

CHAPTER 45

Multidisciplinary Integrated Psychosocial and Palliative Care

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I ask knowledge what it can tell me of life. Knowledge replies that what it can tell me is little, yet immense. Whence this universe came, or whither it is bound, or how it happens to be at all, knowledge cannot tell me. Only this: that the will-to-live is everywhere present, even as in me. I do not need science to tell me this; but it cannot tell me anything more essential. Profound and marvelous as chemistry is, for example, it is like all science in the fact that it can lead me only to the mystery of life, which is essentially in me, however near or far away it may be observed.

Albert Schweitzer (1)

Life and death are intensely personal and social experiences. The way an individual faces life-threatening disease and obtains support from others reflects the shared beliefs and values of the particular culture, society, and treating physicians (2). Over the past several years, it has become clear that the optimal care of patients with progressive motor disorders includes attention to both psychosocial and medical needs. Regardless of the etiologic diagnosis, the goals of psychosocial and palliative care are to enhance, or at least maintain, quality of life by keeping the disruptive features of the disease to a minimum. Such care reinforces independence and self-esteem and enhances useful coping strategies and treatment options while actively identifying and modifying maladaptive behaviors. In progressive and ultimately fatal disorders without known cure or effective therapy, such as amyotrophic lateral sclerosis (ALS), a psychosocial and

palliative model of care can help the patient live and die in a desired manner.

This chapter focuses on the integrated palliative and psychosocial care of patients with ALS; however, the concepts are applicable to other progressive neuromuscular disorders. The essential aspects of the classification, diagnosis, laboratory evaluation, and pharmacotherapy of inherited and acquired ALS are reviewed in preceding chapters in this volume (1–5,31–32,38–40,41–43).

GENERAL CONSIDERATIONS

The psychological and social needs of a patient with progressive motor disorders should be ascertained as soon as possible after the diagnosis is made and discussed with the patient and family, because the emotional reaction to the perceived loss of bodily integrity commences right away. Denial, fear, and anxiety are early reactions, followed later by depression and anger, with an intensity and duration that depend on the severity and rate of progression of the illness, individual character, family variables, and envisioned changes in lifestyle. For several reasons there may be lingering doubts or disbelief of the diagnosis even after a lengthy evaluation. First, the diagnosis of ALS is a clinical one, substantiated by laboratory studies, in particular, electromyography and muscle biopsy, with an accuracy of about 95% in experienced hands. Second, the time to correct diagnosis may lag by weeks or months, with some patients given alternative diagnoses or inaccurate information about the disease. In one study, a third each of 33 patients and their families complained that the diagnosis of ALS was withheld too long or crudely relayed (3). At the time of diagnosis, the leading concern in one half of patients was the prospect of becoming disabled and dependent on others, followed by the uncertainty in the length, mode of progression, and nature of the terminal phases of the illness. Third, neuro-

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ogists can have different styles of communication, with some transmitting their sense of helplessness about the disease and concentrating on the inevitable clinical decline and time to death and others proactively engaging the patient in useful dialogue. Experienced ALS clinicians recommend as many visits as necessary to address personal concerns, treatment expectations, and other questions, because information overload frequently occurs after the initial discussion of the diagnosis.

ALS presents particular challenges because the cause of the disease is not well understood; for the most part, the course of ALS is relentlessly progressive with few stable plateau periods, and approximately 50% of patients die within 3 to 5 years of diagnosis. Its predictable pattern of functional decline permits evaluation of the effects of psychological, social measures, and therapeutic interventions on prognosis and outcome. Measures of psychological well-being were related to outcome in a cohort of 144 patients with ALS in which depression, hopelessness, and perceived stress are more likely to occur in patients with advanced physical disability who are closer to death (4).

An integrated medical and psychosocial approach usually reveals potential medical problems earlier and provides a more effective utilization of resources, particularly in the present managed care environment. The multidisciplinary team includes a neurologist; advanced care or clinical research nurse; psychiatrist; physical and occupational therapists; speech, swallowing, and otolaryngologic specialists; psychologist or psychiatrist; clergy; social worker; respiratory and vocational therapists; and nutritionist. Advanced-trained nurses can provide valuable leadership by virtue of their pivotal position as contact person for patients, their families, and other team specialists.

