

Appendix B

Table of Contents

Psychosocial Care	2
Bereavement in ALS	22
Spirituality and End-of-Life Care in ALS	31
Caregivers	40
Quality of Life Measurement in the Terminal Phase of ALS	58

Psychosocial Care

“The manner in which an individual faces life-threatening disease and obtains support from others reflects the shared beliefs and values of the particular culture, society and treating health care team.” The course of ALS requires constant adaptation of a psychosocial plan of care by health care teams and families, which must be adjusted as the patient’s disease progresses.

INTRODUCTION AND BACKGROUND

Definition of Psychosocial

Psychosocial is formally defined as the psychological and social aspects of one’s being and functioning, which encompasses the emotional, social and intellectual realms. The psychosocial aspect of care of a terminally ill patient is multifaceted making it difficult to define or fully implement.

Currently, studies in ALS related to psychological wellness of patients and caregivers are limited. One study (McDonald, 1995) indicates that patients with psychological distress (low quality of life) are seven times more likely to die sooner than those with psychological well-being (high quality of life), exemplifying the need for sound psychosocial care.

Ideally, health care professionals must gain greater understanding of psychosocial needs, expectations and interventions that apply to ALS patients, their caregivers and additional family members (e.g., children). This type of information is best collected prospectively in order to glean an accurate assessment of what is needed in order to develop interventions and support systems that best meet the needs of ALS patient systems.

Aspects of Psychosocial Care

In considering psychosocial aspects of care for patients with ALS, specific issues appear at different times in the disease progression. For example, readiness of patients and their families to receive and utilize information varies. End-of-life choices and life closure issues, which surface repeatedly as the disease progresses, are paramount to the provision of psychosocial care. Therefore, encouraging discussions and decision making about end-of-life choices promotes quality of life as well as dying.

A review of the literature found few studies about psychosocial aspects of ALS. One study (Sebring and Moglia, 1987) identified psychosocial interventions for patients and caregivers and yet only described psychosocial stressors of caregivers. Further studies and research are needed to identify psychosocial stressors of patients. Once stressors are identified, specific interventions can be recommended to increase patient capacity to adapt, participate in the plan of care and experience overall psychosocial wellness.

Education

Caregivers and patients identify the lack of readily available resources and medical information as a significant stressor. Therefore, it is essential for health care practitioners to develop a cadre of materials and information and to share their knowledge of local and national resources with patients and families. The larger the number of patients with ALS that a health care professional cares for, the more extensive the information and services resources should be. Having ALS-specific literature that includes sources for ALS-relevant information in the form of brochures, phone numbers and Web sites is a starting point for the providers caring for a limited number of ALS patients (Table 1). Larger practices require a greater number and type of resources including staff members who specialize in ALS psychosocial care (e.g., social workers, psychiatrists, psychiatric nurse practitioners and psychologists).

Knowledge of Disease-Related Issues

Patients and caregivers often describe feeling “clueless” about where they are in the disease process. Although prognostication is difficult in ALS, a measure of pulmonary function (vital capacity) can be used, as follows (see Table 2):

- Vital capacity (VC) normally is 80% to 100% predicted (based on age and height).
- Acute respiratory failure related to neuromuscular disease can occur abruptly any time after the VC has declined to 50% or less of predicted values.
- Respiratory failure typically occurs with pulmonary function lower than 20% of predicted values.

One general way to assess illness severity and disease progression is to determine a patient’s capacity to prioritize aspects of their care such as financial planning, medical decision making and active living.

“I must be dying soon since I have been living with ALS for more than 4 ½ years – I read no one lives more than 5 years.”

67 year old man with ALS, vital capacity 82%, no dysphagia

“I only was told I had the disease 6 months ago – I have a lot of time ahead of me before I need to talk about end of life.”
42 year old female, vital capacity 33%, severe dysphagia

Coping

Coping with disease fluctuates as symptoms and concurrent life-cycle events (which continue despite the presence of ALS) change. Studies of coping with ALS have primarily focused on caregivers. Longitudinal studies of patients and caregivers are needed to further identify different types of coping and coping changes during various stages of the disease. Assessing patient coping skills is important when assessing quality of life and dying with ALS. Differentiating between positive versus negative coping mechanisms helps health care providers and caregivers modify treatment plans (Table 3).

Supportive Counseling

ALS patients and family members benefit from education based on psychotherapeutic process. Family therapy helps the family unit to understand their pre-ALS family dynamics and to develop and utilize coping mechanisms. This functional form of short-term therapy facilitates communication and increases understanding of how family units can cope with the disease. A therapeutic process decreases stress and helps to mobilize families to become mutually supportive. Referrals to specific mental health professionals with experience in ALS are ideal, but more realistically what works in facing chronic or terminal illness is addressing a blend of medical issues along with psychotherapy.

Often a patient’s ability to access psychotherapeutic education and services is limited by insurance reimbursement, referral availability, or fear of stigma, or even disinterest in psychotherapy. Referral to an ALS support group can be a more practical intervention. Peer support groups encourage participants to interact with others in similar situations from a perspective of “one who has been or is there” (Table 3).

Case Scenario

Kathy, 42, is a fully employed wife and the mother of a 15-month-old child. Her husband James, who is moderately affected by ALS (e.g., uses non-invasive positive pressure ventilation [NIPPV] and a feeding tube), receives 24 hours of home care each day. During a check-in call, Kathy asks an ALS Nurse Clinician: “How long can he possibly live with this disease? I am worried that I will not be able to sustain much more. I already know that if James decides to be mechanically ventilated, I have no choice but to leave him.” Kathy has been receiving peer counseling from a female ALS

caregiver survivor and counseling from a family psychotherapist for six months (since the time that the care needs of her husband increased). She says: “I feel terrible in saying this, but I hate my life, I hate James and I want it to be over.”

The ALS nurse reassures Kathy that in her experience with other caregivers, Kathy’s statements and emotional responses are not unique. Kathy is tremendously relieved and says she feels less guilty. She is encouraged to continue supportive counseling and peer support and to maintain as much “normalcy” in her life as possible (i.e., work has become a safe haven when Kathy dreads going home to a place occupied by home health aides, medical equipment and family members). The insurance case manager has informed Kathy that she can access respite care at any time — James would be placed into a local nursing home for a few days. Kathy and the ALS nurse discuss respite care as an option to decrease family stress and to give James and her an opportunity to experience how respite could improve their quality of life—especially if he were to choose mechanical ventilation in the future. Kathy is receptive to discussing the “relationship expectations shift” between James and herself as his illness progresses: “If he lived somewhere else, I could visit him and like being with him again—right now I cannot say I like being with him and for that I am most sad.”

Sexuality and Intimacy

Humans are sexual beings by nature and the presence of ALS does not change this as reported by patients and their partners. Though addressed in individual ALS Centers, sexual expression of people with ALS is, on the whole, poorly researched (McDonald, 1994; Caroscio, 1986). Gallagher and Monroe approach sexuality and intimacy from a psychosocial perspective and further identify the need for more research relative to terminal illness and ALS in particular (Borasio, 2000).

Case Scenario

Allison is a 44-year-old woman diagnosed with ALS who is on mechanical ventilation. She is married and has three children. Allison shares that in the past she and her husband had a healthy and active sexual relationship. Due to progressive physical weakness over the past six months, their sexual interaction has transitioned from intercourse to touching—which was mutually satisfying. Shortly after beginning mechanical ventilation, Allison became quite anxious and described periods of suffering in the evenings. She was not able to communicate why she was feeling anxious and distressed. While the physical symptoms of anxiety were well-managed by medication, Allison’s psychic expression of anxiety, distress and suffering persisted. Further exploration with Allison and observation of her daily routine of care showed (albeit subtly) that although her husband was loving, socially interactive and actively involved in overseeing her medical care, physical interaction between them had greatly diminished. When this was raised with Allison, she wept and typed on her laptop, “He no longer touches me. I need to be held. I miss sleeping by his side and feeling his breathing.” She discerned that the presence of a full-time nurse, use of a hospital bed (separate from their queen-sized bed) and the lack of need for her husband to do routine physical care distanced Allison from her husband emotionally and physically. This distance was a loss that produced anxiety and a sense of suffering.

A plan was established for nursing staff to allow for as much private time as the patient wanted with her husband and family. Allison shared her feelings with her husband and requested that he perform a portion of her daily personal care such as body massage. When this plan was implemented, Allison’s distress and anxiety diminished.

Addressing the Needs of Children

The psychosocial effect of ALS on the family unit varies significantly depending on the pre-ALS family dynamics and the progressive nature of the disease. Continual change in physical function provides fodder for ongoing loss, mourning and grief. Though children usually are not directly

involved with care and decision making, the changes in their lives that result from the family member's illness are emotionally charged.

Children can struggle to articulate emotions. Behavior provides clues about how they feel about a parent's or family member's illness. Support must begin long before the family member's death. As described by Gallagher and Monroe (2001), many research studies identify that there is inadequate support for children facing bereavement.

Children need:

- Respect and acknowledgement.
- Information about what is happening and why, and what might happen next. Information (which must be clear, simple and truthful) must be repeated continuously as children struggle with what is happening.
- Reassurance. Children frighten as they watch a parent or relative become dependent and sometimes emotionally labile or irrationally angry. Explanations that help them to understand that they did not cause—nor can they catch—the illness, are helpful. Children need reassurance about practical issues, such as what will happen to the family and who will care for them after the patient dies.
- Appropriate involvement in assisting with the patient's care.
- A chance to share feelings and learn facts from adults who are prepared to share.
- A variety of mediums for self-expression (e.g., drawing, writing, playing games).
- Opportunities to reflect and to remember, to know that life during and after ALS goes on, and that it is all right to have fun.

Reading to or with children about physical illness and disability at an age-appropriate level helps to maintain communication and provides children with a way to express their thoughts and feelings to others (see References and Table 7 in this section of Appendix B). Knowing that they are not alone with their experience is critical for young people. Most children's literature focuses on grief by discussing the death of a loved one. Finding books that relate to loss, aging and the life cycle can be very useful during the period just before the death of a parent or grandparent.

End-of-Life Choices

ALS inevitably results in respiratory failure and death. Patients either choose to allow the natural dying process to occur, or they may choose to have mechanical ventilation introduced at the time of imminent death to thwart the dying process. This decision regarding mechanical ventilation is ideally made in advance of a respiratory emergency by the patient and family unit after thorough education and counseling about the risks and benefits.

It has been well documented in the literature that decision making is not an easy task for some patients and families (Project on Death in America; Albert et al., 1999). Decisions about end-of-life choices are not based on a purely medical point of view. Consideration must be given to financial resources, caregiver support/resources, quality of life and readiness to die.

Helping patients to explore end-of-life choices is difficult for health care professionals, especially for those who have not undertaken this exploration for themselves. Open-ended questions, such as those that follow, make this conversation less threatening and thus easier to discuss in the future:

- Have you experienced the death of others in your life before?
- What was that like for you?
- Have you ever thought about your own death (prior to ALS)?
- What thoughts do you have now?

When patients articulate decisions or feelings about their death, health care providers can help them to explore and document these decisions (e.g., create Advance Directives). Clinicians should provide positive feedback to patients for clarifying their choices/preferences for health care interventions. It is important to reassure patients that making their decisions and choices known supports the health care team's ability to implement medical and psychosocial interventions that are congruent with the patient's wishes.

For example:

- A patient (VC 90%) states he/she does not want invasive ventilation at the end of life.
Providers:
 - Research insurance coverage for hospice coverage and a referral is made when hospice eligibility is met in the future.
- A patient with flailed arms states a desire for a feeding tube in the future.
Providers:
 - Provide continued education about feeding tubes and nutrition in ALS.
 - Refer to gastroenterologist for evaluation and discussion.
 - Identify insurance coverage for tube feed products.
 - Identify caregiver to provide feeds.

Patients who do not want life extension technology benefit from education about the role of hospice (to decrease anticipatory anxiety), an introductory visit by hospice and referral to hospice when eligibility criteria are met (see Table 1). Patients who do want to have life extension technology at the time of respiratory failure benefit from an introduction to the pathway of care for its management, which includes planning for financial and caregiving requirements.

Case Scenario

Mary is a 72-year-old woman with ALS. She has severe dysphagia (such that she has lost 10 pounds and now weighs 78 pounds), general fatigue and a vital capacity of 42% at the time of her first visit with the ALS Center. Mary is comfortable discussing symptom management issues and is receptive to education about feeding tube placement, NIPPV support and end-of-life issues. She refuses both interventions (PEG and NIPPV), though she accepts educational materials about these choices to read at home. During the initial meeting with the ALS clinician, Mary speaks extensively about end-of-life choices.

Mary articulates great satisfaction with her life relative to what she has given and what she has received. Her desire is to have similar control over the choices surrounding the circumstances of her death as she has had in her life. Mary understands that dying from ALS is painful, and that it is possible that she will choke or smother to death. Education about hospice care was given, including how support is provided at the end of life and how patients typically die with ALS. After being educated, Mary became receptive to an introductory visit by hospice.

Mary enters a hospice program two weeks later. Choosing palliative care, she refuses a feeding tube and NIPPV intervention. Mary dies peacefully and in her home two months later, expressing gratitude the day before she died about the choices she has and the control she maintained in her dying process.

End-of-Life Expectations

Once a person has begun to explore choices about the end of life, it is important to discuss expectations. This may be difficult for patients and professionals based upon their individual comfort level with death and dying, cultural and religious beliefs and length of the therapeutic professional relationship. Identifying concepts and principles of palliative care can help patients articulate their expectations. For example:

- I expect to be free of pain.
- I expect to be in my home.
- I expect my family to be there.
- I expect to have continued contact with the ALS team.
- I expect care that promotes my dignity, independence, and control in dying.

Case Scenario

Rose is a 62-year-old woman with ALS who lives with her son, daughter-in-law and granddaughter (age 12). Rose has significant dysarthria and dysphagia with moderate limb weakness, such that she can ambulate with a walker. Requiring assistance with activities of daily living (ADLs), she receives care from a private home health aide for four hours a day, five days per week.

In discussions with health care providers, Rose and her family manifest anxiety about the dying process despite a decision to not extend her life with pulmonary technology (NIPPV or MV) or a feeding tube. Family members seem removed from a caregiving role despite their commitment to and concern for Rose.

In exploring the anxiety about the dying process with the family and Rose, the ALS clinician gains very little information regarding their fears and reticence. However, in private, the daughter-in-law and son express serious concern about their ability to support Rose as she dies due to the recent loss of their 14-year-old child. They also fear leaving their 12-year-old child home alone with Rose in the event the child would be the only person in the home when Rose dies.

Rose and her family agree to increasing formal support at home through hospice or admitting Rose to a nursing home. The son and his wife express relief about having a plan and gratitude for now feeling more capable of supporting Rose.

With the assistance of hospice, Rose remains at home during the next two months, with increased services during afternoons when she would otherwise be alone with her granddaughter. She is later transferred to a nursing home with her full consent. Rose dies with her family at the bedside one month later.

The Dying Process in ALS

Dying with ALS has been documented as a generally painless, comfortable process that varies significantly from other diseases. This is essential information to communicate to patients and their families (see Table 6). The physiologic experience of respiratory failure and carbon dioxide retention resulting in CO₂ narcosis and death can support a peaceful dying process when coupled with increased palliative care. It is important to reinforce to families that the myth of “choking” and “smothering” to death do not typically apply to ALS.

For patients and families to experience anticipatory fear and loss of control as they plan for death is to be expected. Professionals do well to: listen for stated and unstated feelings of family members (not only patients), assess the level and types of anticipation, and determine whether desires have been stated and goals established in the dying process. Whether dying with ALS is a natural experience or follows the withdrawal of mechanical ventilation, consideration of these issues is integral to the development of a care plan.

Because the functional process of dying is rarely discussed in our society, it is important to educate patients and families. Generally, two dynamics operate side-by-side in the final stage of dying, which encompass the unique values, beliefs and lifestyle of the dying person:

On the physical plane, the body begins to shut down until all physical systems cease to function. This is usually an orderly and undramatic progressive series of physical changes rather than a series of medical emergencies that require invasive interventions. Comfort-enhancing measures are most appropriate at this time.

