaining treatment.

**Patients Can Use Advance Directives to Stipulate Their Treatment Preferences**

Advance Directives are written documents available in every state that permit patients, while competent, to communicate how they wish to be treated should they lose their decision-making capacity because of future illness or injury. The directives are of two kinds: the living will (also called a terminal-care document) that outlines which types of therapies will be given and which will not, and the formal appointment of a surrogate decision-maker. The limitation of the living will almost always is that it is written in such general language that its interpretation remains ambiguous in most clinical situations and because, in nearly all jurisdictions, the living will is not activated until the patient is terminally ill, and offers no direction in the setting of incapacity not resulting from terminal illness.

A more useful directive is the appointment of a health care agent, also known as a durable power of attorney for health care. By this appointment, a competent patient can name a friend or relative who the patient trusts and with whom the patient can explain in detail his or her wishes for future types of treatment. The appointment document also provides for annotations or checklists for wishes about specific treatments. Unlike the living will, the health care agent appointment is activated once a patient loses decision-making capacity. Surrogate appointments are more useful than living wills because of their flexibility. Rather than having to anticipate every possible circumstance on a written directive, the agent has the discretion to apply the patient's wishes to novel and unanticipated situations. Patients also should discuss their preferences in detail with their physicians. Most jurisdictions empower health care agents with the same legal authority to consent or refuse on behalf of patients that the patients themselves have. Health care agents make ideal surrogates because they know the patient's values and preferences and are authorized legally to decide on the patient's behalf.

Neurologists have an important role in encouraging their patients to complete Advance Directives. Neurologic patients have a high frequency of developing cognitive incapacity and the need for Advance Directives in advanced stages of illness. It is best for discussions of Advance Directives to take place during regular office visits when neurologists can discuss the benefit to the patient of the directive. Patients with early forms of dementia can be encouraged to complete Advance Directives anticipating the later development of incompetence. The Advance Directive is most useful when it is accompanied by a frank discussion with the patient about treatment preferences to promote patient-centered care. Patients also should be encouraged to have these discussions with family members who are health care agents. Many physicians keep copies of Advance Directive forms that are authorized legally in their states; these can be provided to patients who wish to complete them.

**Patients Have a Right to Refuse Artificial Hydration and Nutrition**

The question of whether patients or their surrogates can refuse artificial hydration and nutrition (AHN) as a component of life-sustaining treatment has been the topic of considerable ethical debate. Although some scholars believe that AHN differs from life-sustaining treatment because
it is essential to life and never should be withheld or withdrawn,[25] [26] most scholars hold that the right to refuse AHN is identical to the right to refuse life-sustaining treatment because both are encompassed in the ethical and legal rights of self-determination.[27] [28] As a matter of practice, the choice to refuse AHN routinely is included in hospital policies on refusal of life-sustaining treatment.

The American legal basis for the right to refuse life-sustaining treatment, including AHN, was the United States Supreme Court 1990 Cruzan ruling.[29] In the Cruzan case, the Supreme Court ruled that there is a constitutional right for all American citizens to refuse unwanted life-sustaining treatment, including AHN. The Court found this right in the liberty rights in the 14th amendment to the Constitution.[30] Thus, there is a strong legal and ethical basis for regarding AHN as a medical therapy that, like other medical therapies, may be consented to or refused by competent patients or by the surrogates of incompetent patients.

Patients Deserve Excellent Palliative Care

The ethical foundation of palliative care is the moral requirement to do good to patients and not harm them.[31] Suffering is a profound harm and represents a principal fear of dying patients. The duty to relieve suffering is a fundamental goal of medical practice.[32] The ethical duty imparts a responsibility to learn and practice the principles of excellent palliative care. Thus, physicians caring for dying patients have an ethical duty to learn the pharmacology of opiates, benzodiazepines and barbiturates, and to learn the palliative treatments of common causes of suffering in dying patients, such as nausea, cough, hiccups, dyspnea, agitation, confusion, anxiety, anorexia, fatigue, constipation and xerostomia.[33]

Nonabandonment is an important ethical duty. Physicians and friends often abandon dying patients as they become sicker.[34] Some physicians develop an overwhelming feeling of failure or impotence when faced with a dying patient. Because of these subconscious feelings they may ignore the needs of the dying patient. The knowledge that the physician improved a dying patient's quality of life should be sufficient gratification to prevent feelings of failure. Other ethical duties include being available for questions and counsel, explaining what to expect during the dying process, and providing emotional, psychological and spiritual support.[35]

Some neurologists fear liability for criminal homicide if the patient dies during palliative treatment using opiates, benzodiazepines or barbiturates.[36] When physicians practice correct palliative care using dosages of medications that are appropriate for the goal of palliation, they incur no liability for homicide even if a patient dies sooner as a result.[37] In the Cruzan case, the United States Supreme Court clarified that appropriate palliative care that culminates in a patient's death is not euthanasia or physician-assisted suicide (PAS);[38] however, if a physician intends to kill a patient by ordering a medication dosage designed to kill that clearly exceeds that required for palliation, plausible grounds could exist for charges of homicide, though prosecutors rarely file such charges.[39]

Physicians are not Required to Provide Physician-Assisted Suicide or Euthanasia

The physician's duty to withhold or withdraw life-sustaining treatment that has been refused validly by a patient does not extend to providing PAS, euthanasia, or any other therapy or act requested by a patient. The absence of a duty to perform assisted suicide or euthanasia requested by a patient is based on the important moral distinction between responding to patient refusals
and requests. Once the preconditions have been satisfied, assuring that a patient's refusal of life-sustaining treatment is valid, physicians must withhold or withdraw treatment because they cannot continue treatment without consent. The physician, however, is not required to respond in the same way by providing PAS or euthanasia at a patient's request. For example, a headache patient may insist on a prescription for an opiate analgesic. Like any request, physicians are not required automatically to fulfill it but should use judgment as to whether or not to fulfill it based on knowledge and experience.

The important moral distinction is based on the premise that a patient's treatment refusal is tantamount to the command "stop doing that to me!" All refusals can be paraphrased to this command. Once valid consent has been withdrawn, there is no ethical or legal doctrine permitting physicians to continue treatment. Indeed, continuing treatment in the setting of explicit valid refusal may constitute battery.

The United States Supreme Court ruled recently that the access of dying patients to PAS is not a constitutionally protected right as is the patient's right to refuse unwanted life-sustaining treatment. The Court found an important legal distinction between treatment refusals and requests and asserted that only refusals are constitutionally protected. In the absence of countervailing state law, requests for PASs are left to the judgment of physicians as would fulfilling any other request.

**Ethical Controversies**

*Futility as a Justification to Discontinue Life-Sustaining Therapy*

The principal remaining controversy in withholding and withdrawing life-sustaining treatment is whether a determination of medical futility constitutes ethically and legally defensible grounds for physicians unilaterally discontinuing life-sustaining treatment in the face of a family's or surrogate's demands to continue it. At its most basic level, a determination of medical futility means that, on the basis of available evidence, a hoped for beneficial effect of treatment will not occur. Because physicians have no ethical duty to prescribe or offer treatments that will not help patients, they may unilaterally refuse to provide or even offer futile therapies despite the demands for them by patients, surrogates or family members.

The seemingly straightforward topic of futility masks an underlying complexity. As reported by Schneiderman et al., medical futility has two components: a quantitative and a qualitative component. The quantitative component measures the probability that a given treatment will produce a desired physiologic effect. The qualitative component measures whether the physiologic effect will offer benefit to the patient. Analogous to medical decision analysis where the force to proceed with a medical intervention is the product of two independent variables, the probability of success of the intervention and its usefulness, the determination of futility is the product of the quantitative and qualitative components. As either component approaches zero, the product approaches zero and futility can be established.

