Applying Holism in the Home Care Environment for Clients With Advanced ALS: A Toolkit for Practice

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Amyotrophic lateral sclerosis (ALS), also known as Lou Gehrig's disease, is a progressive, debilitating disease that targets motor neurons. As motor neurons die, voluntary skeletal muscle becomes weak, atrophied, and eventually paralyzed, to the point of supraquadriplegia with the need of ventilatory support for those who choose it. The purpose of this article is to demonstrate application of the nursing process as a guide to the detailed care of patients with advanced ALS. Applying a model of holism allows the advanced practice nurse (APN) to explore beyond the complex physical needs and technical management by facilitating psychosocial-spiritual growth and the best possible quality of life.

JA was a healthy 36-year-old male and president of his own company when in February 1993, he began to have trouble moving his feet and legs. Initially, he thought it was a bad pair of shoes, but eventually he went to a chiropractor. He was concerned when the chiropractor told him he suspected a larger problem and referred him to a neurologist. Though there is no definitive test for ALS, he had an electromyogram, spinal tap, and muscle biopsy that indicated ALS was "probable." The diagnosis was, of course, devastating to him, his family, and friends; of note, JA stated he felt he had to be strong to support everyone else, and he held onto the belief that he could "beat it."

JA was using a cane by June, and in August was in a wheelchair. He said the fasciculations in his thighs looked like water boiling beneath the skin. By September, he had seen 3 ALS specialists. By November, he was afflicted by complete quadriplegia and was admitted to the hospital in respiratory failure. Noninvasive ventilation was attempted, but failed. A tracheostomy was performed and invasive mechanical ventilation was initiated.

At this time, his face was animated and he was able to speak using a Passy-Muir valve attached to his tracheostomy tube. He was in the hospital for 2 months secondary to left lower lobe atelectasis and resulting pneumonia, while Medicaid and home care arrangements were established. He was sent home with 16 hours of nursing care a day, and a spouse and friend as lay caregivers. His extended family was also a tremendous source of support, but lived out of state. He was home a month when his left leg became swollen, red, and painful. He was readmitted to the hospital and diagnosed with a deep vein thrombosis (DVT) above the knee. During that time in his home before readmission to the hospital, he had 28 different nurses.

Following a 6-week hospitalization and treatment for DVT, he was discharged home in April with a different home care agency and primary nurse. His prescription medications included warfarin for blood clot prophylaxis, sertraline for depression, baclofen for fasciculations, and triamcinolone cream at the tracheal stoma to suppress overgrowth of granulation tissue. He was a partner in his care this time, and one of the first decisions he made was to discontinue his canned tube feedings. He had loved to cook, and not having "real food" was unpalatable to him. A registered dietitian provided aid with nutrient calculations, and the team began experimenting with different menus of whole food to puree, strain, and use in place of canned feedings. By May, he was able to travel by airplane to visit his childhood home.

Over that summer, JA's voice deteriorated rapidly, and by August he was virtually unintelligible. "It's over," he said, as if his mouth were full of marbles. "I can't talk." A hand-held letter board helped supplement speech, with upward eye movement indicating "yes." During this time, Medicaid requested a psychiatric consultation to determine his competence in making resuscitation decisions -- he had never agreed to sign advanced directives, even though his attorney prepared the document. He was determined competent, and chose resuscitation should his heart stop, but was still unwilling to detail any choices in a legal document.

JA was very depressed from the overwhelming losses associated with the disease. His comments, however, spelled out letter-by-letter on the letter board remained colorful and prolific. An assistive technology group was consulted, and augmentative communication software and hardware were purchased for his laptop computer. A switch activated by head movement allowed JA to be able to use a letter board scanning system. His first message was: "Hope is still alive."

Once he was no longer able to turn his head enough to activate the switch, and his eye movements were not reliable, the team tried a biofeedback electromyography (EMG) device that responded to muscle movement so subtle it was only observable by the needle of the EMG monitor. He was able to use this device with a letter board, and eventually just for "yes-no" responses until the fall of the following year. The team had already begun searching for a mind-driven method to communicate that did not require movement. In the meantime, nurses worked to enhance their skills of observation and intuition in lieu of any objective communication. By this time, JA was still determined to live, but chose not to be resuscitated (DNR) should his heart stop.

