

ALS

amyotrophic lateral sclerosis



Completing the Continuum of ALS Care: A Consensus Document

PROMOTING EXCELLENCE IN END-OF-LIFE CARE
A NATIONAL PROGRAM OF
THE ROBERT WOOD JOHNSON FOUNDATION



Completing the Picture of Excellence



In 1997, The Robert Wood Johnson Foundation launched a national program *Promoting Excellence in End-of-Life Care* with a mission of improving care and quality of life for dying Americans and their families. We soon realized that the metaphor of a jigsaw puzzle seemed apt in describing our efforts to expand access to services and improve quality of care in a wide range of settings and with diverse populations. No single approach would suffice - a variety of strategies, models of care and stakeholders are necessary to successfully complete the picture. This monograph represents one aspect of our work and one piece of the puzzle of ensuring that the highest quality of care, including palliative care, is available to all seriously ill patients and their families.

Acknowledgements

This publication was produced by *Promoting Excellence in End-of-Life Care* a national program of The Robert Wood Johnson Foundation, directed by Ira Byock, M.D. Primary authors of this report are:

- Hiroshi Mitsumoto, M.D., D.Med.Sc., ALS Peer Workgroup Chair
- The ALS Peer Workgroup Members

We extend our thanks to:

- The ALS Association for managing the Workgroup, under the direction of Mary Lyon;
- Lisa Bardach, M.S., C.C.C.-S.L.P., for her contributions to the report;
- Barbara L. Massey for assistance with the copyrights;
- Julie Emnett for organizing Workgroup meetings and facilitating group process;
- Starr Pearlman for assisting the Workgroup in writing the report and initial editing;
- Nancy Nelson for assistance editing the report; and
- Blair Pfister, Keila Szpaller and Karyn Collins of the *Promoting Excellence* staff for copy-editing.

About the Artist

As Graphic Artist Manager at The University of Montana Printing & Graphic Services in Missoula for the last 18 years, Neal Wiegert has received awards and recognition for his work. He frequently designs and illustrates for Montana Public Radio, The Maureen and Mike Mansfield Center, UM Intercollegiate Athletics and the UM Alumni Association among many others.

All illustrations © by Neal Wiegert, The University of Montana Printing & Graphic Services, 2004

January 2004

Workgroup Findings and Recommendations to the Field

Table of Contents

2	EXECUTIVE SUMMARY	12	Introduction Defining ALS Issues in End-of-Life Care
3	Introduction		
3	Methods	14	Methodology Establishing the ALS Workgroup and Defining the Aims of the Project The Process Conceptual Considerations in Defining End of Life Resources and Defining Optimal Care
4	Workgroup Findings and Recommendations to the Field Defining End of Life Physicians' Perception of Own Mortality	15	Workgroup Findings and Recommendations to the Field Psychosocial Care Bereavement Spirituality Quality of Life Caregiver Issues Ethics Communication and Decision Making Access to Care Cost of Care Education
4	Psychosocial Care		
4	Bereavement		
5	Spirituality		
5	Quality of Life		
5	Caregiver Issues		
6	Ethics		
7	Communications and Decision Making	26	Symptom Management at the End of Life Respiratory Care Nutrition and Hydration Speech and Communication Depression and Pseudobulbar Affect Insomnia Pain Last Hours
8	Access to Care		
9	Cost of Care		
10	Education	37	REFERENCES
12	REPORT TO THE FIELD	38	ALS PEER WORKGROUP MEMBERS
12	Explanation of the Peer Workgroup Statement of Need The Convening Process	40	APPENDICES (On accompanying CD)

Executive Summary





Introduction

Evidence-based practice parameters in amyotrophic lateral sclerosis (ALS) provide clear management direction on caring for patients during disease progression. However, management direction is also needed specifically for end-of-life care for patients with ALS. Little evidence is available in the published literature identifying optimal management approaches for the dying patient with ALS. Therapeutic direction has been adapted from pain management and other disease states, such as cancer.

Promoting Excellence in End-of-Life Care is a national program of The Robert Wood Johnson Foundation charged with the task of improving the end of life for dying patients. The *Promoting Excellence* program office recognized ALS as a disease that requires special attention at the end of life. Working with The ALS Association (ALSA), an ALS Peer Workgroup was established with the charge of identifying the current state of end-of-life care for ALS patients and establishing recommendations on how to improve the end of life for patients and their families.

Methods

The ALS Peer Workgroup identified topics and issues specific to the end of life in ALS: general psychosocial care, bereavement, spirituality, quality of life, caregiver issues, communication skills, ethics, decision making, symptom management, access to care, costs of care and knowledge/education. For each topic, the Workgroup identified the current state of knowledge, determined what constitutes ideal care and provided recommendations on how to close the gap between current and ideal care. A systematic review of the literature was conducted in order to identify the available evidence for these issues. This evidence review was used as the foundation to define the current state of care, and consensus was used to determine the gaps between current care and optimal care. Recommendations to the field were developed, addressing practice, research, program development and health care policy. These recommendations are based on published evidence and consensus.

Workgroup Findings and Recommendations to the Field

The results of the literature search demonstrated that there were very few studies that specifically address the end of life in ALS patients. It is important to point out that death is at the end of a continuum of care in ALS, and therefore, end-of-life care is heavily influenced and depends on the care provided from the earlier stages of ALS. For this reason, some recommendations are directed at caring for the patient at earlier stages in the disease. The Workgroup used the "Practice Parameter: The Care of the Patient with Amyotrophic Lateral Sclerosis (An Evidence-Based Review)" as the foundation for generating specific recommendations that focus on topics and issues that influence the end of life in ALS (Miller et al., 1999).

Defining End of Life

The Workgroup agreed that palliative care begins at the time of diagnosis for patients with ALS. Initiating end-of-life discussions is difficult for many physicians and health care providers depending on their own experience and philosophy. When to introduce end-of-life discussions with patients and their families is also uncertain. The Workgroup developed six triggers for identifying when end-of-life discussions should be introduced (See sidebar on page 7). Important factors that determine when the patient and family are ready to discuss end-of-life issues include coping skills, depression and anxiety, cultural issues, use of functional assistive devices and physiologic status.

Physicians' Perception of Own Mortality

To engage in end-of-life discussions, physicians need to come to terms with the limitations of medical therapies and to develop an attitude of unconditional acceptance and support for the patient. This requires that health care providers address their personal attitudes toward death and dying, come to terms with their own mortality and take the necessary steps to work through their own personal issues surrounding death and dying.

Psychosocial Care

Current clinical practice focuses predominantly on the patient's medical needs. However, emerging evidence suggests that psychosocial factors significantly influence decisions regarding health care choices, palliative care and attitudes about living and dying with ALS. The analysis of gaps in this area of care indicates that there is a lack of focus, and in some cases, a lack of awareness regarding the importance of addressing psychosocial concerns in patients with ALS. Many physicians feel uncomfortable in approaching patients to offer psychosocial support, and others do not know what type of psychosocial care is needed.

Practice Recommendations

- *Increase awareness of importance for psychosocial care in patients with ALS and their caregivers.*
- *Implement an interdisciplinary team approach to care in ALS that regularly includes a psychosocial evaluation.*

Research and Program Development Recommendations

- *Develop specific training materials to facilitate an interdisciplinary approach to care.*
- *Develop specific training materials to facilitate the goal of increased awareness for psychosocial care (such as vignettes, curricula, care plans or guidelines).*
- *Develop curricula on psychosocial care (including cross-cultural interreligious issues).*

Bereavement

Bereavement in ALS has unique features because both the patient and the family grieve. The patient grieves over the lost function. The caregiver grieves over their loss of independence as the disease progresses. The family grieves in the traditional manner at the time of death. Although there are many resources on bereavement for other diseases, there is limited information about the specific bereavement process in patients with ALS and their caregivers.

Practice Recommendations

- *Offer bereavement support, informally or formally, for the patient and caregiver during the clinic visit.*

Meeting this challenge requires that health care providers review their own attitudes toward death and life, come to terms with their own mortality and take the necessary steps to address their own personal issues surrounding death and dying.

- Offer bereavement support for the caregiver after the death of the patient, including letters of condolence at the time of death.

- Examine spiritual needs of different patient populations (e.g., ALS versus cancer) and different ethnic/religious backgrounds to improve specific spiritual care for different populations of patients.

Research Recommendations

- Determine the scope of bereavement in ALS and whether there are differences between ALS and other diseases, such as cancer.
- Assess whether specialized ALS support groups are needed, and if they confer specific benefits.

Spirituality

Recent reports suggest that addressing spiritual issues is beneficial for terminally ill patients and their caregivers.

Practice Recommendations

- Address the spiritual needs of patients.
- Establish an interdisciplinary team to enhance collaboration with chaplains, spiritual counselors and pastoral workers. Assess the spiritual needs of patients, caregivers and family members repeatedly as the disease progresses.
- Recognize and accept the patient's spiritual and religious attitudes. Educate health care providers regarding cross-cultural religious differences in the approach to death and dying with the intent of improving management of end-of-life issues.

Research Recommendations

- Improve methods and tools for assessing the spiritual needs of patients and caregivers.
- Study the impact of managing spiritual needs on quality of life and longevity of patients, and on the patient-physician relationship.

Quality of Life

Assessing quality of life and psychosocial status and taking steps to improve them have been elusive goals at the end of life. Physicians need a better understanding of the factors that influence quality of life in patients with ALS and their caregivers.

Practice Recommendation

- Use quality-of-life instruments in patients with advanced ALS during the end of life to help detect issues that should be addressed in order to improve the end of life (e.g., McGill QOL or Seiqol-DW patient generated measure of individual quality of life with high acceptance in ALS).

Research Recommendation

- Develop instruments designed to assess quality of life at the end of life. This should be done by considering the following:
 - The physical and psychological comfort of the patient when completing an instrument (eliminate instruments that trigger psychological distress);
 - The time needed to complete the instrument; and
 - The ease of administration to patients with impaired communication abilities.

Caregiver Issues

Patients with debilitating chronic diseases are increasingly cared for at home. According to the North American ALS CARE Database, 63 percent of patients die at home, and thus, the family becomes the principal caregiver. Recent data indicate that caregivers

endure mental distress from caring for patients. Patients and caregivers have high concordance in distress and depression. There are no studies that assess the risk of distress, the burden of caregiving or the support caregivers may require.

Practice Recommendations

- Interview experienced caregivers to identify psychosocial issues that must be addressed.
- Establish support services for caregivers including psychiatric guidance and counseling, support groups that emphasize educational initiatives, hands-on training programs and wellness-promoting behaviors.
- Develop a crisis management system for caregivers in ALS clinics, including an ALS caregiver telephone hotline available 24 hours per day.
- Make spiritual education training and intensive psychosocial support readily available (including home visits), and offer these services to all caregivers during the end-of-life period.

Research Recommendations

- Study whether proactive intensive psychosocial care reduces caregiver burden and distress, enhances caregiver wellness and improves the quality of life of patients.
- Develop an instrument that can quantify caregiver burden and distress.
- Study the medical economics of family caregiving, and expand the evidence base for supporting insurance reimbursement for family caregiver costs.

Policy Recommendations

- Reimburse the cost incurred by physicians who provide supportive care for caregivers.
- Using organized lobbying approaches, improve financial support and reimbursement for the costs assumed in caring for patients with ALS during the end of life (e.g., Medicare reimbursement).
- Improve financial/insurance coverage for psychosocial intervention in the home.
- Encourage hospices to provide universal social work coverage for all families (current policies allow nursing support to families, but many hospices assign social work coverage only to those families deemed at particular risk and do not provide counseling to the majority of caregivers).

Ethics

The ethical basis and legal status of most end-of-life decisions pertinent to ALS have been established. Accepting, forgoing or withdrawing life-sustaining interventions are actions common in ALS, but the decision making involves consideration of values and beliefs that may not have been previously explored by those affected by the disease, including the physicians. Moreover, debates concerning euthanasia and physician-assisted suicide have prominently featured patients with ALS. Thus, ethical issues in caring for patients with ALS are sensitive; many physicians are not comfortable addressing end-of-life issues because of the perceived ethical complexity. One of the most difficult issues surrounding the end of life is physician-assisted suicide. Some physicians, however, have difficulty differentiating accepted clinical practices from physician-assisted suicide and euthanasia. In avoiding topics of euthanasia and physician-assisted suicide, physicians may be avoiding appropriate discussions of good end-of-life care and consequently not providing appropriate palliative care. A gap exists between available guidelines and clinical practice since guidelines are not uniformly implemented across all areas of palliative care in ALS.