CHALLENGE OF PSYCHOSOCIAL CARE

It is in reverence for life that knowledge passes over into experience . . . My life bears its meaning in itself. And this meaning is to be found in living out the highest and most worthy idea which my will-to-live can furnish. . . .

Albert Schweitzer (5)

In this section we consider the psychosocial aspects of the relationships between the patient, family, and health-care team and five specific suggestions to optimize them.

1. *Ensure that the relationship established with the patient and their family is professional, with clearly established boundaries and expectations.*

Healthcare professionals should strive to create as best as possible a warm, welcoming, hospitable environment. To offer hospitality, in the true sense of the word, is to offer an environment that restores the patient's spirit and their physical nature (6). Hospitality and hospital are of the same Latin root, *hospitis*, which means host, guest, or friend (7). Practically speaking, optimal relationships are based on mutual respect, resembling how one would like to be treated if diagnosed with the same condition. Some clinicians believe that an atmosphere of informality may best

help the patient feel at ease. One way to create such informality is to refer to both patients and team members by their first names. This degree of informality is appropriate in patient care if everybody on the team, including the physician, are called by first names. Realistically, it is quite rare for a patient to refer to the physician by his or her first name even when invited to do so. To refer to the patient by their first name conveys a paternalistic posture and lack of gender sensitivity even if it is not intended to do so (8). This style of communication infantilizes the patient and can be perceived as rude and threatening to the patient's dignity. In contrast to those clinicians wishing to promote informality in the clinician-patient relationship, others believe that excessive informality erodes boundaries and leads to confusion (9). With increasing awareness of the need to empower patients, effort has been toward fostering balanced gender-sensitive egalitarian models of team care (10) that convey mutual dignity and respect.

2. *Identify coping strategies and take into account interpersonal communication styles of the patient.*

There are perhaps as many styles of coping as there are individual personalities. It may be useful to elicit information about the ways in which the patient faced challenges that occurred in the past to plan treatment strategies for the present and certainly for the future. There may be underlying emotional, spiritual, intellectual, and cultural factors that interfere with adequate communication, understanding of the disease process, and the successful integration of coping strategies. The patient's reaction to a diagnosis that is terminal is akin to the reaction observed in individuals experiencing trauma. Denial may be observed, or patients may insist that they are coping well, when in fact they may not be. Although this defense mechanism is inordinately useful to the patient (it enables an individual to maintain a sense of competence in the face of actual incompetence [11]), the healthcare team would be wise to recognize that this coping strategy is frequently used. It also has been advised that the healthcare team should not force the acceptance of a frank or poor prognosis or confront denial directly (11). Instead, one should recognize that denial is a common reaction, understand its positive and negative effects, emphasize the patient and family's strengths, and encourage alignment with the medical team. An awareness and understanding of the patient's patterns of coping; current functioning; psychological status, including cognitive change and impairment; and spiritual and cultural orientation can help to facilitate that alignment. Reframing through the utilization of the positive aspects of denial and emphasizing of concerns of safety will orient the patient and family toward reality and help them view the diagnosis of ALS as a challenge and not a crisis situation (12).

3. *Identify coping mechanisms, strategies, and interpersonal communication styles of the patient's family.*

Relationships with the patient's primary caregivers should be actively pursued. This effort may lead to insight

into the system of family interdependence (13) and identification of the members that will be the most influential in effecting change and supporting the use of certain assistive devices. Cultural factors may be important determinants of a successful relationship, as for example when the eldest son of an affected father holds the most influence in a family because of the cultural norms. Language barriers may hamper interpersonal communication, for example, when English is not the primary language spoken at home. In our experience, family members may not openly express helplessness, frustration, anger, fear, and concern about the patient in front of the patient or to all members of the interdisciplinary team. The technique of reframing (14) (again emphasizing the positive aspects of denial and concerns of safety always orienting toward reality) may be useful in ALS family support groups. Group facilitators are mindful that the emphasis is not what is happening to the family but how the family is relating to what is happening (12).

4. *Ensure that an analysis of interdisciplinary treatment goals has been conducted to ascertain how psychosocial support might complement the medical plan of care.*

The provision of psychosocial support can be facilitated or diminished by the behaviors and treatment goals of the interdisciplinary team. It is important for team members to share common goals and establish close communicative ties. Any tendency of the family to manipulate or split the team should be made apparent to all concerned and put into perspective.