On the emotional-spiritual plane, the spirit of the dying person begins to detach from the body, its immediate environment and all attachments. This release has its own priorities, which may include the resolution of unfinished business of a practical nature and receptivity of family members of permission to "let go." These events signify preparation for moving from this existence into the next dimension of life. Family and health care team members participate by being supportive to the patient and encouraging release during this transition.

Case Scenario

Janet is a 52-year-old woman whose sister-in-law is actively dying from ALS. She has been an involved member of the patient's care team while also supporting her brother for the past two years. Though not apparent to the family, during a home visit an ALS nurse realizes that the patient is actively dying. The nurse gently identifies the patient's shallow, patterned breathing (e.g., Cheyne Stokes), unresponsiveness and skin feels cool. Janet and her brother are thankful when the nurse shares the information, but saddened by their impending loss. This is often the paradox of feelings experienced by ALS surviving family members.

During the next hour, as Janet, her brother and the nurse talked at bedside about the patient's accomplishments (an exercise of closure for the family), the patient took her last breath and died. Janet turned to the nurse and said, "That's it? Could it be that peaceful?"

How to be Present with a Dying Person

Providers and caretakers (including hospice nurses, physicians and family members) can feel uncomfortable when a patient begins to die. Families in particular often forget to slow down from their ordinary and sometimes hectic routines to be present, supportive and reflective with their loved one. They may not notice that active dying has begun. Those in attendance, including professionals who address bowel function, blood pressure and medications, must adjust their focus and activities. Following is a list of suggestions for providers and caretakers:

- **Be Yourself**
Relate to the person, not the illness. Bring strength and vulnerability to the bedside. It's okay to cry. People who are dying continue to need intimate, natural and honest relationships. Don't hide behind functions to downplay or avoid a patient's suffering. Express feelings, give permission to die and thank the dying person for time spent.
- **Empathize**
Undivided attention is a great gift. Listen without judgment or an agenda. Be aware of feelings and non-verbal cues. Respect personal truths revealed by the dying person. Be mindful of one's own inner experience and talk with the patients about their discoveries.
- **Show Human Kindness**
Details matter. Placing a cool cloth on a perspiring brow, holding the hand of a frightened patient and listening to a lifetime of stories, convey caring and acceptance. Innate compassion and a capacity to embrace the suffering of another go a long way to comfort the dying person.

- Keep It Simple
Have confidence in the healing power of human presence. Particularly in the final days, slow down and allow room for silence. Reduce distractions. Create a calm and receptive environment (e.g., play soft and comforting music). Honor the spiritual dimensions of dying. Let go of control and be willing to acknowledge ignorance in the face of death. Talk with the patient, who may be unable to reply.

Case Scenario

John is a 57-year-old man with end stage ALS who is cared for by his wife Nancy and supported by hospice. John and his wife have been told that he has limited days to live. Nancy expresses fear and anxiety about her husband's impending death by keeping busy with care-taking tasks. She has not talked with John about what will happen after he dies. The hospice team helps Nancy to find her "voice" with John and to move from focusing on his care to just being present with him. Nancy finds this shift to be very difficult. With support from the hospice social worker, she talks with John about household finances. She conveys that she knows John is dying and that she will always love him. As his body slows down and he prepares to die, John becomes less interactive. Nancy creates a soothing and relaxed environment with soft background music (John loves music) and fresh flowers near his bed (Nancy loves fresh flowers).

Although the decreased need for "tasks" allowed Nancy to slow down, she was unable to sit with John for more than a few minutes at a time because doing so was too painful. She found solace and comfort with reflective, quiet time for herself in the next room.

Withdrawal of Ventilation

The functional, ethical and financial issues related to withdrawal of mechanical ventilation (MV) are well documented in the literature. What is not as well understood or documented are psychosocial aspects of withdrawing MV (specifically in ALS). Further research is needed.

In ALS, withdrawal of MV requires a thoughtful decision long before the mechanical intervention is initiated. Patients who choose MV at the time of respiratory failure in lieu of dying benefit from talking with an ALS team about timing and implications. ALS is progressive even after MV is initiated. Therefore, a person's function continues to deteriorate, placing increased challenges on physical and psychosocial activities of daily living. Documentation of patients' wishes and choices about under what circumstances they would want to have the MV withdrawn are best placed in patients' charts and in advance directives to ensure that patients' wishes about MV withdrawal are followed. A withdrawal request can happen anytime between soon after initiation of mechanical intervention to years later. The number of articles in the literature concerning patients' interest in and tolerance for living with advanced stages of ALS (Cazzolli and Oppenheimer, 1996) is limited and therefore these issues need further investigation. Because the capacity to enhance quality of life is possible with MV and because the current impression of MV by health care professionals is bleak, patients would benefit from further exploration of this intervention as an opportunity.

The timing and process of MV withdrawal is critical due to potential anxiety and suffering of patients and families over such life and death decisions. Initially, an assessment for appropriateness of withdrawal is best accomplished collaboratively with a neurologist and psychiatric mental health specialist (preferably with experience in terminal disease and ALS). Assessing patients for clinical depression and unrelieved pain or suffering are priorities. Every attempt must be made to understand how these issues have previously been addressed and if they impact a decision to withdraw MV. Consultation with an ethicist may help patients, families and health care providers sort out the questions and issues about the patient's physical and psychological status and the family's desires, decisions and plans.

It is helpful to hold family meetings to assess level of consensus and expectations about withdrawal as well as involvement in the patient's process of life closure. Family members work to elicit details from health care providers about the withdrawal plan (i.e., how long it may take and possible physical symptoms). Special attention is given to cultural and religious backgrounds, rituals, pre-morbid family functioning and prior experiences with death.

Regardless of the patient's location (home or hospital), a pathway must be established that adheres to the ANA position statement on withdrawal of MV. This pathway would benefit from:

- Noting that location options for MV withdrawal (home or hospital) are sometimes limited because of organizational constraints, family culture and religion, community resources and availability of personnel.
- Describing to patients and families the process and procedures for increased patient sedation, through an intravenous means, and then lowering the ventilator settings until breathing has ceased. This is ideally accomplished in a timely manner with provision of a high level of patient comfort.
- Specifying to patients and families the amount of time to expect from when the MV is withdrawn until death occurs (can range from minutes to hours).
- Developing specific medical orders for: no vital signs, no IV hydration, no teaching staff evaluations, no EKG monitors, no meals, muted phone ringer and intercom, extra family members to visit and location of room in a quiet place. As the process is initiated, notification of staff not directly involved is good protocol. If the intervention occurs in a hospital, ICU staff is best suited to the tasks. However, ICU settings are often more sterile and less personal than standard hospital rooms or homes.
- Choosing team members in advance that family members want access to before, during or after the withdrawal procedure (chaplain, neurologist, nurse, psychologist, etc.) Discussing plans for the funeral with the family, identifying bereavement resources and providing family members with information about whom to contact with questions in the 48 hours following initiation of MV withdrawal. Follow-up calls to family by health care providers following the patient's death are good practice.

Role of Health Care Team in Psychosocial Care

Understanding the unique onset and progression of ALS is essential to good psychosocial care. Health care practitioners must have knowledge of ALS pathology, prognoses and critical pathways in order to provide optimal management. An understanding of psychosocial resources of patients and families enhances practitioners' holistic care of patients.

Within a context of palliative medicine, the goals of care are to achieve and maintain maximum physical and psychological well-being of patients in the present while planning for the future. Multidisciplinary care is the recommended care strategy (see Appendices D and E of this report). It is essential that patients understand intervention goals offered by the team, which are woven into comprehensive care plans. Counseling and discussions of how care plans benefit patients throughout the disease result in greater patient participation.

It is crucial for members of the health care team to understand patient adaptability so that clinicians can knowledgeably select interventions that promote and enable patients' adaptive capacities. Only in this way can individualized, appropriate care be effectively and economically provided to the patient affecting the quality of their living and dying.

“The team is a flexible blend of skills designed to meet the ever changing needs of the patient and family,” says Elisabeth Earnshaw-Smith.

Health care professionals need to maintain their own physical and psychological wellness, especially because of the intensity of labor and emotions associated with the care of ALS patients. ALS practitioners may benefit from individual or group psychotherapy (the “group” being the ALS team), which helps to decrease burnout and optimize effectiveness with patients. Further studies are needed to explore the effects of psychotherapy on health care providers working with patients with ALS.

The following health care provider “voices” are representative of feelings that clinicians often have when working with ALS patients and their families:

“I go home and cry at night in the shower – I feel powerless at times and do not know how to help them (ALS patients and families) anymore.”

-- ALS Nurse

“I am tired – almost burned out by the demanding nature of dying patients. I am so bad with setting boundaries so it’s my own fault when a patient calls me three times in one day to ask if I am taking care of a prescription. I can’t blame them, though – I would probably do the same thing if it were me.”

-- ALS Physical Therapist

“I know they will all die. I do not have false expectations about them or myself and that preserves my integrity as a professional caregiver and stops me from burning out. All the same, it is difficult to have every patient die slowly before your eyes.”

-- ALS Social Worker

Life Closure

Closure is defined as bringing something to an end. Completing “unfinished business” is very important to a person’s sense of closure and readiness to die. Issues may be functional, financial, spiritual, or psychosocial and will vary, depending upon the culture and family system.

Regardless of religious belief or spirituality, closure can begin months before or in the minutes preceding the actual dying process. Often, a “life review” examines what has been achieved or left behind. Because closure has not been formally researched in ALS, the field would benefit from studies similar to those done with cancer patients.

Open-ended questions facilitate discussion and offer opportunities to identify goals or tasks that can still be met: If you knew that you would die soon, what is left undone in your life?

Providing patients with self-reflective questions can help them address closure issues:

- Who am I?
- How have I used the gift of a human life?
- What do I need to "clear up" or "let go of" in order to be more peaceful?
- What has given my life meaning?
- For what am I grateful?

Case Scenario

Ralph is a 70-year-old man with ALS who is dependent on NIPPV 24 hours per day. He lives with his wife and has two grown children. During his ALS Center visits, Ralph knows that his time is limited because the NIPPV is providing less relief from breathlessness now than it did initially. Ralph is an expressive person who freely discusses his thoughts about dying. He talks about feeling somewhat empty because he never had a grandchild. He fears that he will not leave enough behind for others to remember him. He speaks about the meaning of his life, how he likes to help others, and how his

“payback” is unfinished. Ralph worries about having received care from others during the course of his illness that he cannot “pay back.”

When a call comes to Ralph’s ALS Center requesting participants for the Center’s patient educational video about ALS, Ralph is asked to share his experience of living with ALS. He talks about his diagnosis, his care at the ALS Center and hospice, and his thoughts about how to live with ALS. It pleases him to know that he will live on in this tape. He feels that having ALS was not in vain because he has been given an opportunity to teach others about how to live with ALS. Ralph dies one month after completing his part of the tape.

RECOMMENDATIONS TO THE FIELD

Practice Recommendations

Providers would benefit from:

1. Increased understanding of the principle of psychosocial care as it relates to end-of-life issue;
2. Incorporation of comprehensive psychosocial care and interventions at the end of life for each patient with ALS and their families;
3. Inclusion of a psychosocial expert in the multidisciplinary ALS team; periodic psychosocial education and training for all ALS team members; and
4. Staff, patient and family education that involves a full spectra of psychosocial information.

Research Recommendations

Research studies to investigate, describe and test interventions for:

1. Psychosocial care at the end of life;
2. Components of psychosocial care most pertinent to ALS patients at the end of life,
3. Effects of psychosocial care on dying patients and their families; and potential outcomes for quality of dying;
4. Decision making about end-of-life choices and accompanying psychosocial factors;
5. Decision making about withdrawal of mechanical ventilation and accompanying psychosocial factors; and
6. Effects of caring for ALS patients on health care professionals relative to wellness, burnout and psychosocial factors.

Policy Recommendation

1. Medicare, Medicaid, HMOs and other insurers should reimburse costs for psychosocial care.
2. ALS voluntary health organizations and ALS clinicians should set standards of care for ALS clinics that include:
 - a. Multidisciplinary teams to provide care in organized ALS clinics, and
 - b. Psychosocial care as an integral component of the care provided to patients and families.

REFERENCES

Albert SM, Murphy PM, Del Bene ML, Rowland, LP. “A Prospective study of palliative care in ALS: choice, timing, outcomes.” *J Neurol Sci*, 169: 108-113, 1999.

ANA Position Statement. “Assisted suicide, euthanasia and the neurologist.” *Neurology*, 50; 596-597, 1998.

Borasio GD, Walsh D. *Palliative Care in Amyotrophic Lateral Sclerosis*. Oxford, New York: Oxford University Press, 2000.

Cazzolli PA, Oppenheimer EA. “Home mechanical ventilation for amyotrophic lateral sclerosis: nasal compared to tracheostomy-intermittent positive pressure ventilation.” *J Neurol Sci*, 139 (Suppl): 123-128, 1996.

Caroscio JT. (ed.) *Living and Coping with ALS Psychosocial Impact in ALS: A guide to Patient Care*. New York, NY: Thieme Publishers, 1986.

Longo MB. “Facilitating acceptance of a patients decision to stop treatment.” *Clinical Nurse Specialist*, 7(3): 116-120, 1993.

McDonald ER, Wiedenfeld SA, Hillel A, Carpenter CL, Walter RA. “Survival in Amyotrophic lateral sclerosis. The role of psychological factors.” *Arch Neurol*, 51: 17-23, 1994.

McDonald ER. “Quality of life from the patient’s perspective.” *J Neurosci Nurs*, 27: 222-223, 1995.

Moss AH, Oppenheimer EA, Casey P, Cazzolli PA, Roos RP, Stocking CB, and Siegler M. “Patients with amyotrophic lateral sclerosis receiving long-term mechanical ventilation: advance care planning and outcomes.” *Chest*, 110(1): 249-255, 1996.

Sebring JDL, Moglia P. “Amyotrophic lateral sclerosis: psychosocial interventions for patients and their families.” *Health Soc Work*, 12: 113-120, 1987.

Table 1: General ALS Resources

The ALS Association (ALSA)
27001 Agoura Road, Suite 150
Calabasas, CA 91301
(818) 880-9007
(800) 782-4747
www.alsa.org

MUSCULAR DYSTROPHY ASSOCIATION (MDA)
3300 E. Sunrise Drive
Tucson, AZ 85718
(602) 529-2000
(800) 572-1717
www.als.mdausa.org

World Federation of Neurology in ALS
www.wfnals.org

Online ALS Support Forum:
Bob Broedel's "ALS Digest"
bro@huey.met.fsu.edu

Table 2: Hospice Qualifying Symptomatology:

(A) or (B) must be present

(A) does not require associated symptoms

If just (B), at least 2 other Respiratory Indicators must be present or 1 respiratory and 1 Nutritional Indicator present

(A) FVC \leq 30% of predicted _____

(B) FVC \leq 60% of predicted with a steady decline over past 2-3 months _____

Other Respirator Insufficiency Indicators:

Shortness of Breath _____

Shallow Breathing _____

Paradoxical Breathing _____

Inability to be supine due to diaphragmatic weakness _____

Non-explosive cough _____

Inability to blow nose _____

Breath Support poor as evidenced by accessory muscle use _____

Endurance Poor (excessive fatigue) _____

Significant insomnia related to diaphragmatic weakness _____

Nutritional Insufficiency Indicators:

Excessive oral secretions with dysphagia _____

Nutritional Compromise / Dehydration _____

(With or without feeding tube)

Weight Loss > 10% of body weight, refusing PEG _____

Aspiration or choking (on food or liquid) _____

Standard Requirements:

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

Table 3: National Referrals

Association for Death Education, Counseling
638 Prospect Ave., Hartford, CT 06105.

ADEC is dedicated to improving the quality of death education and death related counseling and caregiving.

Phone: 860-586-7503

Web site: <http://www.adec.org>.

National Hospice and Palliative Care Organization

1901 N. Moore St., Suite 901, Arlington, VA 22209.

NHPCO provides referral services to link individuals with hospices in their local communities, and various educational programs.

Phone: 703-243-5900

Web site: <http://www.nhpc.org>

GriefNet

A comprehensive gateway to bereavement and grief-related resources on the Web, including a support community for people dealing with death, grief and major loss including life-threatening and chronic illness.