For a correct determination of futility, physicians must apply validly obtained outcome data. Such data, however, are not available for many interventions. Physicians should not merely rely on their experiences in such matters because of the biases that such reflection commonly produces, such as last-case bias. Furthermore, patients, families and physicians may disagree on the "usefulness" determination. This disagreement was most graphically portrayed in the
much-discussed case of Helga Wanglie over the benefit to her or her family to continue life-support in a persistent vegetative state (PVS). Her physicians held that continuing life-sustaining treatment to keep her in a PVS was conferring no medical benefit but her husband and family disagreed, believing that her continued life at any quality was beneficial.

Several scholars have characterized the futility debate as a power struggle for authority in which the autonomy of the patient (represented by the surrogate) is pitted against the autonomy of the physician. Physicians should try to cultivate and practice skills of facilitating communication and dispute resolution when faced with futility disputes with families or surrogates. Hospital ethical committees and ethical consultants also can be helpful in mediating and resolving such disputes.

The concept of rationing is relevant to futility disputes. Who should determine the proper use of scarce medical resources? Should patients or surrogates be granted the authority to demand any type of treatment they wish, irrespective of prognosis or cost? What about the impact of these wishes on society? Futility, however, is not solely a rationing issue; the professional integrity of physicians and nurses is relevant. Should physicians and nurses be required to provide certain forms of medical and nursing care they feel is wrong only because some surrogates or families demand it?

The courts have been unwilling to settle this issue in favor of physicians' unbridled authority to dictate the benefit of the therapies they prescribe. In the Wanglie case and the celebrated case of the anencephalic infant Baby K, whose mother insisted on ventilator treatment and other aggressive therapies against the wishes of her physicians, the courts supported the family members who insisted on continued therapy. Only in the highly publicized Gilgunn case did a court find for the physicians who discontinued life-sustaining treatment on a ventilator-dependent elderly woman against the wishes of the patient's daughter. The Gilgunn case, however, was a jury verdict that carries little legal precedential weight.

Some scholars conclude that the only solution to the futility debate is to develop a medical practice standard through a community-wide consensus on policy and then follow that policy. The Houston City-Wide Task Force on Medical Futility and the Bay Area Network of Ethics Committees Nonbeneficial Treatment Working Group are examples of groups of hospitals working jointly to formulate and follow common futility policies. The American Medical Association Council on Ethical and Judicial Affairs also has developed a model policy for resolving futility disputes in dying patients.

Medication Dosage and the Principle of Double Effect

Symptom control is a fundamental goal of palliative medicine and opiates are effective in the control of pain and dyspnea. Chronic opiate therapy is complicated by the development of opiate tolerance. As tolerance develops, larger doses of opiates are necessary to achieve a given level of analgesia and control of dyspnea. Fortunately, the respiratory suppression side effect of opiate therapy is susceptible equally to the development of tolerance. Thus, a patient chronically on high-dose opiates will have no greater respiratory suppression at these higher doses than at lower doses.

The fear that high-dose opiates will suppress respiratory drive and accelerate death is accepted widely but is rare because of the concomitant development of respiratory tolerance.
Nevertheless, occasionally a patient's pulmonary failure is so severe that the small respiratory suppressant effect may become clinically significant. In these cases, it is conceivable that the high-dose opiates necessary to control pain or dyspnea might accelerate death. Is such treatment, therefore, ethically acceptable?

The principal of double effect has been applied to test the morality of actions known to have two morally opposite effects, beneficial and harmful, one intended and the other foreseen but unintended. In this case, a physician prescribed an appropriate dose of morphine to relieve a dying patient's pain or dyspnea, but the patient died sooner than he would have without the morphine. The intent of the physician was to relieve the patient's pain and dyspnea, and the acceleration of death was a foreseen but unintended consequence. The physician, therefore, has performed an act with a double effect. Is the palliation of pain and dyspnea morally justifiable despite accelerating death?

Moral philosophers and theologians have generated criteria to morally justify acts that produce double effects: the act must not be intrinsically wrong; the intended effect must be the good effect, even though the bad effect may have been anticipated; the bad effect must not be the means to create the good effect; the act must be undertaken for a proportionately serious reason; and the good result must exceed the harm produced by the bad effect. The physician's act satisfies the principle of double effect, because giving morphine to relieve suffering is not intrinsically wrong, and it was the desired effect although the risk of death was foreseen. Death was not the means for providing the desired effect. Prevention of suffering during dying is an important goal; for a terminally ill patient, the absence of pain and suffering exceeds the harm of dying sooner.

Sedation in the Imminently Dying

Ordinary palliative care is sufficient to relieve pain and suffering in most dying patients; however, in some states of intractable pain and suffering in patients with far-advanced illness, the only solution may be to sedate the patient to the point of unconsciousness or near-unconsciousness by administering barbiturates or benzodiazepines. This practice, commonly called terminal sedation, but more accurately called sedation in the imminently dying, is controversial.

Analyses of the morality of sedation in patients who are imminently dying, using the principle of double effect, conclude that it is morally permissible when there is no better alternative. Sedating a patient for palliative care is moral. The death of the patient may be a foreseen consequence but is not the intended consequence; only relief of pain or other symptoms is intended. The relief of pain does not require the death of the patient to be effective. The act is performed for a proportionately serious reason, namely to prevent suffering.

Some patients can achieve comfort during light sedation; others require heavier sedation to the point of full unconsciousness. Sedation often is used during the withdrawal of life-sustaining therapy, especially when withdrawing ventilator-dependent patients from the ventilator; however, it is unnecessary and undesirable to prescribe heavy sedation when lesser degrees of sedation are sufficient for symptom relief. Some physicians have performed active euthanasia under the guise of terminal sedation in earlier and less severe stages of illness when sedation was unnecessary for palliative care. In addition to its illegality because it is active euthanasia, such an act is not morally justified by the principle of double effect because the intended consequence is
to kill the patient, and there are alternatives available for palliative care that do not require killing.\[94\]

Another ethical controversy surrounds the use of neuromuscular blocking drugs as part of palliative care when discontinuing ventilators after patients or their surrogates have refused life-sustaining treatment. With ongoing neuromuscular blockades, patients cannot breathe and quickly die, creating a situation that some have likened to active euthanasia. Neuromuscular blocking drugs should not be introduced at the time of removal from the ventilator and usually should be discontinued on withdrawal of the ventilator. The only exceptions are when the patient's death is expected to be rapid and certain, regardless of the use of neuromuscular blocking drugs, and when the burdens to the patient and family from waiting for the drugs to wear off before extubating the patient exceed the potential benefits.\[98\]

**Legalization of Physician-Assisted Suicide and Voluntary Active Euthanasia**

The question of legalizing PAS and voluntary active euthanasia (VAE) continues to be the subject of scholarly and public debates. The reasons for general concern about these issues are clear. Many people believe that the civil rights guaranteed by our society should encompass the right to die when, where and how they choose. Moreover, people fear that their physicians will treat them with more aggressive life-sustaining treatment than they desire, and that they will suffer during terminal illness because they will not receive adequate palliative care. Unhappily, studies of critically ill and dying patients confirm that palliative care is unsatisfactory in many or most settings.\[95\] Some people conclude that legalizing PAS would resolve these concerns. Many believe that most dying patients do not require PAS and the solution lies not in its legalization but in improving palliative care.\[17\]

In PAS, a competent, terminally-ill patient requests the physician to provide the medical means for him/her to commit suicide that the patient subsequently uses. In PAS, the physician is necessary but not sufficient for the patient's suicide because the patient must employ the medical means.\[14\] The most common scenario is a patient requesting a prescription for a lethal dose of barbiturates with instruction on how to take them for a successful suicide. PAS is illegal in most jurisdictions in the United States. In 1997, Oregon legalized PAS by public referendum but only several dozen patients have committed suicide under this law.\[31\] \[89\]

In VAE, a competent, terminally-ill patient asks a physician to kill him or her. The physician's act in VAE is necessary and sufficient for the patient's death. The patient's terminal illness provides only the context for the request but is an unnecessary factor in the death.\[14\] VAE is a violation of criminal law in every jurisdiction in the United States, although prosecutors have exercised discretion in enforcing homicide statutes in some cases because of the physician's compassionate motive. The highly-publicized and later televised euthanasia of an amyotrophic lateral sclerosis (ALS) patient by Jack Kevorkian led to this physician's conviction and incarceration for criminal homicide despite multiple previous exonerations for PAS.