Practice Recommendations

- Implement evidence-based guidelines for terminal care in patients with ALS (Miller et al., 1999).
- Identify and monitor conflicting issues associated with end-of-life care, including conflicts among family members and with health care providers. Intervene early to resolve conflicts and use counseling services, among other support systems, so end-of-life care is agreed upon and a plan is established in advance. Clarify ethical and legal issues with an ethics committee or legal counsel, as necessary.
- Use questions regarding physician-assisted suicide and euthanasia as a trigger to discuss end-of-life care. Health practitioners need to provide explicit assurances of continuity of care and commitment to relieve suffering. This may be one of the most direct and first triggers to discussing end-of-life care. Physician-assisted suicide is not legal in 49 states; clinicians should not feel coerced to provide a lethal prescription.

Research Recommendations

- Investigate the prevalence of the desire to end life, and evaluate interventions that obviate this desire.

- *Examine the impact (outcomes) of the patient's end-of-life decisions on the overall quality of life during the terminal phase of the patient's illness, as well as the impact on the family and caregivers.*
- *Develop a neurology Educating Physicians in End-of-Life Care (EPEC) module and assess the impact of its use on clinicians' knowledge and practice in end-of-life care of ALS.*

Communications and Decision Making

Patient autonomy in end-of-life decisions is the accepted Western paradigm. Although patients or their surrogates have the right to refuse or legally withdraw life-sustaining interventions, many are unaware of their rights. Physicians must skillfully facilitate such communications about decisions regarding life-sustaining interventions. Few studies are available that evaluate clinical practices regarding discussions of sensitive issues such as sedation, pain relief and withdrawal of life support, let alone the impact of these decisions on ALS. Despite the importance of good communication, physician education and training in these skills are largely neglected.

Practice Recommendations

- *Improve communication skills to effectively discuss end-of-life issues with patients with ALS and family members. Identify the physician, or team member, who will take the leadership role in end-of-life discussions, and who will, in turn, communicate the decisions to the other health care providers. This may be done by confidential correspondence in notes in the health records (patient's chart). Health care providers should be prepared to support the patient's decisions, regularly review the patient's decisions and update the patient's chart accordingly.*
- *Establish a partnership between the patient and interdisciplinary team members in the treatment plan during disease progression. This relationship needs to be established prior to initiation of end-of-life discussion. The triggers to end-of-life discussions outlined in the corresponding sidebar represent the most overt prompts to planning care. From the time of diagnosis, clinicians should provide opportunities to discuss all types of care alternatives. Respiratory issues are central to end-of-life decisions. The reasons behind monitoring pulmonary function should be explicitly stated and all options for respiratory care, including symptom management alone, should be clearly explained.*

Six Triggers for Initiating Discussion about End-of-Life Issues

- The patient or family asks—or “opens the door”—for end-of-life information and/or interventions (elicited or spontaneous, verbal or non-verbal)
- Severe psychological and/or social or spiritual distress or suffering
- Pain requiring high dosages of analgesic medications
- Dysphagia requiring a feeding tube
- Dyspnea or symptoms of hypoventilation, a forced vital capacity of 50 percent or less is present
- Loss of function in two body regions (bulbar, arms or legs)

- Adopt a consistent strategy and use it at each visit. The "CLASS" approach is widely accepted, as is the "SPIKES" method of delivering bad news. Present specific choices of medical care as a means of maintaining quality of life and control over health care delivery

Research Recommendations

- Identify decision points (such as changes in respiratory measures) that need to be routinely discussed between the patient and the health care provider. Assemble a list of important decision points and implement prospective studies to assess the impact of standardizing treatment discussion around decision points.
- Develop prospective studies that assess specific areas of communication that are relevant to ALS. Identify optimal timing and ways to communicate the diagnosis, discuss illness progression and approach end-of-life issues.
- Develop overall disease assessment tool(s) for the patient with ALS that help physicians identify specific areas of psychosocial need (e.g., spiritual, social and functional domains).
- Develop prospective studies that evaluate the type and timing of communication and decision-making discussions regarding end-of-life decisions, medical intervention use at the end of life and the utility and durability of Advance Directives.

Policy Recommendations

- Improve training in appropriate communication strategies (e.g., the "CLASS" and "SPIKES" protocol) for end-of-life care and include evaluation of skills in these areas when seeking accreditation (e.g., board certification testing).

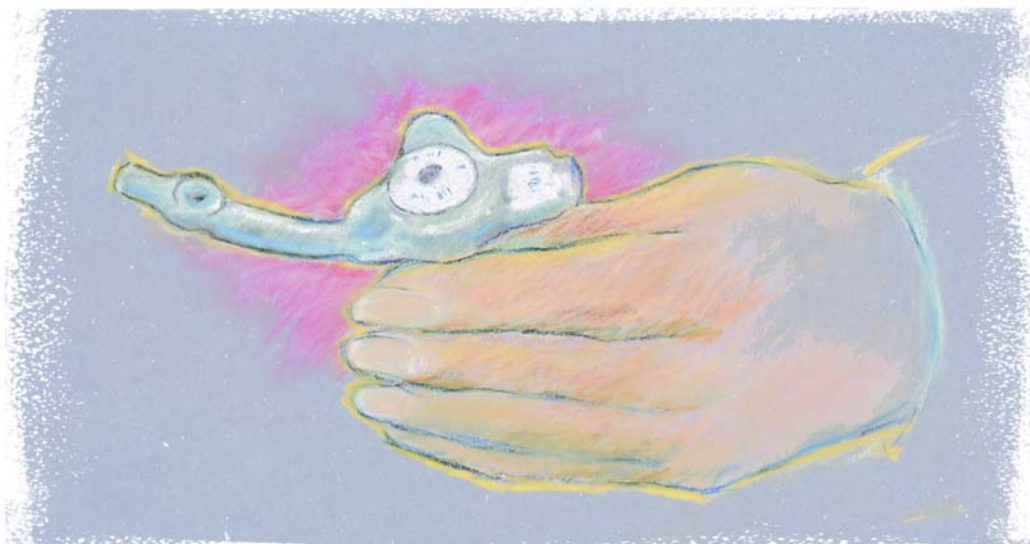
- Incorporate workshops on improving communication and decision-making processes in training programs in ALS and neurology (e.g., continuing medical education at national meetings, board certification courses and medical schools). Develop new ways to disseminate this information (e.g., Internet-based tutorials). Assess the impact of these tools.

- Provide institutional support for improved communication. Provide easy access to medical interpreters, social workers and patient advocates.
- Establish regular debriefings of all involved in mediated decision making as part of quality assurance in health care facilities.
- Integrate monitoring of communication and decision-making practices into the accreditation process of health care facilities.

Access to Care

Basic access to health care should be universal, but it is not. Health care for patients with ALS is inadequate in many areas, especially at the end of life. End-of-life guidelines for hospice care and Medicare do not accurately reflect current standards of care. Limitations in care may be due, in part, to a lack of insurance coverage, lack of availability of knowledgeable physicians or inaccessibility to specialized centers.

Care provision in patients with ALS successively changes from a primary care physician, to a neurologist or an ALS specialist, and finally to a hospice setting. Ideally, patient care is best when maintained by all these providers throughout the course of the disease. However, in current practice several factors



preclude access to specialized pre-terminal and end-of-life care.

Practice Recommendations

- Develop individualized resource lists with the names and contacts of the management team, regional home care agencies, respiratory care companies and hospice services.
- Improve management algorithms for patients with ALS at the end of life:
 - Implement existing algorithms for symptom management;
 - Create management algorithms for supportive care (e.g., skilled home nursing); and
 - Define standard of in-hospital care for patients with ALS.
- Promote cooperation among community physicians, specialty centers and hospices to ensure continuity of care.
- Refer patients to medical centers that provide optimal quality of care for the patient.
- Make a timely referral to hospice.

Research Recommendations

- Create management algorithms specifically for ALS:
 - Evaluate the impact of interdisciplinary versus community care approaches on patient outcomes.
 - Assess barriers to adherence to algorithms and established standards of care processes.
- Study optimal timing of referral to hospice:
 - Develop criteria for appropriate referral to hospice; and
 - Examine the effect of the patient's attitudes and decisions for selecting or not selecting hospice.

Policy Recommendations

- Provide funding for and develop regional Centers of Excellence specific for patients with ALS. Develop the model at these centers for promoting access of effective and optimal care for patients with ALS during the end of life.
- Modify Medicare guidelines for admission to hospice based on specific needs of patients with ALS—include approving use of assistive devices, non-invasive positive pressure ventilation (NIPPV) and physical therapy, among others.

- Establish a comprehensive reimbursement program for patients to cover:

- Hospitalization (i.e., 48-hour hospitalization for percutaneous endoscopic gastrostomy [PEG]),
- Medications during the end of life,
- Assistive devices,
- NIPPV,
- Physical therapy, and
- Home care, hospice care and respite services.

- Develop end-of-life management algorithms for ALS. Specific areas of debate include establishing a policy regarding hospitalization and medications usage and encouraging existing policies to be comprehensive and adequate (e.g., 48-hour hospital stay for PEG placement).
- Institute coverage of case management services to coordinate care across specialties.

Cost of Care

The cost of medical care at the end of life can be high in ALS because many patients require an assistive ventilatory device, use expensive medications, require medical procedures and need hospitalization. Health care insurance, based on Medicare guidelines, does not reimburse for many basic treatments or types of care essential for patients with ALS. These treatments are considered unnecessary or "custodial" rather than "medical" under the current guidelines, thus depriving many patients of much needed care or burdening them with considerable payments for this care.

The results from a 1996 Nationwide Inpatient Sample study showed a 40 percent higher hospitalization cost for ALS as compared to non-ALS patients, and identified nutritional and respiratory morbidity as the most common reasons for admissions. In addition to the expensive direct costs, the indirect costs of informal caregiving of older adults can be staggering.

Most cost studies in ALS are done in association with treatment trials, and there is no analysis or evaluation of indirect costs. Additionally, there are no studies on the costs of end-of-life care, the cost versus quality of care offered by different medical settings and the cost of caregiving on employers.

Practice Recommendations

- Follow evidence-based guidelines for treating terminal phase symptoms.
- Limit medications at the end of life that are normally used for altering disease course.
- Encourage discontinuation of all disease-specific medications (e.g., antioxidants, anti-inflammatory drugs and antiglutamate agents) upon admission to hospice.

Research Recommendations

- Initiate cost-benefit and cost-effectiveness studies for interdisciplinary care at ALS specialty centers and specialty neurology practices.
- Develop a model of management that focuses on cost-effective care for patients with ALS during the end of life.
- Develop a model of cost of care generated by a panel of experts.
- Undertake cost-benefit studies of unplanned hospitalization.
- Evaluate outcomes, including cost effectiveness, quality of life and cost-benefit analysis of home care, hospice, PEG and ventilation.

Policy Recommendations

- Develop comprehensive and equitable policies and reimbursement for in-home care (including ventilation) and long-term care in ALS.
- Emphasize economic benefits to payers of early and timely interventions to reduce unnecessary hospitalizations:
 - Provide a mechanism of funding for in-home care, including reimbursement for family caregivers' services;
 - Increase funding and cost coverage for long-term care and in-home ventilation costs; and
 - Expand hospitalization coverage (to 48 hours) for surgical interventions such as PEG.

Education

In recent years, public awareness of ALS has greatly improved. The American Academy of Neurology (AAN) and voluntary health organizations, such as

ALSA and The Muscular Dystrophy Association (MDA), have contributed significantly to improving the overall education of physicians and policy-makers. However, continued educational programs for patients, families and health care providers need to be improved in the area of end-of-life care in general, but specifically about PEG, NIPPV, treatment of pain/suffering, use of riluzole, other symptom management strategies, home care and long-term care options such as hospice admission criteria and hospice services.

Physician education is still needed regarding enhancement of communication skills and decision making at the end of life, education about the use and subsequent discontinuation of PEG and placement and removal of NIPPV, management of pain, suffering and symptoms at the end of life, and effective use of home care services, long-term care options and hospice services. Frequently, inexperienced health care providers are caring for patients who have highly complex medical and technical needs. Health care providers need to be educated about the importance of hospice care and the Practice Parameter. Treating physicians need to be aware of the importance of the family milieu in caring for patients with ALS. In addition, knowledge of ALS support services available in the community needs to be enhanced. In particular, health care providers need to improve their knowledge and increase the frequency of referral of families to local ALS support services that have the potential to improve quality of life. For example, facilitating access to a local organization that can provide patients with ALS with a communication device can be immensely helpful to patient and family.