ALS has a uniform distribution worldwide, despite differences in climate, geography, race, cultures, and diet. [1] Some clusters of increased incidence have been identified, including the Western Pacific islands. The average age of onset is 55 years, [2] though even children have developed the disease. [3] The prevalence of the disease is between 1 to 7 persons per 100,000 population. [1] In the United States, there are approximately 30,000 people with ALS, with about 5600 new cases diagnosed each year. [2]

ALS manifests differently in each individual. Weakness may begin in the extremities or with speech (bulbar involvement); and paralysis may come very fast --over months, or very slow -- over years. The disease itself does not affect sight, hearing, or the senses of touch, smell, and taste. Cognition is generally intact. The term, "locked-in syndrome" is used to describe people who are conscious and have cognitive function, but are unable to speak or move. [4]

While statistics vary, the ALS Association estimates that half of all people diagnosed with ALS will live at least 3 or more years, 20% will live 5 years or more, and up to 10% will survive more than 10 years. [2] Patients may have the disease for a period of time before a diagnosis can be made.

Research on ALS has increased dramatically in the last 30 years, [1] with a number of etiologic factors explored ( ). Ninety percent to 95% of cases of ALS are sporadic (SALS), occurring without family history; the remaining 5% to 10% of cases are familial (FALS). While a minority of cases, it is hoped that recent discoveries in familial ALS will also shed light on the sporadic form of ALS.

Table 1. Possible Etiologic Factors in ALS [1]

Altered Immunity
• Antibodies to calcium channels • Antineuronal antibodies • Antisprouting antibodies
Disordered Neuronal Metabolism or Function
• Abnormal neurotransmitter function • Abnormal thyrotropin releasing hormone

- Altered axonal transport
- Defects in neuronal membrane structure or function: heat shock proteins/stress proteins
- Defects in the urea cycle
- Disordered calcium, phosphate, and bone metabolism
- Hyperparathyroidism
- Loss of cholinergic receptors
- Loss/dysfunction of androgen receptors

Excitotoxicity

- Alterations in serum and spinal fluid glutamate
- EAAT2 (glutamate transporter) protein deficiency
- Zinc-induced excitotoxicity
- Seed of *Cycas circinalis* (native to Guam)

Genetic Disorders and Abnormalities

- ALS1: Defect in superoxide dismutase gene (SOD1)
- ALS2: Chromosome 2q33
- DNA and RNA abnormalities
- Genetic markers/HLA antigen
- Hexosaminidase deficiency

Infectious or Inflammatory Disorders

- Severe upper respiratory infection
- Syphilis
- Viruses, especially poliovirus, echovirus 7, HIV, and a mutant virus termed stealth virus
- Prions

Heavy Metals and Trace Minerals

- Aluminum
- Copper

- Lead
- Manganese
- Mercury
- Selenium

Nutritional Disorders

- Dietary deficiencies
- Gastrointestinal dysfunction
- Vitamin deficiencies

Physical Injury

- Pneumatic tools
- Prior surgery
- Electrical injuries
- Prior trauma
- Radiation

Toxic Agents or Exposures

- Animal carcasses and hides
- Endogenous "toxins"
- Gasoline
- Household pets
- Spinal anesthesia

Other

- Abiotrophy (premature aging)
- Apoptosis (programmed cell death)
- Malignancy
- Neurotrophic factor deficiencies

- Nitric oxide
- Paraproteinemia
- Altered expression of vascular endothelial growth factor (VEGF)
- Vascular disorders

Type 1 ALS (ALS1) accounts for approximately 20% of familial cases. This autosomal dominant genetic disorder was identified as a defect in superoxide dismutase on chromosome 21 (SOD1) a decade ago. Superoxide dismutase is an antioxidant enzyme found in most cells throughout the body, which acts as a free radical neutralizer to inhibit cell destruction in the brain. [5]

In 2001, a newly identified gene mutation on chromosome 2q33 was deemed responsible for a rare, slowly progressive, early-onset form of the disease, called juvenile inherited ALS (ALS2), found in highly inbred populations of North Africa and the Middle East. [6]

In sporadic ALS, etiopathogenic hypotheses are not mutually exclusive, as one problem may trigger another and it is difficult to ascertain which one came first. [7] Three important etiologic themes are considered here.

