

Appendix E

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Costs, Access to Resources and Health Policy

INTRODUCTION

Experience indicates that as ALS progresses, health care needs become more demanding, yet health care coverage is often less generous. The long-term goal of medical care for ALS patients is to maintain function and quality of life in the most cost-beneficial manner. As the disease progresses and disability rapidly advances, health care providers are faced with the arduous task of meeting the medical needs of patients in an environment that places restrictions on resource utilization. The care providers must understand how to work within the current health care system to meet the needs of patients with ALS in a relatively short time period. The overall objective, therefore, is to start the planning well in advance so as to make medical resources available when patients are most likely to need them.

Cost of Care and Economic Burden of ALS

What are the primary costs associated with end-of-life care?

There are no published prospective studies that assess the total cost of care at the end of life in patients with ALS. Costs associated with late-stage and end-of-life care in ALS come from numerous medical interventions, including percutaneous endoscopic gastrostomy (PEG), non-invasive ventilation (NIPPV) with bilevel positive airway pressure, mechanical ventilation, drug therapy (such as the glutamate antagonist, riluzole and other symptomatic medications), emergency room and urgent care visits, hospitalization, home nursing care and hospice care. For each of these areas, costs vary depending on attitudes and decisions of patients, duration of treatment, complications and co-morbidities.

One prospective study undertaken at the New England Medical Center, Boston, examined the cost of care for patients with ALS, taking into consideration realistic costs (Munsat TL, Riviere M, Thornell B, Andres P, and Lloyd K. "Economic burden of amyotrophic lateral sclerosis in the United States," unpublished data). However, this study represents cost of care prior to the extensive use of NIPPV and riluzole, which are costly treatments being used in a significant proportion of patients. Data on direct and indirect costs and medical resource utilization was collected in 1995 (see Tables 2, 3 and 4) from 71 patients in four ALS Health States (as defined in Table 1). The total average annual cost of ALS care increased from \$5,331 for Health State 1 to \$ 79,591 for Health State 4 (Tables 2 and 3). The major determinant of costs were diagnostic testing in Health State 1, personal expenses in Health State 2 and time lost by caregivers in Health States 3 and 4 (Table 2). The annual patient-borne cost increased from \$1,119 in Health State 1 to \$ 42,681 in Health State 4, and was accounted for largely by time lost from work by caregivers in Health States 3 and 4 (Table 3). The annual cost of home ventilation and nursing was considerable in patients electing respiratory support (Table 4).

Other studies have documented the substantial costs of care across the disease spectrum in ALS patients participating in pharmaceutical industry-sponsored clinical trials (Table 5). A recent hospitalization study derived from a 1996 Nationwide Inpatient Sample (Lechtzin et al., 2001) compared costs in 1600 ALS patients and 5.3 million non-ALS patients. The average length of stay in ALS patients was longer (8.4 days versus 5.4 days), charges were greater (\$19,810 versus \$11,924), and in-hospital mortality was higher (15.4% versus 3.0%) as compared to non-ALS patients. Most common causes of admissions were dehydration and malnutrition in 35.9%, pneumonia in 31.7% and respiratory failure in 24.9% of ALS patients. Only 38.2% of ALS patients were routinely discharged (as compared to 73.4% of non-ALS patients), and 18.8% required home health care (as compared to 7.8% of non-ALS patients). Another 21.3% of ALS patients were sent to a skilled nursing facility, short-term hospital or intermediate care

facility. Thus, ALS patients have lengthy and costly admissions, high in-hospital mortality and few routine discharges.

Pharmacotherapy

Pharmacotherapy is often reserved for management of symptoms (Sufit, 1997), such as sialorrhea, constipation and pain (see section on Symptom Management). One exception is riluzole, a disease-modifying agent that modestly prolongs survival (Bensimon et al., 1994; Lacomblez et al., 1996). For most symptomatic pharmacotherapies there is not an enormous cost burden for oral administration. However, intravenous administration of antibiotics, anxiolytics and narcotics may be associated with increased costs during late-stage disease and the end of life.

The cost of treatment with riluzole is considerable, amounting to \$750-850 per month; however, it allows patients to remain in milder health states wherein the functional status is least impaired for longer, as compared to placebo (Riviere et al., 1998). Consequently, from the societal perspective, its benefits may exceed costs (Ginsberg, Lev, 1997; Tavakoli et al., 1999). In addition to riluzole and symptomatic treatments, vast sums of money are spent by ALS patients in the self-administration of health food supplements, antioxidants and herbal remedies. To date, the value of these therapies is unproven.

PEG

The Practice Parameter guidelines for the management of ALS published recently by the American Academy of Neurology (AAN) Practice Standards Subcommittee encourage the placement of PEG in patients with significant bulbar dysfunction and weight loss (Miller et al., 1999) (see section on Symptom Management). Some studies suggest that early PEG placement may confer improved outcomes (Chio et al., 1999; Kasarskis et al., 1999; Mathus-Vliegen et al., 1994). In 1995, the estimated cost of PEG placement was \$1,500, and an additional \$6,000 per year was required for supplies of enteral nutrition formula and maintenance costs (Klein, Forshew, 1996). Additional costs, usually not considered, include those related to the treatment of complications of the procedure, and the hiring of private aides to do feedings since by law certified home health aides are not permitted to handle PEG feeding. Insertion of PEG late in the disease, as the patient enters a more advanced stage, may not offer the desired clinical benefits and may expose patients to a higher risk of complications of the procedure that may necessitate hospitalization (Miller et al., 1999). As a result, PEG placement has cost implications as the patient enters a more advanced stage of the disease.

NIPPV

Noninvasive ventilation is being increasingly utilized for the treatment of respiratory compromise in ALS, and its use may extend well into the later stages of the disease (Bach, 1995; Sherman, Paz, 1994). Based on an evidence-based review, the AAN Practice Parameter guidelines provide an algorithm for the management of respiratory compromise with NIPPV (Miller et al., 1999) (see section on Symptom Management). NIPPV can reduce costs by decreasing the risk of respiratory infections and by easing the burden on caregivers (Hillberg, Johnson, 1997). NIPPV reduces fatigue, improves quality of life and prolongs survival in ALS patients (Aboussouan et al., 1997 and 2001; Kleopa et al., 1999; Pinto et al., 1995). Unfortunately, there are no studies of the cost associated with use of NIPPV in ALS patients; however, an estimated cost of \$400 per month quoted in a review (Borasio et al., 1998) is considerably lower than the cost of mechanical ventilation (see below) and needs to be confirmed in future studies. Although the cost of renting a Cough Assist device (\$270 to \$330 per month) is less than that of a machine used for NIPPV, insurance coverage is more problematic for the Cough Assist device.

Home Mechanical Ventilation

The costs associated with home ventilation are of significant concern for many patients and may impact patient and family choices regarding treatment. For example, in 1993 to 1994 a comparative study in 75 patients from Ohio, western Pennsylvania, West Virginia and California suggested that the cost of home mechanical ventilation (\$180,000 annually) was about half that of ventilated hospitalized patients

(\$350,000 annually) (Moss et al., 1996). In a study from northern Illinois in 1993, the mean estimated monthly cost of home mechanical ventilation was \$12,771 (range \$2,500 to \$35,000) and the mean annual cost was \$153,252, with the majority of expenses attributable to nursing support (Moss et al., 1993). In another study from Northern California conducted between 1985 and 1992 in 34 ALS patients, the cost of home care for mechanical ventilation was substantial: Equipment rental for ventilatory support was \$20 to \$35 per day, and nursing staff or paid personal caregiver cost ranged from \$64 to \$160 per day (for eight to 16 hours of service) (Oppenheimer, 1993). The expense associated with a licensed nurse ranged from \$200 to \$400 per day, and additional costs accrued from supplies, medications and gastrostomy feeds. Many insurers do not cover the costs of home nursing for ventilated patients. Results from these studies show that the cost of care at the end of life must be taken in the context of *total* care costs, to help in the planning and decision-making processes. Insurers may grant approval for home ventilatory support when health care provision is considered in a relative care-cost scenario. Nevertheless, even when approval is forthcoming for 24-hour nursing for home-ventilated patients such services are often difficult to provide because of a nationwide shortage of experienced nurses. This, in turn, increases the indirect cost of care in lost wages of caregivers who stay at home to provide care for their loved ones.

Where Do Costs Originate for End-of-Life Care?

In order to understand where costs associated with end-of-life care originate, it is important to consider where patients with ALS are spending the end-of-life phase and what treatments they are receiving (Albert et al., 1999). Of the 373 patients who died in the ALS CARE Database between 1996 and 1999, 56% died at home, 19% in a hospital and 7% in a residential hospice facility (Bradley et al., 2001). Home hospice services were utilized by 47% of patients who died at home. Medications to control terminal pain and distress were used by 66% of patients, and 89% reportedly died peacefully. In a retrospective interview study of caregivers who were present at the time of death of 121 ALS patients in Germany, 30% of patients had PEG and 21% were on mechanical ventilation (mainly NIPPV) at time of death (Neudert et al., 2001). In this study, about 30% of patients received narcotics and/or benzodiazepines terminally, and almost 90% were said to have died peacefully. Unfortunately, no published studies have prospectively examined the frequency or cost of hospitalization or hospice use at the end of life in patients with ALS.

What Factors Impact Cost of Care at the End of Life?