Practice Recommendations

- Utilize evidence-based guidelines (Practice Parameter) for care needed during the end of life.
- Educate clinicians, families, caregivers and all stakeholders in ALS about the complexity of care needed during the end of life.
- Educate physicians, using knowledgeable interdisciplinary ALS center team members, regarding Medicare denial, the appeals process, letters of medical necessity for durable medical equipment, Social Security guidelines regarding disability benefits and benefits for war veterans.
- Ensure each ALS health care provider has a list of durable medical equipment and templates for letters of medical necessity in order to reduce denials and facilitate timely insurance coverage of medically necessary services and equipment for patients with ALS.

- Develop tools/models for the education of all health care providers/patients/families by experienced personnel from interdisciplinary ALS centers.
- Ensure timing of referrals to hospice:
 - Educate patients and health care providers regarding criteria and value of hospice; and
 - Educate patients and caregivers regarding hospice guidelines.
- Assess community practices relevant to ALS, especially pertaining to the end of life. This includes conducting regular audits of community and tertiary care practices and identifying specific educational needs between both health care providers and consumers.
- Develop and implement educational intervention initiatives relative to end-of-life practices. The outcome benefits of these educational interventions should be studied to ensure their value in improving the end of life for patients and their caregivers.

Research Recommendations

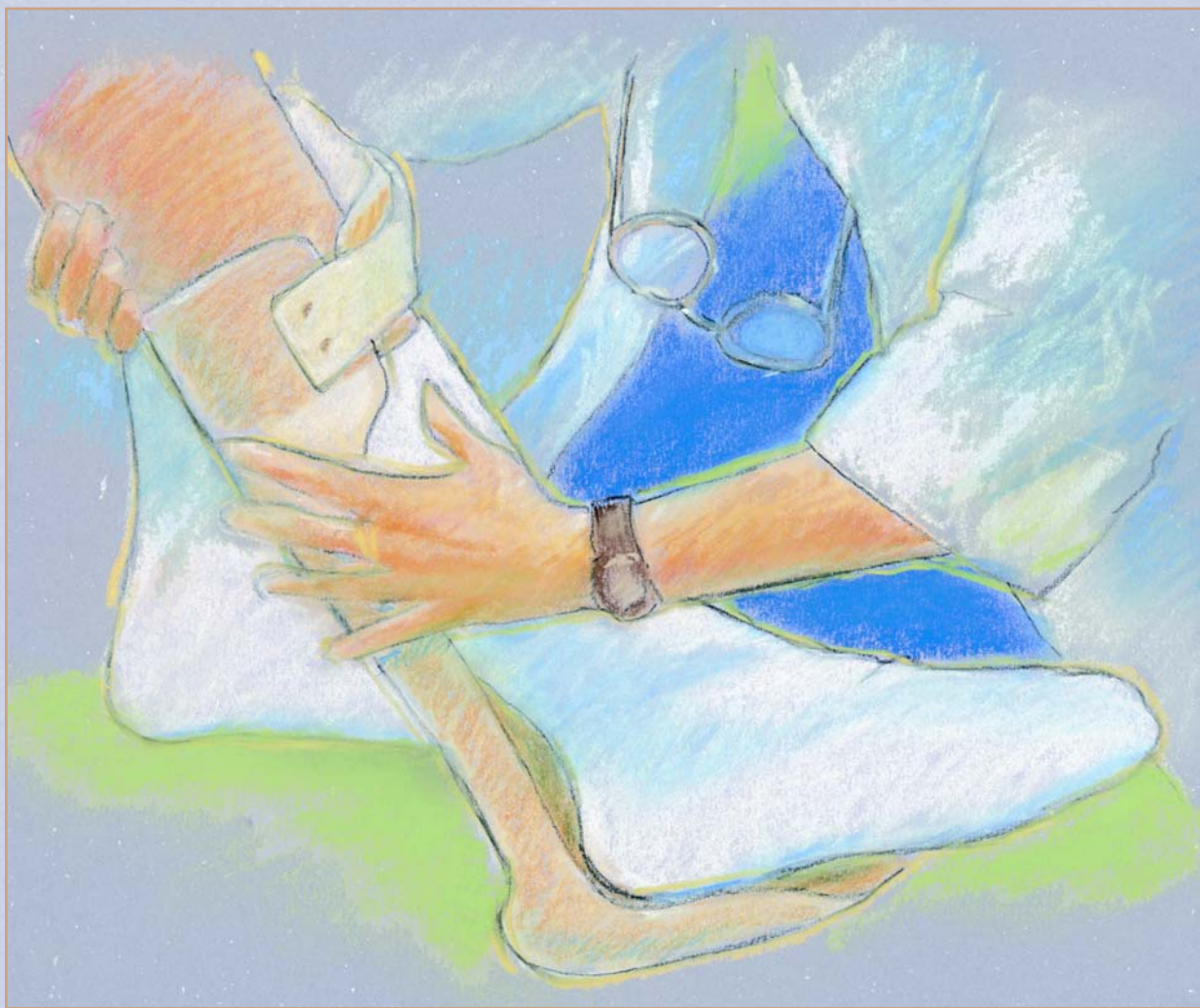
- Identify areas where knowledge among physicians is lacking in caring for patients at the end of life.
- Design new education and training programs to improve the knowledge base of physicians caring for patients with ALS at the end of life.

Policy Recommendations

- Influence health policy agencies, the AAN and other national organizations and foundations to support educational programs on end-of-life care in ALS.
- Increase funding for educational interventions and research surrounding end-of-life issues for patients and their caregivers.



Report to the Field





Explanation of the Peer Workgroup

Statement of Need

Promoting Excellence in End-of-Life Care is a national program of The Robert Wood Johnson Foundation (RWJF) dedicated to facilitating the long-term improvement in health care for dying patients. *Promoting Excellence* developed peer workgroups that engage in ongoing communications among key leaders across clinical disciplines to improve end-of-life care for several specific populations. Amyotrophic lateral sclerosis (ALS) is one of the specific populations identified for improving end-of-life care.

The Convening Process

The *Promoting Excellence in End-of-Life Care* program chose a tactic entitled Peer Workgroups, which consist of small groups of clinicians and researchers who offer high-level clinical experience in specific niches of medicine. *Promoting Excellence* approached The ALS Association (ALSA) to help establish and to manage the ALS Peer Workgroup. The goal of the ALS Workgroup was to provide recommendations on how to improve palliative and end-of-life care specific to patients with ALS. These recommendations are targeted toward multiple audiences including health care providers, health care policy-makers, educators, researchers, patients and families, as well as professional and patient advocacy organizations.

Introduction

Defining ALS

ALS is a progressive neurodegenerative disease that typically results in death from respiratory failure within three to five years from onset of symptoms (Mitumoto et al., 1998). While caregivers, patients and health care providers focus considerable effort on implementing treatments to halt or slow the disease, ALS remains relentlessly progressive. Consequently, because there is no cure for ALS, the major focus of clinical care is providing the highest quality of life through the management of symptoms and emotional or physical suffering (Oliver et al., 2000).

Issues in End-of-Life Care

ALS varies considerably among patients regarding site of onset and rate of progression (Brooks, 1994; Munsat et al., 1988; Ringel et al., 1993). However, the advanced phase of the disease is relatively stereotypic involving decreasing motor function, increasing immobility, increasing respiratory insufficiency and increasing pain. Foregoing active disease treatment, nutrition and hydration, and withdrawing life-sustaining care (such as respirators) are common end-of-life considerations for

patients with ALS. Thus, during the later stages of ALS, most people need their clinician's help with multiple issues, such as psychosocial support, decision making regarding withdrawal of life-sustaining therapies and pain management. However, in reality, end-of-life care for many patients with ALS begins at time of diagnosis. From this point forward, consistent attention to palliative care is paramount to helping patients and their families through the journey of this progressive illness and eventual death.

The primary focus of the ALS Peer Workgroup initiative was to improve end-of-life care for patients and families with ALS so that those affected can experience optimal care and support at the end of life. Patients and caregivers look to health care providers for guidance during this process. Individuals caring for patients with ALS face decisions and challenges that can be overwhelming. The initiative explored resources that should currently be available to the patient and caregivers and who should provide the necessary care. It also explored issues and gaps that have prevented full implementation of what should be standard practice. This initiative also identified barriers that health care providers encounter when caring for patients with ALS.

One of the greater challenges for health care providers in providing palliative care for their patients is coming to terms with the limitations of medical therapies and developing an attitude of unconditional acceptance of and support for patients. Meeting this challenge requires that health care providers review their own attitudes toward death and life, come to terms with their own mortality and take the necessary steps to address their own personal issues surrounding death and dying (Appendix A, Table 1). Beyond exploring mortality issues, other gaps in care exist at the end of life for these patients and their care providers, such as:

- *Clinicians may be reluctant to, or are not familiar with, initiating discussions regarding end-of-life issues.*
- *Clinicians are daunted by the dying patient and believe that they have nothing to offer the patient.*
- *Clinicians and caregivers have inadequate training in the principles and practical application of state-of-the-art pain and palliative medicine.*
- *Health care providers have limited experience with:*
 - *ALS-specific symptom management; this may extend across specialties (e.g., family medicine, internal medicine and general neurology, and also*

across health care providers—nursing, social workers, physicians and residents, among others). This limited experience in symptom management is particularly evident regarding:

- *Use of opioids;*
- *Withdrawal of or failure to provide life-sustaining treatment; and*
- *Provision of psychosocial and spiritual support.*
- *Health care providers often fail to coordinate among different disciplines (respiratory, gastroenterology, neurology, primary care and physical therapy).*
- *Caregivers can be distressed and exhausted.*
- *Patients and their families are given abundant information late in the disease process.*

Methodology

Establishing the ALS Peer Workgroup and Defining the Aims of the Project

The ALS Peer Workgroup consisted of an interdisciplinary panel of experts including neurologists, ALS nurse practitioners, a social worker, a pulmonologist, a palliative medicine specialist, a speech language pathologist, an ethicist (neurologist) and patient advocacy representatives. Based on consensus, the ALS Workgroup defined four areas of care for evaluation and formed small groups to address them:

- *Psychosocial care, bereavement, quality of life, spirituality and caregiver issues;*
- *Ethics, communication and decision making;*
- *Symptom management (respiratory, nutrition, depression and pseudobulbar affect, speech/communication, pain, insomnia and final hours); and*
- *Access to care, cost of care, health policy and knowledge/education.*

The tasks for each of the four groups were to:

- *Identify the current state of care in each of the four areas listed above based on published evidence. When no evidence was available, the state of care was defined by expert consensus (summarized in Appendices B-E);*

- *Determine elements that constitute optimal care based on consensus (summarized in Appendices B-E);*
- *Define the gaps in care between the current state and optimal care;*
- *Provide recommendations on how to close the gaps between current care and optimal care in the realms of clinical practice, research and health policy; and*
- *Identify tools and resources that can help close the gaps (summarized in Appendices B-E).*

The Process

Members of the Workgroup gathered in June 2000 to identify aims, structure and process. Each of the four small groups met by regular conference calls throughout the first year to present and discuss their findings. Half of the Workgroup members met in November 2000 in conjunction with an international ALS meeting. Three of the small groups met for one-day sessions in the fall 2001; the final consensus conference was held in November 2001.

Each small group conducted a systematic review of the literature using Medline. Abstract titles were screened, relevant abstracts were reviewed and key papers were used to support the conclusions made that defined the current state of care. Consensus was reached within each of the four groups to define optimal care for each of the issues addressed. Through consensus, specific recommendations were identified on how to fill the gaps in care. In order to fill these gaps, recommendations were targeted to three different audiences: clinical practice, research and health care policy.

Conceptual Considerations in Defining End of Life

For some health care experts and patients with ALS, palliative care begins at the time of diagnosis. For others, end-of-life discussions and preparations are considerably delayed. Therefore, deciding when to introduce end-of-life discussions with patients and their families can be challenging for clinicians and other health care providers. Additionally, end-of-life care often involves switching from a restorative goal to a more palliative goal, and the timing of this transition can be challenging, even for experienced physicians.

The Workgroup devised an operational definition for end of life to help identify when end-of-life discussions should be introduced to patients and

caregivers. Triggers for initiating discussions about end-of-life care for patients with ALS are:

- *The patient or family asks—or “opens the door”—for end-of-life information and/or interventions (elicited or spontaneous, verbal or non-verbal).*
- *Severe psychological and/or social or spiritual distress or suffering.*
- *Pain requiring high dosages of analgesic medications.*
- *Dysphagia requiring a feeding tube.*
- *Dyspnea or symptoms of hypoventilation, a forced vital capacity of 50 percent or less is present.*
- *Loss of function in two body regions (bulbar, arms or legs).*

The presence of any of these triggers may signal the clinician to initiate discussions regarding the end of life, and if necessary, establish an end-of-life treatment plan with increasing emphasis on palliation. While end-of-life care is an important aspect of palliative care, all patients with ALS need the full range of active palliation from the time of diagnosis. When to focus on the terminal phase of ALS requires professional judgment that takes into account a broad base of information about the patient and family (coping skills, depression and anxiety, cultural issues, use of functional assistive devices and physiologic status, among others).