The term *passive euthanasia* should be abandoned. In former usage, it described the situation in which a physician unilaterally discontinued life-sustaining treatment and the patient then died of his underlying illness; however, this term no longer conforms to the reality of patients dying after removal of life-sustaining treatment. In contemporary *patient-centered* medical practice,\[6\] competent patients and the surrogates of incompetent patients have the ethical and legal rights to refuse further life-sustaining treatment. Physicians must comply with valid treatment refusals in
nearly all cases. It is misleading and incorrect to state that a physician who is required to stop life-sustaining treatment once it has been refused has committed passive euthanasia when the patient later dies of his underlying disease.

The morality of PAS and VAE remain debatable with powerful opinions on both sides. Scholars arguing that PAS and VAE are morally acceptable usually cite justifications based on self-determination and non-malfeasance. PAS advocates believe that respect for patient autonomy and self-determination should encompass the right for a patient to receive a physician's assistance to die at the time of the patient's choosing. Moreover, PAS advocates believe that when physicians cannot successfully cure or palliate dying patients, they have nonmaleficence-based duties to help them die when they request.

Scholars on the other side have expressed equally strong opinions that PAS and VAE are immoral. Physicians should never kill patients or help them kill themselves because doing so harmfully alters the goals of medicine. If physicians are authorized to kill patients or help them kill themselves, they irreversibly damage the patient-physician relationship by diminishing the trust that forms its foundation. PAS opponents hold that physicians have a duty to provide excellent palliative care but not to kill patients or help them kill themselves.

Solely as a matter of public policy and irrespective of the moral issues, it is unwise for society to legalize PAS. First, PAS requests are rare with optimal palliative care. Second, explicitly legalizing PAS in the patchwork health care system, in which palliative care is not available to all dying patients, might preferentially select those patients in the lower socioeconomic classes who lack access to palliative care. Third, it could create the duty to die, a feeling some elderly or terminally-ill patients might develop, in which they feel pressured by circumstance to elect PAS to save their loved ones’ money or prevent emotional distress. The duty to die questions the prerequisite of voluntariness implicit in all proposed schemes of legalizing PAS. Fourth, it changes the traditional role of physicians to the role of assisted killers.

In addition, it is inevitable that despite the restriction of present bills to legalize only PAS and not VAE, once PAS is permitted, the law will later expand to encompass VAE. Why should a legal right to PAS be restricted categorically only to those patients with the capacity to independently perform the act? This restriction unfairly discriminates against terminally-ill patients who lack the physical capacity to commit suicide. As a matter of legal coherence, it is likely that future courts will expand patients' rights to PAS, where legal, to include VAE.

In 1997, the United States Supreme Court ruled that there is no constitutional right for citizens to have PAS on request. The Court overruled the two Federal Circuit Court decisions that found no important distinction between the right to refuse life-sustaining treatment and the right to have PAS on request. The Court affirmed the former right as constitutionally protected but rejected the latter. In so doing, they emphasized the common-sense distinction between positive and negative rights. The only relevant valid constitutional right is the right to be left alone; the right not to receive treatment or any other intervention that a citizen refuses. There is no correlative right to have something one requests, as PAS. This ruling follows the argument made about the important moral distinction between refusals and requests. Physicians almost always must comply with a patient's refusal of life-sustaining treatment, but they are under no obligation to provide a patient's request (for PAS or anything else) unless using the sound clinical judgment they feel medically appropriate.
The experience of legalized PAS in Oregon, where voters in 1997 approved the Oregon Death with Dignity Act is important to review. This law provides terminally-ill patients who have made one written and two oral requests, separated by at least 15 days, the right to receive a prescription for a lethal dose of medications. The physician must inform patients of feasible alternatives, including palliative care, and refer patients to counseling if there is evidence of a psychiatric disorder. According to the Oregon Health Division, the agency supervising and evaluating this program, during 1998, 23 patients received prescriptions for lethal doses of medications, which 15 of the patients used to commit suicide. Thirteen of the 15 patients had cancer. PAS accounted for only six of 10,000 deaths in Oregon that year.\cite{31}

During 1999, 33 persons received, 26 medications used them for suicide, plus another person who had received a prescription in 1998. Cancer was the underlying disease in 17 patients, ALS in four, and chronic pulmonary disease in four. All patients had health insurance, and 21 patients were receiving hospice care. As in 1998, the most common reasons for suicide were loss of autonomy, loss of control of bodily functions, an inability to participate in pleasurable activities and a determination to control the manner of death. Despite the increase in the number of patients committing PAS, they still constituted only nine of 10,000 deaths in Oregon in 1999.\cite{89} A questionnaire study of Oregon physicians disclosed that they granted only one in six patient requests for PAS and that only one in 10 requests eventually resulted in suicide.\cite{39}

During 2000, Oregon physicians wrote 39 prescriptions of which 27 were used to commit suicide. Other than being better educated than other dying patients, the PAS patients were demographically similar. Identical to the findings in 1999, the patients represented only nine of 10,000 deaths in Oregon that year. One patient regurgitated part of the secobarbital dose but died quickly thereafter. One physician was reported to the Oregon Board of Medical Examiners for submitting an incomplete consent form. Compared with 1999, there was a significant increase in the number of patients concerned about being burdens to their family, friends and caregivers.\cite{90}

Critics of the Oregon law correctly point out that despite the legal requirement for physicians to explain feasible alternatives, the law contains no provision requiring physicians to be knowledgeable about palliative care. Moreover, the Oregon law does not require patients to undergo intolerable and unrelieveable suffering, as is required for approval of PAS in the Netherlands. Critics also have attacked the Oregon Health Division because the personnel evaluating the PAS program's success were not knowledgeable about palliative care options. Critics also questioned the objectivity of the report because the Oregon Health Division, the agency responsible for assessing the law's success, has become its principal advocate.\cite{36}

A few conclusions are possible at this early stage of the law's implementation. PAS is used selectively and only rarely in Oregon and few dire problems have occurred. Because PAS is used by less than one-tenth of 1% of dying patients in Oregon, the generalizability of these data to the larger population remains questionable. Clearly, such a rarely used option is not the answer to the problems of the dying patient. Purely from a pragmatic perspective, improving systems of palliative care that benefit all dying patients is a much more useful public policy than legalizing PAS.\cite{81}

The experience of PAS and VAE in the Netherlands also continues to be the subject of scholarly interest. Although PAS and VAE remained illegal under the Dutch Penal Code (until the law was changed in 2001), both have been permissible, and physicians had not been prosecuted since 1985 if they followed a broadly drafted set of judicial guidelines.\cite{100} PAS and VAE are common
modes of death in the Netherlands, accounting for approximately 2.3% of all deaths. Approximately, three-quarters of the cases in which physicians participate are VAE and one-quarter are PAS.\[50\]

The principal criticism of the Dutch practice has been the reported cases of involuntary euthanasia. Despite the fact that involuntary euthanasia remains illegal and that the judicial rules require the consent of the competent patient, numerous reports describe cases of involuntary euthanasia.\[32\] The frequency of involuntary euthanasia was estimated at 0.7% of total deaths in 1995.\[50\] Once the societal taboo against physician assisted killing is lifted, there will be a natural tendency for physicians to expand the "benefits" of euthanasia to incompetent patients in a frightening way.