The first involves free radicals resulting in oxidative stress. Free radicals are highly unstable single electron molecules in pursuit of another electron for pairing, [5] resulting in damage to components of the cell's membranes, proteins, or genetic material. [8] This process, termed oxidation, is what causes metal to rust and apples to turn brown. The brain's first-line defense against free radical attack includes 2 enzymes, superoxide dismutase and glutathione. [5] In ALS, these antioxidant mechanisms fail, resulting in oxidative stress.

Another possible etiology involves the neurotransmitter glutamate. Glutamate, an amino acid present in proteins, is the primary excitatory neurotransmitter in the brain. Those with ALS either produce too much glutamate or have a faulty transport system, resulting in overstimulation of the cell, influx of calcium, and resulting cell death. [8] Riluzole (*Rilutek*), the only drug approved by the US Food and Drug Administration (FDA) to treat ALS, works by blocking the release of glutamate by brain cells. [5]

Finally, neurofilaments are a collection of protein clumps that normally provide structural support for the axon, but when they become tangled, transport of messages is impeded. This axonal strangulation causes the neurons to die. [8]

In ALS, wasting of the muscles (amyotrophic) is due to scarring of the upper motor neurons in the lateral portion of the spinal cord (lateral sclerosis). Two types of motor neurons supply voluntary muscle. Upper motor neurons (UMNs) begin in the brain and extend to the spinal cord. Dysfunction and impairment of UMNs result in loss of dexterity, muscle weakness, spasticity, pathologic hyperreflexia (eg, clonus), pathologic reflexes (eg, positive Babinski), and spastic bulbar palsy (ie, speaking, chewing, and swallowing affected; also spontaneous or unmotivated crying and laughter). Lower motor neurons (LMNs) begin in the spinal cord and extend to the muscles. Dysfunction and impairment of LMNs result in muscle weakness (eg, foot drop), muscle atrophy, hyporeflexia, muscle hypotonicity or flaccidity, fasciculations, muscle cramps, and truncal muscle weakness.

The holistic framework is a concept primary to the practice of nursing, but traditionally avoided by medical professionals since it cannot be externally measured or controlled. [9] Margaret Newman asserts in her model, *Health as Expanding Consciousness*, that every person in every situation, no matter how disordered and hopeless it may seem, is part of the universal process of becoming more of oneself, of finding greater meaning in life, and of reaching new heights of connectedness with other people and the world. [10] This is significant in ALS, a diagnosis that seems devoid of hope.

Newman also theorizes that as the nurse fully engages in the experience with the client, both mutually participate in a process of expanding consciousness. [9] This process can be profound for persons with ALS and their caregivers. It is meaningful that Margaret Newman's mother had ALS.

In the holistic framework, body, mind, and spirit are one: separating them aids in defining things, but in reality they are a dynamic whole. Nursing practice requires exploration of all avenues to help people, and nurses are in a unique position to bridge the gap between traditional, Western healthcare and complementary therapies. [11] Barnum describes healing not just as the curing of symptoms, but a blending of technology with caring, love, compassion, and creativity. [12]

What makes physical care so challenging in advanced ALS is the absence of other indicators of change -- the verbal and physical response to care. The physical changes are much more subtle, and the nurse is more likely to detect changes when there is a connection with the patient that allows a rhythm to unfold. Daily inspections included in the assessment are crucial if potential problems are to be avoided. Particular challenges of patients with ALS who are locked-in follow. provides a blueprint for supplies in establishing and maintaining a safe home care environment.

Table 2. Homecare Supply Set-Up and Maintenance

Item	Comments
Respiratory	
Ventilators (2)	1 at the bedside; 1 for wheelchair and backup
Bacterial filters (3 sets)	For ventilators
External 12-volt battery	For portable ventilator
Battery charger	For portable ventilator
Tracheal cuff pressure gauge	Pressures < 25 cm/H_2O decrease risk of complications
Percussor	Use for chest physiotherapy
Pulse oximeter	Continuous bedside monitoring
Suction machines (3)	1 at the bedside; 1 as backup; 1 portable (battery powered)
Low-pressure backup alarm	Backup for ventilator alarm
Compressor for nebulizer (2)	1 at bedside; 1 as backup
O_2 condenser	Continuous at the bedside
O_2 tubing (3)	1 25 ft for travel, the rest 6 ft
E-Tanks (number depends on use)	Backup O_2, travel
Heat and humidity source	Example, Hudson Heater, Concha columns, with Concha water; for portable, need heat and moisture exchanger, sometimes call an "artificial nose"
Manual resuscitation bags (2)	1 immediately available at all times; 1 behind ventilator as backup
Ventilator circuits	2 rotate for weekly cleaning (return circuit behind ventilator once dry); 1 for wheelchair