Resources and Defining Optimal Care

Results from the evaluation of the currently available resources and the defining optimal care exercise were used to identify the gaps in end-of-life care, and for developing practice, research and policy recommendations for the field. The detailed results from the review of the currently available resources and definitions of optimal care are presented in Appendices B-E.

Workgroup Findings and Recommendations to the Field

Psychosocial Care

Identification of Existing Resources

Psychosocial care encompasses a range of issues relevant to patients, their families and caregivers and health care providers. Although current practice focuses predominantly on the patient’s specific

Some Components of Psychosocial Care in ALS

- **Bereavement***
- **Spirituality***
- **Education**
- **Children and family needs**
- **End-of-life expectations**
- **How to be present with a dying person**
- **Role of health care provider/team**
- **Life closure**
- **Quality of life***
- **Caregivers***
- **Sexuality and intimacy**
- **End-of-life choices**
- **Withdrawal of ventilation and other support**
- **Supportive counseling**
- **Coping mechanisms**

*The workgroup selected these areas to review in this project. They are reviewed for: defining the current state of care, defining optimal care, assessing gaps in care and providing recommendations.

medical needs, psychosocial care actually may be the central factor that underlies the decisions regarding health care choices and attitudes about living and dying with ALS. A recent study published in the *New England Journal of Medicine* reports that seriously ill patients care about how burdensome the treatment is, their quality of life and likely outcomes (Fried et al., 2002).

Psychosocial care includes a number of different aspects of life for the patient with ALS, only some of which are listed in the corresponding side bar. Psychosocial care underlies many decisions relative to caring for patients with ALS, including ethics, communications, symptom management and accessing resources and cost issues. Within each of the respective sections of this document, it is important to recognize that psychosocial components influence management and treatment decisions. Tools available for addressing many of these psychosocial issues are listed in Section 1 of Appendix B, Tables 1-7.

Identification of Existing Gaps

Despite its general influence across all areas of the patient's life, psychosocial care is understudied in ALS. There are few resources that provide direction for health care providers, patients and families regarding the psychosocial management of patients with ALS. As a result, there is a lack of focus, and in some cases a lack of awareness, regarding the importance of addressing psychosocial concerns in these patients. Many health care providers do not know how to approach patients to offer psychosocial support, and others do not know what type of psychosocial care would be beneficial.

Recommendations to the Field

Practice Recommendations

- Increase awareness of importance for psychosocial care in patients with ALS and their caregivers.
- Implement an interdisciplinary team approach to care in ALS that regularly includes a psychosocial evaluation.

Research and Program Development Recommendations

- Develop specific training materials to facilitate an interdisciplinary approach to care.
- Develop specific training materials to facilitate the goal of increased awareness for psychosocial care (such as vignettes, curricula, care plans or guidelines).
- Develop curricula on psychosocial care (including cross-cultural interreligious issues).

Bereavement

Identification of Existing Resources

Bereavement occurs in many medical conditions with a loss of a body part or function. Onset of bereavement in patients with ALS is unique because there is a progressive loss of strength and function, with no stable time for adaptation to these losses. For some patients, bereavement starts at the time of diagnosis. Others grieve as they lose their ability to function and lead a normal life. Both the patient and caregivers grieve the loss of their future together.

For caregivers, bereavement is also progressive, as they grieve the loss of life as they knew it. Therefore, caregivers grieve for the loss of function in patients, and also grieve the loss of their own independence. Little is known about the process of bereavement in the caregivers of patients with ALS. A detailed overview of the current practices and existing resources about bereavement at the end of life is summarized in Appendix B.

Identification of Existing Gaps

Little published information exists to guide patients with ALS, caregivers and health care providers through the bereavement process. The majority of information available on bereavement is not specific to ALS, and few books include chapters devoted to bereavement.

Issues in the bereavement process that may be unique to ALS have not been well studied. Little is known about how to provide support and services for the caregivers of patients with ALS as they experience this type of bereavement. For example, it is unclear how to optimally work with caregivers and help them come to terms with resentment or guilt, which often surface during the grieving process.

Recommendations to the Field

Practice Recommendations

- Offer bereavement support, informally or formally, for the patient and caregiver during the clinic visit.
- Offer bereavement support for the caregiver after the death of the patient, including letters of condolence at the time of death.

Research Recommendations

- Determine the scope of bereavement in ALS and whether there are differences between ALS and other diseases, such as cancer.
- Assess whether specialized ALS support groups are needed and if they confer specific benefits.

Spirituality

Identification of Existing Resources

The literature published to date suggests that spiritual care is an integral component of palliative care (Appendix B). Addressing spiritual issues with patients is one that is personal, varied and has only recently been considered beneficial for terminally ill patients and their caregivers. Studies suggest that health care providers may encourage this aspect of care, and that they can be aware of the importance of spirituality and its impact on patients and caregivers.

It is likely that spiritual/religious attitudes influence decisions surrounding ongoing medical management and choices about end-of-life issues (e.g., implementing Advance Directives, use of technological interventions, withdrawing life support, physician-assisted suicide, autopsy and disposal of the body). The spiritual beliefs of the health care providers also can significantly impact their own comfort levels in managing certain end-of-life decisions (i.e., withdrawal of ventilation and use of opioids). Despite its influence, spirituality is almost never discussed with regards to supervision of health care providers.

Identification of Existing Gaps

Addressing spiritual issues of patients with ALS should be standard practice for health care professionals in ALS. Additionally, spiritual needs of caregivers and patients' families should be addressed, as these individuals will play a role in the patient's end-of-life decisions. Unfortunately, these concerns are rarely addressed—recognition is lacking that the role of spiritual/religious attitudes is relevant to end-of-life issues and palliative care in ALS. Many physicians and health care providers are not aware that spirituality has a significant impact on patients with terminal illnesses, such as ALS. Several tools address spiritual needs, and these are generally underutilized in caring for patients. Specific language and tools are needed to help initiate discussions on spirituality, because little work has been done on how to make

spiritual care meaningful for patients with ALS. An interdisciplinary team that includes addressing spiritual care of patients may be of significant value to patients and their families.

Recommendations to the Field

Practice Recommendations

- *Address the spiritual needs of patients.*
- *Establish an interdisciplinary team to enhance collaboration with chaplains, spiritual counselors and pastoral workers. Assess the spiritual needs of patients, caregivers and family members repeatedly as the disease progresses.*
- *Recognize and accept the patient's spiritual and religious attitudes. Educate health care providers regarding cross-cultural religious differences in the approach to death and dying with the intent of improving management of end-of-life issues.*

Research Recommendations

- *Improve methods and tools for assessing the spiritual needs of patients and caregivers.*
- *Study the impact of managing spiritual needs on quality of life and longevity of patients, and on the patient-physician relationship.*
- *Examine spiritual needs of different patient populations (e.g., ALS versus cancer) and different ethnic/religious backgrounds to improve specific spiritual care for different populations of patients.*

Quality of Life

Identification of Existing Resources

Quality-of-life instruments are available for numerous diseases and can be helpful in assessing treatment satisfaction and overall patient well-being. The practical clinical goal is to use quality-of-life information to help identify areas of concern for patients in the terminal phase of ALS. Quality-of-life instruments can be used in a proactive manner so that resources, treatment modifications and lifestyle changes can all be implemented to address specific concerns. This initiative identified 19 different instruments that are available for assessing quality of life (See Appendix B).

Identification of Existing Gaps

Although clinicians are conscious of improving quality of life in patients with ALS and their caregivers, assessing quality of life and taking steps to improving it have been elusive. Most currently available quality-of-life instruments have not been validated in patients with ALS. An instrument to assess quality of life in ALS must evaluate the psychosocial status of patients during the end of life. Additionally, a better understanding is needed of factors that influence quality of life in patients with ALS and their caregivers.

Recommendations to the Field

Practice Recommendation

- *Use quality-of-life instruments in patients with advanced ALS during the end of life to help detect issues that should be addressed in order to improve the end of life (e.g., McGill QOL or Seiqol-DW patient generated measure of individual quality of life with high acceptance in ALS).*

Research Recommendation

- *Develop instruments designed to assess quality of life at the end of life considering the following:*
 - *The physical and psychological comfort of the patient when completing an instrument (eliminate instruments that trigger psychological distress);*
 - *The time needed to complete the instrument; and*
 - *The ease of administration to patients with impaired communication abilities.*

Caregiver Issues

Identification of Existing Resources

In the current health care system in the United States, patients with chronic debilitating diseases are increasingly cared for at home, except for acute medical complications that occur during the disease course. According to the North American ALS CARE Database, 63 percent of patients die at home (Bradley et al., 2002), and thus, informal family caregivers (most often spousal caregivers) become the principal caregivers in this setting. Little psychosocial or medical attention is paid to caregivers. Often caregivers are suffering and mentally distressed from

caring for patients. Although there are patient support groups, there may be a limited number and lack of awareness about formal caregiver support groups. Available resources and support programs for caregivers are presented in Appendix B.

Identification of Existing Gaps

Patients and caregivers have high concordance of distress and depression. How caregivers are managing their own care during the end of life of patients with ALS is understudied. Some caregivers do well, despite long days of extensive caregiving, yet others suffer severely from this challenging task. There are no data that assess characteristics of caregivers that might increase their risk of experiencing distress and burden from caregiving. Additionally, there are no studies that systematically evaluate the type of support caregivers may require over the long term in caring for patients with ALS.

Although this initiative focuses on caregiver wellness during the patient's end of life, most practice recommendations are appropriate when initiated in the early stages of the disease.

Recommendations to the Field

Practice Recommendations

- Interview experienced caregivers to identify psychosocial issues that must be addressed.
- Establish support services for caregivers including psychiatric guidance and counseling, support groups that emphasize educational initiatives, hands-on training programs and wellness-promoting behaviors.
- Develop a crisis management system for caregivers in ALS clinics, including an ALS caregiver telephone hotline available 24 hours per day.
- Make spiritual education training and intensive psychosocial support readily available (including home visits), and offer these services to all caregivers during the end-of-life period.

Research Recommendations

- Study whether proactive intensive psychosocial care reduces caregiver burden and distress, enhances caregiver wellness and improves the quality of life of patients.
- Develop an instrument that can quantify caregiver burden and distress.

- Study the medical economics of family caregiving, and expand the evidence base for supporting insurance reimbursement for family caregiver costs.

Policy Recommendations

- Reimburse the cost incurred by physicians who provide supportive care for caregivers.
- Using organized lobbying approaches, improve financial support and reimbursement for the costs assumed in caring for patients with ALS during the end of life (e.g., Medicare reimbursement).
- Improve financial/insurance coverage for psychosocial intervention in the home.
- Encourage hospices to provide universal social work coverage for all families (current policies allow nursing support to families, but many hospices assign social work coverage only to those families deemed at particular risk and do not provide counseling to the majority of caregivers).

Ethics

Identification of Existing Resources

The ethical basis and legal status of end-of-life decisions pertinent to ALS were recently reviewed (Bernat, 2001) and specifically addressed in the Practice Parameter (Appendix F). Yet ethical issues in caring for patients with ALS are sensitive, and many health care providers are not comfortable with ethically charged decision making.

One of the most difficult discussions surrounding the end of life relates to physician-assisted suicide. Both anecdotal reports and systematic studies indicate a high level of interest in physician-assisted suicide and euthanasia in patients with ALS (American Academy of Neurology Statement 1998). Physician-assisted suicide is illegal in most U.S. jurisdictions, and euthanasia is illegal in all U.S. jurisdictions and in most parts of the world. Some health care providers, however, have difficulty differentiating accepted clinical practices (medication use for symptom relief, including sedation and palliative medicine) from physician-assisted suicide and euthanasia (Carver et al., 1999). In avoiding topics of euthanasia and physician-assisted suicide, physicians may be avoiding appropriate discussion of good end-of-life care and consequently not providing appropriate palliative care, including medications. Continued

discussions are needed among health care providers, the patients and caregivers regarding end-of-life issues specific to palliative care.

Identification of Existing Gaps

There is a gap between knowledge and practice regarding ethical and legal practices at the end of life. Guidelines published in medical journals do not penetrate all areas of clinical practice. The reasons for the lack of implementation of treatment guidelines and the difficulty of putting theory into practice are not known.