Medical professional societies in the United States and most other countries outlaw PAS and VAE. For example, the practices have been condemned by the American Medical Association,\[9\] the American College of Physicians,\[6\] the American Psychiatric Association,\[10\] the American Academy of Neurology,\[5\] the American Geriatric Society,\[7\] and the British Medical Association,\[103\] among other societies.

When faced with a dying patient's request for PAS or VAE, the physician should ask why the patient has made such a request.\[70\] The physician should consider a patient's poor quality of dying as a medical emergency and act accordingly to improve the quality of dying.\[77\] This action includes redoubling all efforts to address suffering and relieve it, search for and treat depression, try to enhance the integrity of the family unit, arrange for spiritual and psychologic support, and invite hospice assistance and counseling.\[18\]\[35\] If all these measures do not change the patient's wish for PAS or VAE, physicians can inform patients that they may simply refuse all enteral and parenteral hydration and nutrition.

**Patient Refusal of Hydration and Nutrition**

Refusal of eating and drinking by dying patients who choose to die sooner has been practiced in numerous cultures throughout recorded history. Scholarly attention to this topic over the past decade has been motivated by attempts to find a socially-acceptable avenue for suffering, dying patients who wish to die more quickly, without risking the potentially harmful changes resulting from legalizing PAS and VAE.\[68\] Dying patients may be informed that they can refuse all hydration or nutrition. This practice is called "patient refusal of hydration and nutrition" (PRHN).\[14\]

The ethical and legal basis of PRHN is the right to refuse treatment and the right to be left alone based on respect for autonomy and self-determination. Unlike PAS and VAE, its implementation requires no change in current law. Caring for the patient dying of PRHN produces no potentially harmful alteration in the physician's role, as could occur by legalizing PAS and VAE. The physician's principal role remains one of palliation.\[14\]

Fears that patients dying of dehydration would suffer terribly are not confirmed by hospice data on the quality of death of dying patients without food or water.\[59\] The terminally-ill patient has an involution of hunger and thirst.\[60\] If a well-meaning but poorly-informed physician attempts to hydrate or nourish such patients, the treatment may increase suffering by producing pulmonary edema, nausea and vomiting.
Physicians have an ethical duty to the patient using PRNH to ensure no suffering by providing adequate analgesia and mouth care. These types of palliative care are provided routinely by experienced hospice nurses. Several studies have been published describing individual cases and series of patients who have died peacefully using PRNH.

Summary

Ethical and legal principles underlie the provision of palliative care. Patients have an ethical and legal right to refuse life-sustaining treatment that is grounded in the doctrine of informed consent. Lawful surrogate decision-makers are authorized to consent for or refuse life-sustaining therapy on behalf of incompetent patients. Patients can employ Advance Directives to help assure that their treatment preferences are followed should they later become incompetent. Although they may feel psychologically different to the physician, there are no important ethical or legal distinctions between withholding and withdrawing treatment once it has been refused by the competent patient or the lawful surrogate decision-maker. The right to refuse life-sustaining therapy encompasses the right to refuse artificial hydration and nutrition. Physicians are not required to provide physician-assisted suicide or voluntary active euthanasia that is requested by patients but should use the opportunity to identify sources of patients' suffering and to enhance their palliative care.

Several areas remain controversial. Should physicians be allowed to cite "medical futility" to justify unilateral discontinuation of life-sustaining therapy? Can the principle of double effect adequately defend the provision of sedation in the imminently dying? Should physician-assisted suicide or voluntary active euthanasia be legalized? Should dying patients be permitted to stop eating and drinking or is that action tantamount to suicide? A uniform consensus on the answers to these questions remains to be developed.

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   Abstract

   Citation

   Full Text

   Full Text

   Abstract

   Full Text


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Role of Communication Skills

Effective symptom control is impossible without effective communication. The most powerful analgesics are of little value if health care professionals do not have an accurate understanding of the patient's pain and other symptoms, and an accurate assessment requires effective communication skills. Almost invariably, communication is a central part of the therapy. Occasionally, it is the only constituent. It often requires greater thought and planning than a drug prescription, and unfortunately, is commonly administered in subtherapeutic doses.

There is no lack of published literature concerning the emotional and psychosocial needs of the dying patient, and the importance of communication as a major component of the delivery of all medical, and, particularly, palliative care (a useful resource is currently available). There is also some published works on the obstacles and deficiencies in communication between the dying patient and the health care professional. There is less general medical literature that provides detailed practical assistance for the palliative care practitioner in improving his or her communication skills. The major objective of this article is to remedy that omission and provide an intelligible and coherent approach to communication in a palliative care setting between health care professionals and their patients.

It is undeniable that in our society any conversation about death and dying is awkward and difficult, and even more so when it occurs between a doctor and patient. Some of that awkwardness is social and originates in the way society views death. Other sources of difficulty originate with the patient and other sources include the health care professional, because professional training, although it prepares physicians to treat sick people, or paradoxically may lead to loss of touch with human skills when the medical treatment of the disease process fails. Improving the techniques in communication may make it easier for the health care professional to overcome or reduce the impact of some of these obstacles.

Communication Skills as Teachable Techniques

In the last two decades, clinicians have become increasingly aware of the need for improved communication skills, but it has been difficult to define and test techniques that can be acquired by practitioners. In the late 1970s and early 1980s, it was widely believed that communication
skills were intuitive, almost inherited, talents (you either have the gift or you don't). This was coupled with the belief that somehow the physician would be able to feel or sense what the patient was experiencing, to divine what the patient wanted, and then be able to respond intuitively in an appropriate way. This belief alienated a large number of health care professionals who found the whole topic (as taught at that time) excessively touchy-feely, intangible and amorphous, with no guidelines that could lead even a highly motivated practitioner to improve his or her skills.

In the last fifteen years, researchers and educators have shown that communication skills can be taught and learned (and retained over years of practice) and that they are acquired skills like any other clinical technique and not inherited or granted as gifts.\[2\] \[11\] \[26\]

The following sections of this article describe two practical protocols that can be used by any health care professional to improve communication for breaking bad news: a basic protocol, the context listening acknowledge strategy summary (CLASS) protocol that underlies all medical interviews, and a variation of that approach, the setting perception invitation knowledge emotions strategy summary (SPIKES) protocol. Both protocols have been published in greater detail elsewhere in textbook\[6\] form and also in fully illustrated form with over 40 different videotaped patient scenarios in CD-ROM and video formats.\[5\] The reader is encouraged to review some of this video material, because written descriptions of communication techniques are limited and of less educational value than practical demonstrations.

**Class: A Practical Protocol for Effective Communication**

Although there are probably many ways of summarizing and simplifying medical interviews, few are practical and easy to remember. The five-step basic protocol for medical communication set out here, bearing the acronym CLASS, is easy to remember and use. Furthermore, it offers a straightforward technique-directed method for dealing with emotions. This is of crucial importance, because a recent study showed that the overwhelming majority (over 85%) of oncologists feel that dealing with emotions is the most difficult part of any clinical interview.\[1\]

In brief, the CLASS protocol identifies the following five main components of the medical interview as essential and crucial: Context (the physical context or setting), Listening skills, Acknowledgement of patient's emotions, Strategy for clinical management and Summary (Display Box 1).