Web site: <http://griefnet.org>.

E-mail: griefnet@griefnet.org

ALS Support Groups: see MDA, ALS Association resources

Online ALS Support Forum: Bob Broedel's Digest (apply at: bro@huey.met.fsu.edu)

Table 4: National Hospice and Palliative Care Organization

1901 N. Moore St., Suite 901, Arlington, VA 22209.

NHPCO provides referral services to link individuals with hospices in their local communities, and various educational programs.

Phone: 703-243-5900

Web site: <http://www.nhpc.org>

Choice In Dying

200 Varick Street, New York, NY 10014.

Provides information about Advance Directives (Living Wills and Durable Powers of Attorney for Health Care). Call for state-specific forms.

Phone: 800-989-WILL.

Web site: <http://www.choices.org/>

Hospice Referral Key Points

- Utilization of an ALS-specific hospice referral form (see attached).
- Before referral, call to establish contact with hospice and frame out the issues related to this patient.
- Use and establish hospices repeatedly in certain catchment areas to develop their knowledge and experience with ALS.
- Establish contact after the evaluation by hospice is done to further discuss plan of care; standing hospice orders for meds and treatments and plan for contact every 2 weeks between the hospice and ALS team (initially and then every month).
- Emphasize with the hospice that their services are a compliment to ALS team and not in lieu of ALS team.
- Clearly identify to the hospice the pre-morbid functioning of the family (previously elicited by psychosocial evaluation) and the present challenges that are being and need to be addressed in collaboration with the hospice.
- Discuss with the hospice (provide written material) on the signs and symptoms of ALS death and how they will and do differ from cancer patients.
- Describe and discuss in detail the use of standing hospice medications and how they are used in ALS versus cancer patients and what to expect from the medications.
- Discuss and identify the role of NIV as a comfort measure (albeit in some minority a life-extending measure) and how to titrate it to improve comfort and then to allow death to occur comfortably.
- Encourage the hospice to set up a different billing rate with the respiratory company providing the NIV so that it does not financially drain the budget for that patient.
- Discuss the role and use of the PEG as non-life-extending measure and how to titrate its use to allow death to occur comfortably.
- Identify the pathway of whom to call on the ALS team for emergencies or questions (as hospices are most used to the traditional medical practice of one doctor and not a multidisciplinary team).
- Discuss the potential of autopsy donation at the time of death and how that would be facilitated.

Table 5: Signs and Symptoms of Approaching Death (Adapted from Hospice of Central Florida)

Signs and Symptoms of Approaching Death – Non ALS (Adapted from the Hospice of Central Florida www.hospicecares.org)	Signs and Symptoms of Approaching Death in ALS (M. L. Del Bene: Columbia University Eleanor and Lou Gehrig MDA/ALS Center)
<p>Skin and Temperature Changes The person’s hands and arms, feet and then legs may be increasingly cool to the touch, and at the same time the color of the skin may change. This is a normal indication that the circulation of blood is decreasing to the body’s extremities and being reserved for the most vital organs.</p>	<p>Infrequently do we see overt changes in color or temperature of the skin in the days preceding death – as the circulatory system is not affected in ALS.</p> <p>Cyanosis of the lips, toes and fingers may be seen in the hours before death.</p>
<p>Sleeping The person may spend an increasing amount of time sleeping, and appear to be uncommunicative or unresponsive and at times be difficult to arouse. This normal change is due in part to changes in the metabolism of the body.</p>	<p>ALS patients will describe feeling more fatigued but may not actually sleep more in the days or hours before death. Somnolence is more prevalent in ALS.</p>
<p>Excessive sleeping may occur several days before death and then a semi-comatose state for the 72 hours before death.</p>	<p>Somnolence is seen as a primary indicator of respiratory insufficiency.</p> <p>There is not typically a protracted period of comatose state before death in ALS. An extra nap or need to rest for a “little while” is often the only indicator that the patient was about to die.</p>
<p>Disorientation Confusion about the time, place, and identity of people surrounding him or her, including close and familiar people often occurs. This is also due in part to the metabolism changes.</p>	<p>Disorientation in the days before death is rare in ALS. The experience of fatigue makes interactions and communications difficult. Most often though, ALS patients are alert and oriented up until death.</p>
<p>Incontinence The person may lose control of urine and/or bowel matter as the muscles in that area begin to relax.</p>	<p>Not a typical pattern in ALS. Usually a patient has decreased urine output due to the natural dehydration process and decreased bowel movement decrease due to decreased intake and decreased peristalsis with the dying process.</p>
<p>Congestion The person may have gurgling sounds coming from his or her chest as though marbles were rolling around inside. These sounds may become very loud (terminal congestion). This normal change is due to the decrease of fluid intake and an inability to cough up normal secretions.</p>	<p>Rarely do ALS patients have terminal congestion given the natural dehydration process.</p> <p>Oral secretions may be present – either thick or thin.</p>
<p>Excessive pulmonary secretions may fill the oropharyngeal cavity.</p>	

Restlessness

The person may make restless and repetitive motions such as pulling at bed linen or clothing. This often happens and is due in part to the decrease in oxygen circulation to the brain and to metabolism changes.

This does not usually occur with ALS patients since carbon dioxide retention promotes sedation and somnolence.

Urine Decrease

The person's urine output normally decreases and may become tea colored referred to as concentrated urine. This is due to the decreased fluid intake as well as decrease in circulation through the kidneys.

With dehydration urine color and amount may change, though it is often so subtle that it is not recognized prospectively as a sign of dying.

Fluid and Food Decrease

The person may have a decrease in appetite and thirst, wanting little or no food or fluid. The body will naturally begin to conserve energy, which is expended on these tasks.

ALS patients may have a subtle change in food and fluid intake. It is so subtle though that it is not recognized prospectively as a sign of dying.

Energy expenditure with eating is often too extensive, and intake is limited to conserve energy.

Breathing Pattern Change

The person's regular breathing pattern may change with the onset of a different breathing pace. A particular pattern consists of breathing irregularly, i.e., shallow breaths with periods of no breathing of 5 to 30 seconds and up to a full minute. This is called Cheyne-Stokes breathing.

This patterning of breathing is less common in ALS. As shallow breathing results from diaphragmatic weakness and thus prohibits stridor and Cheyne Stokes breathing. Breathing is shallow (possibly with periods of apnea) and not presented as a struggle.

The person may also experience periods of rapid shallow pant-like breathing. These patterns are very common and indicate decrease in circulation in the internal organs.

It is usually unobservable to the untrained professional that the patient is even in the very last stage of the disease and is about to die.

The person may also have rapid deep breathing patterns that appear to be effortful and difficult (stridor).

Sign of pre-respiratory failure is often a subtle pulling of the neck muscles, with little to no movement of the chest or abdominal wall (and prior to that the shoulder and other accessory muscles and paradoxical breathing).

Subtle changes of accessory muscle use in the shoulders and neck are the indicators for impending death rather than heavy, congested breathing seen in the non-ALS dying person.

Table 6: Tool for Optimizing Psychosocial Care in ALS
Maura Del Bene

1. Clarify and normalize the impact of the illness on the family and patient.
 - Provide clear and concise information on the disease through written materials and resources.
 - Listen actively to their perception of their experience of the disease.
 - Clarify and support how they are feeling.
2. Empower the family to believe in their ability to problem solve and manage.
 - Instruct on symptom management.
 - Give a clear description of the multidisciplinary team members, their roles and how, when to contact them.
 - Demonstrate and assist the family on problem solving techniques.
 - Give feedback on their performance.
3. Assess the families' definition of the disease.
 - Reframe the patient's narrative to be hopeful, helpful and positive.
 - Normalize the situation.
4. Identify resources inside and outside the family.
 - Emotional support, referrals to community clinicians, books and internet resources.
5. Encourage family and patient to connect with community resources to decrease isolation.
 - Support groups, peer support, home care agencies, medical suppliers, religious community connections.
6. Provide clear and concise medical information and medical resources.
 - Written literature on ALS, manuals on the multidisciplinary team interventions, educational materials related to disease progression and interventions, ALS mailing lists.
7. Identify the functioning patterns of the family as they existed before the illness and know that they will be the same throughout the disease.
 - Identify the strengths and weakness of the family unit and assist the family in understanding the manner in which the family members can support each other.
 - Refer to a psychotherapist for family counseling (proactively).
8. Encourage the family and patient to plan for the future while the patient is able to participate.
 - Advanced directive, tissue donation, use of specialized equipment for speech synthesis, environmental control, wheeled mobility, bilateral forearms orthoses.

Table 7: Children's Bibliography

(Let's Talk About) Feeling Angry (Sad, Frustrated etc.) [series on feelings for ages 3-6]
by Maggie Smith
Scholastic Trade

The Fall of Freddie the Leaf [ages 4-8]
by Leo F. Buscaglia
Holt Rinehart Winston; ISBN: 0805010645

Lifetimes: The Beautiful Way to Explain Death to Children [ages 4-8]
by Bryan Mellonie
Bantam Doubleday Dell Pub (Trd Pap)
ISBN: 0553344021

I'll Always Love You [ages 4-8]
by Hans Wilhelm
Crown Pub; ISBN: 0517572656

Sad Isn't Bad: A Good-Grief Guidebook for Kids Dealing With Loss [ages 4-8]
by Michaelene Mundy
Abbey Press; ISBN: 0870293214

Nana Upstairs, Nana Downstairs [ages 4-8]
by Tomie De Paola
Puffin; ISBN: 0698118367

How It Feels When A Parent Dies [ages 4-8]
By Jill Krementz
Alfred A. Knopf, New York
ISBN: 0394758544

BEREAVEMENT IN ALS

INTRODUCTION

Bereavement is a natural part of death and dying and is therefore an important topic for patients with ALS, their caregivers and family members. Portions of the process are familiar, but the spectrum of bereavement in ALS may be broader than appreciated. The process of bereaving is most frequently used in the context of death, and focuses on the loss of individuals. However, during the ALS disease process the patient, caregiver and family members can also mourn both the loss of the patient's physical function and their respective independence.

Objective

This section reviews the spectrum of bereavement in ALS and shows how bereavement is part of the disease process. It is important that health care providers have a thorough understanding of the bereavement process to help guide patients and family members to better understand their feelings. This can lessen psychological stress and lead to a better sense of well-being.

Methods

This section includes a literature search of bereavement in ALS in journal articles and books. In addition, broad concepts of bereavement have been obtained from sources related to other diseases. Since little is published about bereavement in relation to ALS, the experience of colleagues working with ALS patients has been sought.

WORKGROUP FINDINGS

Definitions

Several terms are associated with bereavement. Bereavement is a process that begins when something is lost or someone dies. Grief is the feeling of sadness associated with the loss. Mourning is the expression of sorrow and grief. In this section on bereavement, these terms will be used interchangeably.

Spectrum of Bereavement in ALS

Bereaving is commonly used in the context of the death of an individual. However, in chronic diseases where death is expected from the outset, bereaving can start with knowledge of a fatal prognosis at time of diagnosis, which is often the case with ALS.

While grieving is often associated with those left behind, patients also grieve the loss of abilities. Similarly, caregivers and family members can grieve the loss of their independence as they devote more time to patient care.

Bereavement takes into account the age of affected individuals, and is influenced by historical events and issues that have occurred in a family. Accordingly, bereavement can follow a number of pathways and may travel through many stages.

Bereavement extends beyond grief and mourning to affect many aspects of physical, social, emotional and psychological health including loss of social support and function, physical symptoms, change of hopes and changes in financial status (McMurray, 2000).

Stages of Bereavement

While the process of bereavement is incompletely understood, it can be divided into stages (McMurray, 2000).

PROCESS	MANIFESTATIONS
Accepting reality of loss	Numbness, disbelief
Working through pain of grief	Struggling to understand, yearning, searching
Adjusting to environment without patient	Depression, anguish, apathy, fatigue
Emotional relocation, moving on	Making sense of experience with themselves, socially

Grief takes many forms. With a progressive illness like ALS, family members have time to prepare for the death of the patient. The bereavement process, therefore, differs from sudden and unexpected death.

Anticipatory grief may start with the diagnosis, and can be magnified by the imagination and inaccurate or limited knowledge about the manner of death in ALS. A succession of losses occurs with disease progression, and each impending loss of function elicits a period of bereavement in a recurrent pattern.

Bereavement occurring in stages can help caregivers and families prepare for a patient's death and can lessen the intensity and duration of bereavement after death. Progressive loss of a patient's functional abilities leads to loss of caregiver independence and caregivers can experience physical and emotional relief when their patient dies. Relief can counterbalance grief from loss at death (Murphy, 2001).

Bereavement must be considered within a family context. Previous intra-family relationships influence anticipatory and final bereavement. Sometimes, illness brings family members together, and sometimes, estranged family members return to the fold. A united family is likely to be stronger than a divided one, and reuniting family members is an important part of preparing for the end of life.

Multiple generations provide additional support within families. For example, grandchildren provide a sense of balance between aging and death and youth and promise. Furthermore, conveying to children that infirmities and death are natural parts of the life cycle can help them deal with subsequent deaths of family members and friends (see Vignette 1).

Responses to death can vary among religions and cultures, and inquiring about customs can be a means of opening the topic of bereavement. Health care providers must consider the impact of customs and religion upon patients and their families.

Family financial and social resources impact bereavement. Because division of property can generate strife, discussions about property distribution can reduce conflict during the bereavement process. Financial loss during the illness can leave families bitter and financially struggling. While there are little data on the overall financial impact of care on ALS, the cost of home ventilator care has been studied with the finding that financial burdens cause hardship and bitterness and guilt (Moss et al., 1993; Bromberg et al., 1996). Social resources and support may be available through support groups, governmental agencies and bereavement support groups.

Bereavement and Health Care Providers

Providing health care to ALS patients can be emotionally taxing, and providers often form strong attachments to patients. When several patients die over a short period of time, providers are likely to suffer. Health care providers must also face their mortality as they prepare patients and families for

death. They must also learn to manage emotional involvement with patients and seek counseling to address repeated losses, as needed (Murphy, 2001).

Resources Available to Patients and Families

ALS has unique features for patients and families that distinguish it from other progressive and fatal diseases such as cancer, which holds hope for remission. What is known about bereavement in general is not directly relevant to ALS. While the differences are important, general literature on bereavement *is* useful background information. Many books have been published about grieving and bereavement, and there are organizations that offer a variety of services. Table 1 includes books and support organization listings.

Discussions in the literature on bereavement in ALS primarily occur in connection with hospice care (Thompson & Murphy, 2001). Special stresses placed on the patient and caregiver are being considered in the literature (Oliver & McMurray, 1993; Thompson & Murphy, 2001).

Resources Needed for Patients and Families

Bereavement should begin at the time of diagnosis, and extend through the course of the disease to a period after death. There is growing recognition that end-of-life care must be tailored to the chronic time course of ALS (Lynn, 2001). Although a structured approach to death and dying is preferable, both unstructured and structured programs should be available.

Unstructured programs include an increased awareness of the bereavement process and stages, and addressing issues as they arise. Structured programs focus on specific issues and include formal organizations with the goal of helping the family achieve a satisfactory closure at death and through the bereavement period.

Unstructured Programs

Open acknowledgement of the bereavement process is essential. This includes an understanding of the stages so that cues can be recognized. Providing appropriate information to allay fears is reassuring. Being available as a partner with the patient, spouse and family during the process is important. Follow-up at the end closes the health care provider's role in the process.

Acknowledgement of the Process

At the time of diagnosis, describing the course of ALS helps patients and families come to terms with the progressive nature of the illness and impending death. The perspective that "death is a consequence of living" can be helpful.

Anticipation of Stages of Bereavement

Acquiring knowledge about stages of bereavement as they could relate to stages of disease progression is aided by reading, attending lectures and participating in discussions (Curry, 1990; Fitzgerald, 1994; James and Friedman, 1998). Health care providers must recognize their own feelings about loss of function, death and grief (Cassidy, 1992). Patients and families can most easily discuss their feelings when providers seem more comfortable with these topics.