Recommendations to the Field

Practice Recommendations

- *Implement evidence-based guidelines for terminal care in patients with ALS (Miller et al., 1999).*
- *Identify and monitor conflicting issues associated with end-of-life care including conflicts among family members and with health care providers. Intervene early to resolve conflicts and use counseling services, among other support systems, so end-of-life care is agreed upon and a plan is established in advance. Clarify ethical and legal issues with an ethics committee or legal counsel, as necessary.*
- *Use questions regarding physician-assisted suicide and euthanasia as a trigger to discuss end-of-life care. Health practitioners need to provide explicit assurances of continuity of care and commitment to relieve suffering. This may be one of the most direct and first triggers to discussing end-of-life care. Physician-assisted suicide is not legal in 49 states; clinicians should not feel coerced to provide a lethal prescription.*

Research Recommendations

- *Investigate the prevalence of the desire to end life, and evaluate interventions that obviate this desire.*
- *Examine the impact (outcomes) of the patient's end-of-life decisions on the overall quality of life during the terminal phase of the patient's illness, as well as the impact on the family and caregivers.*
- *Develop a neurology Educating Physicians in End-of-Life Care (EPEC) module and assess the impact of its use on clinicians' knowledge and practice in end-of-life care of ALS.*

Communications and Decision Making

Identification of Existing Resources

Patient autonomy in end-of-life decisions, as in all medical decision making, is the accepted Western paradigm. Although patients or their surrogates can legally and ethically withdraw or not initiate life-sustaining interventions (including hydration and nutrition), many are not aware of their rights or do not exercise their rights. Voluntary organizations (e.g., The ALS Association) offer resources that can help decision making surrounding end-of-life issues.

The extent to which the individual is actually autonomous will vary widely. The cultural context of the individual will influence the acceptance of the prevailing societal standards. Exploring the cultural background, particularly the spiritual beliefs of the individual and family, is essential. Where major differences are detected in perceived morality of the decisions and in other areas such as disclosure of medical information, advice from appropriate authorities in the patient's community and from the consultative ethics team in the medical institution should be sought prior to a crisis.

Communication of information among health professionals and the patient and family is required for making treatment decisions. Clinicians impart knowledge of, and experience with, all stages of ALS. The patient with ALS and the family impart their values and experiences that will guide their decisions. Strategies to effectively communicate complex and difficult information have been promulgated; effective communication strategies are advocated in managing patients and families with ALS (Appendix C). A recent brief review summarizes the "CLASS" and "SPIKES" protocols, which detail effective communication approaches for health care providers and their patients (Buckman, 2001). The EPEC Program provides training to educators who promote dissemination of improved end-of-life care, including improving communication techniques.

Decision making in ALS differs from other life-threatening diseases in that life-sustaining measures can prolong life while having little or no impact on disease progression. Decisions made by default (or in emergent situations) may not be congruent with previously unstated goals or values of the patient and family. Unless specifically addressed, and in sufficient detail, placement of feeding tubes, institution of respiratory support and referral to hospice can be

omitted, or undertaken haphazardly. The role of denial in delaying decision making, and the correlates of denial have never been specifically evaluated. In the literature, there are some guidelines regarding timing of interventions (such as percutaneous endoscopic gastrostomy [PEG] placement) and when to initiate discussions regarding starting interventions; however, there is little information on how to approach patients who delay decision making surrounding end-of-life issues. Additionally, clinical practice varies widely with regards to what to do when patients delay decision making, when discussion should begin, and how to approach patients who delay decision making.

A particularly difficult communication and decision-making situation is one of a "locked in" patient on a ventilator with no Advance Directives. This circumstance reflects a failure of advance care planning because there had been ample opportunity earlier to ask how the patient wished to be treated in this circumstance and to have avoided the present predicament. At this point, clinicians should try to establish a communication system with whatever movement response the patient retains. But assuming the patient is so de-efferented that no communication system is possible, the patient would require a surrogate decision-maker. The patient lacks the capacity to make a health decision, not for the usual reason of cognitive impairment, but because of an absence of communication (somewhat similar to that of an aphasia patient). The surrogate should be appointed through the usual means and be instructed to decide by the usual standards.

Identification of Existing Gaps

Communication Between Clinicians and Patients/Families/Caregivers

Few studies evaluate clinical practices regarding discussion of sensitive issues such as sedation, pain relief and withdrawal of life support (including ventilation, nutrition and hydration), let alone the impact of these decisions on those affected by ALS, their caregivers and on medical practitioners. The clinicians' attitudes about the diagnosis, available therapies and perceived quality of life in patients with ALS during the end of life need to be better understood in order to assess how physicians influence patient attitudes and choices.

In ALS, no studies address the impact and efficacy of communication strategies or the validation of specific approaches to communication and decision

making. Communication of the diagnosis is felt to be unsatisfactory by the majority of those surveyed, suggesting that effective communication remains elusive for many in clinical practice.

For some patients, conflicts arise from differences in values, and these conflicts may extend to either family members or health care providers. These conflicts should be resolved prior to entering the end-of-life phase, yet systematic approaches on how to resolve ethical conflicts are lacking.

The understanding of how and when to create a predefined plan detailing the end-of-life process is not available. For example, the individual and family must be prepared to prevail in their end-of-life choices when encountering under-informed medical staff. Legally binding documents (e.g., durable medical power of attorney) and ideally, on-call or back-up health care providers should be arranged in advance and available to the family. Decisions are not immutable; the evolution of choices and the influences that lead to changes in decisions must be better understood.

Communication Between Clinicians in Patient Care Decision Making

Management of ALS involves many clinicians, whether formally structured into an interdisciplinary team or more loosely networked within a community. Communication between the different clinicians is often problematic, as knowledge of the course and prognosis of ALS is varied and the values and experiences of each clinician are not normally stated. Often, the patient does not have a single physician who takes the leadership role in initiating end-of-life discussions. One of the primary responsibilities for this lead physician's role also is to communicate to other health care providers what the management plan is and what the patient's decisions are surrounding the end of life.

Recommendations to the Field

Practice Recommendations

- *Improve communication skills to effectively discuss end-of-life issues with patients with ALS and family members. Identify the physician, or team member, who will take the leadership role in end-of-life discussions, and who will, in turn, communicate the decisions to the other health care providers. This may be done by confidential correspondence in notes in the health records (patient's chart).*

Health care providers should be prepared to support the patient's decisions, regularly review the patient's decisions and update the patient's chart accordingly.

- Establish a partnership between the patient and interdisciplinary team members in the treatment plan during disease progression. This relationship needs to be established prior to initiation of end-of-life discussion. The triggers to end-of-life discussions outlined on page 7 represent the most overt prompts to planning care. From the time of diagnosis, clinicians should provide opportunities to discuss all types of care alternatives. Respiratory issues are central to end-of-life decisions. The reasons behind monitoring pulmonary function should be explicitly stated and all options for respiratory care, including symptom management alone, should be clearly explained.
- Adopt a consistent strategy and use it at each visit. The "CLASS" approach is widely accepted, as is the "SPIKES" method of delivering bad news. Present specific choices of medical care as a means of maintaining quality of life and control over health care delivery.

Research Recommendations

- Identify decision points (such as changes in respiratory measures) that need to be routinely discussed between the patient and the health care provider. Assemble a list of important decision points and implement prospective studies to assess the impact of standardizing treatment discussion around decision points.
- Develop prospective studies that assess specific areas of communication relevant to ALS. Identify optimal timing and ways to communicate the diagnosis, discuss illness progression and approach end-of-life issues.
- Develop overall disease assessment tool(s) for the patient with ALS that help physicians identify specific areas of psychosocial need (e.g., spiritual, social and functional domains).
- Develop prospective studies that evaluate the type and timing of communication and decision-making discussions regarding end-of-life decisions, medical intervention use at the end of life and the utility and durability of Advance Directives.

Policy Recommendations

- Improve training in appropriate communication strategies (e.g., the "CLASS" and "SPIKES" protocol) for end-of-life care and include evaluation of skills in these areas when seeking accreditation (e.g., board certification testing).

- Incorporate workshops on improving communication and decision-making processes in training programs in ALS and neurology (e.g., continuing medical education at national meetings, board certification courses and medical schools). Develop new ways to disseminate this information (e.g., Internet-based tutorials). Assess the impact of these tools.
- Provide institutional support for improved communication. Provide easy access to medical interpreters, social workers and patient advocates.
- Establish regular debriefings of all involved in mediated decision making as part of quality assurance in health care facilities.
- Integrate monitoring of communication and decision-making practices into the accreditation process of health care facilities.

Access to Care

Identification of Existing Resources

Basic access to health care should be universal, but it is not. For an overview of programs currently in place that address resource availability and access to care, refer to Appendix E.

Identification of Existing Gaps

There are many areas where the current system of availability of and access to health care for ALS patients and families is inadequate, both with advanced disease and at the end of life. Guidelines for hospice care and Medicare do not accurately reflect current knowledge and practice regarding the disease. Gaps in effective care may be due mainly to lack of insurance coverage and lack of knowledgeable care providers/facilities.

Lack of continuity of care across the ALS disease spectrum (from primary care to ALS centers/specialty neurology practitioners to hospice) is largely the result of an absence of case management services. When there is no single person ensuring that all health care issues are being addressed and providers are communicating with one another, patient care is often fragmented, needs are often overlooked or efforts are duplicated. Use of an interdisciplinary team approach is clearly needed.

The following are areas for which there is inadequate coverage or a need exists for guidelines or equipment.

Health Care Provider Services

There is a need for continuing:

- Physical therapy (range of motion and massage) to decrease spasticity and pain;
- Respiratory therapy/respiratory technician services for advice on use of suction machines, ventilators and adjustment of non-invasive positive pressure ventilation/ventilator settings;
- Home social work services for education about availability of resources and counseling regarding loss/grief issues;
- Home health aides for custodial care that may involve bathing and transferring;
- Bereavement programs for caregivers; and
- Long-term care insurance coverage.

Home Nursing Care

There is no coverage of unskilled nursing for PEG feeding. There are no available ALS-specific guidelines regarding use of pulse oximetry or non-invasive positive pressure ventilation (NIPPV) settings, and nutritional care/feeding.

Ventilator Patients

There is a dire need for coverage of in-home care of patients with ALS using NIPPV, tracheostomy or mechanical ventilation. Uncovered areas include: skilled nursing, home health aide, respiratory therapist (which is most often covered in the price of rental equipment), physical therapist, technical support and back-up ventilator and power source. Even when insurance approves in-home skilled nursing services, many families are unable to find trained staff due to the severe nursing shortage nationwide. Further, only a small number of facilities exist nationwide that provide residential in-patient, end-of-life palliative care specifically targeted for patients with ALS.

Admission to Hospice

Medicare and other insurers lack correct information regarding markers for the end of life in ALS. As such, the hospice referral guidelines are inappropriate and hospice intake forms do not ask the questions that accurately reflect the ALS patient's condition. Thus, Medicare criteria for admission to hospice are

too restrictive. Consequently, patients with ALS are being referred to hospice too late in their disease course and are thus denied the supportive services that hospice offers.

Recommendations to the Field

Practice Recommendations

- *Develop individualized resource lists with names and contacts of the management team, regional home care agencies, respiratory care companies and hospice services.*
- *Improve management algorithms for patients with ALS at the end of life:*
 - *Implement existing algorithms for symptom management;*
 - *Create management algorithms for supportive care (e.g., skilled home nursing); and*
 - *Define standard of in-hospital care for patients with ALS.*
- *Promote cooperation among community physicians, specialty centers and hospices to ensure continuity of care.*
- *Refer patients to medical centers that provide optimal quality of care for the patient.*
- *Make a timely referral to hospice.*

Research Recommendations

- *Create management algorithms specifically for ALS:*
 - *Evaluate the impact of interdisciplinary versus community care approaches on patient outcomes; and*
 - *Assess barriers to adherence to algorithms and established standards of care processes.*
- *Study optimal timing of referral to hospice:*
 - *Develop criteria for appropriate referral to hospice; and*
 - *Examine the effect of the patient's attitudes and decisions for selecting or not selecting hospice.*

Policy Recommendations

- *Provide funding for and develop regional Centers of Excellence specific for patients with ALS. Develop the model at these centers for promoting access of effective and optimal care for patients with ALS during the end of life.*

- *Modify Medicare guidelines for admission to hospice based on specific needs of patients with ALS—including approving use of assistive devices, NIPPV and physical therapy, among others.*
- *Establish a comprehensive reimbursement program for patients to cover:*
 - *Hospitalization (i.e., 48-hour hospitalization PEG placement),*
 - *Medications during the end of life,*
 - *Assistive devices,*
 - *NIPPV,*
 - *Physical therapy, and*
 - *Home care, hospice care and respite services.*
- *Develop end-of-life management algorithms for ALS. Specific areas of debate include: establishing a policy regarding hospitalization and medications usage and encouraging existing policies to be comprehensive and adequate (e.g., 48-hour hospital stay for PEG placement).*
- *Institute coverage of case management services to coordinate care across specialties.*

Cost of Care

Identification of Existing Resources

The cost of medical care is especially high in ALS because many patients require respiratory support. Medications such as riluzole are expensive and can be discontinued during the final stages of life. Procedures, such as placement of PEG or tracheostomy, also incur surgical costs. These are only a few of the direct costs associated with care, and the indirect costs (lost work income and out-of-pocket expenses) also escalate management costs for patients with ALS (Appendix E).