<table>
<thead>
<tr>
<th>The CLASS Protocol</th>
</tr>
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<tbody>
<tr>
<td>C--Physical CONTEXT or setting</td>
</tr>
<tr>
<td>L--LISTENING skills</td>
</tr>
<tr>
<td>A--ACKNOWLEDGE emotions and explore them</td>
</tr>
<tr>
<td>S--Management STRATEGY</td>
</tr>
<tr>
<td>S--SUMMARY and closure</td>
</tr>
</tbody>
</table>

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C: Context (or Setting)

The context of the interview means the physical context or setting, of which there are five major components: arranging the space optimally, body language, eye contact, touch and introductions. A few seconds spent establishing these features of the initial interview may save many minutes of frustration and misunderstanding later (for the health care professional and the patient). These rules are not complex but they are easy to forget during stressful moments.

Spatial Arrangements

Try to ensure privacy for the patient. In a hospital setting, if a private room is not available, the practitioner can draw the curtains around the bed. In an office setting, the practitioner can shut the door. Next, get any physical objects out of the way. Move any bedside tables, trays, or other obstacles away from you and the patient. Ask for the television or radios to be turned off for a few minutes. If you are in an office or room, move your chair so that you are next to the patient, not across the desk. There is evidence that conversations across a corner occur three times more frequently than conversations across the full width of a table.\(^1\) Clear any clutter and papers away from the area of desk nearest to the patient. If you have the patient's chart open, make sure you look up from it and do not talk to the patient while reading the chart. If you find any of these actions awkward, state what you are doing (e.g., "It may be easier for us to talk if I move the table and you turn the television off for a moment").

Then, the most important component of organizing the physical context is sitting down. This is an almost inviolable guideline. It is virtually impossible to assure a patient of a physician's undivided attention and intent to listen seriously if the physician remains standing. Only, if it is impossible to sit, should a physician hold an interview while standing. Anecdotal impressions suggest that when the doctor sits down the patient perceives the period of time spent at the bedside as longer than if the doctor remains standing.\(^1\) Thus, not only does the act of sitting down indicate to the patient that he or she has control and that the physician is there to listen, but it also saves time and increases efficiency. Before starting the interview itself, take care to get the patient organized if necessary. If the patient has just been examined, allow or help him or her to dress and restore a sense of modesty.

It is also important to be seated at a comfortable distance from the patient. This distance (sometimes called the body buffer zone) seems to vary from culture to culture, but a distance of two to three feet between the doctor and patient usually serves the purpose for personal conversation.\(^1\) This is another reason why the doctor who remains standing at the end of the bed appears remote and aloof.

The height at which a doctor sits also can be important; normally, a doctor's eyes should be level approximately with the patient's eyes. If the patient is already upset or angry, a useful technique is to sit so that the doctor is below the patient's eye level, with the doctor's eyes at a lower level. This often decreases the anger. It is best to try to look relaxed, particularly if you do not feel that way.

Make sure that, whenever possible, you are seated closest to the patient and that any friends or relatives are on the other side. Sometimes relatives try to dominate the interview, and it may be important for you to send a clear signal that the patient needs privacy.
In almost all oncologic settings, it is important to have a box of tissues nearby. If the patient or relative begins to cry, it is important to offer tissues, which not only gives overt permission to cry but also allows the person to feel less vulnerable when crying.

**Body Language**

Try to look relaxed and unhurried (despite the fact that you are probably feeling the opposite). To achieve an air of relaxation, sit down comfortably with your feet flat on the floor. Let your shoulders relax. Undo your coat or jacket if you are wearing one, and rest your hands on your knees (often termed in psychotherapy, the neutral position).

**Eye Contact**

Maintain eye contact for most of the time the patient is talking. If the interview becomes intense or emotionally charged, particularly if the patient is crying or is angry, it is helpful to the patient if you look away (to break eye contact) at that point.

**Touching the Patient**

Touch also may be helpful during the interview if a non-threatening area is touched (e.g., hand or forearm), you are comfortable with touch, and the patient appreciates touch and does not withdraw. Most physicians aren't taught specific details of clinical touch during training. Physicians are, therefore, likely to be ill at ease with touching as an interview technique until they gain experience. Nevertheless, there is considerable evidence (although the data are “soft”) that touching the patient (particularly above the patient's waist to avoid misinterpretation) is of benefit during a medical interview. It is likely that touching is comforting during distress and should be encouraged, with the proviso that the health care professional is sensitive to the patient's reactions. If the patient is comforted by the contact, the physician can continue; if the patient is uncomfortable, the physician should stop. Touch can be misinterpreted (as lasciviousness, aggression or dominance) so be aware that touching is an interviewing skill that requires extra self-regulation.

**Commencing the Interview**

Make sure the patient knows who you are and what you do. Many practitioners make a point of shaking the patient's hand at the outset, although this is a matter of personal preference. Often the handshake tells you something about the family dynamics and the patient. Frequently, the patient's spouse also extends his (or her) hand. It is worthwhile to make sure that you shake the patient's hand before the spouse's (even if the spouse is closer) to demonstrate that the patient comes first and the spouse (although an important member of the team) comes second (Display Box 2).

<table>
<thead>
<tr>
<th>The Elements of Physical Context</th>
</tr>
</thead>
<tbody>
<tr>
<td>ARRANGEMENT: Introductions, sitting down, placement of patient, appropriate distance</td>
</tr>
<tr>
<td>BODY LANGUAGE: Drop shoulders, sit comfortably and attentively</td>
</tr>
</tbody>
</table>
**EYE CONTACT:** Maintain eye contact except during anger or crying (not when hot)

**TOUCH (OPTIONAL):** Touch patient's hand or forearm if you and the patient are comfortable with touch

**INTRODUCTIONS:** Tell the patient who you are and what you do

---

**L: Listening Skills**

As dialogue begins, the health care professional should show that he or she is in listening mode. (For a general review of interviewing skills see Lipkin et al.[15] ) The four most essential points are as follows: the use of open questions, facilitation techniques, clarification, and the handling of time and interruptions (Display Box 3).

---

**Fundamental Listening Skills**

Use your listening skills and techniques to show that you are an effective listener.

Four Fundamental Listening Skills:

1. **Open Questions**
   Questions that can be answered in any way (e.g. "how are you/what did that make you feel?")

2. **Facilitating**
   Pausing or silence when patient speaks
   Nodding, smiling, saying "hmm mm/tell me more about that"
   Repetition (i.e., repeating one key word from the patient's last sentence in your first sentence)

3. **Clarifying**
   Making overt any ambiguous or awkward topic

4. **Handling Time and Interruptions**
   With pagers and telephones, acknowledge the patient who is with you as you answer.
   Tell the patient about any time constraints and clarify when the discussion will resume.
Open Questions

Open questions are questions that can be answered in any manner. The questions do not direct the respondent or require him or her to make choices from a specific range of answers. In taking the medical history, most of the questions are, appropriately, closed questions (Do you have any difficulty with fine hand movements? Do you have any numbness or tingling?). In therapeutic dialogue, when the clinician is trying to be part of the patient's support system, open questions are an essential way of finding out what the patient is experiencing as a way of tailoring a support system for the patient. Hence, open questions (What did you think the diagnosis was? How did you feel when you were told that?) are mandatory parts of the non-history therapeutic dialogue.