The diagnosis and treatment of ALS generate a significant economic burden on patients and families. Cost of medical treatments can deplete financial resources in a short time, often forcing patients and caregivers to make decisions regarding medical care based on the actual cost of care. Financial burdens associated with ALS are not restricted to patients and their families; the economic burden is passed on to insurers and ultimately to society as increased premiums or restrictions in coverage.

The incidence of ALS in the community is low; few clinics have protocols in place for the diagnostic work-up and on-going treatment of patients with ALS. The ALS Practice Parameters are the best available benchmark in this regard (Miller et al., 1999). Differences in management philosophy and in physician practices among medical centers, and regional variations in reimbursement policies, lead to a wide variance in treatment regimes in the United States. This lack of a standard of care significantly impacts the cost of care and the allocation of available resources.

For example, during the diagnostic work-up extensive neuroimaging tests (MRI scans and CAT scans) are undertaken, and unnecessary laboratory tests are ordered (e.g., heavy metal determination in urine; serum GM1 antibodies) by some clinics, both of which drive up the cost of care for these patients early in the disease process. Ultimately, these high initial costs have a negative impact on overall resources available for end-of life care, at a time when the need for supportive medical care is often very high.

Different treatment practices among 39 large centers in North America were evident in a recent study of the use of NIPPV in ALS patients (Cedarbaum et al., 2001). This study indicated a range of utilization

from none at all to that in 40% of patients. In another survey of 20 ALS centers reporting on a combined experience in 2,357 patients, use of NIPPV varied from 0% to 50%, with a mean of 15% (Melo et al., 1999).

The popularity of large multidisciplinary clinics for the care of ALS patients has increased in recent years, despite the relative absence of clear outcomes data (Chio et al., 2001; Corr et al., 1998). Nevertheless, this trend and some recent preliminary data from the ALS CARE Database may suggest advantages of care in multidisciplinary centers over that in single-practitioner or single specialty treatment clinics (Rosenfeld, 2000). The existence of current multidisciplinary clinics is threatened because of the high cost of comprehensive care and inadequate reimbursement. Additional financial or staff support is available to some clinics from organizations such as The ALS Association (ALSA) and the Muscular Dystrophy Association (MDA), but this support is meager. Thus, studies of the effects of the mode of care delivery on cost and outcomes are desperately needed.

Conclusions On Cost of Care At the End of Life

The results from these studies collectively suggest that the costs associated with care of ALS are substantial, and increase exponentially as the severity of the disease progresses. However, significant differences in costs accrued at the end of life depend on site of care delivery, patient choices and biases toward selected procedures, level of education of patients, awareness of treatment options among patients and family caregivers, rate of progression of disease and financial planning. Consequently, although the focus of this initiative is toward issues of cost and resources at the end of life, financial planning for decisions made at the end of life needs to be initiated early in the disease process, preferably shortly after diagnosis. If not, decisions to accept medical interventions may not be based on an educated assessment of the pros and cons of the treatment or procedure, but rather on the economic constraints the intervention poses on patients and family members.

Specific studies on how to reduce costs and maintain or improve quality of life during the end of life are few, yet clearly the direct costs of home care, hospitalization and respiratory support, and the indirect cost of lost time at work (by patients and caregivers), may account for a significant portion of the economic burden associated with ALS.

Tools and Resources

What Resources are Currently Available for ALS Patients and Their Families?

A list of not-for-profit organizations, along with the support they provide, is given in Table 6.

Government Agencies

Medicare and Medicaid are the major government sponsored programs, and are discussed later. Other federal programs available nationally include medical care services through the Department of Veterans Affairs and CHAMPUS.

State and Local Level Resources

As with national level resources, there are both ALS-specific and general resources, including government agencies, at the state and local levels.

ALS-Specific Resources

ALS patients and families can access ALS-specific services available at local ALSA chapters and through local support groups sponsored by ALSA and MDA. These chapters help provide equipment loans, home visits, caregiver training and respite care that may service an entire state or a portion of a state. However, not every state has an ALSA chapter and the level of service varies by chapter. At the present time, one of the main goals of ALSA is to more consistently standardize the availability of services across chapters. In addition, MDA operates regional and local offices that provide patient services coordinators who work directly with patients and families to help coordinate appropriate care.

Currently the available clinical care resources for patients with ALS include ALSA's network of 19 Certified ALS Centers that provide excellence in ALS clinical care in 14 states, and 25 MDA/ALS Centers available in 22 states.

General Resources

In cases where neither ALSA nor MDA is able to provide technological support services that facilitate communication at the end of life, the Assistive Technology Act (AT Act) programs can be accessed. The AT Act was enacted to improve access to assistive technology assessments and devices through state-based programs. There are AT Act grant-funded programs throughout all 50 states and six territories. Program services may vary from state to state.

The Rehabilitative Engineering and Assistive Technology Society of North America (RESNA) provides assistance to each AT Act-sponsored program and is a clearing house for information on each program operating throughout the United States and six territories. Equipment purchases may or may not be covered by private health plans; Medicare does cover devices if they are designed as tools dedicated only for communication purposes.

Other Programs

Other state-level government resources include the Department of Aging and the Department of Mental Health. Program availability and scope will differ from state to state. The State of California Department of Mental Health offers families who have a family member affected by brain injury or neurological disease, including ALS, an excellent array of services through nine regional California Resource Centers located throughout the state. Services include counseling, support groups, respite care, caregiver training and education, counseling and legal and financial consultations.

Each state and local area throughout the country also may offer a myriad of resources that are unique and limited to a specific area. Obviously, it is beyond the scope of this document to list every possible resource. Therefore, known available national, state, and local level organizations should be looked upon as clearing houses for information on other available resources, even though it cannot be assumed that the

information will be all inclusive. Availability of, and access to, the Internet is critical for organizations and families to learn about other programs and services that may not be widely known.

Access to Resources

Access to resources depends on their existence in a geographic region, on the ability of patients to physically get to care (or have care come to them), and on their financial resources to cover the cost of care. As previously indicated, due to the rarity of the illness, there is a dearth of knowledgeable ALS specialists across the country. Patients who do not live in major urban environments may have no specialty care available. Previous sections covered treatment modalities and the costs of care; below we will look at policies defining the coverage of these costs and eligibility for obtaining available services.

Influence of Attitudes and Acceptance of Interventions on End-of-Life Care

At the end of life, patients generally choose one of two routes – hospice/palliative care or mechanical ventilation. Physician attitudes and recommendations strongly affect choices, attitudes and acceptance of end-of-life interventions by patients. Studies looking at these influences have only just begun, yet are needed as they may be among the primary determinants of ensuring the optimal scenario for patients and families during the end of life. For example, 65% of ALS patients who were interviewed said they wanted respiratory support (Rabkin et al., 2000). However, the North American ALS CARE Database shows that among 2,018 patients, 13% of whom had severe respiratory impairment and were not ventilator-dependent, only 28% were using NIPPV (Bradley et al., 2001). Of the 38% with FVC < 40% of predicted at the time of enrollment, only 9% were using NIPPV and 4% had received tracheostomy or ventilation. Patients exhibiting depression and distress express a desire for modest intervention with NIPPV; however, NIPPV is likely to be refused by patients who are older, are more depressed and report more pain with lower quality of life (Rabkin et al., 2000).

When interviewed about desires for accepting measures that might prolong life, 28% to 42% of ALS patients were in favor of PEG (Albert et al., 1999; Rabkin et al., 2000), and when followed for >12 months, were more likely to have undergone the procedure (largely in association with tracheostomy) as compared to those who were initially opposed to it (48% versus 8%) (Albert et al., 1999). About 20% of patients preferred PEG but rejected tracheostomy, and less than 2% of patients endorsed tracheostomy without PEG (Albert et al., 1999). In the ALS CARE Database 20% of the 2,018 patients had significant dysphagia (ALSFRS swallowing scores of zero, one, or two), of whom 30% underwent PEG; this included 12% of patients with FVC < 50% of predicted (Bradley et al., 2001). Patients exhibiting depression and distress are less likely to express a desire for invasive measures such as PEG, tracheostomy or cardiopulmonary resuscitation (Rabkin et al., 2000).

The attitudes, behavior and knowledge of the treating neurologists have a significant bearing on end-of-life care. For example, a recent AAN survey showed that 75% of ALS patients being cared for by neurologists specializing in ALS had completed Advance Directives, compared with only 55% to 57% of patients under the care of general neurologists and neuro-oncologists (Carver et al., 1999). In the same survey, discussions about “Do Not Resuscitate” orders were more frequent between patients and their ALS neurologists (75%) than with general neurologists (57%) or neuro-oncologists (55%). Neurologists specializing in caring for ALS patients were also less likely to administer very large doses of IV morphine to end the patient’s life as compared to general neurologists or neuro-oncologists.

*What Factors Influence Resource Accessibility at the End of Life?**Health Coverage*

An investigation into policies regarding coverage for ALS care reveals a fractured and inconsistent system. The medical community is just beginning to define practice parameters for all aspects of care for ALS patients. The ALS CARE Database shows that there is no one standard of practice across the country, and access to specialty care varies considerably. Without a strongly articulated standard of care, supported by data and research confirming the efficacy of such care, insurance companies have been left to develop their own policies regarding acceptable services and appropriate coverage.