Identification of Existing Gaps

There are little data on the cost of care and hospitalization for patients with ALS. Most studies are done in association with treatment trials, and there is no analysis or evaluation of indirect costs (e.g., loss of income).

Additionally, there are no studies of costs associated with end-of-life care, no prospective studies comparing cost-to-quality of different care delivery

modes (e.g., community versus tertiary care) and no analysis of assessing the cost of caregiving to employers.

Recommendations to the Field

Practice Recommendations

- *Follow evidence-based guidelines for treating terminal phase symptoms.*
- *Limit medications at the end of life that are normally used for altering disease course.*
- *Encourage discontinuation of all disease-specific medications (e.g., antioxidants, anti-inflammatory drugs and ant glutamate agents) upon admission to hospice.*

Research Recommendations

- *Initiate cost-benefit and cost-effectiveness studies for interdisciplinary care at ALS specialty centers and specialty neurology practices.*
- *Develop a model of management that focuses on cost-effective care for patients with ALS during the end of life.*
- *Develop a model of cost of care generated by a panel of experts.*
- *Undertake cost-benefit studies of unplanned hospitalization.*
- *Evaluate outcomes, including cost effectiveness, quality of life and cost-benefit analysis of home care, hospice, PEG and ventilation.*

Policy Recommendations

- *Develop comprehensive and equitable policies and reimbursement for in-home care (including ventilation) and long-term care in ALS.*
- *Emphasize economic benefits to payers of early and timely interventions to reduce unnecessary hospitalizations:*
 - *Provide a mechanism of funding for in-home care, including reimbursement for family caregivers' services;*
 - *Increase funding and cost coverage for long-term care and in-home ventilation costs; and*
 - *Expand hospitalization coverage (to 48 hours) for surgical interventions such as PEG.*

Education

Identification of Existing Gaps

Education of patients, families and health care providers needs to be improved in the area of end-of-life care in general, but specifically about PEG, NIPPV, treatment of pain/suffering, use of riluzole, other symptom management strategies, home care and long-term care options such as hospice admission criteria and hospice services. Frequently, inexperienced health care providers are caring for patients with ALS in whom the highly complex medical and technical needs require considerable experience. Health care providers need to be educated about the importance of hospice care and the Practice Parameter. Treating physicians need to be aware of the importance of the family milieu in caring for patients with ALS. In addition, knowledge of ALS support services available in the community needs to be enhanced. In particular, health care providers need to improve their knowledge and increase the frequency of referral of families to local ALS support services that have the potential to improve quality of life. For example, facilitating access to a local organization that can provide patients with ALS with a communication device can be immensely helpful to patient and family.

Recommendations to the Field

Practice Recommendations

- Utilize evidence-based guidelines (Practice Parameter) for care needed during the end of life.
- Educate clinicians, families, caregivers and all stakeholders in ALS about the complexity of care needed during the end of life.
- Educate physicians, using knowledgeable interdisciplinary ALS center team members regarding Medicare denial, the appeals process, letters of medical necessity for durable medical equipment, Social Security guidelines regarding disability benefits and benefits for war veterans.
- Ensure each ALS health care provider has a list of durable medical equipment and templates for letters of medical necessity in order to reduce denials and facilitate timely insurance coverage of medically necessary services and equipment for patients with ALS.
- Develop tools/models for the education of all health care providers/patients/families by experienced personnel from interdisciplinary ALS centers.

- Ensure timing of referrals to hospice:
- Educate patients and health care providers regarding criteria and value of hospice; and
- Educate patients and caregivers regarding hospice guidelines.

Research Recommendations

- Identify areas where knowledge among physicians is lacking in caring for patients at the end of life.
- Design new education and training programs to improve the knowledge base of physicians caring for patients with ALS at the end of life.
- Assess community practices relevant to ALS, especially pertaining to the end of life. This includes conducting regular audits of community and tertiary care practices and identifying specific educational needs among both health care providers and consumers.
- Develop and implement educational intervention initiatives relative to end-of-life practices. The outcome benefits of these educational interventions should be studied to ensure their value in improving the end of life for patients and their caregivers.

Policy Recommendations

- Influence health policy agencies, the AAN and other national organizations and foundations to support educational programs on end-of-life care in ALS.
- Increase funding for educational interventions and research surrounding end-of-life issues for the patient and their caregivers.

Symptom Management at the End of Life





The following sections summarize recommendations on symptom management in ALS and medical management during the last days and hours of life.

Respiratory Care

Identification of Existing Resources

Respiratory insufficiency represents one of the most critical issues for the majority of patients with ALS. The Practice Parameter reviews assessment of respiratory insufficiency and provides practice recommendations for management and is included in Appendix F. However, in addition to the clinical questions addressed in the Practice Parameter, there are several other questions in respiratory care that are critical in evaluating end-of-life care for patients with ALS. These include:

- *What is the ideal way to manage patients who do not have a plan regarding respiratory ventilator support? Some patients cannot make a decision and will by default end up on ventilatory support. In discussing the initiation of non-invasive or invasive ventilation, the ethics and logistics of withdrawal of these interventions should be part of that discussion.*
- *How can health care providers work optimally with those patients who have arrived unexpectedly at the decision for respiratory support (e.g., those where monitoring was not performed, or who were unable to make a timely decision, or in whom the diagnosis was made concurrently with the detection of respiratory failure)? For many, the decision-making process in these patients is deferred to health care providers.*
- *While withdrawal of ventilator support is legal and ethical, studies need to be done to assess the optimal way to do this in practice.*
- *What is the best way to care for patients who change their minds regarding ventilator support? Advance Directives stipulating no ventilator support may be insufficient if not accompanied by a concrete plan of management of respiratory symptoms. This may include overt involvement of palliative care or hospice, education and counseling regarding emergency services and the risk of emergency intubation by emergency medical training staff or emergency room staff.*
- *What is the best way to manage expectations and raise quality-of-life issues for patients opting for ventilatory support? The quality of life of those on ventilators needs to be studied to see if there are differences in those with planned and unplanned transitions to ventilator status.*

Clearly, the management of respiratory issues in ALS is complex and involves a number of areas of concern outside of the decision regarding initiation and discontinuation of respiratory support. A review of the published evidence, existing resources and helpful tools that address many of these questions is included in Appendix D.

Identification of Existing Gaps

Despite the near universal occurrence of respiratory failure in the patient with ALS, management of respiratory care at the end-of-life period has not been well studied. Several areas remain unexplored, particularly those relative to the use of non-invasive ventilatory support and the current availability of smaller portable invasive ventilators and other mechanical devices. Management of distressing symptoms and signs of choking, aspiration,

stridor and dyspnea have not been systematically analyzed. The effects of medications that may offer therapeutic benefits on respiratory parameters also require systematic analyses.

Despite the strong relationship between respiratory dysfunction and death in ALS, great variability exists in the diagnosis and treatment of respiratory failure. Similarly, there is a lack of uniformity in the timing of NIPPV and invasive ventilation (IV). This substantial variability from patient to patient suggests that for a subset of patients respiratory care is suboptimal. Invasive procedures, such as placement of a PEG tube, have not always been performed with an understanding of the pulmonary status of the patient. This oversight carries significant morbidity and mortality risks.

Another gap is that for some health care providers the use of permanent ventilation is not a favorable treatment option for a variety of reasons. Physicians' opinions regarding respiratory support limitations and benefits might influence patients who may have a desire to stay alive and choose to have appropriate tertiary support (or vice versa, where patients choosing not to use respiratory support may be persuaded or influenced to start respiratory therapy).

Respiratory support is very expensive, and inadequate financial resources, including payment from third party payers, may influence the type of care patients with ALS receive. For some patients, funds are provided, but these are usually insufficient to cover all the costs of care—leaving a large financial burden on caregivers and families.

Recommendations to the Field

Practice Recommendations

- *Adhere to Practice Parameter recommendations for respiratory care for patients with ALS.*
- *Increase awareness and improve sensitivity and compassion among physicians for patients' needs for management of respiratory symptoms and their needs for respiratory assistive devices, such as NIPPV and In/Exsufflator.*

Research Recommendations

- *Conduct prospective studies to determine whether NIPPV improves quality of life and prolongs survival for patients with ALS. Assess how the use of respiratory devices facilitates or interferes with meeting the patient's goals for the end of life.*
- *Identify the following:*
 - *Whether early ventilatory support alters prognosis;*
 - *The optimal timing of ventilatory intervention; and*
 - *Better noninvasive methods of ventilatory support.*
- *Study patients' wishes and feelings about living with a ventilator and identify how much the insufficiency of current insurance coverage interferes with patients' wishes to live with a ventilator.*
- *Study methods of withdrawing both invasive and noninvasive respiratory support in ALS and assess how respiratory symptoms should be managed clinically.*

Policy Recommendation

- *Current Medicare criteria for NIPPV are too strict. Lobby Medicare to liberalize the criteria allowing NIPPV. Pressure Medicare to lower the barrier for reimbursement for NIPPV and tracheostomy.*

Nutrition and Hydration

Management of nutritional care for patients with ALS becomes more challenging with disease progression as patients lose motor function, particularly the ability to feed themselves and to swallow. As a result, non-oral routes of providing nutrition and hydration are implemented. Guidelines on how to monitor the nutritional state and provide nutritional support are reported in the Practice Parameter. The literature reviewed in Appendix D suggests that patients are very reluctant to proactively accept PEG. When the decision is finally made to move forward with a PEG, the disease severity often is more advanced and the benefits of PEG are less evident. The consensus is that PEG should be placed earlier in the disease process—yet consensus defining the optimal time for PEG placement has not been established.

Identification of Existing Gaps

One gap is addressing the patient's reluctance to proactively accept PEG. Within the medical community there may be considerable variation on how rigorously PEG is recommended. Additionally, as the illness progresses and patients approach the end of life, decisions need to be made regarding withdrawal of nutritional and hydrational support, yet optimal timing in the context of end-of-life care is not known.

Recommendations to the Field

Practice Recommendations

- *Proactively discuss the importance of nutritional care with patients with ALS, including earlier initiation of PEG.*
- *Adhere to the recommendations made for nutritional care in the Practice Parameter recommendations.*
- *Discuss Advance Directives regarding PEG and hydration, including self-determination of withdrawing nutritional and hydrational support during the end of life.*

Research Recommendations

- *Prospectively investigate survival benefits and quality-of-life benefits of PEG.*
- *Assess nutritional needs during the end of life in patients with and without PEG.*

Policy Recommendation

- *Increase coverage for supplemental medical equipment needed for feeding patients with ALS. This includes feeding tubes, special equipment for eating and drinking and nutritional supplements.*

Speech and Communication

Identification of Existing Resources

Communication becomes increasingly challenging with progression of the disease, ultimately resulting in the patient's inability to use speech or motor movements. Therefore, establishing communication strategies early in the course of illness will improve communication at the end of life. There are many different assistive devices to aid in communication, and these are listed in Appendix D.

Identification of Existing Gaps

Some patients may not have access to a speech-language pathologist (SLP), particularly one with expertise in augmentative and alternative communication (AAC). Speech-language pathologists help direct patients to appropriate technology to aid in communication. Many health care providers are unaware of the technology available to help with speech and communication.

Some speech-language pathologists may not have adequate experience or specific training and preparation to handle patients with ALS in need of augmented communication. There is a lack of adequate funding to pay for the evaluation, to purchase the device and to train the patient to use the device. The current documentation process for funding augmentative communication devices is too lengthy and complicated and generally falls outside the limited time patients with ALS have to use such devices. The use of augmentative devices at the end of life has not been studied.