Facilitation Techniques

Silence

The first and most important technique in facilitating dialogue between a patient and clinician is silence. If the patient is speaking, don't talk over him or her. Wait for the patient to stop speaking before you start your next sentence. This, the simplest rule, is the rule most often ignored and is most likely to give the patient the impression that the doctor is not listening.

Silences also have other meanings. They can, and often do reveal the patient's state of mind. Often, patients fall silent when they have feelings too intense to express in words. A silence, therefore, means that the patient is thinking or feeling something important, not that he or she has stopped thinking. If the clinician can tolerate a pause or silence, the patient may express the thought verbally later.

If you need to break the silence, a helpful way to do so is ask: "What were you thinking about just then?" or "What is making you pause?" or similar questions.

Other Simple Facilitation Techniques

Once you have encouraged the patient to speak, it is important to demonstrate that you are hearing what is being said. Hence, in addition to silence, dialogue may be facilitated by using any or all these facilitation techniques: nodding, pausing, smiling, using responses such as "yes," "mmm hmm," tell me more. In addition to these responses, it is valuable to use repetition as a conscious and deliberate facilitation technique. To demonstrate that you are really hearing what the patient is saying, employ one or two key words from the patient’s last sentence in your first sentence. ("I just feel so lousy most of the time." "Tell me what you mean by feeling lousy."). Reiteration means repeating what the patient has told you but in your words, not the patient's words. ("Since I started taking those new tablets, I've been feeling sleepy." "So, you're getting drowsy from the new tablets?") Repetition and reiteration confirm to the patient that you have heard what has been said.

Clarifying

As the patient talks, it is tempting for the clinician to go along with what the patient is saying, even when the exact meaning or implication is unclear. This may quickly lead to serious obstacles in the dialogue; hence, it is important to be honest when you do not understand what the patient means. Several different phrases can be used ("I'm sorry. I'm not quite sure what you meant when you said..." "When you say...do you mean that...?"). Clarification gives the patient
an opportunity to expand on the previous statement or to emphasize some aspect of the statement when the clinician shows interest in the topic.

**Handling Time and Interruptions**

Clinicians have poor reputations for handling interruptions by the telephone, pager or other people. Often the clinician may appear to abruptly ignore the patient and respond immediately to the telephone, pager or a colleague. This frequently is interpreted as a snub or an insult to the patient. If it is not possible to hold all calls or turn off the pager, then it is at least worthwhile to indicate to the patient that you are sorry about the interruption and will resume the interview shortly. ("Sorry. This is another doctor I must speak to briefly. I'll be back in a moment." "This is something quite urgent about another patient. I won't be more than a few minutes.") The same is true of time constraints ("I'm afraid I have to go to the emergency room now, but this is an important conversation. We need to continue this tomorrow morning.")

**A: Acknowledgment (and Exploration) of Emotions**

**Empathic Response**

The empathic response is a useful technique in an emotionally charged interview and is frequently misunderstood by students and trainees.

The empathic response has nothing to do with a physician's personal feelings. If the patient feels sad, the physician is not required at that moment to feel sad. It is simply a technique of acknowledgment, showing the patient that you have observed the emotion he or she is experiencing. It consists of the following three steps:

1. Identifying the emotion the patient is experiencing;
2. Identifying the origin of that emotion;
3. Responding in a way that tells the patient that you have made the connection between steps 1 and 2.

Often the most effective empathic responses follow the format of "You seem to be...." or "It must be...." (e.g., "It must be distressing for you to know that all that therapy didn't give you a long remission," or "This must be awful for you.") The objective of the empathic response is to demonstrate that the physician identifies and acknowledges the emotion the patient is experiencing, and by doing so the physician is legitimizing it as an item on the patient's agenda. In fact, if the patient is experiencing a strong emotion (e.g., rage or crying) a physician must acknowledge the existence of that emotion or all further attempts at communication will fail. If strong emotions are not acknowledged in some way, the physician will be perceived as insensitive and this will render the rest of the interaction useless.

The empathic response is a physician's acknowledgement of what the other person is experiencing. It has nothing to do with a personal view (or judgement) of the situation nor how the physician herself would react if facing these circumstances. A physician does not have to feel the same emotion that the patient is experiencing or agree with their viewpoint. The physician simply is observing what the other person is feeling and bringing that emotion into the dialogue between the two (Display Box 4).
Acknowledgment of Emotions: The Empathic Response

Acknowledging the emotional content of the interview is the fundamental skill to being sensitive and supportive.

The Empathic Response:

1. Identify the emotion;
2. Identify the source of the emotion; and
3. Respond in a way that shows you have made the connection between steps 1 and 2 (e.g., "that must have felt awful/this information has obviously come as quite a shock")

The empathic response is a technique or skill, not a feeling. It is not necessary for the physician to experience the same feelings as the patient or agree with the patient's view or assessment.

S: Management Strategy

There are several useful techniques to ensure that a physician constructs a management plan the patient will follow. The following are useful guidelines:

1. Determine the optimal medical strategy. In your mind (or aloud), define the ideal management plan.

2. Assess (in your own mind or by asking the patient) what are his or her expectations of condition, treatment, and outcome (summarize this in your mind or clarify and summarize aloud if needed). Be wary if there is a marked mismatch between the patient's view of the situation and the medical facts. You will need to work harder to make the plan appear logical and acceptable to the patient if there is a marked discordance between the patient's view and reality. Propose a strategy, bearing in mind your conclusions from steps 1 and 2.

3. Assess patient's response (e.g., make note of what stage of forming an action plan the patient is in [e.g., the stages are often defined as the precontemplation, contemplation, implementation, or reinforcement phases]). Acknowledge the patient's emotions as they occur and continue in a contractual fashion to arrive at a plan that the patient has bought into and will follow (Display Box 5).

Strategy Management

A reasonable management plan that the patient understands and will follow is better than an ideal
plan that the patient will ignore.

Think what is best medically, then--

- Assess the patient's expectations of condition, treatment, and outcome (summarize this in your mind or clarify and summarize aloud if needed)

- Propose a strategy

- Assess the patient's response (e.g., what stage of action is the patient in: the precontemplation, contemplation, implementation or reinforcement phase)

- Agree on a plan (as detailed as possible)

S: Summary

The summary is the closure of the interview. In gynecologic oncology, the relationship with the patient is likely a continuing one and a major component of the patient's treatment. The closure of the interview is an important time to emphasize that point.

It is relatively straightforward to cover three areas in the summary (Display Box 6).

### Summary and Closure

Ending the interview has three main components:

1. A precis or summary of the main topics you have discussed;

2. "Any important issues or questions that we should be discussing?" (Even if the physician does not have time to discuss questions in this interview, they can be on the agenda for the next);

3. A clear contract for the next contact.

The areas are a precis or reiteration of the main points covered in the dialogue, an invitation to the patient to ask questions, and a clear arrangement for the next interaction (a clear contract for the next contact). This particular part of the interview is not necessarily long but does require considerable focus and concentration.

### Spikes: A Variation of Class for Breaking Bad News

Among the various types of medical interviews, breaking bad news is special and important for both parties in the clinician-patient relationship. Reviews, chapters, and a textbook have been published on this subject. In other words, the degree or intensity of bad news is the gap between the patient's expectations of the future and the medical reality. In gynecologic oncology, bad news is common during many stages in a patient's history: at the time of initial diagnosis, recurrence, disease...
progression and failure of therapy, clinical deterioration, new complications, treatment of related effects, and change from therapeutic to palliative care. It is necessary to establish a protocol that will function in all of these circumstances.

The SPIKES protocol has been designed specifically for these purposes and will allow the physician to assess the patient's expectations before going on to share the information (Display Box 7).