Furthermore, insurance providers are separated into federal programs (Medicare), federal programs that are state-administered (Medicaid), and private insurers and HMOs. While the policies governing Medicare are fairly uniform across the country, Medicaid eligibility and coverage varies widely from state to state. Private insurers/HMOs may provide different levels of coverage depending on the size of the employer/purchaser pool or the cost of the policy (i.e., durable medical equipment [DME] may be covered by one policy, but may not be included in another less expensive policy). In brief, there are as many different policies for ALS coverage as there are different insurances.

Following is a brief explanation of the various types of insurance coverage for the care of ALS patients; it will not, however, include a description of all coverage.

Medicare

Medicare is a federal health insurance program begun in 1965 to provide health coverage for eligible persons who are 65 years of age or older, or for those with specific disabilities. Medicare is administered by the Centers for Medicare and Medicaid Services (CMS) (previously known as the Health Care Financing Administration or HCFA), which is an agency of the Department of Health and Human Services (DHSS). Medicare is administered solely by the federal government and is available to everyone who meets the requirements, regardless of income. Medicare provides coverage for medical care and hospice care for many ALS patients.

Until last year, patients with ALS were required to wait 24 months after qualifying for Social Security Disability Insurance (SSDI) before they were eligible to be covered by Medicare. In late 2000, the Medicare Waiver Act was enacted making it possible for eligible ALS patients to waive the 24-month waiting period and begin receiving benefits concurrently with their SSDI. However, for patients who are eligible for Medicare under the new Medicare Waiver Act, their private insurance may provide better end-of-life care benefits under an existing health plan than Medicare. Once an individual receives Medicare benefits, Medicare becomes the primary insurance if the individual is neither working nor covered under a group health plan provided by an employer (of either the patient or a working family member) of greater than 100 employees. Thus, the decision to enroll in the Medicare option should be made after a careful comparison of the different health plans available to ALS patients. Some patients may, therefore, wish to forego Medicare if they are able to remain on their private insurance programs. Accessing private health plan benefits is often done through a working spouse's health plan or COBRA.

Medicare coverage is divided into two parts: Part A and Part B. Part A covers inpatient hospital care, inpatient care in a skilled nursing facility (for 100 days only following an acute care admission), home health care (skilled care or care by home health aides in conjunction with skilled care on an intermittent basis), and hospice care. Part B covers physician services (other than for routine care), outpatient hospital care, diagnostic tests, ambulance services and durable medical equipment (DME). Medications are not covered under either Part A or Part B.

Medicare claims under Part A are processed by private insurance organizations called fiscal intermediaries contracted by the federal government. There are FIVE fiscal intermediaries across the country that divide claims according to geography (Table 7). Claims for DME are processed by agencies

called DME Regional Carriers (DMERCs) according to the state of residence of the patient and the services provided. The alacrity with which these intermediaries respond and the accuracy of their knowledge about Medicare guidelines varies substantially, as does the accuracy of understanding of the Medicare Policy Manual. DMERCs may deny coverage where the policy manual would indicate it exists. Also, funds can be subsequently re-directed from a general special resource even though coverage was initially denied. Full knowledge of Medicare justification is, therefore, helpful when submitting claims, as only by knowing the policies can a provider adequately advocate for patients. However, although the policy manual is available on the CMS Web site, the information is difficult to find.

Although most insurers will determine coverage prior to provision of a DME, Medicare requires that DME be delivered to the patient before a claim is considered. This puts both the patient and the vendor in a difficult position: If Medicare does not approve coverage, the patient must either return the device (often not possible if it is a highly customized device) or pay out of pocket.

Medicaid

To be eligible for Medicaid, applicants need to prove their financial need for health care benefits. While the federal government sets guidelines, each state designs its own program within the limits of federal laws and regulations. Generally, the cost of the program is shared by the federal government (under DHSS and CMS), the state (through the state's Department of Insurance), and the municipality.

All Medicaid programs are mandated to provide services that are relevant to ALS, including: physician services, hospital inpatient services, outpatient hospital services, laboratory tests and X-rays, skilled nursing facility services, home health services, medical transportation services and nurse practitioner services (other services are mandated, but less relevant for the ALS population). The amount of these services provided may vary (i.e., New York State provides 24-hour home attendant care to patients living alone; other states provide for limited hours).

Medicaid benefits may vary across states. As not all providers accept Medicaid payment, ALS patients in some areas may be limited to receiving care from non-ALS specialists. Furthermore, in many states only one service can be billed to Medicaid per day; therefore, if patients are evaluated in a multidisciplinary clinic as is often the case in ALS centers, only one health care discipline will be reimbursed for services unless a unified bill is submitted.

Despite these limitations, Medicaid is useful to many patients with ALS. Medicaid covers prescription costs, pays for most assistive devices, includes a hospice benefit and covers the cost of home care and ventilation support. However, many ALS patients do not meet the financial requirements of Medicaid since they are often at the peak of their earning potential when diagnosed. Medicaid examines both monthly income and assets for eligibility. It may be possible to transfer funds out of an ALS patient's name in order to meet Medicaid asset requirements (many states have a waiting period of three years following such transfers before being considered Medicaid eligible). Income is more difficult to shelter, but patients under the age of 65 may shelter their income in a supplemental needs trust, thus making themselves eligible for Medicaid. Such transfers should not be done without the guidance of a reputable Elder Care Attorney, as the legal ramifications vary from state to state and the regulations change frequently.

Private Insurances/Preferred Provider Organizations (PPOs)/ Health Maintenance Organizations (HMOs)

There are no federal or state guidelines mandating what services private insurers must provide. Each insurer is free to develop policies according to the needs and demands of their customers and their own need to make a profit. Some insurers will cover a percentage of specialty care, even if it is out-of-network. Other HMOs will pay nothing toward the cost of specialty care, regardless of whether therapists trained in ALS exist within their network. Historically, almost half of ALS patients have private

insurance, but this proportion is likely to change with the recent passage of the Medicare Waiver Bill for all ALS patients.

DME coverage is dependent upon the policy; if DME is included in the policy, many insurers require the submission of a letter of medical necessity and justification before deciding on coverage. The criteria insurers use to determine claims are often, but not always, consistent with Medicare criteria. A full knowledge of Medicare justification is, therefore, helpful when submitting claims. Most insurers will determine coverage prior to provision of a device. Generally, each state's Department of Insurance is expected to have developed regulations governing the time frame in which insurance companies must respond to claims, the ways in which they must set up appeals procedures, etc. These state regulations, however, are not consistent among states. Given the disparity of available insurance policies, it is impossible to make blanket statements regarding needs of ALS patients that may or may not be covered by insurance.

What are the Effects of Managed Medicine on ALS Care at the End of Life?

In order to contain spiraling health care costs, managed care programs began appearing about 12 years ago. These programs promised to reduce costs to insurers by reviewing the need for requested care. Managed care was also viewed as being attractive to consumers as their out-of-pocket costs were reduced to a small co-payment for services. However, patients must be diligent in reviewing policies; although the general out-of-pocket costs may appear beneficial, some managed care contracts place strict limits on reimbursement for DME that improve the quality of life of patients (such as power wheelchairs and transfer aids). Furthermore, managed care policies may not cover out-of-network providers. Thus, if the ALS care team is not in-network, patients will have to choose between bearing the full cost of care or receiving treatment from providers who are unfamiliar with their condition. In the latter event, a request can be made for case managers who can be educated regarding the special needs of ALS patients.

More recently, Medicare and Medicaid have turned to managed care to reduce costs to the national system. Many ALS patients, familiar with the benefits of managed care, opt into Medicaid managed care programs offered by several states in order to continue receiving coverage for pharmaceuticals and to avoid higher co-payments. However, patients must be aware that their ALS care providers may not have contracts with the Medicare managed care company (even if the providers do have contracts with the same company under a non-Medicare policy). Patients, once again, must choose between paying for care by an ALS specific care team, or receiving less costly care from providers who are unfamiliar with the disease.

Managed Medicare is actually very restrictive and patients need to know these limitations. Physicians have had difficulty getting services for patients because of the lack of availability of either standard protocols or case managers who can lobby on behalf of patients in order to get support for their medical needs.

Hospice or Ventilation? Are They Covered?

Hospice

Hospice is a program of multidisciplinary support specifically designed for patients with limited life expectancy who want comfort and quality to be the focus of care. Hospice programs within the United States require a life expectancy not exceeding six months. Patients also must request a natural death, without the life extending intervention of tracheal intubation. The care of the family, the mind and spirit are paramount in achieving the goals of comfort, dignity and patient autonomy at the end of life. The number of individuals receiving hospice care is growing by 10% to 20% annually (Christakis, Escarce, 1996).

Hospice is a concept of care that is delivered in a variety of settings (home, hospital or free-standing facility) by an interdisciplinary team of health care professionals and trained volunteers. Within the United States, 85% of hospice patients are diagnosed with cancer. Yet, in the wake of increased awareness and utilization of hospice in the past 10 years, there is a growing need to define hospice criteria for non-cancer diagnoses.

Although the first hospice program in the United States began in 1974, Medicare incorporated hospice services in 1983. The Medicare policy for determining end-of-life status in ALS became effective on Sept. 30, 1998 (The Medicare News Brief, 1998) (Table 8). The identification of the need for hospice care in ALS patients is the responsibility of the treating medical team. Deciding when and how to begin hospice care is complex. The multifaceted issues that need to be comprehensively assessed by the medical team greatly impact the timing of hospice discussion and referral.