Providing Appropriate Information

It is important at different points in the disease to review the manner of death, especially at the end of life because the actual manner of death creates fear in patients and families. Worry that they will choke or that death will be painful is common. A review of patient deaths from ALS indicates that death was peaceful in almost all cases (Neudert et al., 2001).

Being a Partner in the Process

It is important for patients and families to feel supported. In the past, a diagnosis of ALS was associated with abandonment by the health care profession because “there is nothing that can be done, no return appointment is necessary, put your affairs in order.” The development and proliferation of ALS clinics in the United States over the past 10 to 15 years has changed this paradigm. However, there is a substantial difference between providing physical care and providing psychological care. Many health care providers still do not understand the importance of their role in the grieving process. Letters of gratitude from families are an indication of the importance of this role.

Follow-up

It is easy for health care providers to distance or step out of the process when death occurs. However, formal bereavement begins at the time of death. A personal phone call and follow-up card or letter is extremely important to the family. Omitting this type of closure can leave a long-lasting gap (Bedell et al., 2001). Attendance at funerals or memorial services provides opportunities for the whole family to come to closure with the health care providers.

Structured Programs

Structured bereavement programs are most effective when combined with personal elements of unstructured programs and are tailored to individual patient and caregiver needs.

Written Handouts

Written handouts about death and bereavement can help patients and caregivers, and family members, especially those who cannot attend clinic visits. The amount and type of information given to families, patients and caregivers must be adjusted to meet individual family needs. Handouts raise awareness of feelings and fears that patients and families often experience about the dying process. Adequate time to discuss issues raised in written materials with providers is important.

Availability of Professional Psychological Support

Bereavement is a set of complex psychological processes that may not be clearly and fully recognized by the family or health care providers. Social workers, psychologists and psychiatrists are helpful in understanding bereavement. Bereavement training for the health care providers by experienced psychological staff can improve the skills of health care providers.

Support Groups

Formal bereavement support groups help family members reach closure. ALS may have special issues that are not routinely addressed in bereavement groups for other disorders, such as cancer. Individuals with experience, specialized education and knowledge about grieving usually lead support groups. Within support groups, grieving individuals receive support from other group members. Some support groups have an annual “remembrance” event or “memorial service,” when those who have passed away during the year are remembered.

Continued Involvement

Continued involvement with the ALS community helps some family members. Involved family members whose routines are halted with the patient’s death may benefit from helping other families with ALS. Receiving ALS clinic newsletters and invitations to continue with or lead local support groups can be therapeutic.

Defining the Gaps

Most information about bereavement is not specific for ALS. A review of ALS literature reveals little information about bereavement. Accordingly, more ALS-specific information on bereavement is warranted. Specific gaps are listed below.

Understanding How Bereavement in ALS Differs from Bereavement in Other Progressive Fatal Diseases

Clinical features that differentiate ALS from other progressive and fatal disorders include uncertainty about the disease's origin as well as a uniform progressive course that has no available treatment or hope of temporary remission. ALS patients lose strength and function without organ failure. It is not clear whether ALS clinical issues require a unique approach to bereavement.

Understanding Psychological Aspects of Bereavement in ALS Patients

A unique characteristic of ALS is the continual loss of strength and function, thus allowing little stability or time to adapt. All patients with progressive fatal diseases grieve the loss of being with their family. How these factors are addressed to ease a patient's burden requires further study.

Grieving by the ALS patient for losses can affect quality of life. Acceptance of losses may represent a natural process, and has been called a "response shift," defined as a resetting of expectations over the course of the disease (Wilson, 1999). Incomplete response shifts may be associated with lower quality of life, somatic complaints, feelings of anger and so forth (Wilson, 1999). The ability of patients to grieve may be linked to their response shift.

Understanding Psychological Aspects of Bereavement in ALS Caregivers

ALS caregivers must provide more patient care with disease progression while losing their own independence. Grieving over loss of the patient's function and grieving over loss of their independence may create conflict within the caregiver. For example, they may experience relief from the burden of care and a return of independence at death. Limited data exists to substantiate the negative impact of progressive care on caregivers (Bromberg et al., 2001). Little data are also available about how to address conflicting feelings of caregivers in the context of bereavement.

Bereavement Management by Support Groups, National Organizations and Hospice

There is a growing use of hospice and palliative care for ALS patients (Oliver, Borasio, Walsh, 2000). It is not clear how hospice can best be used to help patients and families during the course of the disease. Although principles underlying hospice are clear, the low prevalence of ALS leaves most hospice caregivers unfamiliar with its unique aspects at the end of life. An assessment of hospice experience with ALS patients and familiarity of hospice workers with disease-specific management will improve caring for this small population of patients.

National ALS organizations have few specific publications on grieving and bereavement for the patient and caregiver and family. Few regional ALS support groups offer specific bereavement support.

RECOMMENDATIONS TO THE FIELD

Practice Recommendations

Clinics are strongly encouraged to offer bereavement support, informally or formally, addressing the grieving process for patients, caregiver and family during the course of the disease and at the time of death.

Research Recommendations

GATHERING BASIC DATA

1. Determining the scope of bereavement in ALS and differences compared to other diseases, such as cancer, can be accomplished with focus groups of ALS and cancer patients and caregivers. Further exploration of bereavement models might encourage integration of ALS-specific concerns.

BEREAVEMENT SUPPORT GROUPS

2. There appear to be fewer bereavement support groups for ALS compared to other diseases. The frequency of use of bereavement support groups by ALS caregivers is not known. Unique caregiving features related to ALS require disease-specific bereavement support groups. Studies are needed to assess the usefulness of specialized support groups.

RAISE AWARENESS OF THE BEREAVEMENT PROCESS

3. As information about ALS bereavement becomes available, dissemination of specific recommendations in the form of booklets for patients and caregivers can be effective.

REFERENCES

EPEC Participant's Handbook, Education for Physicians on End-of-Life Care (The EPEC Project). © EPEC Project, The Robert Wood Johnson Foundation, 1999.

American Hospice Foundation. www.americanhospice.org/index.htm, 101.223.0204.

Bedell SE, Dadenhead K, Graboys TB. "The doctor's letter of condolence." *N Engl J Med*, 344: 1162-1164, 2001.

Bromberg MB, Forsheo DA, Iaderosa S, McDonald ER. "Ventilator dependency in ALS: management, disease progression, and issues of coping." *J Neuro Rehab*, 10: 195-216, 1996.

Casarett D, Kutner JS, Abrahm J. "End-of-Life Care Consensus Panel." *Ann Intern Med*, 134: 208-215, 2001.

Cassidy S. *Sharing the Darkness: The Spirituality of Caring*. Orbis Books, 1992.

Lynn J. "Serving patients who may die soon and their families." *JAMA*, 285: 925-932, 2001.

McMurray A. "Bereavement." In, *Palliative Care in Amyotrophic Lateral Sclerosis (Motor Neuron Disease)*, Oliver D, Borasio GD, Walsh D (eds). Oxford: Oxford University Press, 2000.

Moss AH, Casey P, Stocking CK, Roos RP, Brooks BR, Seigler M. "Home ventilation for amyotrophic lateral sclerosis patients: outcomes, costs, and patient, family and physician attitudes." *Neurology*, 43: 438-443, 1993.

Murphy NM. "Mourning." In *Amyotrophic Lateral Sclerosis. A Guide for Patients and Families, 2nd edition*. Mitsumoto H, Munsat TL (eds). New York: Demos Medical Publishing, 2001.

Neudert C, Oliver D, Wasner M, Borasio GD. "The course of the terminal phase in patients with amyotrophic lateral sclerosis." *ALS* (in press).

Oliver D, McMurray N. "Bereavement – whose responsibility?" *Pal Med*, 7 (Suppl 2): 73-76, 1993.

Thompson B, Murphy NM. "Hospice Care." In *Amyotrophic Lateral Sclerosis. A Guide for Patients and Families, 2nd edition*. Mitsumoto H, Munsat TL (eds). New York: Demos Medical Publishing, 2001.

Wilson I. "Clinical understanding and clinical implications of response shift." *So Sci Med* 45: 1577-1588, 1999.

Table 1: Resources That Are Available to the Patient and Family**Books**

Curry CL. *When Your Spouse Dies*. Ave Maria Press, Notre Dame, IN, 1990

Fitzgerald H. *The Mourning Handbook*. A Fireside Book, Simon & Schuster, NY, 1994.

James JW, Friedman R. *The Grief Recovery Handbook*. Harper Perennial, NY, 1998.

National Organizations

National Family Caregivers Associations offers publications and bereavement kits available directly to caregivers (National Family Caregivers Associations: www.nfcacares.org; 800.896.3650).

American Hospice Foundation and the Hospice Foundation of America offer booklets on understanding grief (Hospice Foundation of America: www.hospicefoundation.org/; 800.854.3402.).

Last Acts® offers booklets that address different aspects of grief (Last Acts: www.lastacts.org/section/resources/).

Vignettes

1. Leaving a Book –Taking on a New Activity:

Mrs. H was losing the use of her hands. She had several grandchildren under the age of 6 who visited her frequently. She used to bake and tell them stories. They accepted her weakness without question and enjoyed their time with Grandmother.

She decided to put her stories into a book. In addition to making up a story about little children, she wanted to illustrate the book. Although she had no previous experience in art, she took up watercolor painting. The result was a simple story with a colorful illustration for each page.

Clinical Points:

Working on a project that will be lasting.

Accepting the cycle of life and death.

Expressing feelings for a person (*i.e.* grandchild) with overflow to others (*i.e.* children).

Taking on a new skill or using an existing one (*i.e.* painting).

2. Making a List of Good and Bad Points:

Mr. P, along with a close family, assumed full care of Mrs. P. After she died, Mr. P wrote a list of good and bad points about the disease.

Good points: Enjoying time with patient. For example, spending time (showered together, shampooed hair, applied lotion to her body).

Bad points: Loss of function (limbs, speech), change in personal relationship (no longer passionate) and social relationships (no longer desires to go out socially).

Clinical Points:

Realizing there can be good and difficult times.

Expressing the items in formal manner (written list) with offer to review with clinical team.

Spirituality and End-of-Life Care in ALS

INTRODUCTION

Preservation of human dignity at the end of life cannot be disconnected from attention to spiritual needs of the dying. For this reason, in its definition of Palliative Care, the World Health Organization gives the same level of importance to the management of spiritual care, psychosocial care and physical care (WHO, 1990). However, health care providers often neglect patients' spiritual needs, even in the context of end-of-life care.

WORKGROUP FINDINGS

Definition of Spirituality

The word "spiritual" has several implications and all but escapes definition. There is often confusion between the words "religious" and "spiritual" (Doyle, 1992). Sykes (2000) defines "spiritual" as "the need to find within present existence a sense of meaning," which may or may not involve a religious framework. Most men and women seek this "sense of meaning," particularly as they approach death. Of necessity, there is significant overlap between religious, spiritual and psychological issues, the extent of which varies greatly between individuals.

It is important to point out problems that arise when spirituality and religiousness are confused. Spirituality, being centered on the meaning of life, may not be assessed by monitoring adherence to defined formal practices, such as church going or praying. Conversely, data on frequency of attendance at religious services need not necessarily correlate with spiritual well-being. This distinction must be kept in mind, because patients also tend to confuse the two terms.

Role of the Physician

Physicians tend to have numerous excuses for not attending to the spiritual aspect of care, including lack of specific competence, inadequate training and insufficient time, among others (Doyle, 1992). However, data from recent surveys indicate that 70% of cancer patients want to discuss spiritual issues with their physicians (Balducci, Meyer, 2001). The problems of professional boundaries and patient-physician relationships have recently been reviewed (Post et al., 2000). It is important to remember that spiritual care for patients and families can only be administered by those individuals, regardless of their profession, who have been taking very good spiritual care of themselves and who have a firm spiritual standing.

Role of the Spiritual Counselor or Chaplain

Collaboration with chaplains and spiritual counselors is of paramount importance. Cross-cultural differences in spiritual attitudes and needs with patients of diverse ethnic and religious backgrounds must be acknowledged and responded to in a sensitive way. This requires a basic understanding of the different ways death and dying are viewed in the major religious traditions throughout the world. Hospital or hospice chaplains have often received specific training in these issues and usually have contacts with ministers of several faiths who can be called in when needed.

Role of the Family

Spiritual care is not limited to patients, but should encompass the whole family as a means of preventing problems during bereavement. Some of the initial research data indicate that bereavement in relatives after an ALS patient's death may be particularly severe and prolonged (Martin, Turnbull, 2000), most likely due to the huge burden of care in the months preceding death. On the other hand, strategies to reduce the impact of loss of a loved one through appropriate counseling are available

(McMurray, 2000). Importantly, the acknowledgment that the process of bereavement in ALS starts immediately after diagnosis is communicated. This initial grief is often referred to as anticipatory grief; therefore, callous delivery of the diagnosis may affect the psychological adjustment to bereavement (Ackerman, Oliver, 1997). This is true for patients and relatives.

Identification of Existing Resources

A recent study of 46 individuals with ALS (Murphy et al., 2000) indicated that those with a higher degree of spirituality were:

- more likely to have completed advance directives,
- less likely to have gastrostomy; less fearful of death and dying,
- less likely to participate in support groups, and
- more likely to choose a natural death over life-prolonging mechanical ventilation.

In this study, the Beck Hopelessness Scale was significantly related to levels of religiousness ($p=0.04$) and spirituality ($p=0.05$). The authors concluded, "...although religion and spirituality may not offer a cure, they may ease adaptation to a grave diagnosis and the transitions of life that occur in the process of dying." Several cases in the literature support these results that spiritual practice can enhance patients' ability to cope with the disease (Borasio, 2001).

Models

Kellehear (2000) developed a theoretical model of spiritual needs in palliative care based on a review of palliative care literature. Three sources of transcendence, also considered to be building blocks of spiritual meaning, were identified: the situational; the moral and biographical; and the religious. This model has implications for future theory, research and practice.

Assessment Tools

CLINICAL

FICA, a recently developed spiritual assessment tool, is a simple set of questions that assesses patients' spiritual needs (Puchalski, Romer, 2000). A full description of the FICA is included in Table 1. Although the clinical efficacy of this instrument has not been formally validated in ALS, it represents a simple and workable screening tool that provides physicians and other health care providers with a non-threatening approach to addressing spiritual issues with patients. It is important for physicians and other health care providers to note that discussion regarding spirituality may be interpreted as an indication of impending death. This may elicit a defensive response, and therefore, continued discussions on this topic will require particular sensitivity. For these reasons, initial assessment of spiritual needs is better performed early rather than late in the disease course, with repeat assessments following as needed.

A second example of a spirituality assessment tool is shown in Table 2: Taking a Spiritual Inventory. This spirituality inventory is a helpful tool that can also be completed by health care professionals for themselves, providing an important element to their own training in spiritual care.

RESEARCH

Several scales and questionnaires relating to spiritual and existential issues are available in the literature. Among these, the Spiritual Well-Being Scale has been used in a pilot study specifically involving patients with ALS (Dal Bello-Haas, 2000). Other published scales include: the SELT-M (van Wegberg et al., 1998), the Spiritual Involvement and Beliefs Scale [SIBS] (Hatch et al., 1998), and the Life Evaluation Questionnaire (Salmon et al., 1996). In addition, the SEIQoL-DW (Hickey et al., 1996) provides information on existential domains that are relevant to quality of life of individual patients (Neudert et al., 2001). Finally, qualitative research tools can provide invaluable information on this highly personal dimension of quality of life.

Defining the Gaps

- Goal 1: Recognition of spiritual care as an integral component of palliative care. Addressing spiritual issues during routine care of patients with ALS as part of standard practice for professionals involved in the care of these patients.
- Goal 2: Inclusion of spiritual care of patients and families as part of a multidisciplinary team approach.
- Goal 3: Improvement in the evidence base for spiritual care interventions.