Recommendations to the Field

Practice Recommendations

- *The ALS Peer Workgroup calls on the AAN Quality Standards Subcommittee to initiate the second Practice Parameter Guideline in ALS and address management of communication impairment in ALS.*
- *Improve content of existing Web sites and educational materials about communication for patients and families, including printed materials such as brochures and booklets.*
- *Educate health care providers about what resources are available for patients and their families (facilities, reimbursement and health care coverage). This can be done at national meetings (courses and lectures), independently sponsored courses and conferences.*
- *Establish training curricula accredited through ALSA/MDA that require a minimal level of expertise among clinic staff in assistive technology use and other communication initiatives.*
- *Require augmentative and alternative communication (AAC) expertise of interdisciplinary ALS clinics (by the supporting patient advocacy organizations—ALSA and MDA).*
- *Create regional centers that could provide AAC evaluation, loans of devices and post-placement training of patients and families.*
- *The American Speech Language and Hearing Association's (ASHA) Special Interest Division on Augmentative and Alternative Communication should create a list of certified speech-language pathologists (SLPs) with a minimum number of years of experience in the provision of services to the AAC population and proof of continuing education credits in AAC.*

Research Recommendations

- *Identify problems of acquisition and utilization of AAC.*
- *Define the impact of inability to communicate on quality of life and interaction with health care providers. Determine the effectiveness of AAC intervention for individuals with ALS and its impact on quality of life.*
- *Determine whether AAC prevents or reduces caregiver burden and to what extent. Examine if the effect of AAC intervention makes a positive change in the domains of functional limitation, disability and societal limitations according to the World Health Organization (WHO) framework.*

Despite the importance of good communication, physician education and training in these skills are largely neglected.

Policy Recommendations

- *Third-party payers must provide adequate reimbursement for speech-language pathologists to evaluate and train patients who need assistive communication devices.*
- *Third-party payers other than Medicare and Medicaid, especially HMOs, must add assistive communication devices as a covered benefit.*
- *Third-party payers must streamline documentation requirements for assistive communication devices.*

Depression and Pseudobulbar Affect

Identification of Existing Resources

Behavioral distress in ALS is common and presents a number of different signs and symptoms, including depression and anxiety. With progression of motor neuron loss, pseudobulbar affect occurs in a group of patients with ALS. It presents as sudden onset of excessive laughing or crying, with no relevant trigger. The Practice Parameter reviewed the current management recommendations for patients with onset of these issues, which usually occur prior to the end of life. The few reports on the prevalence and management of behavioral symptoms at the end of life are reviewed in Appendix D.

Identification of Existing Gaps

Currently, there are no standardized algorithms available that instruct how and when health care providers should screen for behavioral comorbidities (e.g., anxiety, depression) at the end of life in ALS. With progression of the disease, testing for depression is more challenging and tools adapted for end-of-life screening for depression are lacking.

The clinical benefits of treating depression and pseudobulbar affect are not well studied in ALS. For example, the role of psychostimulants (methylphenidate) for depression at the end stages of other diseases (cancer, HIV/AIDS) offer therapeutic benefit; but, possible benefits of psychostimulants in ALS at the end of life have not been studied. Additionally, as the disease progresses and likelihood of depression and pseudobulbar affect (or other behavioral effects) increases, there are no protocols for preparing the patient and family for these changes that might occur.

Recommendations to the Field

Practice Recommendation

- *Screen for signs and symptoms of depression and anxiety in patients with ALS; offer treatment (e.g., SSRIs) and counseling for depression and anxiety, especially at the end of life.*

Research Recommendations

- *Examine the epidemiology, cause and impact of depression during the end of life.*
- *Investigate and standardize the optimal approach to screening and diagnosing depression, anxiety and pseudobulbar affect in patients with ALS during the end of life.*
- *Investigate effective therapies for treatment of depression, anxiety and pseudobulbar affect during the end of life in patients with ALS.*

Insomnia

Identification of Existing Resources

For the patient and family, insomnia is a disconcerting symptom, and its cause varies. Some patients experience shortness of breath, and others are anxious. One recent study suggests that insomnia is prevalent and can be severe in the last month of life (Ganzini et al., 2002).

Identification of Existing Gaps

Both in clinical practice and in the medical literature, there is inadequate evidence on the diagnosis, incidence, prevalence and management of insomnia in patients with end-stage ALS. Similarly, there is neither evidence on the safety and efficacy of medical interventions for insomnia, nor on factors other than respiratory insufficiency that may contribute to insomnia.

Common pharmacological intervention for insomnia includes the use of analgesics, hypnotics or sedatives, which could possibly hasten death. The use of such pharmacological therapies needs evaluation, and the impact of such pharmacotherapies on mortality needs to be assessed.

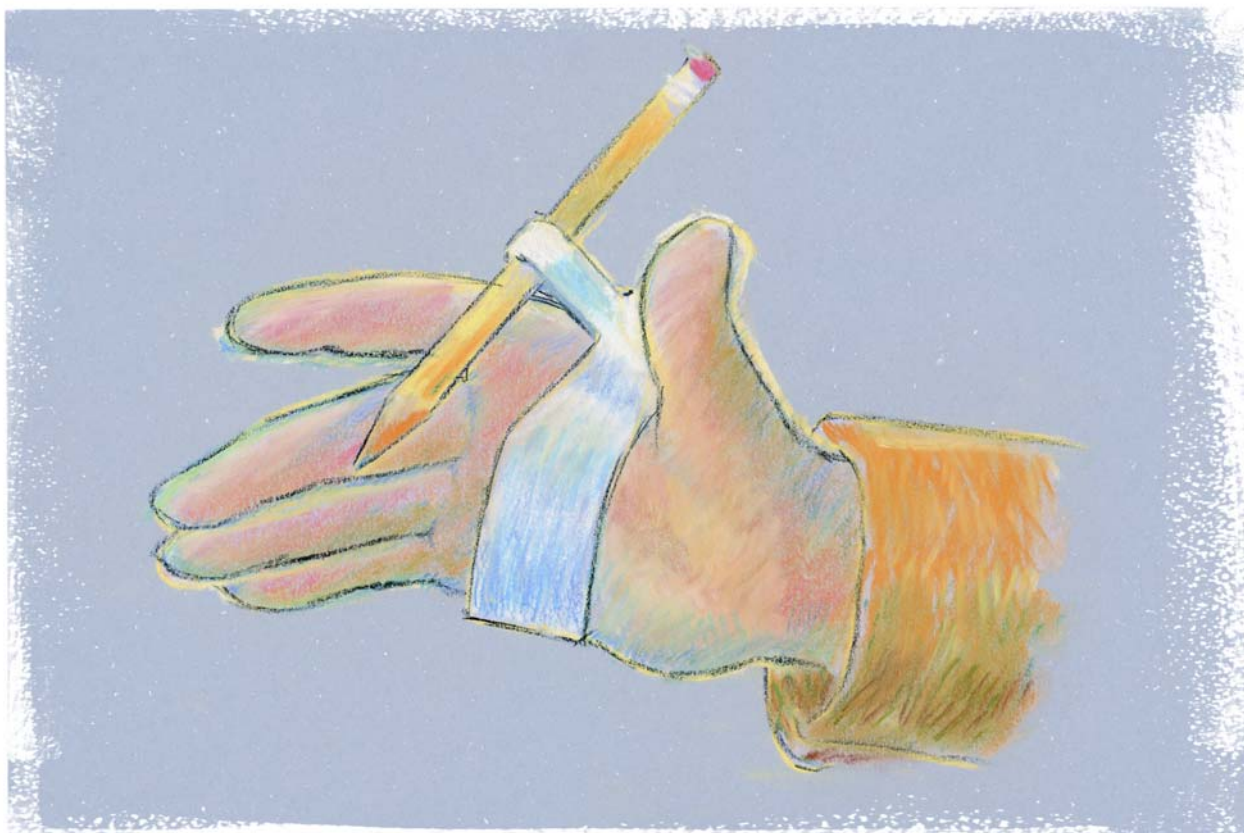
Recommendations to the Field

Practice Recommendations

- *Assess insomnia in patients with ALS during each health care visit and include symptom surveillance in home care. The origin of insomnia should be precisely determined and therapy directed at the underlying cause (Miller et al., 1999).*
- *Inquire about signs and symptoms of insomnia, particularly during the end of life.*
- *Use appropriate sedative-hypnotics in patients with insomnia due to respiratory insufficiency.*

Research Recommendations

- *Define the incidence and causes of insomnia in ALS, and study the relationship of insomnia to quality of life at the end of life.*
- *Investigate (prospectively) the optimum management of insomnia in ALS patients at the end of life.*
- *Develop evidenced-based algorithms for management of insomnia, particularly in patients who choose not to use NIPPV.*



Policy Recommendation

- Educate health care providers about not withholding sedative-hypnotics in patients with respiratory insufficiency.

Pain

Identification of Existing Resources

There is widespread misperception that pain is not a significant problem at the end of life in ALS. This may originate from literature that emphasizes the diagnostic criteria, including absence of sensory findings. Thus, the health care provider may not address pain complaints. Also, patients may not have the ability to communicate the nature or origin of their pain. A current review of management of pain in ALS is summarized in Appendix C and in the Practice Parameter.

Identification of Existing Gaps

There are no standardized tools to measure severity of pain specifically in patients with ALS. Also, the impact of pain on the patient with ALS, especially for patients at the end of life, has not been studied. Many health care providers managing pain in patients with ALS may not be pain specialists and may not be aware of the WHO pain management guidelines (Appendix D). One of the most challenging tasks for health care providers is the appropriate use of opioid medications for patients in pain at the end of life. Many health care providers are uncomfortable with using high levels of opioids, and there are no specific protocols for medication use in this population of patients at the end of life.

Recommendations to the Field

Practice Recommendations

- Follow established WHO guidelines for pain management, including liberal use of opioids and narcotic analgesics.
- Utilize nonpharmacological interventions in the routine management of patients with pain, including range of motion exercises, frequent repositioning, therapeutic mattresses, relaxation and diversion.
- Establish a method of quantitating symptom severity prior to the loss of verbal communication.

Research Recommendations

- Develop ALS-specific pain protocols, and determine how to evaluate pain in anarthric patients.
- Define specific pain syndromes in ALS, including the frequency and nature of pain and the severity of different syndromes.
- Define the impact of pain on insomnia, depression and quality of life at the end of life.
- Develop pain protocols specific to patients with ALS.

Last Hours

Identification of Existing Resources

The right to discontinue both invasive ventilation and nutritional support is legal and morally valid in the U.S. Every U.S. citizen has the right to stop ventilator therapy and any other type of life-sustaining therapy, as determined by the U.S. Supreme Court in the 1990 Cruzan ruling. The norms and accepted standards are to follow the valid treatment refusal of a competent patient. In ALS, this often requires establishing a communication system with the patient and proactively obtaining Advance Directives concerning discontinuing ventilator support in patients choosing permanent ventilation or NIPPV. However, the issues surrounding the discontinuation of ventilatory support in patients with ALS in practice can be controversial and difficult to manage (Appendices C and D). For this reason, the establishment of Advance Directives is an important step in guiding the decision-making processes surrounding the last hours of life.

Identification of Existing Gaps

To date, current treatment algorithms on withdrawing respiratory support at the end of life are available for some other disease states, but not specifically for ALS. Many clinicians and health care providers are neither experienced nor comfortable with the clinical management of patients with ALS during the last days of life. The impact of the last few hours has not been well studied. Another gap is that the legal and ethical guidelines surrounding withdrawal of respiratory support are not clear, especially in those patients who become "locked-in" after years of ventilator support and have no Advance Directives regarding discontinuing support.

Legal and ethical guidelines surrounding withdrawal of respiratory support are not clear.

Recommendations to the Field

Practice Recommendations

Management of Patients on Ventilatory Support:

- Discuss with patients and families when to withdraw ventilatory support and what to expect at this difficult time. Review Advance Directives with the patient and family. Attempt to establish the basis or withdrawal of ventilation prior to initiating ventilation. Discuss the following issues prior to discontinuing respiratory support:
 - The expected manner and time course of death;
 - Medications that will be used to manage symptoms; and
 - Possible use of sedation.
- Maintain comfort and a physician presence at the bedside for termination of ventilatory support. All arrangements should be in place prior to the removal of support:
 - All family members wishing to be present should be nearby;
 - All cultural or religious rituals should be discussed, planned and implemented;
 - The location should be prepared, if possible (for example, peaceful lighting and music if desired); and
 - If in a home or nursing home setting, all potentially useful medications and suction should be readily available.
- Parenteral administration of necessary medications will provide a more rapid onset of action (unless there is already an indwelling IV in place, the subcutaneous route is preferred):
 - It is ethically appropriate to sedate to unconsciousness, but as noted in the Practice Parameter, muscle-paralyzing agents should not be used;
 - If oxygen is not already in use, it should not be instituted at this time. If oxygen is being used, flow should not be increased and discontinuation should be considered. For patients with dyspnea, oxygen may be used to alleviate symptoms; and
 - Once comfort has been obtained, positive expiratory pressure can be discontinued, followed by conversion to a T-piece.