Hospice Guidelines in ALS

In 1996, the National Hospice and Palliative Care Organization (NHPCO) developed hospice admission criteria for ALS; these criteria were meant to be a tool for hospice personnel unfamiliar with ALS, and not as concrete eligibility rules. The NHPCO hoped the guidelines would expand hospice access for patients with less common diseases, such as ALS, by providing hospices with a means to evaluate these patients. Unfortunately, the guidelines have come to limit hospice access for some patients. The NHPCO has not revised the guidelines since their inception in 1996 amidst concern that any revisions may further limit access to hospice care. According to the NHPCO, these initial guidelines were modified by Medicare Fiscal Intermediaries to use as rules determining payment. Local hospices often use these guidelines as rules, not because the NHPCO has published them, but because they know Medicare uses them to determine payment.

ALS historically has been a red flag for Medicare hospice claims. According to the NHPCO, ALS patients stand out as having higher “lengths of stay” on hospice than most other diseases; for this reason, Medicare is particularly stringent about these criteria and hospices are particularly reluctant to use their own judgment and go beyond the NHPCO guidelines.

Local Medical Review Policies

There are five different Medicare entities throughout the country called Fiscal Intermediaries, who evaluate Medicare claims in their geographic region. Each Fiscal Intermediary is free to develop its own Local Medical Review Policies (LMRP) for coverage of specific claims (such as for hospice in ALS). If patients meet the LMRP criteria, they are presumed appropriate and the claim for coverage is paid no matter how long they are on hospice under Medicare (as long as they continue to meet the criteria). If patients do not meet the LMRP criteria, the hospice claim for payment is not automatically denied; however, the hospice must provide additional documentation to explain why that patient should be considered appropriate. Thus, local care providers may receive a favorable response toward a hospice referral by providing supportive information beyond that initially requested.

It is important to note that LMRPs use a variation of the NHPCO guidelines to determine eligibility. However, they also indicate clearly that the disease varies from one ALS patient to another, and that patients who do not meet the LMRP criteria will be considered for hospice if the physician can document why a particular patient has a six-month life expectancy.

Implementation of Hospice Referral Guidelines in ALS

The unique manner in which individuals live and die with ALS presents a challenge to health care practitioners in determining life expectancy. Despite the newly recognized and established Medicare criteria for determining prognosis of ALS, this challenge is still present. The current Medicare criteria for admission to hospice are not sensitive to the range of involvement and handicaps faced by ALS patients. In effect, the Medicare criteria place concrete parameters of disease progression or terminality of

symptoms in ALS as they would occur in other disease processes. The truth is that the terminal stage of ALS bears no similarity to that in other diseases, such as cancer. This manner of predicting life expectancy in ALS creates an environment of minimizing end-of-life care rather than enhancing the likelihood of eligibility for hospice.

The Medicare hospice referral criteria are difficult to apply practically in ALS due to the variability of symptoms in the late stages of the disease. They consider severity of respiratory involvement, nutritional compromise, and rapidity of progression of the disease (Table 8). Medicare criteria for admission to hospice are applied at the end of life in several diseases. Studies in other diseases have indicated that the majority of patients are referred late in the course of a terminal illness; the median stay on hospice is estimated to be about 36 days (Christakis, Escarce, 1996).

There is only one prospective study of hospice in ALS, which included 90 patients at Columbia-Presbyterian Medical Center, New York. This study was done between 1995 and 1997 and used FVC criteria and respiratory symptoms as the basis for referrals to hospice (Table 9) (Del Bene et al., 1998). Admitting FVC was 20% to 40% of predicted in 32% of patients, 40% to 79% of predicted in 26% of patients, and unknown in 42% of patients. Mean survival was six months in patients with an FVC \geq 40%, and five months in those with an FVC $<$ 40% of predicted. A comparison of the content of the current Medicare “impaired breathing capacity” criteria with symptoms experienced by ALS patients in the pre-terminal and terminal stages of their disease shows that the criteria are often misunderstood and poorly applied to patients with ALS (Table 10). These results suggest that hospice referrals based primarily on FVC criteria result in close adherence to the six-month benefit period and ensure patient access to the comprehensive benefits of hospice care. Furthermore, these results emphasize the importance of FVC and the presence of respiratory symptoms over other criteria for admission to hospice.

The hospice benefit is generally limited to 210 days. Some insurance providers (such as Medicare) will extend the duration of this coverage if the patient continues to actively decline during this time. Others, however, will give only a 210-day lifetime benefit. Due to the difficulty of accurately predicting the prognosis, ALS patients on hospice who do not deteriorate significantly should be advised to consider coming off hospice, so as to reserve their benefit for a time in the future, when they might need it. In the hospice referral study at Columbia Presbyterian Medical Center, New York, 76 of the 90 patients died on hospice, and four were decertified due to the length of stay potentially exceeding the six-month benefit period because of a plateau of disease progression (Del Bene et al., 1998).

In order to take full advantage of a patient’s medical coverage, it is vitally important that physicians understand the Medicare guidelines for hospice admission as well as the manner in which hospices are reimbursed for care. Hospices generally receive a per diem payment, rather than a per service payment, from which they must provide all services covered under the hospice benefit, including physician visits, nursing visits, social work intervention, medications, home health aide salary and durable medical equipment. Consequently, any expensive custom-made equipment, such as wheelchairs and augmentative communication devices, must be obtained prior to referral as hospices cannot afford to cover them from their limited per diem payment.

In general, poor timing of referring patients to hospice has been ascribed to the desire of physicians to preserve hope by postponing the referral. For patients with an ultimately terminal disease like ALS, hope is continuously offered through management of symptoms aimed at preserving quality of life. Nevertheless, timely referral of patients with ALS to hospice services is important in order to maximize the benefit of comprehensive psychosocial end-of-life care and improve the quality of their living and dying.

Ventilation

Guidelines covering the costs associated with mechanical ventilation vary. Many insurers do not cover nursing home care (unless it is step-down care following an acute hospital admission). Some will provide limited coverage of home care for ventilated patients; this coverage is frequently so limited, however, that patients opt against mechanical ventilation as their families are unable to assume either the remaining care or the cost of private help.

Medicare coverage is vastly insufficient for those patients opting for mechanical ventilation. The major cost for ventilated patients is that of nursing care, to manage the ventilator and maintain patient comfort and safety. Medicare does not cover the cost of home nursing, nor does it pay for nursing home care if patients cannot remain at home.

Unless patients have Medicaid, the out-of-pocket costs are much higher if they choose mechanical ventilation over hospice care. Consequently, financial considerations are the driving force behind the decisions of many patients regarding their health care at the end of life.

How can ALS Patients Receive Reimbursement or Coverage for Assisted Ventilation During End-of-Life Care?

There is very little published information on the coverage for assisted ventilation, particularly during the end of life, and there are no data available on the cost of NIPPV support. In an older study of home mechanical ventilation in which all patients with ALS (n=19) reported health coverage, 47% had private insurance only, 37% disclosed private insurance and Medicare coverage and 5% each reported exclusive Medicare, Medicaid or VA insurance (Moss et al., 1993). Insurance covered a mean of 83% of the gross monthly costs of ventilation, and in 63% of patients all costs were reimbursed. Medicare covered the least costs, and reimbursed no nursing costs related to ventilation; 68% of patients and families incurred out-of-pocket monthly expenses of \$100 to \$7,200.

Long-Term Care

A major shortcoming of current funding and availability of adequate services for patients with ALS is long-term nursing care—both home-based and institution-based. There is a broad gap between the needs and availability of care for patients with ALS and their families. This problem is compounded when patients use mechanical ventilation, as there is a shortage of home care providers who are knowledgeable about ALS care in general, and ventilator care in particular. When home-based care is not an option, finding a skilled nursing facility licensed for ventilator care also is very difficult. Skilled nursing facilities for ventilator-dependent patients are few, necessitating institutionalization of ALS patients in some cases. These institutions may be distant from family and other care providers.

For instance, a 52-year-old ventilator-dependent ALS patient in Colorado Springs, Colo., had to be moved to the closest available long-term care facility in Pueblo, Colo., 50 miles away when his family caregivers were no longer able to provide 24-hour care, and a suitable home care provider could not be found. This relocation was extremely distressing for the patient and his family.

Even when adequate skilled nursing care is available, the cost of accessing care can be prohibitive. Health insurance plans limit covered home care benefits, and Medicare currently only provides care if the beneficiary is completely homebound. Many patients with ALS present with the disease in the prime of their lives and most have not purchased long-term care insurance. Long-term care insurance policies are helpful if patients already possess them, but cannot be purchased once the diagnosis of ALS is made and patients have knowledge that they may be needed in the future. This is a serious problem for many families. When patients with ALS need hospice but are not eligible, as determined by Medicare criteria, their quality of life and that of their family caregivers often is adversely affected.

Impact of Lack of Resources on Patients and Caregivers

The lack of availability of resources for end-of-life care has consequences on choice of care and medical decision making, and ultimately impacts negatively on patients, caregivers and society. The inability to access and pay for much needed care adversely affects the psychological wellness of patients and caregivers by lowering their quality of life and escalating burden. Society is burdened with expensive medical costs at the end of life, when timely early and much-needed interventions are overlooked, unavailable or denied.