RECOMMENDATIONS TO THE FIELD

Practice Recommendations

1. Recognize that spiritual care is an integral component of palliative care. Addressing spiritual issues during routine care of patients with ALS should be standard practice for professionals involved in the care of these patients.
 - a. Provide formal health care and physician education and initiate these early in the training process, starting at the undergraduate level and continuing through postgraduate and residency training programs.
 - b. Develop a specific curriculum on the spiritual care of patients with ALS for training purposes; strengthen curriculum with inclusion of cross-cultural and inter-religious issues.
 - c. Identify key support literature regarding the value of spirituality in palliative care to educate health care professionals (a provisional list can be found in Table 3). Use as a resource with patients and families.
 - d. Educate team members on cross-cultural religious differences in approach to dying and death to help providers interact and work more effectively with patients and families.
 - e. Establish close collaborations with ministers of various religions practiced within the surrounding community to make further resources and support available, when needed.
2. A multidisciplinary team approach that includes spiritual care of patients is recommended for the benefit of patients and families, with the following specifics:
 - a. Training in spiritual care is given to all health care professionals involved in end-of-life care of ALS patients.
 - b. Assessment of spiritual needs is performed repeatedly as the disease progresses (a patient might not be interested in this aspect of care earlier in the course of the disease, but this may change over time).
 - c. Spiritual needs of caregivers and family members, which may differ from those of patients, are best addressed separately.
 - d. Close collaboration with chaplains, spiritual counselors, and pastoral workers is a critical component of care: Every multidisciplinary ALS care team would benefit from having or being linked to spiritual counselors. However, patients may request spiritual care from any member of the team.
3. Recognize that the role of spiritual and religious attitudes is relevant to patient and family decisions surrounding medical management and end-of-life issues (i.e., Advance Directives, termination of life support, PAS, autopsy, disposal of the body).

Research Recommendations

1. Improvement in the evidence base for spiritual care interventions would include:
 - a. Research projects aimed at improving assessment of spiritual needs. Such studies would establish an epidemiological database, including a possible relationship between religious/spiritual attitudes and Quality of Life (QOL)/longevity in ALS (similar studies have been done in cancer patients).
 - b. Evaluation of efficacy of spiritual interventions on the quality of life and longevity of patients with ALS in studies that include assessing the role of chaplains or pastoral workers.

- c. Research on different spiritual needs of varying patient populations (e.g., ALS versus cancer) and ethnic/religious backgrounds to identify specific approaches that work with different patient populations.
- d. Additional validation studies on FICA as a spiritual needs assessment tool for patients with ALS. Better understanding of the role of scales (such as the SEIQoL-DW or the ALS-specific McGill) to assess individual patient spiritual needs and well-being.
- e. Assessing the effect of inquiry about spiritual needs on patients' quality of life (Hawthorne effect) and the relationship between patients and physicians.

Conclusions

Cicely Saunders once wrote: "It is not the worst thing for patients to find out that they have lived and are now going to die; the worst thing is to find out that they haven't lived and are now going to die." Obviously, no end-of-life care can solve life's dilemma completely. However, appropriate spiritual counseling helps patients come to terms with missed opportunities. If spiritual counseling does not help and patients end their lives in utter despair, it is important to remember that it is not the doctors' or health care providers' "fault." It is also not their merit if some patients, like John in the case vignette, accept their disease and impending demise with serenity (importantly, he started his "work" early in the disease). The professional's task is to remove obstacles that impede patients' ability to come to terms with their illness and death. Health care providers may facilitate this process through gentle counseling, when appropriate and possible. In this respect, end-of-life care is similar to the task that midwives perform at the beginning of life – that of removing obstacles and allowing a natural process to unfold (Vaughan, 1996). Professionals involved in end-of-life care may benefit from thinking of themselves as "midwives for dying."

REFERENCES

- Ackerman GM, Oliver D.** "Psychosocial support in an outpatient clinic." *Palliat Med.* 11: 167-168, 1997.
- Balducci L, Meyer R.** "Spirituality and medicine: a proposal." *Cancer Control* 8: 368-376, 2001.
- Borasio GD.** "Meditation and ALS." In *Amyotrophic Lateral Sclerosis: A Comprehensive Guide to Management*, Mitumoto H and Munsat T, eds, New York: Demos Medical Publishing, 2001.
- Dal Bello-Haas V, Andrews-Hinders D, Bocian J, Mascha E, Wheeler T, Mitumoto H.** "Spiritual well-being of the individual with amyotrophic lateral sclerosis." *Amyotroph Lateral Scler Other Motor Neuron Disord.* 1(5): 337-41, 2000.
- Doyle D.** "Have we looked beyond the physical and psychosocial?" *J Pain Symptom Manage*, 7(5): 302-11, 1992.
- Hatch RL, Burg MA, Naberhaus DS, Hellmich LK.** "The Spiritual Involvement and Beliefs Scale. Development and testing of a new instrument." *J Fam Pract*, 46(6): 476-86, 1998.
- Hickey AM, Bury G, O'Boyle CA, Bradley F, O'Kelly FD, Shannon W.** "A new short form individual quality of life measure (SEIQoL-DW): application in a cohort of individuals with HIV/AIDS." *BMJ*, 313: 29-33, 1996.
- Kellehear A.** "Spirituality and palliative care: a model of needs." *Palliat Med*, 14(2): 149-55, 2000.

Martin J, Turnbull J. “Lasting impact, and ongoing needs, in families months to years after death from ALS.” *Amyotr Lat Scler*, 1(Suppl. 3): S14-S15, 2000.

McMurray A. “Bereavement.” In *Palliative Care in Amyotrophic Lateral Sclerosis*. Oliver D, Borasio GD, Walsh D (eds). Oxford: Oxford University Press, 2000.

Murphy PL, Albert SM, Weber C, Del Bene ML, Rowland LP. “Impact of spirituality and religiousness on outcomes in patients with ALS.” *Neurology*, 55: 1581-1584, 2000.

Neudert C, Wasner M, Borasio GD. “Patients' assessment of quality of life instruments: a randomized study of SIP, SF-36 and SEIQoL-DW in patients with amyotrophic lateral sclerosis.” *J Neurol Sci*, 191: 103-109, 2001.

Post SG, Puchalski CM, Larson DB. “Physicians and patient spirituality: professional boundaries, competency, and ethics.” *Ann Intern Med*; 132(7): 578-583, 2000.

Puchalski C, Romer AL. “Taking a spiritual history allows clinicians to understand patients more fully.” *J Palliat Med*, 3: 129-137, 2000.

Salmon P, Manzi F, Valori RM. “Measuring the meaning of life for patients with incurable cancer: the life evaluation questionnaire (LEQ).” *Eur J Cancer*, 32A(5): 755-760, 1996.

Sykes N. “End-of-life care in ALS.” In *Palliative Care in Amyotrophic Lateral Sclerosis*, Oliver D, Borasio GD, Walsh D (eds.) Oxford: Oxford University Press, 2000.

van Wegberg B, Bacchi M, Heusser P, et al. “The cognitive-spiritual dimension--an important addition to the assessment of quality of life: validation of a questionnaire (SELT-M) in patients with advanced cancer.” *Ann Oncol*, 9(10): 1091-1096, 1998.

Vaughan C. “Teach me to hear mermaids singing.” *BMJ*, 313: 565, 1996.

World Health Organization. Cancer pain relief and palliative care. Report of a WHO Expert Committee. World Health Organ Tech Rep Ser. 804: 1-75, 1990.

Table 1: Spiritual Assessment Tool

An acronym, which can be used to remember what to ask in a spiritual history, is:

F: Faith or Beliefs**I: Importance and influence****C: Community****A: Address**

Some specific questions you can use to discuss these issues are:

- F:** What is your faith or belief?
 Do you consider yourself spiritual or religious?
 What things do you believe in that give meaning to your life?
- I:** Is it important in your life?
 What influence does it have on how you take care of yourself?
 How have your beliefs influenced your behavior during this illness?
 What role do your beliefs play in regaining your health?
- C:** Are you part of a spiritual or religious community?
 Is this of support to you and how?
 Is there a person or group of people you really love or who are really important to you?
- A:** How would you like me, your health care provider to address these issues in your health care?

General recommendations when taking a spiritual history:

1. Consider spirituality as a potentially important component of every patient's physical well-being and mental health.
2. Address spirituality at each complete physical exam and continue addressing it at follow-up visits if appropriate. In patient care, spirituality is an on-going issue.
3. Respect a patient's privacy regarding spiritual beliefs; don't impose your beliefs on others.
4. Make referrals to chaplains, spiritual directors or community resources as appropriate.
5. Be aware that your own spiritual beliefs will help you personally and will overflow in your encounters with those for whom you care to make the doctor-patient encounter a more humanistic one.

Table 2: Taking a Spiritual Inventory
by Kathleen Dowling Singh

The fact of death is the great mystery and the great truth that illuminates our lives. To face our own imminent death is to examine our lives with an urgency and honesty we may never have felt before.

A spiritual assessment is a helpful practice as we move close to dying. Such an assessment seems to arise naturally in the course of the profound psychological and spiritual transformations of dying. Since we all share the same human condition, many terminally ill people report asking themselves the same questions. These are many of the questions that those who have had a near-death experience report that they have been asked. They are questions that pierce through the frivolousness at the surface of life and confront us with the value and significance this precious gift of a human life offers.

It is not too late to take stock of our lives, even in the last weeks and days of terminal illness. And for those of us in the midst of life, in the apparent safety and security of our health, it is not too early. No matter how much time we have left to live, the answers to the following questions, voiced in the quiet honesty of our own hearts, provide direction to the rest of our living.

Who have I been all this time?

How have I used my gift of a human life?

What do I need to "clear up" or "let go of" in order to be more peaceful?

What gives my life meaning?

For what am I grateful?

What have I learned of truth and how truthfully have I learned to live?

What have I learned of love and how well have I learned to love?

What have I learned about tenderness, vulnerability, intimacy, and communion?

What have I learned about courage, strength, power, and faith?

What have I learned of the human condition and how great is my compassion?

How am I handling my suffering?

How can I best share what I've learned?

What helps me open my heart and empty my mind and experience the presence of Spirit?

What will give me strength as I die? What is my relationship with that which will give me strength as I die?

If I remembered that my breaths were numbered, what would be my relationship to this breath right now?

Who am I?

Kathleen Dowling Singh, Ph.D. is a former hospice worker, a transpersonal psychologist and the author of *The Grace in Dying: How We Are Transformed Spiritually As We Die* (HarperSanFrancisco, 1998). She lectures frequently on the spiritual dimensions of dying.

Copyright © 2000 Educational Broadcasting Corporation

Table 3: Recommended Books on Spirituality and End-of-Life Care

Albom, M. *Tuesdays with Morrie: An Old Man, a Young Man, and Life's Greatest Lesson.* New York, New York: Doubleday, 1997.

Cassidy, S. *Sharing the Darkness: The Spirituality of Caring.* Maryknoll, New York: Orbis Books, 1991.

De Mello, A. *One Minute Wisdom.* Garden City, NY: Doubleday, 1986

Nhbat Hanh, T. *The Miracle of Mindfulness: A Manual on Meditation (revised edition).* Boston: Beacon Press, 1987

Kabat-Zinn, J. *Full Catastrophe Living: Using the Wisdom of Your Body and Mind to Face Stress, Pain, and Illness.* New York: Deltacort Press, 1990.

Kearney, Michael. *Mortally Wounded: Stories of Soul Pain, Death, and Healing.* New York: Scribner, 1986.

Kushner, HS. *When Bad Things Happen to Good People.* Boston: GK Hall, 1981.

Longaker, C. *Facing Death and Finding Hope: A Guide to the Emotional and Spiritual Care of the Dying.* New York: Doubleday 1997.

Rinpoche, S. *The Tibetan Book of Living and Dying.* San Francisco: Harper San Francisco, 1992.

Table 4: Case Vignette (adapted from Borasio, 2001)

John had been a successful manager before contracting ALS at the age of 48. The first time I saw him (when he came to our ALS clinic to ask about enrollment in a clinical study), he conveyed a calmness and a peace of mind that was remarkable. At that time (almost four years into the disease), John was completely dependent on outside help and unable to move his arms or legs. His speech was almost unaffected. When I asked him to elaborate on his feelings about the disease, he said:

"You know, at the beginning I was shattered. My business, my career, my life plans – I had to give it all up. It was terrible. I went into big-time depression. Suicide seemed a reasonable option, since I did not want to become a burden to my wife and family. At that point, a friend pointed out that Buddhist teachings and meditation might help me. I was quite skeptical, but willing to try anything, and I gave it my best shot. Now I am totally dependent on others for help (the thing I dreaded most), but I would say, strange as it may seem to you, that my quality of life is actually better than before the disease started. Then, I didn't have time for anything, I was constantly rushing and stressed. Now, I have time for myself, and what is even more important, I can make use of this time and live in it."

One possible reaction to this account might be to doubt John's mental sanity. How could anybody with a clear mind say that he is happier with ALS than without? Yet John showed absolutely no signs of psychosis or delusions. He was perfectly at ease and one could sense that he was actually trying to make everybody else in the room feel comfortable, too. On one occasion, he made it clear that he was indeed not feeling "happier" in the common sense of the word. He was painfully aware of his physical disability, his breathing problems requiring non-invasive ventilation at night, and fear of losing speech. But he said, "that is exactly what it is all about: awareness. At least now I am aware of what is happening to me, while before I wasn't. Therefore, now I can enjoy even little pleasures much more than before."

John was one of the first ALS patients in his country (Germany) to receive non-invasive ventilation (NIV). While on NIV for almost five years, he helped many other ALS patients overcome their fears of NIV (and of PEG as well) by talking to them about his experience, either in person or by phone. John stopped NIV voluntarily when ventilation became necessary for most of the day, since he did not want to go onto tracheostomy. We arranged for him to spend his last days in a hospice to ensure appropriate palliative medication to relieve terminal dyspnea. To the surprise and awe of the entire hospice team, after saying goodbye to his wife and family, John simply slipped into sleep, went from there into coma and died peacefully within a few hours. Through his death, John spoke to the world around him: the collaboration of our ALS clinic with that hospice (which had been reluctant to accept ALS patients – especially if ventilated) has been cooperative ever since.

Caregivers

INTRODUCTION

Background and Objectives

Traditionally, patients are believed to be health care providers' primary responsibilities. Health care providers have not historically thought of the family caregiver as someone for whom they had responsibility. Some negative presumptions about families are derived from this Western medicine's approach of focusing on individual patients (e.g., codes of ethics, training and practice) (Levine, Suckerman, 1999). Family members act as advocates, provide or manage care, serve as trusted companions through illness and death, and make decisions on behalf of incompetent patients (Levine, Suckerman, 1999). The issues of family members who care for people with ALS have not been well studied.

ALS is a relentlessly progressive neuromuscular disease that leads to severe impairment and loss of mobility, speaking, feeding, swallowing and eventually breathing. In the current health care system in the United States, patients with debilitating chronic diseases are increasingly cared for at home, except when acute medical complications occur during the disease course. According to the North American ALS CARE Database, 56 percent of patients die at home and thus, family caregivers (most often spousal caregivers) are the principal care providers in this setting.

The burden of caring for patients with ALS is high. Krivickas and colleagues assessed home care status in a cross-sectional study of more than 70 patients with ALS. They found that primary caregivers spend a median of 11 hours per day caring for patients – despite home care assistance (Krivickas et al., 1997). Primary caregivers often feel physically (42%) or psychologically (48%) unwell. Formal home care for patients with ALS is often inadequate and too late to relieve the burden placed on family caregivers. Another study of eleven family caretakers of seven ventilator-dependent ALS patients revealed that caregivers were heavily burdened, and their outside activities were severely limited (Gelinas et al., 1998). Goldstein et al., (1998) found that caregivers demonstrated symptoms of anxiety and depression (the latter correlating with aspects of patients' functional impairments). Facing this reality, health care professionals can best meet caregivers' needs by educating, encouraging skills acquisition, establishing partnerships with families and communicating regularly.

To summarize, the ALS Peer Workgroup analyzed the medical literature and summarized current knowledge surrounding broad caregiver issues. Resources for caregivers are included, and a concept of “ideal” caregiving and caregiver status has been developed and is recommended as the standard of care. Recommendations are provided to fill gaps between current practice and ideal standards of care for patients with ALS and their caregivers.

Methods

Medline search on "caregiver" from 1999-2001 identified 740 published reports, 132 abstracts of which were subsequently reviewed. Of those reviewed, 34 reports that discussed the caregiver's role at the end of life are summarized below.