5. *Ensure that the coping strategy and interpersonal communication of health professionals are continually self-assessed and open to outside counsel.*

Healthcare professionals should be aware of their own behaviors. Just as the patient and family are asked to meet the diagnosis of ALS as a continual challenge, so should the healthcare team. One manner of self-assessment is to ask a set of related questions. Have our behaviors promoted problem-solving and positive adaptation or dependency in patients with ALS and other progressive motor illness? Whose psychological needs are being met—the patient's or ours? Are we being realistic in our behavioral self-assessment? Have we been creative in our approach to alternative treatments? Do we need supportive guidance from a psychiatric nurse clinician, psychologist, or psychiatrist in responding to a patient and their family, particularly when psychological dysfunction is evident? Are we aware of our own psychological functioning, dysfunctioning and maladaptive patterns? Have we promoted collegiality and collaboration among all our team members?

INTEGRATED PALLIATIVE CARE

To act as one-caring, then, is to act with special regard for the particular person in a concrete situation. We act not to achieve for ourselves a commendation but to protect or

enhance the welfare of the cared-for. Because we are inclined toward the cared-for, we want to act in a way that will please him. But we wish to please him for his sake and not for the promise of his grateful response to our generosity.
Nel Noddings (15)

Palliative therapy should be offered at all stages of ALS to promote confidence, encourage independence, reduce the burden of physical handicaps, and sustain relationships with family, friends, and colleagues.

The gradual loss of ambulation is a nearly universal feature of progressive motor disease that leads to consultation with physiatry and physical and occupational therapy. These professionals, as well as other members of the interdisciplinary team, will guide the patient in their increasing reliance on assistive devices to maintain independence. In ALS, leg weakness and spasticity are the causes of gait difficulty. Mild to moderately affected patients derive benefit from a cane or walker. Ankle-foot orthoses and other bracing maneuvers may improve balance, preserve energy, promote safety, and avert fatigue that might otherwise preclude the participation of some patients in social activities. When frequent falls occur, a wheelchair may be necessary. Contemporary lightweight chairs are easy to operate and are portable. Self-propelled larger units offer the potential for continued independence even in advanced disease, but they are more expensive and heavier than manually propelled ones.

Communication impairments resulting from dysarthria, anarthria, and dysphonia are challenges for patients with ALS and their healthcare providers. Speech difficulty leads to a sense of isolation, enhances preexisting dysfunctional communication styles, and may limit the ability to communicate basic needs, such as suctioning or repositioning. Consultation with an experienced speech pathologist is essential early in the diagnosis before problems in communication become overtly apparent. Under normal circumstances, speech is possible through the combined action of the lips, tongue, palate, and larynx. Bulbar weakness and spasticity lead to a mixed pattern of dysarthria. Hyperadduction of the vocal cords leads to elevated laryngeal resistance in exhalation and a raspy voice. Flaccid weakness of one or both vocal cords causes a breathy hypernasal voice due to escape of air into the nasal pharynx; in addition, there may be slow strained vocalization with poor pronunciation of consonants.

Management of bulbar symptoms in patients diagnosed with ALS begins with speech, language, and otolaryngologic assessments. It may be helpful to educate patients with bulbar ALS in oromotor exercises for mild impairments and to encourage early intervention for evaluation of augmentative aids. Verbal communication can be prolonged in tracheostomized patients as long as speech is intelligible, in spite of respiratory dependency, by cuffless tubes or intermittent positive pressure ventilation. Computer-assisted aids and electronic communication systems are useful for maintaining communication to

family, friends, and the healthcare team and in allowing the patient to actively participate in the decision-making process even late in the illness.