Conclusions on Availability and Accessibility of Resources

In most cases, the state resources listed in Table 6 are not specifically targeted for the ALS population, although patients with ALS and families can utilize them. Due to the fact that many issues surrounding the ALS experience are unique to this disease, many available resources may not meet the needs of the ALS community when they are accessed. However, this is likely to be the case with many rare diseases. Therefore, instead of creating new ALS-specific resources, it would be prudent to facilitate education of policy-makers, program administrators and service delivery personnel about ALS. In addition to education, the ALS community needs to become more informed about potential avenues available for funding and services. Hopefully, through a multi-level educational strategy, the utilization and quality of existing resources will be improved and the gaps narrowed.

Identification of Existing Gaps

All patients with ALS should be provided with consistent care and educational opportunities leading up to, and including, the end of life. Providing patients access to ALS specialty centers that have established treatment processes and protocols will help reduce variability in the quality of care and methods of care, especially during the end-of-life phase.

There are many areas where the current system of availability and access to health care of patients with ALS and families is not sufficient or complete. These shortcomings are evident in the advanced late and end-of-life stages of the disease.

Medicare guidelines for adaptive equipment and hospice care are too stringent and are unlikely to change unless supportive data from the literature is available. The guidelines are perhaps not as strict for private insurers. Some examples of these gaps in support are given below.

1. Health Care Provider Services

Lack of coverage of:

- regular physical therapy (range of motion and massage) to prevent or minimize effects of spasticity and pain;
- regular occupational therapy for assessment of hand function and activities of daily living;
- regular respiratory therapy/respiratory technician services for advice on use of suction machine, cough assist device, NIPPV and adjustment of NIPPV settings; and
- home social work services for education about availability of resources.

2. Durable Medical Equipment (DME)

- no coverage for important DME (e.g., adaptive equipment to improve function and independence such as hand devices, walkers, wheelchairs, phone grips, resting hand splints, cups with two handles and reachers);
- considerable delay in processing of applications for DME ;
- frequent denials for DME considered medically necessary;
- denial of augmentative communication devices by insurers despite availability of coverage by Medicare; and

- no coverage of larger repository for home oxygen use (e.g., an oxygen tank, which usually only lasts for one hour, is covered but is not enough for doctors office visits).

3. Home Evaluations

- no consistent coverage for safety, modifications and equipment needs;
- lack of standardized forms for evaluations by physical therapists, occupational therapists and home health aides;
- lack of standardization of home social worker evaluations (which can be beneficial for Advance Directives, hospice planning and assessment of stress in the family).

4. Home Adaptations

- no consistent coverage for provisions of access into home (e.g., ramps);
- no consistent coverage for adaptations inside the home (grab bars, raised toilet seat, lift chair, Hoyer lift for access to bathroom, chair lift for access to upstairs bedroom);
- no coverage for panic alert button if not living alone (for when spouse or caregiver is out of home) (covered by Medicaid if living alone, but not covered by private insurers);
- no coverage of environmental controls.

5. Home Nursing Care

- no coverage of unskilled nursing for PEG feeding;
- no available ALS-specific guidelines regarding pulse oximetry, PEG feeding, NIPPV settings;
- no guidelines for care of PEG, nutritional care and feeding, and NIPPV;
- no available guidelines on management of pain, pre-terminal distress and terminal psychological distress.

6. Hospital Admissions

- no existing guidelines for admissions for PEG;
- no prospective studies of costs associated with hospitalization.

7. Education of Patients, Families and Health Care Providers

- insufficient knowledge among patients, families and health care providers about end-of-life care, PEG, NIPPV, invasive ventilation and treatment of pain and suffering.

8. Health Care Delivery

- lack of coordination of care (fragmented care across the spectrum from primary care to ALS center/specialty neurology practitioner to hospice);
- lack of continuity of care (many physicians do not understand hospice reimbursement and therefore refer when patients still need costly care that hospice does not cover);
- lack of protection regarding HMOs not insuring or discontinuing coverage for older patients;
- no data on end-of-life care from community neurologists or individual neurologists to compare with excellent data from large centers.

9. Ventilator Patients

- no dedicated funding for patients with ALS using NIPPV or mechanical ventilation (including skilled nursing, home health aides, respiratory therapist, physical therapist, technical support, back-up ventilator and power source);
- insufficient knowledge of providers about non-invasive and invasive ventilation, PEG and treatment of pain and suffering.

10. Admission to Hospice

- lack of sensitivity and knowledge regarding the severity of disability of patients with ALS, which influences Medicare criteria for admission to hospice;
- no ALS-specific guidelines for justification of referral and admission to home hospice or in-patient hospice;
- lack of availability of Medicare hospice patient health forms that are specific for patients with ALS (which would include detailed demographic data, specific symptomatic issues such as respiratory compromise, nutrition, depression and disability).

11. In-Patient Hospice

- lack of Medicare funding for in-patient hospice, which is currently available for other diseases, such as cancer. (Note: In-patient hospice is available to anyone on a hospice program with in-patient services. The long-term residential care for people with other diseases is usually provided NOT through a hospice program, per se, but through palliative hospitals, such as Calvary in the New York City area.)

12. Caregiver Issues

Lack of:

- adequate counseling and respite services;
- resources for distress management and bereavement.

Identification of Gaps in Cost Data Regarding End-of-Life Care in ALS

- no prospective studies have been done on the cost of care and hospitalization of patients with ALS (most studies have been done in association with treatment trials);
- no analysis or evaluation of indirect costs; most data collected focuses exclusively on direct costs;
- no studies specifically on the costs associated with end-of-life care;
- no prospective studies comparing cost-to-quality of distinct care delivery modes (e.g., community versus tertiary care); and
- no analysis has been done on assessing the cost of caregiving to employers.

Identification of Gaps in Knowledge of Health Care Providers

- insufficient knowledge and education of health care providers about the use, pros and cons of PEG, NIPPV, invasive ventilation and home care regarding patients with ALS;
- lack of knowledge among health care providers about Medicare guidelines for hospice care;
- lack of dissemination of information about ALS Practice Parameters and other guidelines and recommendations for caring for patients with ALS (such as severe pain, respiratory failure and severe disability);
- lack of a clear understanding of long-term care options (e.g., home hospice care);
- lack of knowledge in patients and family members about PEG, NIPPV, invasive ventilation, home care;
- inexperienced health care providers caring for the highly technical and medical needs of ALS patients; and
- lack of knowledge about use of riluzole and other medications (such as those needed for symptom management) especially at the end of life.

RECOMMENDATIONS TO THE FIELD FOR IMPROVING END-OF-LIFE CARE IN ALS

	Gaps/Barriers	Recommendations	Audience
I. ACCESS TO QUALITY CARE			
Practice	<p><u>Diagnostic and Management Algorithms</u></p> <ul style="list-style-type: none"> - Lack of practitioners with ALS diagnostic and treatment experience - Lack of formalized standard of care - Lack of standardization of home skilled nursing evaluations - No guidelines for admission for PEG placement 	<p><u>Diagnostic and Management Algorithms</u></p> <ul style="list-style-type: none"> - Implement existing algorithms for diagnosis and symptom management to standardize care - Create algorithms where they do not exist (i.e., skilled home nursing) - 48-hour hospital stay for PEG placement 	<p>HC Providers</p> <p>HC Providers and case managers</p> <p>Neurologists and payers</p>
	<p><u>Centers of Excellence</u></p> <ul style="list-style-type: none"> - Lack of coordination of care - Patient dissatisfaction with fragmented care, inadequate education, and physicians attitude toward caring for ALS patients - Lack of continuity of care between community physicians, specialty centers and hospice 	<p><u>Centers of Excellence</u></p> <ul style="list-style-type: none"> - Develop Centers of Excellence Program - Promote timely referral to specialty center - Create close cooperation between community physicians, specialty centers and hospices to ensure continuity of care 	<p>ALSA</p> <p>MDA</p> <p>AHQQR</p> <p>HC Providers</p> <p>HC Providers</p>
	<p><u>Timing of Referral to Hospice</u></p> <ul style="list-style-type: none"> - Lack of clear guidelines for timing of referral to hospice 	<p><u>Timing of Referral to Hospice</u></p> <ul style="list-style-type: none"> - Emphasize consideration of referral to hospice (Table 11) 	<p>Payers</p> <p>HC Providers</p>

	Gaps/Barriers	Recommendations	Audience
Research	<u>Diagnostic and Management Algorithms</u> <ul style="list-style-type: none"> - Lack of evidence regarding outcomes of various patient care models - Lack of evidence regarding barriers to adherence to existing practice guidelines 	<u>Diagnostic and Management Algorithms</u> <ul style="list-style-type: none"> - Evaluate impact of multi-disciplinary versus community care approach on patient outcomes - Assess barriers to adherence 	HC Providers HC Providers
	<u>Timing of Referral to Hospice</u> <ul style="list-style-type: none"> - Insufficient evidence regarding appropriate criteria for hospice referral - No data on ethnicity / culture on access to care 	<u>Timing of Referral to Hospice</u> <ul style="list-style-type: none"> -Further studies to evaluate criteria for appropriate referral to hospice - Prospective studies of patient attitudes and decision choices for hospice through standardized tools 	Payers Neurologists/Payers
Policy	<u>Diagnostic and Management Algorithms</u> <ul style="list-style-type: none"> - No guidelines for hospitalization (e.g., PEG placement) - Lack of coverage of ALS- specific DME, home evaluations, home modifications and some professional services 	<u>Diagnostic and Management Algorithms</u> <ul style="list-style-type: none"> - Develop comprehensive policy regarding hospitalization and DRGs - Advocate for coverage of 48-hour hospital stay for PEG placement - Increase funding for ALS- specific care 	Neurologists/ Payers Payers Payers
	<u>Centers of Excellence</u> <ul style="list-style-type: none"> - Lack of coordination of care 	<u>Centers of Excellence</u> <ul style="list-style-type: none"> - Fund Centers of Excellence 	ALSA MDA Payers
	<u>Timing of Referral to Hospice</u> <ul style="list-style-type: none"> - Lack of clear guidelines for timing of referral to hospice 	<u>Timing of Referral to Hospice</u> <ul style="list-style-type: none"> - Modify Medicare guidelines of admission to hospice based upon specific needs of ALS patients 	Payers