Medical literature about caregivers in ALS is limited, and in fact, only four papers have been published. The remaining reports address caregivers' roles as reported for other diseases such as cancer, dementia and aging and senescence.

WORKGROUP FINDINGS

Identification of Existing Resources

Establishing an Individual End-of-Life Care System

In the United States, the health care system for the chronically ill has become progressively dependent on family caregivers (Levine, Suckerman, 1999). Yet there is no formal system or process that helps caregivers provide informal and daily care to sick family members, so they must learn to cope with caregiving independently. By contrast, Japan has a new care and management network for patients with chronic neurodegenerative disorders including ALS, which was developed at the local community level (Kimura, 2001).

In the United States, principal medical facilities and services for end-of-life care are available extensively and include: acute and chronic medical care facilities, ALS centers or ALS clinics, formal home care services, and hospice services. However, services and facilities often operate independently with few or no links to other resources, making it necessary for patients and families to seek services one at a time. At many ALS centers or clinics, health care providers coordinate services but if patients/caregivers want services independent of those facilities, coordination of care becomes quite difficult. There are no structural models that patients/caregivers can easily rely upon at any level of medical care, so it can be difficult to obtain second opinions or ALS clinic consultations. Health care insurance coverage and HMO guidelines complicate medical decision making and treatment options that influence end-of-life care. The community-based system of coordinated services that is being tested in Japan presents a good model for other countries, including the United States.

Financial Issues

Interviews with 988 terminally ill patients (across all diseases) and 893 caregivers found that 35% of patients needed substantial care and reported economic burden, with 10% of household income spent on health care (Emanuel et al., 2000). Patients with substantial care needs were more likely to consider euthanasia or physician-assisted suicide. Caregivers were more likely to have symptoms of depression and to report that caring for these patients interfered with their lives. The study also reported that caregivers of patients whose health care providers listened to patient and caregiver needs experienced less burden, suggesting that an empathic approach to care ameliorates some burden associated with ALS care (Emanuel et al., 2000). Despite the fact that family caregivers are forced to provide care, there is no financial support.

The economic value of caregivers' efforts was studied in a home care environment for patients on ventilators (Sevick et al., 1997). Depending on the figure used to estimate cost of long-term care and on methods used to calculate caregiver value, home care was generally assessed as more expensive for at least 4.6% of ventilator-assisted individuals and for as many as 36.7% of other patients. Home care costs for ventilator-dependent patients is often more expensive than hospital care. The economic value on caregiver effort is assumed through indirect costs such as the economic impact of reduced work hours, lost income of caregivers who forego employment or educational opportunities and lost income of patients on ventilators who are no longer able to work.

Respite Service Availability

In the United States, respite has not been extensively utilized despite the fact that Medicare provides respite for patients/caregivers during the end of life. Braithwaite (1998) reported a negative relationship between intimacy and love and likely future use of respite services. An absence of intimacy and love was a significant predictor of past use of respite care (Braithwaite, 1998). The study suggests that where intimacy and love are low, the use of respite care goes up. Hanson et al., (1999) recommend that the concept of respite care be expanded to include caregiver information, skills training, education and emotional support.

McNally et al., (1999) pointed out that although respite care provides immediate relief from demands of caring, it often fails to facilitate the maintenance of socially supportive relationships, which reduce strain after respite ends. To address this problem, a more care-centered approach to provision and evaluation of respite service is recommended.

Social customs and societal health care system regulations influence the utility and provision of respite care. In Japan (Kimura, 2001), hospital admission is easily available and patients with ALS use hospital admission primarily to allow respite for family caregivers. Use of hospitalization for respite purposes occurred more frequently with ALS than with other neurodegenerative disorders. The Kimura study also suggests that in the U.S. (and other countries), education and health care policy changes influence acceptability and frequency of respite use.

Educational Programs

Identifying services within local communities and organizing programs specifically for ALS patients and caregivers is challenging (Ham, 1999). There are seminars and print and Internet materials about ALS and how to manage the symptoms of the disease. Such ALS-specific programs are facilitated by organizations such as The ALS Association and the Muscular Dystrophy Association, however more emphasis needs to be placed on developing and delivering educational programs and services to family caregivers. Family caregivers need information and skills in providing physical and psychological care, supervising other caregivers and in taking care of their own physical and psychological needs.

Cultural Competence

Patients in large cosmopolitan areas come from varied cultural and ethnic backgrounds that can drive their responses to life's challenges and especially family crises. Patient and family cultural customs are important and, at times, health care providers are not familiar with specific customs (Dedier et al., 1999). Such a knowledge base enhances competence in management of clinical care and social needs of people from diverse populations. The challenge lies in designing programs that educate medical teams about equitable environments for patients of differing backgrounds, customs and practices.

Caregiver Burden and Distress

The burden of care leads to stress. As stated earlier, more than 40% of primary caregivers feel physically and psychologically unwell, and manifest symptoms of anxiety and depression. In ALS, depression correlates with functional impairment (Goldstein et al., 1998; Emanuel, 2000). Gelinias and colleagues found that care of ventilator-dependent patients was particularly associated with burden and limitations on caretaker activities (Gelinias et al., 1998).

In contrast to these notions, Rabkin et al., (2000) interviewed 56 patients with ALS and 31 caregivers and found that neither patients nor caregivers displayed significant psychopathology (depressive disorders or scores on symptom scales). Among caregivers, "perceived caregiver burden" was significantly associated with finding positive meaning in caregiving. Although perhaps counterintuitive, it is important to note that the higher the degree of burden caregivers perceive, the higher their sense of positive meaning they find from the caregiving experience. Concordance between patient and caregiver distress was high, suggesting that attending to mental health needs of caregivers alleviates distress in patients (Rabkin et al., 2000). These results also suggest that clinical depression or significant depressive symptomatology in caregivers is not an inevitable or a common outcome of a life-threatening illness, even in the presence of major disability.

The ALS CARE Database highlights the current status of caregiver burden as shown in Table 1. Data indicate presence of a significant burden in many ALS caregivers during all stages of the disease. Specifically, a positive correlation exists between increased activities of care performed by caregivers

and caregiver burden. Community health and home health nurses can reduce caregiver burden by assessing the caregiving environment and implementing early interventions (Faison et al., 1999).

In another study of caregiver kinship groups (husbands, wives and daughters), mental and physical health status did not differ, even though a one-person variable (i.e., age) and appraisal variables (i.e., worry and burden) differed significantly (Sparks et al., 1998).

Awareness of Dying in Family Caregivers

Developing an awareness of dying is a gradual process characterized by uncertainty and anguish for family caregivers. Health care providers contribute to the uncertainty and agony caregivers may experience as they develop awareness of their dying loved one through limited, or absent, communication. Family caregivers attempt to cope by hoping, pretending, or preparing for death (Yates and Stetz, 1999). Nurses and social workers are a valuable resource of information and support to family caregivers. Supportive interventions are based on sensitivity to the experience of family caregivers, which include helping caregivers maintain hope, sustain social relationships and make preparations for the death. Further research is needed on how family caregivers use these strategies to manage awareness of the dying process (Yates and Stetz, 1999).

Psychological Mechanisms of Distress in Caregiving

Perceived “strain in caregivers” of patients with ALS correlates with a loss of intimacy in their relationships, as predicted by patients’ increased physical, cognitive, behavioral and communication disabilities. Changes in patients’ social performance also correlates with the extent to which caregivers feel the illness affects other areas of their lives (Goldstein et al., 1998).

In an interview-based study involving 54 caregivers, Grand and colleagues explained mechanisms of caregiver stress as evaluated by negotiation between patients and caregivers (Grand et al., 1999). That is, negotiations are based on caregiver-patient exchanges that exemplify the “theory of the gift” (giving, receiving, and giving back [Mauss 1950]). When the balance is perceived to be “too negative,” stress results. Caregivers experience stress when they feel trapped in a negative cycle of giving, receiving and giving back without a margin for negotiation (Grand, 1999). Stress is reported when caregivers continue to give without receiving.

Yates and colleagues (1999) also explored relationships between caregiving stressors and caregiver well-being, using a personal interview with 204 disabled elder/informal caregivers. The survey, which was based upon random sampling from 5,855 disabled elderly/informal caregivers, indicated that caregiving stressors led indirectly to caregiver depression (related to hours of care provided and caregiver perception of role overload). Regardless of the level of primary stressors (need for care), caregivers with high levels of mastery or emotional support were at lower risk for depression. These results are valuable to the design of proactive caregiver interventions (Yates et al., 1999).

Pre-existing Physical Problems and Mortality in Caregiver

Caregivers for cancer patients who have physical problems are at risk for psychological morbidity, which may have a delayed onset (Jepson et al., 1999). These results suggest that a more comprehensive assessment of caregivers’ physical problems and other major life stressors would enhance research efforts. Individuals with perceived lower subjective burden practice more health-promoting behaviors than those with higher subjective burden scores (Sisk, 2000).

Schulz and Beach (1999) suggest that among elderly spousal caregivers, those who experience mental or emotional strain have a higher mortality risk. However, no studies exist that report the same for caregivers of patients with ALS. Older married couples benefit from evaluation as a unit, relative to health status and caregiving demands in the home environment (Schultz, Beach, 1999). ALS

clinicians must address physical well being of spouses who assume major caregiver roles during end-of-life care.

Spousal Caregivers

The past relationship of middle-aged and elderly married couples influences psychological distress when one spouse assumes a caregiver role (Williamson et al., 1998). If the past relationship was communal (i.e., characterized by mutual concern for and responsiveness to one another's needs), restriction of activity is predicted by loss of intimacy and affection (rather than by the severity of symptoms in patients), which in turn, is a predictor of caregiver depression. Among caregivers in *less communal* relationships, activity restriction was predicted by severity of patient symptoms (rather than by loss of intimacy and loss of affection).

Allen and colleagues (1999) showed differences between wives and husbands in the selection of spouses as caregivers. Wives are only one-third as likely to select their husbands as caregivers. Spouses who consider their mates to be confidants are three times more likely to select their spouses as caregivers. Although gender role norms are key to caregiver selection, the intimacy inherent in the caregiving role renders an emotionally close marriage an important criterion in the selection of spouse as caregiver (Allen et al., 1999).

According to Rudd et al., (1999), spousal caregivers of patients with dementia mourn the loss of their living but demented spouses as if they had already died. In this study, wives providing more caregiving expressed significantly more anger than caregivers offering less care. The “place of care” and “gender of caregiver” were the most powerful predictors of the four psychological states of grief: anxiety, sadness, anger and guilt.

Male caregivers constitute nearly 30% to 36% of all caregivers. However, the vast majority of research centers on female caregivers. Attention to the experience of husbands as caregivers is important because husbands play a major role in caring for older women with chronic illnesses. It is predicted that husband caregivers will increase in numbers in the future (Kramer and Lambert, 1999).

Husbands who assume a caregiving role experience significant changes in household responsibilities, social integration, marital relationship and well-being. Further investigations are recommended to better understand what changes in household tasks mean to older husband caregivers and what role these changes play in his well being (Kramer and Lambert, 1999).

Family Caregivers

Dautzenberg et al. (1999) studied the distress of middle-aged daughters who provide assistance to elderly parents. Most distressed were women *not* performing any other major social role – such as wife, mother or businesswoman for example – suggesting that the lack of social roles rather than the multiplicity of roles is associated with distress. The caregiver role might even reduce distress when women have other roles in addition to caregiver.

Sparks et al., (1998) investigated predictors of mental and physical health by interviewing 151 caregivers (55 wives, 43 husbands and 53 daughters) of family members with dementia. They found that these three caregiver groups were not homogeneous. Study results suggest that among caregiver kinship groups, mental and physical health as outcomes of stress do not differ, even though one person variable (age) and appraisal variables (worry and burden) differ significantly (Sparks et al., 1998).

A complex psychological relationship develops when one spouse is in need of caregiving from the other. Well-being of the caregiving spouse can be influenced by gender of the caregiver as well as the

past and current marital relationship. In ALS, there is only one study investigating how relationships between partners influence caregiver burden and distress (Goldstein et al., 1998), as reviewed above.

Improving Coping in Caregivers

A study of caregivers of patients with ALS indicates that satisfaction with formal services and the number of social groups to which care providers belonged correlated with caregivers' self-predicted ability to cope in the future (Goldstein et al., 1998). Caregivers of patients whose physicians listened to patients' and caregivers' needs had fewer burdens. Through empathy, health care providers may ameliorate some burdens (Emanuel, 2000). Caregivers who used religious or spiritual beliefs to cope with caregiving had better relationships with care recipients, which in turn, was associated with lower levels of depression and role submersion (Chang et al., 1998).

Berg-Weger and Tebb (1998) developed the following scale for "activities of living" to assess the well-being of caregivers:

Activities of Well-Being

- getting the house work done
- attending social events
- asking for support from friends and family
- laughing

Basic Needs

- eating a well-balanced diet
- attending to medical and dental needs
- feeling appreciated by others
- expressing anger

Use of Formal Home Care

Satisfaction with formal home care services correlates with caregivers' self-predicted future ability to cope with ALS (Goldstein et al., 1998). While it seems plausible for the use of formal services to alleviate a caregiver's sense of overload, the use of services does not alleviate caregiver overload or depression in this study. Formal services supplement care rather than reduce the amount of informal care. While formal services might become necessary as a partner becomes more disabled, use of such services appears to have little or no effect on caregiver well-being (Yates et al., 1999). When working with ALS patients, the real challenges range from inadequate home care or services that come too late to finding ways to relieve the burden placed on family caregivers (Krivickas et al., 1997).

Another study supports consideration of care recipient and caregiver characteristics when predicting utilization of formal services in the home. Houde (1998) showed that utilization of formal services became less frequent as hours of informal care increased. Care recipient characteristics found to be important predictors for use of formal services included: residence in elder housing, recent hospitalization, gender, limitations in daily living activities, receipt of Medicaid services, age, quantity of informal care and number of household members. Caregiver characteristics that predicted use of formal services included: difficulty getting around outside, rearrangement of work hours and bowel and bladder care performed by female caregivers. The results support the development of interventions that supplement the home caregiving process.

Support of caregivers is especially important, given expected demographic and social trends that signify an increased need for care of elders and a decrease in available family caregivers. In order for caregiving relationships to be maintained over the long term, health care policy must support informal

caregivers. Research that guides the development of public policy and approved medical interventions that support caregivers is needed (Houde, 1998).

Formal home care health services pose many issues. For example, patients and caregivers do not often have choices in the selection of designated home care nurses. At times, home care agencies change personnel without telling patients and caregivers. In addition, home care nurses may not be knowledgeable about ALS or end-of-life issues (Krivickas et al., 1997). Extensive education about ALS, palliative and end-of-life care of home care and hospice care agency staff is recommended.

Disease Knowledge

While several publications in the medical literature on ALS exist, patients and caregivers appear to have limited knowledge and access to this information. This is likely due to the psychological burdens and physical demands that ALS places on the patient and their caregiver. Time and effort are focused on coping with the symptoms of the disease and activities of daily living. Patients and caregivers are eager to obtain support literature and other information regarding the disease process, coping with ALS, research directions, and crisis management as observed in support groups and at ALS clinics. Health care providers and ALS organizations need to be more proactive in reaching patients and caregivers with end-of-life information, education and supportive programs and services.

An Ideal Caregiving and Caregiver Status in ALS

Four fundamental issues influence the well-being of caregivers: empowerment, resentment, guilt and limited time/isolation. Table 2 addresses key factors that influence caregiver wellness and distress.

Based on these factors, ideal care and support for caregivers includes:

- Individualized end-of-life care programs available to caregivers of patients with ALS;
- Access to accurate and immediate knowledge about ALS and end-of-life care;
- Support systems for caregivers; and
- Psychosocial care and coping support.

Individualized End-of-Life Care Programs Available to Caregivers of Patients with ALS

When caregivers have early access to well-established end-of-life care systems, their burden and concerns are reduced. By participating in end-of-life care programs, caregivers will have the necessary tools to cope with the disease and the ensuing end-of-life issues.