Optimal management of dysphagia and nutritional requirements is important in the psychosocial and physical well-being of patients with ALS and other progressive neuromuscular disorders. Dysphagia precedes ventilatory difficulty in three fourths of patients with ALS and is present in virtually all others late in the illness (16). Normal swallowing requires the coordinated function of structures of the oral cavity, pharynx, larynx, and esophagus. Chewed food moves posteriorly in the oral cavity through constrictor muscles and other pharyngeal spaces to the esophagus where peristaltic movement carries it past the gastric sphincter and into the stomach. Alterations in smell, taste, and fear of aspiration and respiratory weakness can contribute to the occurrence of weight loss even before overt dysphagia is present. Weakness of lip, cheek, lingual, neck muscles, hyperactive pharyngeal gag and cough reflexes, dyspnea, spinal hyperlordosis, and balance difficulty due to axial weakness can all impair the early phase of swallowing; esophageal weakness and dysmotility compromise lower esophageal function. The clinical evaluation includes a review of clinical symptoms and signs of dysphagia and inspection of the nasopharynx, larynx, and esophageal paths by fiberoptic and video fluoroscopic studies. Liquids are generally more difficult to swallow than solids. Pooling of liquids and secretions may be found along the vallecula and pyriform sinuses or in the laryngeal vestibule, increasing the likelihood for aspiration.

Even the treatment of mild dysphagia includes dietary counseling, oromotor exercises, and positioning devices for the head and trunk. With bulbar involvement, aspiration can be improved by the management of secretions, abnormal breathing patterns, assisted coughing or chest physical therapy, oropharyngeal suctioning, and percutaneous endoscopic gastrostomy (PEG) placement. Nasogastric tubes are typically not used due to local irritation and an offensive appearance. The PEG is the most used procedure for dysphagia management due to its appeal. The ease of implementation, low risk for individuals with a forced vital capacity of 50% of predicted or greater (16), and minimal anesthesia are factors that contribute to PEG utilization. PEG may prolong survival in ALS (17), particularly before weight loss becomes too great, but its impact on the quality of life is still unknown (18,19). Its routine use runs counter to the view that death due to starvation or malnutrition in ALS is a painless, final, merciful act and is one of many options to be considered for prolongation of life (20–23).

Respiratory symptoms inevitably occur in all patients with ALS, often in association with an ineffective cough, difficulty in clearing secretions, and in the aspiration of fluids or food. An astute clinician will recognize the signs of impending respiratory insufficiency, including agita-

tion, lethargy, orthopnea, poor cough, increased use of accessory muscles, diminution of the volume of speech, and disturbed sleep. Pulmonary consultation can provide helpful information regarding respiratory muscle function to the ALS clinician. Pulmonary muscle function tests are the most reliable and sensitive measures of respiratory strength capacity and life expectancy and are optimally performed every 3 to 6 months. As vital capacity approaches 50% of predicted capacity, noninvasive intermittent positive-pressure aids should be introduced. Some patients decided early in the course of their illness to pursue tracheostomy and are comfortable considering life assisted with ventilation, knowing that they may be unable to move and, at some point, unable to communicate.

The decision to proceed with endotracheal intubation or indwelling tracheostomy should be discussed as openly and supportively as possible with the patient and family members in advance of impending emergencies to remain in compliance with patient preferences for medical decision making. Similarly, documentation with respect to advance directives, healthcare proxy, or, in the case of some states, durable power of attorney, should be completed and placed in the patient's chart and copies distributed to appropriate team members. The optimal situation for home ventilatory support includes adequate financial resources and psychosocial and medical support systems, including proximity to a clinic or hospital for the treatment of complications or emergencies (24).

Drooling is a vexing problem in ALS that is associated with oropharyngeal and lower lip muscle weakness, faulty containment and overflow of secretions, but usually not hypersalivation. Early in the disease, patients report a small pool of saliva on the pillow case upon awakening or excessive secretions from the mouth requiring frequent dabbing of facial tissue. Beyond the embarrassment and social isolation it causes, drooling is associated with a heightened risk of aspiration. Medications such as tricyclic antidepressants and atropine-like drugs, with potent anticholinergic effects, reduce salivation by blocking parasympathetic outflow; however, they also have the potential for urinary retention, confusion, and hallucinations.

Care for the mental health of patients with ALS is a steady challenge (25,26). It includes recognition and treatment of clinically significant depression, anxiety, lability of mood, and dementia. The seemingly healthy adjustment to serious illness often includes the denial of depression and anxiety that may be helped through referrals to psychiatry, psychology, social work, or pastoral care and with pharmacotherapy. Observations suggest that treatment for depression has an impact on denial and facilitates adaptive coping mechanisms.

Anxiety disorders and obsessive thought disorders associated with ALS have been less well studied, but experience suggests that they might also be amenable to counseling and antianxiety drugs. Lability of mood, leading to