Supplementary Comments

- Policy should be changed to provide more reimbursement for multidisciplinary care. One model would be to have “bundled care” that covers multidisciplinary care at specialty centers which have a multidisciplinary team. Policy decisions will need to be made to help determine who pays for this care given the assumption that the research is done to establish that multidisciplinary care improves outcomes.
- Changes need to be made to hospice admission guidelines. The current guidelines are too stringent for patients with ALS, and many patients do not qualify under the current regulations. (Table 9).
- Current hospice benefits do not allow adequate home health care. Create informative documents that detail the level of home care coverage that should be provided by the state.
- Inpatient hospices are not really a viable option in some communities. One might consider that more facilities be made available or that all hospice organizations provide beds for patients. Specifically, the level of care that hospices should provide should be defined in detail. For example, each patient with ALS should be allowed:
 - 5 days/month, every month in respite (nursing home or facility);
 - 20 hours per week of home health aide (these regulations may differ state to state).
- There is no pathway that guides the patients and families regarding “what can be done and when” so that patients and family members are prepared during the course of the illness for the costs and other burdens of illness. A detailed algorithm or flow chart needs to be created that directs the family members and their health care providers in guiding them through issues regarding coverage, availability to care, etc.
- It is important that health care for ALS be provided to all patients, despite differences in culture, attitudes, etc. Studies are needed to assess how different patients and subgroups of patients need and have access to care, attitudes about types of care, and support provided in the health care system.

	Gaps/Barriers	Recommendations	Audience
II. COST OF CARE			
Practice	<u>Diagnostic and Management Algorithms</u> <ul style="list-style-type: none"> - In-home care not adequately funded - Long-term care not reimbursed - In-home ventilation costs not adequately covered 	<u>Diagnostic and Management Algorithms</u> <ul style="list-style-type: none"> - Provide mechanism of funding for in-home care - Enhance availability of funding for long-term care - Expand coverage for in-home ventilation costs 	Payers HC Providers
	<u>Timing for Hospice Referral</u> <ul style="list-style-type: none"> - Cost of care is an issue; hospice resources may be capped and thus patients on riluzole cannot be accepted. This is complicated, with legal issues, as hospice should not deny care - Insufficient data on cost-benefit of riluzole at the end of life - Patients on hospice cannot get customized devices 	<u>Timing for Hospice Referral</u> <ul style="list-style-type: none"> - Clinicians may consider discontinuing riluzole upon admission to hospice, re-evaluate the use of riluzole, and determine who pays - Cost-benefit studies of riluzole at the end of life are needed - Educate physicians about coverage - Allow patients to draw from standard coverage for DME in addition to hospice coverage 	HC Providers Payers Neurologists Payers
Research	<u>Diagnostic and Management Algorithms</u> <ul style="list-style-type: none"> - Lack of evidence regarding cost/benefit of ALS care 	<u>Diagnostic and Management Algorithms</u> <ul style="list-style-type: none"> - Develop model of cost of care generated by panel of experts (Table 12) - Conduct studies to evaluate cost-benefit of riluzole at the end of life - Undertake prospective cost-benefit studies of unplanned hospitalization - Conduct prospective studies to evaluate outcomes including cost effectiveness, QALY, and cost-benefit analyses (of home care, hospice, PEG, ventilation) 	Neurologists Neurologists/ Payers Neurologists/ Payers Neurologists/ Payers AHQR

	Gaps/Barriers	Recommendations	Audience
Policy	<p><u>Diagnostic and Management Algorithms</u></p> <ul style="list-style-type: none"> - Lack of coverage for comprehensive ALS care <ul style="list-style-type: none"> - Lack of adequate coverage of in-home nursing care, both custodial and skilled - Lack of coverage of long-term care (home care or in nursing facility) - Lack of adequate coverage of in-home ventilation costs (respiratory therapist, equipment) 	<p><u>Diagnostic and Management Algorithms</u></p> <ul style="list-style-type: none"> - Emphasize economic benefit to payers of early and timely interventions to reduce unnecessary hospitalizations - Establish funding for Centers of Excellence to provide appropriate, cost-effective neurological care to ALS patients - Develop comprehensive policies regarding in-home care (including ventilation) and long-term care 	<p>Payers</p> <p>Payers</p> <p>Payers</p> <p>Payers</p>

Supplementary Comments

- Issues that need to be addressed in clinical practice, research and health care policy needs include:
 - Cost coverage for supplies, O₂, NIPPV;
 - Reimbursement of NIPPV in hospice; and
 - Patients on hospice can be eligible for placement of PEG.
- Hospices are usually reluctant to reimburse for riluzole, even though they are required to pay for all prescribed medications. Consequently, some hospices refuse patients who are taking riluzole. This needs to be addressed in a health care policy.
- Do physicians have the right to withdraw riluzole from patients on hospice? In ALS Health State 4, the cost of care goes up three-fold as compared to Health State 2. There is no evidence that riluzole helps at the end of life. Funds may be better spent if reallocated to other treatment and support.
- If patients are willing to pay for riluzole, or if their private insurers are prepared to reimburse, should they be able to continue while on hospice? This controversy needs to be resolved at a policy level.
- If differences exist between states, a uniform policy that defines the standard of care and benefits for patients with ALS is needed.

	Gaps/Barriers	Recommendations	Audience
III. KNOWLEDGE/EDUCATION			
Practice	<p><u>Diagnostic and Management Algorithms</u></p> <ul style="list-style-type: none"> - Lack of adequate understanding and follow-through on appeals process of insurance denials - Lack of understanding of disability benefits - No consistent approach to patient/family/HC Provider education 	<p><u>Diagnostic and Management Algorithms</u></p> <ul style="list-style-type: none"> - Educate physicians regarding Medicare denial, the appeals process, Letter of Medical Necessity (LMN) for DME, disability benefits - Ensure each ALS HC Provider has a list of DME and standard copies of LMN - Develop educational tools/models for all HC Providers/patients/families 	<p>HC Providers</p> <p>HC Providers</p> <p>Patients/HC Providers</p>
	<p><u>Timing of Referrals to Hospice</u></p> <ul style="list-style-type: none"> - Tardy referrals to hospice - Lack of knowledge in hospice personnel about progression of ALS - Poor understanding of hospice reimbursement policies 	<p><u>Timing of Referrals to Hospice</u></p> <ul style="list-style-type: none"> - Educate patients and HC providers regarding criteria and value of hospice - Educate patients and caregivers regarding hospice guidelines - Convene joint workshops between hospice and health care providers 	<p>Patients/Families</p> <p>Payers/Hospice Personnel</p> <p>HC Providers/Hospice</p>
Research	<p><u>Management Algorithms</u></p> <ul style="list-style-type: none"> - Lack of data regarding care patterns, particularly from audits rather than surveys - Absence of audits and examinations assessing educational gaps - Lack of data regarding educational intervention methods 	<p><u>Management Algorithms</u></p> <ul style="list-style-type: none"> - Conduct audits of community and tertiary care practices - Conduct interviews or assessments of educational needs - Develop and implement educational intervention methods - Evaluate effectiveness of various educational intervention methods 	<p>HC Providers</p> <p>Payers</p>
Policy	<ul style="list-style-type: none"> - Lack of funding for educational interventions and investigation of intervention effectiveness 	<ul style="list-style-type: none"> - Increase funding for educational interventions and research regarding education 	<p>ALSA</p> <p>MDA</p>

Supplementary Comments

- There are inconsistent opinions between medical and hospice health care providers regarding what is “good for the patient” (likely due to a lack of familiarity). Differences in how to approach patient care may be improved through educational consensus conferences and recommendations detailing patient care and specifically defining protocols. Additionally, specific guidelines on management of patients with ALS in hospice may help improve and unify the approach to treatment according to accepted good clinical practice as based on clinical evidence or expert consensus.

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Table 1: Four Composite States of Health in ALS**Stage 1 (MILD)**

1. Recently diagnosed
2. Mild deficit only in one of the three regions (speech, arm, leg)
3. Functionally independent in speech, upper extremity, activities of daily living and ambulation

Stage 2 (MODERATE)

1. Mild deficit in all three regions, OR
2. Moderate to severe deficit in one region while the other two regions are normal or mildly affected

Stage 3 (SEVERE)

1. Needs assistance in 2 or 3 regions
2. Speech is dysarthric and/or patient needs assistance to walk and/or needs assistance with upper extremity functions and activities of daily living

Stage 4 (TERMINAL)

1. Non-functionality of at least two regions and/or moderate use or non-functionality of the third region

From: Riviere et al., 1998

Table 2: Total Average Annual Cost for each ALS Health State

Type of Cost	State 1 (\$US)	State 2 (\$US)	State 3 (\$US)	State 4 (\$US)
Personal Expenses	718	2,167	4,785	5,086
Non-medical Expenses	25	153	4,288	362
Medical Equipment	297	910	3,211	5,708
Transportation Expenses	8	163	119	20
Workdays lost by Patient	31	216	33	0
Workdays lost by Caregiver	40	328	1,460	0
Time lost by Caregiver*	0	1,191	16,196	42,681
Hospitalization	5	0	18	17,994
MD Fees	1,094	658	582	300
Allied Health Consultations	176	146	252	946
Investigations	2,268	59	60	14
Medical Procedures	0	0	33	5,037
Medication	669	636	736	1,443
Total	5,331	6,627	31,773	79,591

*Assessed on the basis of the number of hours lost by a caregiver in caring for a patient and the hourly salary of a home aide.