Management of Patients Not on Ventilatory Support:

- The same principles stated above should be used, except that medications should be initiated in a more gradual fashion since there is no specific event to anticipate:

- *The physician should be readily accessible for medication adjustments;*
- *Discontinue use of NIPPV completely, if patients are using NIPPV;*
- *Appropriate medications for sedation may be given around-the-clock if distress recurs frequently. All other unnecessary medications should be stopped;*
- *All monitoring (vital signs, oximetry) should be discontinued. If hospitalized, no further laboratory testing or X-rays should be done; and*
- *Oxygen should not be started but may be used to treat signs of dyspnea.*

Research Recommendations

- *Develop treatment and management protocols/algorithms on how to manage pain and withdrawal of ventilatory support specifically in patients with ALS at the end of life.*
- *Examine how patients die in a natural setting, as compared to those patients on ventilatory support.*

Policy Recommendation

The ALS Peer Workgroup calls on health care policy-makers to investigate how to ensure that patients can die at home or in a hospice setting, with or without ventilatory support. In achieving this goal, the patients' comfort should be a priority and their dignity maintained. Establishing these guidelines will ensure that physicians, patients' families and caregivers are comfortable with the interventions employed at the end of life.

References

- Annas, GJ. "Nancy Cruzan and the right to die." *New England Journal of Medicine*, 323: 670-673, 1990.
- Bernat JL. "Ethical and legal issues in palliative care." *Neurologic Clinics*, 19(4): 969-987, 2001.
- Bradley WG, Anderson FA, Bromberg MB, Yadollah H, Miller RG and Ross M. "Changes in management of ALS since Publication of AAN Practice Parameter: Evidence from the ALS Patient Care Database." *Neurology*, 58:A78, 2002.
- Brooks BR, Lewis D, Rawling J, et al. "The natural history of amyotrophic lateral sclerosis." In: *Motor Neuron Disease*, Williams AC (ed.). London: Chapman and Hall Medical, 1994.
- Buckman R. "Communication skills in palliative care: A practical guide." *Neurologic Clinics*, 19(4): 989-1004, 2001.
- Cancer Pain Relief and Palliative Care*, 1990. World Health Organization, Technical Report Series 804:1-75.
- Carver AC, Vickrey BG, Bernat JL, Keran C, Ringel SP and Foley KM. "End-of-life care: A survey of US neurologists' attitudes, behavior, and knowledge." *Neurology*, 53(2): 284-293, 1999.
- Education for Physicians on End-of-Life Care (The EPEC Project). © EPEC Project, The Robert Wood Johnson Foundation, 1999.
- Fried TR, Bradley EH, Towle VR and Allore H. "Understanding the treatment preferences of seriously ill patients." *New England Journal of Medicine*, 346(14): 1061-1066, 2002.
- Ganzini L, Johnston W and Silveira M. "The final month of life in patients with ALS." *Neurology*, 59: 428-431, 2002.
- Miller RG, Rosenberg JA and Gelinas DF, et al. "Practice parameter: The care of the patient with amyotrophic lateral sclerosis (an evidence-based review)." *Neurology*, 52(7): 1311-1323, 1999.
- Mitsumoto H, Chad DA, Pioro EP (eds). *Amyotrophic Lateral Sclerosis*. Philadelphia: FA Davis, 1998.
- Munsat TL, Andres PL, Finison L, Conlon T and Thidodeau L. "The natural history of motoneuron loss in amyotrophic lateral sclerosis." *Neurology*, 38: 409-413, 1988.
- Oliver D, Borasio GD and Walsh D. *Palliative Care in Amyotrophic Lateral Sclerosis*. Oxford University Press, 2000.
- Ringel SP, Murphy JR, Alderson MK, et al. "The natural history of amyotrophic lateral sclerosis." *Neurology*, 43: 1316-1322, 1993.
- Roethlisberger FJ, Dickson W. *Management and the Worker*. Cambridge, Mass.: Harvard University Press, 1939.



ALS Peer Workgroup Members

Hiroshi Mitsumoto, M.D., D.Med.Sc. Chair

Wesley J. Howe Professor of Neurology
Director, The Eleanor and Lou Gehrig
MDA/ALS Research Center
Division Head, Neuromuscular Disease
College of Physicians & Surgeons
Columbia University
Neurological Institute
New York, N.Y.

Stanley H. Appel, M.D.

Director, MDA/ALS Research and Clinical Center
Chair, Department of Neurology
Baylor College of Medicine
Houston, Texas

Joshua O. Benditt, M.D.

Professor of Medicine
Pulmonary and CCM
University of Washington Medical Center
Seattle, Wash.

James L. Bernat, M.D.

Neurology Section
Dartmouth-Hitchcock Medical Center
Lebanon, N.H.

Gian Domenico Borasio, M.D., DipPallMed

Interdisciplinary Palliative Care Unit and
Department of Neurology
Munich University Hospital - Grosshadern
Munich, Germany

Mark Bromberg, M.D., Ph.D.

Director, Neuromuscular Program and
Motor Neuron Disease Clinic
Department of Neurology
University of Utah
Salt Lake City, Utah

Ira Byock, M.D.

Director
Promoting Excellence in End-of-Life Care
The University of Montana
Missoula, Mont.

Alan C. Carver, M.D.

Mount Sinai Medical Center
Department of Neurology
New York, N.Y.

Lora L. Clawson, M.S.N., C.R.N.P.

Johns Hopkins University School of Medicine
Baltimore, Md.

M. L. (Maura) Del Bene, M.S., R.N., N.P.-P.

Nurse Practitioner
The Eleanor & Lou Gehrig
MDA/ALS Research Center
Columbia University
New York, N.Y.

Wendy Johnston, M.D., F.R.C.P.C.

Associate Professor, Neurology
University of Alberta
Edmonton, Alberta Canada

Edward J. Kasarskis, M.D., Ph.D.

Department of Neurology
University of Kentucky and VA Medical Centers
Lexington Ky.

Susan B. LeGrand M.D., F.A.C.P.

Director of Education
The Harry R. Horvitz Center for Palliative Medicine
The Cleveland Clinic Foundation
Cleveland, Ohio

Mary Lyon, R.N., M.N.

Vice President, Patient Services
The ALS Association
Calabasas Hills, Calif.

Raul N. Mandler, M.D.

Professor of Neurology
Director, The ALSA Certified Center
The George Washington University
Washington, D.C.

Jane McCarthy, M.Sc., M.P.H.

Director of Affiliate Patient Services (Former)
The ALS Association
Calabasas Hills, Calif.

Robert G. Miller, M.D.

Chairman, Department of Neurology
California Pacific Medical Center
San Francisco, Calif.

Theodore L. Munsat, M.D.

Director, Neuromuscular Research Unit
 New England Medical Center
 Professor of Neurology and Pharmacology
 Tufts University School of Medicine
 Boston, Mass.

Daniel S. Newman, M.D.

Harry J. Hoenselaar ALS Clinic
 Department of Neurology
 Henry Ford Hospital
 Detroit, Mich.

Robert L. Sufit, M.D.

Professor, Department of Neurology
 The Feinberg School of Medicine
 Northwestern University
 Chicago, Ill.

Rup Tandan, M.D., F.R.C.P.

Professor of Neurology
 University of Vermont College of Medicine
 Burlington, Vt.

Andrea Versenyi, C.S.W.

Patient Services Director
 The ALS Association Greater New York Chapter
 New York, N.Y.

Visit the comprehensive Web site of *Promoting Excellence in End-of-Life Care* for reports from other workgroups dedicated to long-term changes to improve health care for dying people and their families: <http://www.promotingexcellence.org> or contact:

Ira Byock, M.D., Director

Promoting Excellence in End-of-Life Care
 The University of Montana
 1000 E. Beckwith, Missoula, MT 59812
 Phone: 406/243-6601
 Email: ibyock@aol.com

Jeanne Sheils Twohig, M.P.A., Deputy Director

Promoting Excellence in End-of-Life Care
 The University of Montana
 1000 E. Beckwith, Missoula, MT 59812
 Phone: 406/243-6602
 Email: TwohigJS@mso.umt.edu

Appendices (on enclosed CD)

Appendix A

Better Spiritual Care: Facing Death and Finding Hope

Appendix B

*Psychosocial Care, Bereavement, Spirituality,
Caregivers and Quality of Life*

Appendix C

Ethics, Communication and Decision Making

Appendix D

Symptom Management

Appendix E

Access to Care, Cost of Care and Health Policy

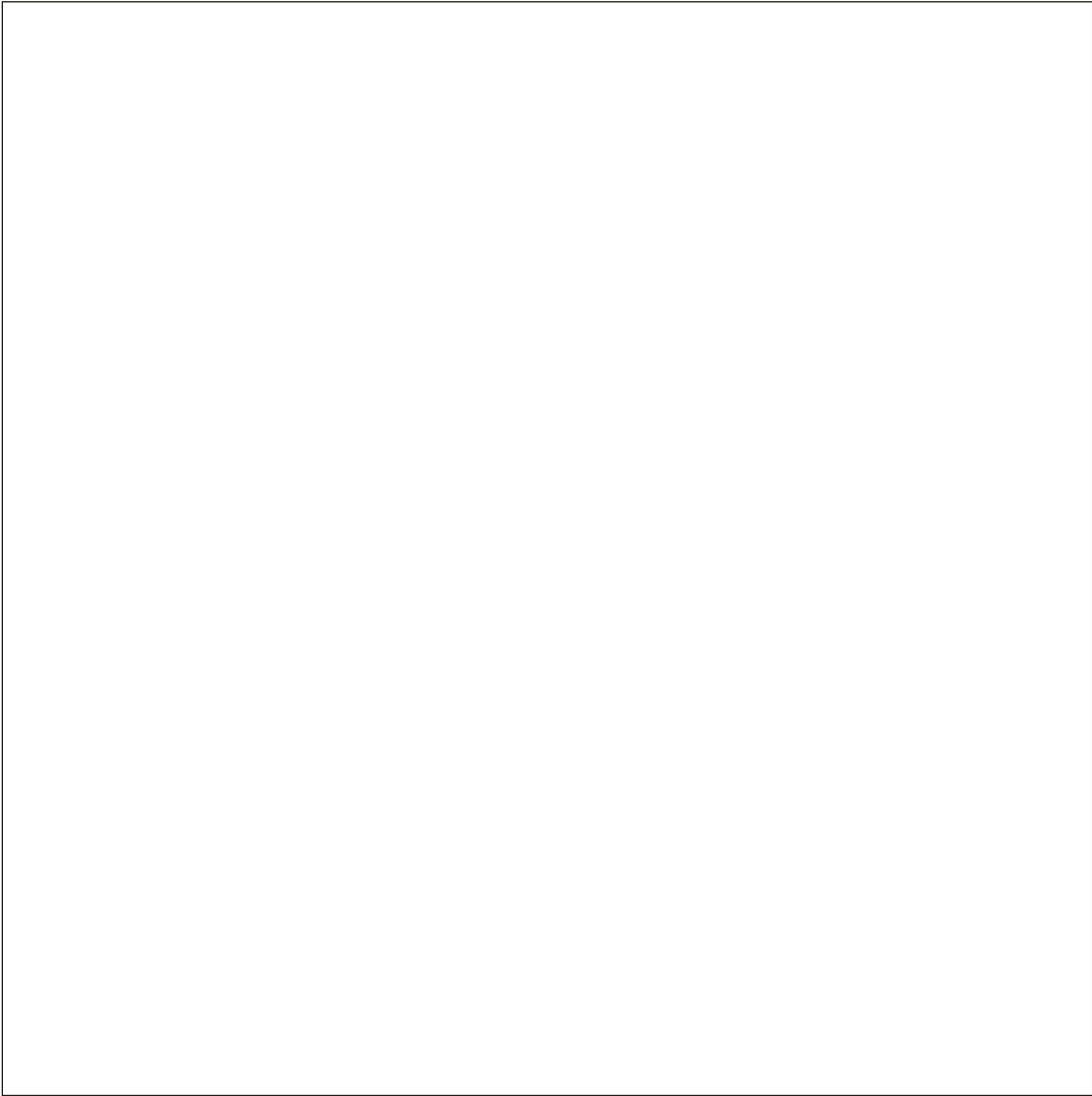
Appendix F

AAN Practice Parameter (Miller et al., 1999)

Appendix G1 and G2

Published Abstracts 2001-2002

Presentation Slides 2001-2002



**The ALS Report and Appendices are available at
www.promotingexcellence.org**



PROMOTING EXCELLENCE IN END-OF-LIFE CARE

A NATIONAL PROGRAM OF
THE ROBERT WOOD JOHNSON FOUNDATION

1000 East Beckwith Avenue
Missoula, MT 59812
406-243-6601 Phone
406-243-6633 Fax
excell@selway.umt.edu
www.promotingexcellence.org