<table>
<thead>
<tr>
<th>The SPIKES Protocol for Breaking Bad News: A Variant of the Basic CLASS Approach</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>S:</strong> SETTING (= CONTEXT + LISTENING SKILLS)</td>
</tr>
<tr>
<td><strong>P:</strong> Patient's PERCEPTION of condition and seriousness</td>
</tr>
<tr>
<td><strong>I:</strong> INVITATION from patient to give information</td>
</tr>
<tr>
<td><strong>K:</strong> KNOWLEDGE--explaining medical facts</td>
</tr>
<tr>
<td><strong>E:</strong> EXPLORE EMOTIONS AND EMPATHIZE as patient responds</td>
</tr>
<tr>
<td><strong>S:</strong> STRATEGY AND SUMMARY</td>
</tr>
</tbody>
</table>

**S: Setting (= Context + Listening Skills)**

In the SPIKES protocol, for the sake of simplification, the first two steps of the CLASS protocol, context and listening skills (see above) have been combined into one step, setting.

**P: Patient's Perception of the Situation**

A useful and important principle of breaking bad news is to find out what the patient already knows or suspects before continuing to disclose information ("Before you tell, ask") (Display Box 8).

<table>
<thead>
<tr>
<th>Patient's Perception of Condition</th>
</tr>
</thead>
<tbody>
<tr>
<td>Ask patient to say what he or she knows or suspects about the current medical problem (e.g., &quot;What did you think when...&quot; or, “Did you think it might be serious&quot;).</td>
</tr>
<tr>
<td>As patient replies, listen to the level of comprehension and vocabulary.</td>
</tr>
<tr>
<td>Note any mismatch between the actual medical information and the patient's perception of it (including denial).</td>
</tr>
</tbody>
</table>

The wording a physician uses to find out how much the patient already understands is a personal choice. ("Before I go on to tell you about the results, why don't you tell me what you've been thinking?" "When you first developed unsteadiness while walking, what did you think was going
on?" "Did you think this was something serious?" or "What did the referring medical team tell you about your medical condition?"

As the patient replies, pay particular attention to his or her vocabulary and comprehension of the subject. When a physician starts giving information, it is helpful to start at the patient's level of knowledge.

I: Getting a Clear Invitation to Share News

Next, try to get a clear invitation from the patient to share the information. Studies show the majority of patients want full disclosure. There has been a steady increase in the proportion of physicians who give honest information since Oken's study\cite{22} in 1961 that showed that 95% of surgeons did not disclose to patients a cancer diagnosis. Eighteen years later, Novack's study\cite{21} showed a dramatic reversal of this proportion. The proportion of the patients who wanted to be informed, according to the Jones' study\cite{13} in 1981 showed that 50% of (British) patients wanted to know. Since then studies put the proportion of patients who want to know at above 90\%\cite{18} (see the excellent review by Northouse\cite{20}) (Display Box 9).

<table>
<thead>
<tr>
<th>Invitation from the Patient to Share Information</th>
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<tbody>
<tr>
<td>Find out from the patient if he or she wants to know the details of the medical condition or treatment (e.g., &quot;Are you the sort of person who...?&quot;).</td>
</tr>
<tr>
<td>Accept the patient's right not to know (but offer to answer questions if the patient wishes later).</td>
</tr>
</tbody>
</table>

Concealing information or lying to the patient is likely to be unsatisfactory. The phrase used to obtain a clear invitation is again a matter of personal choice and judgment ("Are you the sort of person who'd like to know exactly what's going on?" "Would you like me to go on and tell you exactly what the situation is and what we recommend?" or "How would you like me to handle this information? Would you like to know exactly what's going on?").

K: Knowledge (Explaining the Medical Facts)

Having obtained a clear invitation to share information, it is time to explain the medical facts and be aware of (and sensitive to) the patient's reaction to that information. In other words, sharing knowledge and responding to emotions occur simultaneously.

The most important guidelines for sharing medical facts are the following (Display Box 10):

<table>
<thead>
<tr>
<th>Knowledge: Explaining the Medical Facts</th>
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<tbody>
<tr>
<td>Bring the patient towards a comprehension of the medical situation, filling in any gaps</td>
</tr>
<tr>
<td>Aligning: Use language intelligible to the patient, and start at the level he or she used</td>
</tr>
</tbody>
</table>
Appendix C: Ethics, Communication and Decision Making

Explain information gradually.

Check the reception: Confirm that the patient understands what you are saying after each significant chunk.

Respond to the patient's reactions as they occur.

Explore denial (if present) using empathic responses (e.g., "It must be very difficult for you to accept the situation").

- Start at the patient's level of comprehension and use vocabulary that was used when the patient told you what he or she understood about the situation (This is called aligning).
- Use layman's English, avoiding the technical jargon of the medical profession (Medspeak).
- Explain the facts gradually, and never talk for more than a few minutes at a time.
- Check that the patient understands what you've said before going further (use phrases such as "Do you follow what I'm saying?" "Is this clear so far?" "Am I making sense so far?")
- Use a narrative approach to make sense of what has occurred. Explain the sequence of events and how the situation seemed as events unfolded ("When you became short of breath we didn't know whether it was just a chest infection or something more serious. So that's when we performed the chest radiograph.")
- Respond to all emotions expressed by the patient as they arise.

E: Emotions (Exploration and Empathic Response)

The acknowledgment of emotions is even more important in an interview about bad news than it is in most other interviews.

As in the acknowledgment step of the CLASS protocol, it is important to address the emotions expressed in the interview. It is also worth pointing out that the empathic response can be used just as effectively to acknowledge a physician's feelings if they are becoming intense. ("I'm finding this upsetting, too." "I'm finding this difficult to tell you, but I must explain that the latest MR image showed...").

The value of all empathic responses is that you are making an observation that is almost unemotional in itself, about an issue that is heavily charged with emotion (whether the patient's or physician's emotions). This is why an empathic response calms a stressful moment and facilitates exploration of the situation without causing more anxiety.

S: Strategy and Summary

Close the interview with a management strategy and a summary as in the strategy and summary steps in the CLASS protocol above.
Dealing with Hope and False Hopes

A frequently heard response from patients and clinicians alike is "You can't take away hope." Frequently, this line is used by clinicians as an excuse for not telling the patient the truth. Often the rationale behind this has more to do with protecting the clinician from discomfort than protecting the patient.

Using the above protocol, it is possible to give important information in a tactful manner, supporting the patient (relying on acknowledgment and exploration of the patient's feelings), and avoiding concealment or dishonesty.

Clinicians are more likely to create trouble for themselves if they promise cure when it is not possible or express unrealistic hopes. Supporting the patient and reinforcing realistic hopes are parts of the foundation of a therapeutic relationship. The central issue is not whether to tell the truth or not (physicians have moral, ethical and legal obligations to do so if that is what the patient wants) but how the truth is told. Insensitive and ineffective truth telling may be just as damaging and counter-productive as insensitive lying. In practice, the protocols discussed here will enable a physician to tell the truth at a patient's pace and in a way that will enable the physician to recruit and reinforce instead of diminishing the patient's coping strategies to face the situation.

Summary

This review has been brief, setting out the main guidelines and hints and tips in point-form only. There is a great amount of valuable practical material already published.

Like any clinical intervention, effective communication requires motivation to be successful. If a physician is motivated to be a good clinical communicator, in addition to his or her other skills, effective is achievable. Some of it depends on having a basic strategy for the task, and the protocols presented here should help. The rest is largely a matter of being aware of the effect of what a physician says and does on the patient and family. To some extent, there is a great deal of courtesy and common sense mixed in with the specific strategies.