Table 3: Average Patient-Borne Annual Cost Per ALS State of Health

Type of Resource Used	State 1 (\$US)	State 2 (\$US)	State 3 (\$US)	State 4 (\$US)
Personal Expenses	718	2,167	4,785	5,086
Non-medical Expenses	25	153	4,288	362
Medical Equipment	297	910	3,211	5,708
Transportation Expenses	8	163	119	20
Workdays lost by Patient	31	216	33	0
Workdays lost by Caregiver	40	328	1,460	0
Time lost by Caregiver*	0	1,191	16,196	42,681
Total	1,119	5,128	30,092	53,857

***Assessed on the basis of the number of hours lost by a caregiver in caring for a patient and the hourly salary of a home aide.**

Table 4: Annual Direct and Indirect Costs

	State 1	State 2	State 3	State 4	Home Vent.	Home Nurse
Direct	5,260	4,892	14,084	86,910	80,950	36,910
Indirect	71	1,735	17,689	42,681	42,681	94,848
Total	5,331	6,627	31,773	79,591	123,631	131,758

Table 5: Cost of ALS Care in Published Studies

Source	Cost Statistics	Clinical Outcomes	Psychosocial/QoL Issues	Other Information
Moss et al., <i>Neurology</i> 1993	Average annual cost for home ventilation (HV): \$153,252; nursing expense major cost factor		Family and caregivers reported major burdens	Based on study of 19 patients in North Illinois using HV (16/19 on 24-hr. HV)
Sevick et al., <i>Chest</i> 1996	Figures based on direct and indirect monthly cost to maintain Ventilator Assisted Individuals at home; HV w/LPN: median cost= \$5,406 mean cost= \$7,642; HV w/RN: Median cost= \$5,911 Mean cost= \$8,596		Development of programs/policies to support home placement need to further investigate ability/willingness of families to accept cost and responsibility of care	Used Katz index to measure costs
Klein & Forshew <i>Neurology</i> , 1996	PEG surgery=\$1500 PEG Supplies \$6,000/yr; NIV= \$5,220/yr Invasive Ventilation= \$199,500/yr (w/16/hrs/d nursing)			Cost analysis of ALS restricted to major survival issues: adequate nutrition and respiration
Woolley et al., "Health resource utilization and the cost of care in ALS," 1997	FVC (0-30%) =\$2,436/mo FVC (30-60%) =\$1,512/mo Partially dependent= \$816/month Dependent=\$2,117/month			Costs derived using prospectively obtained data from large BDNF clinical trial (1135 patients)

Source	Cost Statistics	Clinical Outcomes	Psychosocial/QoL Issues	Other Information
Woolley et al., "Caregiver quality of life in ALS: relationship between patient health status and caregiver health-related QoL, role satisfaction, and workforce participation," 1997	Costs in \$ not measured, but 20% caregivers reduced work hours; 10% quit job; 10% reported hiring professional help to give care (out of pocket expense); all of these actions would result in financial burden		Sense of reward for caring for loved one was not correlated with disease progression; caregiver physical and mental SF-36 summary scores and other items declined as patient health declined	Data on 800 caregivers over 9 months obtained as part of BDNF clinical trial
Ginsberg/Lev <i>Pharmacoeconomics</i> , 1997	Assuming 3-yr life expectancy, gain in longevity resulted in financial burden of \$757 on health services; overall resource benefits to society 1.28:1; total benefits to society 2.89:1	Average increase in survival was 3 months; average delay in hospitalization was 3 months		Israeli cost benefit analysis study of riluzole in 200 patients; costs reported in US \$

Source	Cost Statistics	Clinical Outcomes	Psychosocial/QoL Issues	Other Information
Messori et al., <i>Pharmacoeconomics</i> , 1999	Poor cost-effectiveness ratio calculated; more selective use could improve ratio and survival benefit	Average increase in survival was 2-3 months		Cost-effectiveness of riluzole study; limitation of study is that economic information was estimated indirectly
Tavakoli et al., <i>J Drug Assessment</i> , 1999	Larger proportion of riluzole-treated patients will stay in milder health states; cost per unadjusted additional life-year gained was British Pound Sterling 8,587 over expected lifetime.	Necessary to treat 3 patients for 3 years to obtain one additional life-year		Modeling of long-term cost-effectiveness of riluzole treatment in 954 European placebo-controlled study patients
Munsat et al., <i>J Drug Assessment</i> 1998 (UK data-costs quoted are in British Pound Sterling)	Hospitalization major cost driver at mild, severe, and terminal stages of ALS; physician; remuneration costs major factor in moderate stage; Primary analysis: Health State (HS) 1= 1185.20; HS2=799.60; HS3=1697.78; HS4=3127.51			Only direct medical costs were considered

Source	Cost Statistics	Clinical Outcomes	Psychosocial/QoL Issues	Other Information
Ackerman et al., <i>Pharmacoeconomics</i> , 1999	IGF-I treatment most cost-effective in patients either in earlier stages or progressing rapidly. Overall cost per QALY gained by IGF-I therapy versus placebo was \$67,440; for subgroups in early stages was \$43,197 and for patients progressing rapidly was \$52,823	Cost-effectiveness of IGF-I therapy in ALS compared favorably with treatments for other chronic diseases such as AIDS and end-stage renal disease		Posthoc analysis of data from North American placebo-controlled IGF-I study in 177 patients

Table 6: Resources and Web Site Addresses for National, State and Local Organizations*Resources Currently Available for Patients with ALS and their Families?*

The following is a summary of resources available to help finance, or provide free of charge, services related to end-of-life care for patients with ALS and their caregivers. The list is segmented into national, state and local level resources. Some resources are publicly-funded programs, while others are private for-profit and not-for-profit organizations. Contact information for some of the important resources mentioned are included at the end of the table. The resources listed here are well-established agencies and organizations that have proven track records as being consistently valuable and available.

ALS-Specific Non-Profit Organizations

For ALS specific information, it is best that patients and families contact both The ALS Association (ALSA) and the Muscular Dystrophy Association (MDA). Both have national toll-free phone numbers and extensive Web sites regarding available information, services and resource lists for a variety of support programs, clinical care providers and vendors that provide equipment to enhance quality of life. Both Associations may also have local support groups, loan closets for durable medical equipment and affiliated multi-disciplinary clinics for comprehensive care.

Other ALS-specific organizations that can assist families with services often associated with end-of-life needs include Ride for Life, Inc. (RFL) and the Les Turner ALS Foundation. RFL is a nonprofit, grassroots organization that grants limited funding to individuals to pay for respite care and legal counseling pertaining to end-of-life related decisions (e.g., estate planning). The Les Turner ALS Foundation can be accessed nationally for information, but their direct services are limited to the Greater Chicago Area.

There is considerable disparity in resources available in different regions of the country – for example, significantly greater resources are available to patients and families in the Greater Chicago and Philadelphia and other metropolitan areas as compared to more rural states.

Non-ALS Specific Nonprofit Organizations

Several other long-standing national level non-profit agencies are available to provide assistance to patients with ALS and their families at the end of life.

- The National Organization for Rare Diseases (NORD) can assist families with networking, patient services, medication assistance and an Internet-based medical equipment exchange.
- The National Family Caregivers Association (NFCA) offers a variety of supportive and educational materials for family caregivers.
- The National Hospice and Palliative Care Organization (NHPCO) is a professional membership organization that engages in patient advocacy, promotes research, provides professional and public education and makes referrals.
- The National Hospice Foundation (NHF) engages and informs the public about the quality of end-of-life care that hospice provides. Consumer brochures are available to assist families with choosing hospice care, communicating end-of-life wishes and understanding the Medicare Hospice Benefit.
- The National Association for Home Care (NAHC) is not a direct resource for patients and families as it is a trade association, but its mission is to help health care provider members better serve the end-of-life care needs of their patients.
- The Visiting Nurses Association of America (VNAA) is a national organization with 200 community-based visiting nurse agencies (VNA) in 40 states that can link families to local services. The VNAA also has available educational manuals for family caregivers. Each affiliated agency's service may vary by locale.
- The Plan for Life program, which is part of Ride For Life, offers funding for legal fees associated with communicating end-of-life decisions.

- Compassion in Dying offers counseling and advocacy geared toward improving pain and symptom management, increasing patient empowerment and self-determination and expanding end-of-life choices to include aid-in-dying for terminally ill adults.