Components of the Individualized End-of-Life Program:

- *Identification of the Neurologist and/or Primary Care Physician*
All patients with ALS have specific doctors such as ALS neurologists and/or primary care physicians who need up-to-date information and knowledge about end-of-life care in ALS.
- *Identified Care Sites*
Patients and caregivers select care sites, which most often are the patients' homes. ALSA Centers or ALS clinics are best suited to coordinate health care needs via outpatient care. Local Muscular Dystrophy Association (MDA) Clinics also function as ALS Centers. When such clinics are not available or easily accessible, private neurologists or family physicians provide necessary care or coordinate selection of care sites. Hospitals that admit patients on an emergency basis must be clearly identified.
- *Other Available Professional Services*
Physical therapy, occupational therapy, speech pathology, physical medicine, pulmonary medicine, and orthotics are necessary services for patients with ALS. Many services are available in local communities. ALS Center providers and neurologists caring for patients with ALS utilize referral networks to arrange for services.

- *Home Care System Services*
ALS Centers, ALS Clinics, or MDA Clinics have full knowledge of geographic distribution of home care services. Health care teams review pros and cons of home care services and help to arrange for them, as needed.
- *Hospice System Available*
ALS Centers and the health care teams have full knowledge of geographic distribution of hospice services and facilities. ALS Center medical staff (e.g., neurologists and nurses) usually knows members of community-based hospice service medical staff. Within hospice centers, medical recommendations and doctors orders respond to end-of-life conditions effectively and efficiently. Hospice centers also provide in-service educational programs for ALS caregivers.
- *Palliative Care Available*
Ensuring optimal palliative care requires several basic steps, including preparation of Advance Directives. Advance Directives may be modified over time, so periodic reviews are recommended. As death approaches, a terminal caring plan is initiated according to a predetermined series of steps. Hospice is contacted for recommendations about arranging the plan and initiating appropriate palliative care.

Access to Accurate and Immediate Knowledge on the Disease and End-of-Life Care

Effective Communication Established with Health Care Professionals

In an ideal clinical setting, the caregiver's concerns and questions are readily answered by the treating neurologist and the patient's nurses. This is one of the most difficult goals to achieve in busy clinical practices. Changes in the health care system are needed to allow patients access to health care providers. For example, patient and caregiver education and other psychosocial efforts (spirituality, counseling, etc.) are highly dependent on very busy health care providers. Additionally, health care providers need to accept the parameters of care for patients with ALS, which includes answering questions and discussing issues related to dying with ALS.

Literature And Other Educational Materials Available

Educational materials for patients with ALS and their families are available in written and audiovisual form through national organizations such as The ALS Association. Materials are also distributed to doctors' offices and even when not displayed, are sometimes available upon request. Caregivers are encouraged to read materials from sources mentioned above or libraries and to access Web site information. Books on spirituality and psychosocial issues relative to ALS are sometimes available at local churches, synagogues or other places of worship.

Educational Programs

As part of independent or support group meetings, educational programs organized by ALS Centers or voluntary health organizations are held periodically. Hands-on training about specific caregiver issues (bathing, PEG management, hydration, physical therapy and exercise) is provided by ALS Centers, ALS clinics or voluntary organizations.

Support Systems for Caregivers

Legal and Financial Support Available

In an ideal world, financial and legal issues would not be of concern to caregivers of ALS patients. The following support would be offered:

- Medical payers reimburse family caregivers' loss of wages due to caregiving at the patients' end of life;
- Home care is reimbursed;
- Voluntary disease organizations (such as The ALS Association and Muscular Dystrophy Association) provide support volunteers and extensive informational and medical literature to family caregivers; and
- Within physicians' offices and local ALS Association organizations, caregivers and patients receive a list of experts who help them with legal and financial concerns.

Available Respite

Respite provides patients and caregivers with a break, which can help assuage guilt and resentment as well as time for the caregiver to recover physical stamina.

Additional Support System is Implemented

Caregivers benefit from knowledge of and access to other people available to provide care for patients with ALS. These individuals include siblings, dependents, neighbors, close friends or community volunteers. Caregivers also benefit from lists of home aide agencies that provide additional support services.

Psychosocial Care and Coping Support

Caregiver Guidance and Counseling

The caregiver's "burden" is legitimate and often substantial. Psychosocial care proactively benefits caregivers in need of help. To assess needs, full or partial interviews can be repeated as patient conditions worsen. When issues increasing caregiver burden are identified, guidance and counseling can be initiated. For example, health-promoting exercise or programs such as yoga, religious studies, or therapy can be encouraged. It is important for caregivers to consult their primary care physicians about underlying medical diseases or general health concerns. Physicians and nurses and other members of the interdisciplinary team caring for patients with ALS function as strong advocates for caregivers.

Bereavement and Spiritual Care

Health care professionals who care for patients with ALS and caregivers benefit from spiritual training which, provides comfort as well as opportunity for discussion about spiritual care and bereavement.

Caregiver Support Groups (Independent of Patient Support Groups)

Caregiver support groups, which function independently from patient support groups, are helpful and routinely available. Caregiver support groups advocate for physical and mental health-promoting behavior.

Crisis Management

When caregivers are overwhelmed by physical and emotional crises, a 24-hour telephone hotline would be of service. Experienced nurses, physicians and others who specialize in ALS would provide crisis/urgent counseling as well as medical and psychosocial support.

RECOMMENDATIONS TO THE FIELD

The operational definition of end-of-life care for patients with ALS varies from patient to patient based upon the need and acceptance of such care. Although Workgroup recommendations focus on caregiver wellness during the end of life for patients with ALS, most recommendations are more effective when initiated in the early stages of the disease, as we have indicated.

Practice Recommendations

1. Establish Individual Care Systems with the following characteristics, services and resources: ALS Clinics should create master lists of allied health care professionals involved in the care of ALS patients. Such lists include physical therapists, occupational therapists, physiatrists, pulmonologists, respiratory therapists, gastroenterologists, psychiatrists, psychologists, orthotists, speech and language pathologists, nutritionists, social workers and counselors.

Similarly, a list of all outpatient rehabilitation centers, home care services, hospice services and respiratory care services should be identified. ALS Clinics should develop effective networks that focus on individual patient/caregiver needs. Importantly, patients' primary care physicians are situated at the center of these care networks. Good care reduces caregivers' burden.

Support Systems

1. ALS Centers identify legal and financial experts who provide advice to patients/caregivers.
2. ALS Centers provide regional lists of home-aid services.
3. Education and the use of respite, an underutilized resource, must be promoted.

Disease Knowledge

1. Educational resource information including guidebooks, pamphlets, videos, etc., is distributed to patients, caregivers and health care providers.
2. Palliative Care Handbooks about ALS palliative care are updated and disseminated to patients, families and providers.
3. Services that respond to questions 24 hours per day and all-day counseling are available via electronic or telephone communication systems at ALS Centers or regional ALS clinic networks.

2. Caregiver's Coping and Psychosocial Care

The following recommendations are based on the patient/caregiver unit concept: Potential obligations related to medical interventions (sometimes accompanied by intrusive questions) must be fully addressed with caregivers before decisions are made to move forward with such interventions:

1. Experienced physicians, nurse practitioners or psychologists interview caregivers when family members with ALS are evaluated.
2. Caregiver assessment instruments need to be developed. Specific items to be assessed are shown in Table 3. Assessment includes use of a caregiver burden scale.
3. Potential medical diseases are addressed with appropriate medical consultation.
4. Psychosocial issues that interfere with caregiving are addressed.
5. Designated psychologists/psychiatrists are available to provide guidance and counseling to caregivers, as part of psychosocial care.
6. Independent caregiver support group meetings are established that provide support while emphasizing educational initiatives, hands-on training and wellness-promoting behaviors.
7. Spiritual education training is given to ALS health care professionals, home care providers and hospice professionals.

8. Crisis management systems are developed at ALS Centers for caregivers. ALS caregiver telephone hotlines are available for 24 hours per day.
9. Readily available intensive psychosocial support, including home visits, are offered to caregivers during the end-of-life period.

Research Recommendations

1. Study ALS caregiver demographic characteristics to better identify caregiver issues, unmet needs, and suggested practice tools and methods to resolve these.
2. Investigate predictive value of high-risk caregivers who develop high degrees of caregiver burden and psychosocial distress.
3. Develop assessment instruments that identify high-risk caregivers for caregiver burden and distress.
4. Study whether proactive intensive psychosocial care reduces caregiver burden and enhances caregiver wellness.
5. Investigate the impact of care from caregivers on patient quality of life.
6. Investigate the concept of a patient/caregiver unit. Study the medical and economic advantage of treating patients and caregivers as a unit.
7. Investigate reasons and impediments for use of respite.
8. Study medical economics of informal family caregiving. Expand the evidence base to support insurance reimbursement for family caregiver costs.
9. Investigate the need for an ALS-specific caregiver burden scale. If the need is sufficient, develop the scale.
10. Develop a compilation of family caregiver experiences, biographies, and support literature for caregivers (a “companion ” book to *Journey With ALS: Personal Tales of Courage and Coping with Lou Gehrig’s Disease*).

Policy Recommendations

More efforts are needed to improve public and private financial support for costs of caring for patients with ALS during the end of life. Voluntary health organizations, such as The ALS Association and other patient advocate groups, can take a proactive role in educating and raising awareness of those in policy-making positions in government at all levels and private payers about the importance of optimal patient and caregiver care at the end of life. Specific changes needed include:

1. Medicare and other insurance carriers reimburse informal family caregiver costs.
2. Medicare and other insurance agencies reimburse home care nursing, home aides and respite services as patients’ disabilities increase and especially during the end of life.
3. Insurance companies reimburse lost income of caregivers at patients’ end of life, including unemployment or temporary leave of absence.
4. Insurance companies reimburse costs of medical and psychosocial treatment of caregivers during patients’ end of life.

REFERENCES

Allen SM, Goldscheider F, Ciambrone DA. "Gender roles, marital intimacy, and nomination of spouse as primary caregiver." *Gerontologist*, 39(2): 150-158, 1999.

Berg-Weger M, Tebb SS. "Caregiver well-being: A strengths-based case management approach." *J Case Manag*, 7(2): 67-73, 1998.

Braithwaite V. "Institutional respite care: breaking chores or breaking social bonds?" *Gerontologist*, 38(5): 610-617, 1998.

Chang BH, Noonan AE, Tennstedt SL. "The role of religion/spirituality in coping with caregiving for disabled elders." *Gerontologist*, 38(4): 463-470, 1998.

Dautzenberg MG, Diederiks JP, Philipsen H, Tan FE. "Multigenerational caregiving and well-being: distress of middle-aged daughters providing assistance to elderly parents." *Women Health*, 29(4): 57-74, 1999.

Dedier J, Penson R, Williams W, Lynch T Jr. "Race, ethnicity, and the patient-caregiver relationship." *Oncologist*, 4(4): 325-331, 1999.

Donnelly JM, Kornblith AB, Fleishman S, et al. "A pilot study of interpersonal psychotherapy by telephone with cancer patients and their partners." *Psychooncology*, 9(1): 44-56, 2000.

Emanuel EJ, Fairclough DL, Slutsman J, Emanuel LL. "Understanding economic and other burdens of terminal illness: the experience of patients and their caregivers." *Ann Int Med*, 132(6): 451-459, 2000.

Faison KJ, Faria SH, Frank D. "Caregivers of chronically ill elderly: perceived burden." *J Community Health Nurs*, 16(4): 243-253, 1999.

Friedman RH, Stollerman JE, Mahoney DM, Rozenblyum L. "The virtual visit: using telecommunications technology to take care of patients." *J Am Med Inform Assoc*, 4(6): 413-425, 1999.

Gelinas DF, O'Connor P, Miller RG. "Quality of life for ventilator-dependent ALS patients and their caregivers." *J Neurol Sci*, 160(suppl 1): S134-136, 1998.

Goldstein LH, Adamson M, Jeffrey L, et al. "The psychological impact of MND on patients and carers." *J Neurol Sci*, 160: S114-121, 1998.

Grand A, Grand-Filaire A, Bocquet H, Clement S. "Caregiver stress: a failed negotiation? A qualitative study in South West France." *Int J Aging Human Dev*, 49(3): 179-195, 1999.

Ham RJ. "Evolving standards in patient and caregiver support." *Alzheimer Dis Assoc Disord*, 13(suppl 2): S27-35, 1999.

Hanson EJ, Tetley J, Clarke A. "Respite care for frail older people and their family carers: concept analysis and user focus group findings of a pan-European nursing research project." *J Adv Nurs*, 30(6): 1396-1407, 1999.

Houde SC. "Predictors of elders' and family caregivers' use of formal home services." *Res Nurs Health*, 21(6): 533-543, 1998.

Jepson C, McCorkle R, Adler D, Nuamah I, Lusk E. "Effects of home care on caregivers' psychosocial status." *Image - the Journal of Nursing Scholarship*, 31(2): 115-120, 1999.

Kimura I. "Annual report of the research committee of medico-welfare network construction for supporting severely disabled patients with specific diseases." *Ministry of Health and Welfare in Japan Grant-in-Aid Annual Report*, 2001.

Kramer BJ, Lambert JD. "Caregiving as a life course transition among older husbands: a prospective study." *Gerontologist*, 39(6): 658-667, 1999.

Krivickas LS, Shockley L, Mitsumoto H. "Home care of patients with amyotrophic lateral sclerosis (ALS)." *J Neurol Sci*, 152: S82-89, 1997.

Levine C, Suckerman C. "The trouble with families: toward an ethic of accommodation." *Ann Int Med*, 130: 148-152, 1999.

McNally S, Ben-Shlomo Y, Newman S. "The effects of respite care on informal carers' well-being: a systematic review." *Disabil Rehabil*, 21(1): 1-14, 1999.

Noonan AE, Tennstedt SL. "Meaning in caregiving and its contribution to caregiver well-being." *Gerontologist*, 37(6): 785-794, 1997.

Rabkin JG, Wagner GJ, Del Bene M. "Resilience and distress among amyotrophic lateral sclerosis patients and caregivers," *Psychosom Med*, 62: 271-279, 2000.

Rudd MG, Viney LL, Preston CA. "The grief experienced by spousal caregivers of dementia patients: the role of place of care of patient and gender of caregiver." *Int J Aging Hum Dev*, 48(3): 217-240, 1999.

Schulz R, Beach SR. "Caregiving as a risk factor for mortality: the Caregiver Health Effects Study." *JAMA*, 282(23): 2215-2219, 1999.

Sevick MA, Bradham DD. "Economic value of caregiver effort in maintaining long-term ventilator-assisted individuals at home." *Heart Lung*, 26(2): 148-517, 1997.

Sisk RJ. "Caregiver burden and health promotion." *Int J Nurs Stud*, 37(1): 37-43, 2000.

Sparks MB, Farran CJ, Donner E, Keane-Hagerty E. "Wives, husbands, and daughters of dementia patients: predictors of caregivers' mental and physical health." *Sch Inq Nurs Pract*, 12(3): 221-234; discussion 235-238, 1998.

Ushikubo M, Kawamura S, Inaba Y, Shima C, Nakamura T. "[Characteristics of home care patients with intractable neurological diseases (Nanbyo) in Tokyo]." *Nippon Koshu Eisei Zasshi*, 45(7): 653-663, 1998.

Williamson GM, Shaffer DR, Schulz R. "Activity restriction and prior relationship history as contributors to mental health outcomes among middle-aged and older spousal caregivers." *Health Psychol*, 17(2): 152-162, 1998.

Yates ME, Tennstedt S, Chang BH. “Contributors to and mediators of psychological well-being for informal caregivers.” *J Gerontol B Psychol Sci Soc Sci*, 54(1): P12-22, 1999.

Yates P, Stetz KM. “Families' awareness of and response to dying.” *Oncol Nurs Forum*, 26(1): 113-120, 1999.

WORKGROUP PRODUCTS AND TOOLS

Table 1:
Responses to Question # 21 of ALS Patient CARE Database Caregiver Burden

Based on responses of 2,688 caregivers at the time of patient enrollment.

The numbers following each statement below indicate the total percentage of these caregivers who answered: moderately, a lot, or a great deal.