Finally, clinicians should be aware that communication tasks are important in the relationship with a patient. As one family member once said: "Doctors doing this job badly will never be forgiven; doctors doing it well will never be forgotten."

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Neurology, Pain, and Palliative Medicine: A 2002 Working Road Map

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In the last quarter of the 20th century neurologists evolved from diagnosticians to providers of therapeutic approaches to neurologic disease. Despite extraordinary advances in many areas of neurologic practice, most patients cared for by clinical neurologists do not have curable disorders. Competent neurologic practice is therefore all about improving the quality of life of neurologically compromised patients. In the evolving field of palliative medicine, with best practice and evidence-based guidelines, neurologists need educational resources to incorporate state-of-the-art symptom control and supportive therapies into their everyday care of patients. To date, the scope of neurologic education has not included detailed coverage of pain management, symptom control and competent care of the dying patient and family. In the past decade, several steps have been taken to assure that neurologists have the tools to manage pain and other symptoms effectively from the time of diagnosis to the time of death.

The American Academy of Neurology (AAN) Ethics and Humanities Subcommittee has published a series of position statements highlighting the responsibilities of neurologists in caring for chronically ill, critically ill and dying patients. These documents are helping to define competency in caring for patients with advanced diseases of the nervous system (see Addendum).

In an AAN-sponsored survey of Academy members in 1996, a sizable gap was identified between established legal, medical and ethical guidelines for the care of patients with advanced neurologic disease and the beliefs and practices of those surveyed. For example, 37% of neurologists surveyed thought it illegal to administer analgesics in doses that risk respiratory depression in dying patients with amyotrophic lateral sclerosis (ALS), and 40% believed they should obtain legal counsel when considering withdrawing life-sustaining treatment. (1) In a survey of practicing AAN-affiliated neurologists only 30% felt adequately trained to diagnose and 20% to treat pain disorders. Ninety-one percent believed that practicing neurologists need more pain education. (2) An examination of the end-of-life care content among fifty major medical textbooks, including the leading neurology texts, demonstrated that the neurology textbooks ranked in the lowest quartile in end-of-life coverage. (3) Bernat et al. has cited the lack of data on the physiology of death, role models of mature physicians with expertise in palliative care and reimbursement of physicians who care for the dying as critical barriers toward the delivery of competent end-of-life care for patients dying of neurologic disease. (4)

The Report of the Quality Standards Subcommittee of the American Academy of Neurology on the care of the patient with ALS outlines a rigorous scientific approach toward patient care, and serves as a model for the development of guidelines and scientifically valid measurement tools in other areas of neurologic practice. (5) In a retrospective study of patients with ALS, Oliver
reported dyspnea (88%), dysphagia (88%), pain (76%), weight loss (68%) and dysarthria (60%) as some of the most common symptoms experienced by patients at or near the end of life. (6) Voltz et al., have reported similar percentages underscoring the need for all neurologists who care for patients with ALS to be competent practitioners of palliative medicine.(7)

The Residency Review Committee in conjunction with the Accreditation Council of Graduate Medical Education requires that, “The resident must receive instruction in appropriate and compassionate methods of end-of-life palliative care, including pain relief and psychosocial support and counseling for patients and family members about these issues.”(8) To help U.S. neurology residency programs meet this accreditation standard, the Graduate Education Subcommittee of the AAN recently unanimously endorsed a proposal by Drs. David Weissman and Alan Carver to integrate end-of-life care into neurology residency training. During the 2001-2002 academic year, 30 neurology residency programs will send representatives to a seminar on palliative care training in neurology and learn how to enhance the teaching and delivery of end-of-life care in their respective programs and institutions.

Other initiatives include the development of an ALS Workgroup that, with the support of Promoting Excellence in End-of-Life Care, is creating guidelines and practical assessment tools for neurologists who care for patients with ALS and other neuromuscular diseases. An International Working Group in Palliative Care in Neurology has formed and is developing a core curriculum and textbooks for neurologists. New texts such as Palliative Care in Amyotrophic Lateral Sclerosis,(9) Hospice Care for Patients with Advanced Progressive Dementia,(10) and the November 2001 volume of Neurologic Clinics devoted to Palliative Care (11) are available to educate health care professionals and specifically neurologists in a comprehensive approach for patients with advanced, incurable neurologic diseases. The development of the EPEC (Education for Physicians on End-of-Life Care) curriculum (12) and establishment of a yearly half-day course at the annual meeting of the American Academy of Neurology offer neurologists the opportunity to assure that their clinical competency includes up to date knowledge of pain and palliative medicine.

Addendum: AAN Position Statements Related to End-of-Life Care


References

Appendix C  Ethics, Communication and Decision Making


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OBJECTIVE: The American Academy of Neurology (AAN) surveyed the attitudes, behavior, and knowledge of its members regarding care at the end of life. Three groups of AAN members were surveyed: neuro-oncologists, ALS specialists, and a representative sample of U.S. neurologists. METHODS: The survey presented two clinical scenarios involving end-of-life care. Neurologists were asked a series of questions to assess their knowledge of existing medical, ethical and legal guidelines; their willingness to participate in physician-assisted suicide (PAS) or carry out voluntary euthanasia (VE); and their general attitudes regarding end-of-life care. RESULTS: Neurologists support a patient's right to refuse life-sustaining treatment, but many believe that they are killing their patients in supporting such refusals. Thirty-seven percent think it is illegal to administer analgesics in doses that risk respiratory depression to the point of death. Forty percent believe they should obtain legal counsel when considering stopping life-sustaining treatment. One half believe that PAS should be made explicitly legal by statute for terminally ill patients. Under current law, 13% would participate in PAS and 4% would carry out VE; if those procedures were legalized, 44% would participate in PAS and 28% in VE. Approximately one third believe that physicians have the same ethical duty to honor a terminally ill patient's request for PAS as they do to honor such a patient's refusal of life-sustaining therapy. CONCLUSIONS: There is a gap between established medical, legal and ethical guidelines for the care of dying patients and the beliefs and practices of many neurologists, suggesting a need for graduate and postgraduate education programs in the principles and practices of palliative care medicine. Many neurologists would participate in PAS and carry out VE if legalized.
Patients' Assessment of Quality of Life Instruments: A Randomised Study of SIP, SF-36 and SEIQoL-DW in Patients with Amyotrophic Lateral Sclerosis.

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The evaluation of quality of life (QoL) plays an increasingly important role in clinical research and drug trials in ALS. However, most of the scales employed so far are based on a fixed external value system, and may therefore not reflect the patients' subjective perception of QoL accurately. In addition, many ALS patients complain about the psychological distress inflicted by QoL questionnaires which focus on functional status, as they constantly remind patients of their deterioration. We therefore asked 42 ALS patients to assess, using visual analogue scales, their subjective perception of the validity of three QoL instruments as well as the emotional distress caused by them. The scales were: the Sickness Impact Profile (SIP), the Short Form 36 (SF-36), and the Schedule for the Evaluation of Individual QoL-Direct Weighting (SEIQoL-DW). Patients were examined at least three times at two-month intervals. The SIP was filled out by all patients, the SF-36 and the SEIQoL-DW were assigned at random. The validity of the SEIQoL-DW was rated higher than that of the SIP (p<0.001) and of the SF-36 (p<0.001). The SIP imparted a higher emotional distress to patients than the SEIQoL-DW (p=0.005), with a trend in the same direction for the SF-36 (p=0.082). The most frequently mentioned QoL-relevant domains in the SEIQoL-DW were family (100%), health (53%), and profession (50%). These results should prompt further discussion and investigation on the most appropriate way to assess QoL in patients with ALS.

Publication Types:
- Clinical Trial
- Randomized Controlled Trial
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