National Organizations:

The ALS Association (ALSA): www.alsa.org

18 ALSA Centers, 64 Chapters and Free-Standing Support Groups

Caregiving: Solutions, Support, Relief: www.thirdage.com

Compassion in Dying: www.compassionindying.org

Disease Management Association: www.dmaa.org

Durable Medical Equipment Regional Carrier (DMERC) Regional Medical Review Policy (RMRP) on respiratory assist devices: www.cignamedicare.com/

DMERC processing claims for DME, prosthetics, orthotics and supplies: www.adminastar.com/

Family Caregiver Alliance: www.caregiver.org

Health Insurance Portability and Accountability Act (HIPAA): can be accessed through multiple sites.

www.aspe.hhs.gov/admsimp

www.wedi.org

www.wpc-edi.com

International Alliance of ALS/MND

Muscular Dystrophy Association of America (MDA): www.mdaua.org

22 ALS Centers and 230 MDA Clinics

Medicare: <http://www.cms.gov/>

National Alliance for Caregivers: www.caregiving.org

National Coalition on Health Care: www.nchc.org

National Family Caregivers Association: www.nfcacares.org

National Hospice and Palliative Care Organization (NHPCO): www.nhpc.org

National Hospice Foundation (NHF): www.hospiceinfo.org

National Association for Home Care (NAHC): www.nahc.org

National Organization for Rare Diseases (NORD): www.rarediseases.org

National Partnership for Women and Families: www.nationalpartnership.org

Paralysis Resource Center (ALS Association assisting the Christopher Reeve Paralysis Foundation) – for people living with paralysis. This will be a national coordinating facility providing library, Web site, and educational materials.

Patient Assistance Programs by Pharmaceutical Companies

Practical Tips for Living with ALS

Rehabilitative Engineering and Assistive Technology Society of North America (RESNA) (source for Assistive Technology Act: text and listing of nationwide Assistive Technology programs):

www.resna.org

Technical Assistance Project

1700 North Moore Street, Suite 1540

Arlington, VA 22209-1903

Phone: 703/524-6686 Fax: 703/524-6630 TTY: 703/524-6639

Today's Caregiver Magazine on-line: www.caregiver.com

Well Spouse Foundation: www.wellspouse.org

Veterans Administration Program

Visiting Nurse Associations of America (VNAA): www.vnaa.org

Visiting Nurse Society

Wheelchair-accessible Van Dealers: www.ride-away.com

World Federation of Neurology: <http://www.wfnals.org/>

State Organizations:

ALSA Chapters (equipment loans, respite care services)

ALSA Certified Centers (clinical services, education, support services)

Colorado

Older Women's League (OWL) Respite Fund

Department on Aging (or the State Agency on Aging - other states may give it a different name). They can all be accessed through the Administration on Aging, which is a DHHS agency. <http://www.aoa.gov/>

Department of Mental Health [e.g., California Resource Centers - statewide system of regional resource centers serving brain impaired adults (ALS is included as one of the qualifying diseases), families and caregivers. Provide information, counseling, support groups, respite care funding, free legal and financial consultations and educational workshops for caregivers. May be referred to by a different name in other states.

South Carolina site from where other states' DMH sites could be accessed:

http://www.state.sc.us/dmh/usa_map.htm

Local Organizations:

ALS Association Centers of Excellence, Support Groups and Chapters can be accessed through the National ALS Association Web site address: www.alsa.org

Muscular Dystrophy Association ALS Centers of Excellence and Support Groups can be accessed through the MDA Web site address: www.mdaua.org

Table 7: Web Sites for the Fiscal Intermediaries

1. Associated Hospital Services of Maine (CT, ME, MA, NH, RI and VT) <http://www.ahsmedicare.com/>
2. United Government Services (MI, MN, NJ, NY, PR, VI, and WI)
<http://www.ugsmedicare.com>
Does not presently have an ALS-specific policy

Table 8: Medicare Criteria for Determining Hospice Eligibility for ALS

Patients will be considered in the terminal stage of ALS (life expectancy of 6 months or less) if they meet the following criteria. (Must fulfill 1, 2, or 3).

1. Patient must demonstrate critically impaired breathing capacity
Critically impaired breathing capacity as demonstrated by all of the following characteristics occurring within 12 months preceding the initial hospice certification:
 - i. Vital capacity (VC) less than 30% of normal
 - ii. Significant dyspnea at rest
 - iii. Requiring supplemental oxygen at rest
 - iv. Patient declines artificial ventilation

2. Patient must demonstrate rapid progression of ALS
Rapid progression of ALS as demonstrated by all of the following characteristics occurring within 12 months preceding the initial hospice certification:
 - i. progression from independent ambulation to wheelchair or bed-bound status
 - ii. progression from normal to barely intelligible or unintelligible speech
 - iii. progression from normal to pureed diet
 - iv. progression from independence in most or all activities of daily living (ADL) to needing major assistance by caretaker in all ADL.

3. Patient must demonstrate critical nutritional impairment
Critical nutritional impairment as demonstrated by all of the following characteristics occurring within 12 months preceding the initial hospice certification:
 - i. recurrent aspiration pneumonia (with or without feeding tubes)
 - ii. upper urinary tract infection (e.g., pyelonephritis)
 - iii. sepsis
 - iv. recurrent fever after antibiotic therapy

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Table 9: ALS Hospital Referral Criteria at Columbia-Presbyterian Medical Center
New York, New York

Either (1) or (2) must be present

If (2) is selected, there must be at least TWO accompanying symptoms identified.

- (1) FVC \leq 30% of predicted
- (2) FVC \leq 60 % of predicted (with a decline over the past 2 months of 30%)
 - (a) inability to lie supine
 - (b) inability to blow nose or sneeze
 - (c) compromised cough
 - (d) moderate to severe dysphagia*
 - (e) uncontrolled weight loss
 - (f) use of accessory muscles of respiration

Patients with PEG must have an FVC of \leq 30%.

Table 10: Respiratory Assessment at the End of Life

Medicare Criteria	ALS Center Interpretation of Medicare Criteria
<p>1. Critically impaired breathing capacity</p> <ul style="list-style-type: none"> • Vital Capacity (VC) less than 30% of normal • Significant dyspnea at rest • Requiring supplemental oxygen at rest • Patient declines artificial ventilation 	<ul style="list-style-type: none"> • FVC less than 50% results in increased potential for respiratory failure. Equation must factor in nutritional status and mobility status for complications • Dyspnea with ALS may present as use of accessory muscles, paradoxical breathing, sleep disturbance and inability to lie supine • Paradoxical effects of supplemental oxygen (in the absence of primary pulmonary disease) may result in increased retention of carbon dioxide and therefore not usually indicated • NIV use in terminal stages is for symptom management. Artificial but non-invasive ventilation. Promotes symptom control and may affect longevity

Used with permission of ALS Center at Columbia-Presbyterian Medical Center, New York.

Table 11: Amyotrophic Lateral Sclerosis Hospice Referral Form

Patient's Name: _____
 Address: _____
 Telephone Number: _____

Primary Caregiver: _____
 Primary Contact Person: _____

Physician's Name: _____
 Social Security #: _____ Insurance: _____

DOB: _____ Ethnicity: _____

Primary Language: English Other _____

Diagnosis	Onset Date	Prognosis
(Primary): _____	_____	_____
(Secondary): _____	_____	_____

Current Medications: _____

Pulmonary Function Tests
 FVC _____ % of Predicted
 _____ Liters

Date Completed _____

Clinical Course:
 Slowly Progressive _____
 Rapidly Progressive _____

Qualifying Symptomatology: (A) or (B) must be present as core to other respiratory indicators

(A) FVC 30% of predicted _____
 (B) FVC 60% of predicted with a rapid decline over 2-3 months _____

Shortness of Breath _____
 Shallow Breathing _____
 Paradoxical Breathing _____
 Inability to be supine due to diaphragmatic weakness _____
 Non-explosive cough _____
 Inability to blow nose _____
 Excessive oral secretions with dysphagia _____
 Nutritional Compromise / Dehydration _____

Prognosis of Six Months or Less	YES	NO
Patient Election of Hospice Services	YES	NO
Patient Election of DNR Status	YES	NO
Patient Refusal of Invasive/ Biomedical Intervention	YES	NO
Patient is Home Care Eligible	YES	NO

Additional Presenting Symptomatology

Dysphagia	Mild	Moderate	Severe
Weight Loss	_____ lbs		
Feeding Tube	Yes	No	
Aspiration	Yes	No	Unknown
Dysarthria	Mild	Moderate	Severe
Intelligibility	Poor	Fair	Good
Breath Support	Labored	Moderate	Comfortable
Excessive Oral Secretions	Yes	No	(Intermittent/Constant)
Mobility Status	Independent	Assistance	(1 or 2 persons)
Endurance	Poor	Fair	Good
ADLs	Independent	Mod Assist	Full Assist
Labile Mood	Yes	No	
Depressed Mood	Yes	No	
Cognitive Changes/Dementia	Yes	No	
DME/Augmentative Aides Utilized			
Suction Machine	Yes	No	To be ordered _____
	Yankauer	Catheter	
Bi-Pap	Yes	No	
	Daytime	Nighttime	All Day
Augmentative Communication Device(s)	Yes	No	

Walker	Yes	No	To be ordered _____
Wheelchair	Yes	No	To be ordered _____
Hospital Bed	Yes	No	To be ordered _____

Past Medical History

Physician Responsibility

_____ I will continue as primary physician

_____ I refer this patient to the care of the hospice designated physician

Patient management issues should be triaged through a multidisciplinary ALS Center.
 Please direct all inquiries, orders, and signature requests to the attention of:

 Physician's Signature

 License Number

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