Added tension to your life	48%
Restricted the vacation activities and trips you take	44%
Decreased the time you have to yourself	43%
Increased your anxiety about things	39%
Decreased the time you have to spend in recreational activities	38%
Reduced the time you have to do your own work and daily chores	35%
Increased the nervousness and depression you have concerning your relationship with him/her	33%
Decreased the time you have for friends and other relatives	30%
Increased the stress in your relationship with him/her	26%
Decreased the money available to meet the rest of your expenses	25%
Restricted personal privacy	20%
Caused you to neglect other important relationships (e.g., children, parents)	15%
Increased the number of unreasonable requests made of you	15%
Increased attempts by him/her to manipulate you	13%
Affected your ability to attend to your personal medical needs	12%
Increased demands made by him/her that are over and above what he/she needs	10%
Added to your feelings that you are being taken advantage of	8%

These data are based on patients who were enrolled by neurologists at 97 clinical sites participating in the North American ALS Patient CARE Database through May 2001. Rhonda Montgomery developed the Caregiver Burden Questionnaire.

Table 2: Factors Influencing Caregivers' Wellness

<p>A. Establishing an Individual Care System Primary physician and ALS neurologist identified</p> <ul style="list-style-type: none"> • Care site identified • Other professional services available • Home care system available • Hospice system available • Palliative care available 	<p>B. Support System Legal and financial concerns Additional support system</p> <ul style="list-style-type: none"> • Other family members available • Friends available • Volunteers available <p>Home aid service Respite service available</p>
<p>C. Disease Knowledge Effective communication established with health care professionals</p> <ul style="list-style-type: none"> • Literature and other educational materials available • Educational sessions • Questions and concerns answered 	<p>D. Psychosocial Care and Coping Caregiver wellness assessment Caregiver guidance and counseling Caregiver support group available (independent of patient support group) Crisis management Spirituality Bereavement</p>

Table 3: Caregiver Assessment for Caregiver Wellness

Relationship and the degree of communication with patient's physician
 Relationship to patient (and to children, siblings, parents, friend, same-sex partner, others)
 Man or woman
 Working caregiver
 Patient's functional status (caregiver's assessment for patient's ALSFRS)
 Location of caregiving (home, alternate site)
 Caregiver's physical condition
 Preexisting medical and mental health
 For spousal caregiver

- Marital status (financial, decision making, intimacy, mutual trust, other factors)
- Current marital status
- Spousal depression

 Support system

- Availability of other family member(s)
- Availability of friends or others

 Participation in social activities
 Participation in religious activities
 Spiritual and existential beliefs
 Priority of caregiving issues
 Priority of caregiver's own life expectation
 Caregiver burden scale
 Caregiver quality of life measurement

Supportive Web Sites and Contact Information

Organization	Web and Telephone Contact
The ALS Association	www.alsassociation.org 818-880-9007
Muscular Dystrophy Association	www.mdaua.org 800-572-1717
Home Care Companions	www.homecarecompanion.com 415-824-3269
National Family Caregivers Association	www.nfcacares.org 800-896-3650
Family Caregiver Alliance	www.caregiver.org 415-434-3388

Table 4: Books and Book Chapters for Caregivers

Bolen JS. *Close to the Bone: Life-Threatening Illness and the Search for Meaning*. New York: Scribner, 1996.

Center L. "One day at a time: the experience of an ALS caregiver." In *Palliative Care in Amyotrophic Lateral Sclerosis*. Oliver D, Borasio GD, Walsh D, (eds.) Oxford: Oxford University Press, 2000.

Cobb AK, Reckling JB, Fernengel KJ. "The ALS Caregiver's Experience: An Impelled Journey." In *Amyotrophic Lateral Sclerosis. A Comprehensive Guide to Management*, Mitsumoto H, Norris FH Jr, (eds.) New York: Demos, 1994.

Mitsumoto H, Munsat TL. *Amyotrophic Lateral Sclerosis. A Guide For Patients and Families*. New York: Demos, 2001.

Murphy NM. "Mourning and ALS." In *Amyotrophic Lateral Sclerosis. A Comprehensive Guide to Management*, Mitsumoto H, Norris Jr FH, eds. New York: Demos, 1994.

Pollin I. *Taking Charge: Overcoming the Challenges of Long Term Illness*. Times Books, 1994.

Remen RN. *Kitchen Table Wisdom. Stories that Heal*. New York: Riverhead Books, 1994.

Sankar A. *Dying at Home: A family Guide to Caregiving*. Baltimore: Johns Hopkins University Press, 1991.

Strong M. *Mainstay: For the Well Spouse of the Chronically Ill*. North Hampton, MA: Bradford Books, 1997.

Vorst J. *Necessary Losses*. New York: Simon and Schuster, 1986.

QUALITY OF LIFE MEASUREMENT IN THE TERMINAL PHASE OF ALS

INTRODUCTION

The Utility of Assessing Quality of Life in Terminally Ill ALS Patients

Quality of life instruments, available for numerous diseases, are helpful in assessing treatment satisfaction and overall patient well-being. The practical clinical goal is to use quality of life information to identify areas of patient concern in the terminal phase of ALS. Quality of life instruments can be used proactively such that resources, treatment modifications and lifestyle changes are implemented during treatment to address specific concerns.

In measuring quality of life of patients with ALS, the focus is on the patient as the instrument attempts to:

- Identify needs from the patient perspective in the physical, psychosocial and spiritual domains.
- Identify the nature of patient concerns in order to prioritize time and resources of health care providers and family.
- Provide assurance and feedback to health care providers and families that patient needs are being met.
- Facilitate communication among patients, families and health care teams.
- Identify patients' level of satisfaction with completing "life's work."
"Life's work" – different for every patient – can be very difficult to assess clinically. According to Calman's hypothesis, a "good quality of life exists when the hopes of an individual are matched and fulfilled by experience" (Calman et al., 1984). A review of one's life is likely to be intense as death approaches, especially for an ALS patient who typically has no cognitive impairment.

Methods

The ALS Peer Workgroup evaluated how health care teams assess quality of life during the end of life of patients with ALS. A literature search identified quality of life instruments available for use relative to ALS and other diseases. Specific information on assessing patient quality of life was reviewed to identify what is still needed to achieve optimal quality of life for patients with ALS.

WORKGROUP FINDINGS

Identification of Existing Resources

Nineteen health-related instruments are detailed below. Few of these instruments have been specifically designed or validated for use with ALS patients. None focus on patients with ALS during the end of life.

Identifying What is Ideal

Measurement of quality of life in terminally ill patients with ALS is particularly challenging – and to some, unachievable – due to the paralytic and debilitating nature of the disease. Communication poses formidable challenges for investigators and patients, especially those with significant physical disabilities (which in extreme cases involve patients who have no ability to vocalize or move their hands). Yet, these are the very patients for whom a study about quality of life would be most important. The ultimate goal of health care teams is to provide treatment and care that enhances patient quality of life. To achieve this, quality of life must be routinely assessed.

Practically, assessing quality of life of terminally ill patients with ALS allows providers and caregivers to judge the efficacy of specific palliative care interventions. Using validated and structured instruments to assess quality of life assures evaluation of major domains of care.

In theory, quality of life instruments are “vital signs” taken before and after measurements of specific interventions. A potential negative aspect of using structured instruments is that care can become regimented (i.e., patient needs are technically “met” but without compassionate person-to-person contact).

Information collected from quality of life tools identifies future research arenas and improves care for ALS patients. McDonald and colleagues showed that psychological factors influence survival (McDonald et al., 1994). However, little work has been reported on evaluating quality and effectiveness of palliative care in the terminal phase of ALS.

Quality of life assessments of caregivers and health care providers identify issues that pose impediments to effective delivery of care to terminally ill ALS patients.

Assessment of patient quality of life requires more than one instrument as instruments assess different parameters. Because increasing physical limitations of terminally ill ALS patients interfere with their ability to communicate, data collection is best focused on attaining a limited number of clearly identified goals.

Although information from quality of life assessments is helpful to health care teams, administration of quality of life instruments must not interfere with patients’ psychosocial health (i.e., lengthy instruments or those that focus only on negative aspects of ALS, such as physical limitations). Instruments should be validated by test/retest examinations of patients and examiners.

RECOMMENDATIONS TO THE FIELD

Practice Recommendations

Providers would benefit from:

1. Using quality of life instruments with ALS patients and family members during the end of life.
2. Using quality of life instruments (McGill Quality of Life or SEIQoL-DW) with patients who have advanced ALS to assess the effect of care and management on quality of life when appropriate.
3. Monitoring and if necessary, measuring quality of life of caregivers and health care providers of ALS patients to detect their need for support.
4. Educating patients, caregivers and health care providers about the value of assessing quality of life during the terminal phase of ALS.

Research Recommendations

Researchers could benefit the field by:

1. Examining quality of life instruments in the terminal phase of ALS and developing new or refining existing quality of life instruments that best assess quality of life at the end of life, taking into account the following:
 - a. Patient’s physical comfort when completing an instrument,
 - b. Time needed to complete an instrument,
 - c. Ease of administration to patients with impaired communication abilities, and
 - d. Psychological distress associated with an instrument.

Policy Recommendations

1. Encourage NIH and other federal funding sources to designate research funding to create ALS-specific quality of life instruments for end-of-life care.

REFERENCES

- The ALS CNTF treatment study (ACTS) phase I-II Study Group.** “The Amyotrophic Lateral Sclerosis Functional Rating Scale. Assessment of activities of daily living in patients with amyotrophic lateral sclerosis.” *Arch Neurol*, 53(2): 141-147, 1996.
- Bergner M, Bobbitt RA, Carter WB, Gilson BS.** “The Sickness Impact Profile: development and final revision of a health status measure.” *Med Care*, 19(8): 787-805, 1981.
- Byock IR, Merriman MP.** “Measuring quality of life for patients with terminal illness: the Missoula-VITAS quality of life index.” *Palliat Med*, 12(4): 231-44, 1998.
- Calman KC.** “Quality of life in cancer patients—an hypothesis.” *J Medical Ethics*, 10: 124-127, 1984.
- Cedarbaum JM, Stambler N, Malta E, et al.** “The ALSFRS-R: a revised ALS functional rating scale that incorporates assessments of respiratory function.” *J Neurol Sci*, 169(1-2): 13-21, 1999.
- Cohen SR, Mount BM, Bruera E, Provost M, Rowe J, Tong K.** “Validity of the McGill Quality of Life Questionnaire in the palliative care setting: a multi-centre Canadian study demonstrating the importance of the existential domain.” *Palliat Med*, 11(1): 3-20, 1997.
- The EuroQol Group.** “EuroQol – a new facility for the measurement of health-related quality of life.” *Health Policy*, 16(3): 199-208, 1990.
- Frisch MB, Cornell J, Villanueva M, Retzlaff PJ.** “Clinical validation of the Quality of Life Inventory. A measure of life satisfaction for use in treatment planning and outcome assessment.” *Psychol Assess*, 4: 92-101, 1992.
- Ganzini L, Johnston WS, McFarland BH, Tolle SW, Lee MA.** “Attitudes of patients with amyotrophic lateral sclerosis and their care givers toward assisted suicide.” *N Engl J Med*, 339(14): 967-973, 1998.
- Gelinas DF, O’Connor P, Miller RG.** “Quality of life for ventilator-dependent ALS patients and their caregivers.” *J Neurol Sci*, 160: S134-136, 1998.
- Hickey AM, Bury G, O’Boyle CA, Bradley F, O’Kelly FD, Shannon W.** “A new short form individual quality of life measure (SEIQoL-DW): application in a cohort of individuals with HIV/AIDS.” *BMJ*, 313(7048): 29-33, 1996.
- Hunt SM, McEwen J.** “The development of a subjective health indicator.” *Sociol Health Illn*, 2(3): 231-246, 1980.
- Jenkinson C, Fitzpatrick R, Brennan C, Bromberg M, Swash M.** “Development and validation of a short measure of health status for individuals with amyotrophic lateral sclerosis/motor neuron disease: the ALSAQ-40.” *J Neurol*, 246(Suppl 3): 16-21, 1999.
- Jenkinson C, Fitzpatrick R.** “Reduced item set for the amyotrophic lateral sclerosis assessment questionnaire: development and validation of the ALSAQ-5.” *J Neurol Neurosurg Psychiatry*, 70(1): 70-73, 2001.
- Kaplan RM, Bush JW, Berry CC.** “Health status: types of validity and the index of well-being.”

Health Serv Res, 11(4): 478-507, 1976.

McDonald ER, Wiedenfeld SA, Hillel A, Carpenter CL, Walter RA. “Survival in amyotrophic lateral sclerosis. The role of psychological factors.” *Arch Neurol*, 51(1): 17-23, 1994.

McGuire D, Garrison L, Armon C, et al. “A brief quality-of-life measure for ALS clinical trials based on a subset of items from the sickness impact profile. The Syntex-Synergen ALS/CNTF Study Group.” *J Neurol Sci*, 152: S18-22, 1997.

O’Boyle CA, McGee H, Hickey A, O’Malley K, Joyce CR. “Individual quality of life in patients undergoing hip replacement.” *Lancet*, 339(8801): 1088-1091, 1992.

Shields RK, Ruhland JL, Ross MA, Saehler MM, Smith KB, Heffner ML. “Analysis of health-related quality of life and muscle impairment in individuals with amyotrophic lateral sclerosis using the medical outcome survey and the Tufts Quantitative Neuromuscular Exam.” *Arch Phys Med Rehabil*, 79(7): 855-862, 1998.

Simmons Z, Bermer BA, Robbins RA, Walsh SM, Fisher S. “Quality of life in ALS depends on factors other than strength and physical function.” *Neurology*, 55(3): 388-392, 2000.

Ware JE Jr, Sherbourne CD. “The MOS 36-item short-form health survey (SF-36). I. Conceptual framework and item selection.” *Med Care*, 30(6): 473-483, 1992.

Table 1: Health-related Quality-of-Life Instruments Available and Used with Patients with ALS

ASSESSMENT INSTRUMENT	KEY CITATION [CITATIONS IN ALS PATIENTS]	ADVANTAGES	DISADVANTAGES	COMMENTS
Instruments that Quantify Health Status				
Health Indices: A single, global score of health and well-being				
EuroQOL	Health Policy 16:199-208 (1990)	Visual analogue scale		Not ALS-Specific
Health Profiles: Measurement of multiple domains of health status				
Sickness Impact Profile (SIP)	Med. Care 19:787-805 (1981) N Engl J Med 339:967-73 (1998) J Neurol Sci 169:13-21 (1999)	Well-validated	Depressing effect on ALS patients Lengthy (136 items) No pain measures	Not ALS-Specific Used in the IGF-I, BDNF and Xaliproden trials
SIP/ALS-19	J Neurol Sci 152(suppl):S18-S22 (1997) Neurol 55:388-92 (2000)	Short subset of SIPdesigned for ALS		Used in the ALS CARE database
SF-36/SF-12	Med. Care 30:473-83 (1992) Arch Phys Med Rehab 79:855-62 (1998)	Well-validated	May lack sensitivity to change in clinical setting	Not ALS-Specific
ALSFRS ALSFRS-R	Arch Neurol 53:141-7 (1996) J Neurol Sci 169:13-21 (1999)	Well-validated in ALS patients Yields prognostic information	Some responses may reflect clinical practice and not stage of ALS (e.g., use of BiPAP)	Used in many ALS clinical trials
McGill QOL	Palliat Med 11:3-20 (1997) Neurol 55:388-92 (200)	Individualized measure of physical problems Well-validated	Only limitations on physical functioning are individualized	Not ALS-Specific
ALS Quality-of-Life index (ALSQLI)	J Neurol Sci 160(Suppl 1):S134-S136 (1998)			
ALSAQ-40	J Neurol 246(Suppl 3):16-21 (1999)	ALS-specific scale	Mostly physical functioning	
ALSAQ-5	J Neurol Neurosurg Psychiat 70:70-3 (2001)	ALS-specific scale Validated against ALSAQ-40 (shorter version)		
Individual QOL Measurement				
SEIQoL-DW	Br Med J 313:29-33 (1996) J.Neurol.Sci. 191: 103-109 (2001)	Individualized measure Uses colored pie chart for weightings May facilitate patient-physician communication	Favored by ALS patients over SIP and SF-36	Not ALS-Specific