complete (3)	
5 gallon buckets with lids (2)	1 for ventilator cleaning solution; 1 for rinse
Red Rubber Catheters (RRC) (case)	Use for clean suctioning technique; clean nightly; change weekly
Quart containers with flip-top (2)	1 for RRC/sterile water/vinegar; 1 for sterile water rinse
Suction hoses (4)	2 rotate for weekly cleaning; 1 for portable; 1 for backup
Yankauer suction (3)	2 rotate for weekly cleaning, wash nightly; 1 for portable; 1 for backup
Tracheostomy tube (3)	Backups for monthly changes and emergencies; include a tube 1 size smaller
Trach tube holder (case)	Example: Dale
Nebulizer kits (6)	Rinse after each use; clean 3x per week
Bronco Saline	NS for nebulizer
Normal Saline vials (box)	Best for suctioning when needed for tenacious secretions
General	
2x2 IV sponges, sterile (6 boxes)	G-tube stoma care
2x2 sponges, clean (2 packages)	Eye care, stoma care
4x4 sponges sterile (2 boxes)	Wound care and miscellaneous
4x4 drain sponges, sterile (6 boxes)	Trach care
6" Cotton-tipped swabs (2 boxes)	Trach and g-tube care
Alcohol swabs (2 boxes)	
Cleaning solution for ventilator circuit	Example: Control III
G-tube (3)	Change monthly and as needed
Gloves (by box or case)	All sizes to fit staff and caregivers; consider powderless, nonlatex
Absorbent pads	Deluxe grade for comfort and absorbency
Nonpetroleum lubricating jelly	Tube changes
Fire extinguishers and smoke detectors	1 near patient, others in rooms such as kitchen

While some people with ALS on ventilators maintain eye movement for years, complete paralysis is also possible. The eyes may appear frozen open, or unable to open at all, though lagophthalmos (incomplete closure of the lid) may occur. If a consistent protocol is not instituted, dryness of the membranes, infection, exposure keratitis, corneal ulceration, and scarring can occur, causing some physicians to stitch the eyelids shut. This is completely avoidable with meticulous preventive care. The sclera needs to remain moist at all times. A lubricating ointment applied every 4 hours on a regular, set schedule decreases the prevalence of corneal abrasions in patients paralyzed or heavily sedated, [13] making taping unnecessary. Some patients have reported success with lubricating inserts, such as *Lacriserts* (hydroxypropyl cellulose), placed in the conjunctival sac once per day.

Nurses may have worked with mechanical ventilators for years, but have usually shared this responsibility with a respiratory therapist. In the home, the nurse is solely responsible, and must be knowledgeable and confident about the machine itself, the complete circuit configuration, the ventilator settings and their implications, and emergency interventions.

The supply company leasing the equipment will schedule routine ventilator checks and maintenance. In the home, it is good practice to have a backup manual resuscitator bag and complete circuit immediately available, with a backup ventilator nearby. Ventilator checks can be included in the documentation flow sheet competed every 2 hours around the clock.

Peak inspiratory pressure sustained over a predetermined level requires immediate implementation of a pulmonary hygiene protocol, such as increased chest physiotherapy, nebulization treatments, and suctioning. Bedside oximetry provides continuous, real-time assessment data. Clear breath sounds should be the goal, and this requires routine clearance of the airway. An alternative to suctioning for some people with ALS is routine use of a mechanical insufflation-exsufflation (MI-E) device such as the CoughAssist ^{TM}, which helps clear secretions by applying positive pressure to the airway and then cycling rapidly to negative pressure. The high expiratory flow produced from the lungs simulates a cough. [14] Studies demonstrate efficacy of the device, with the need for suctioning eliminated in some patients. [15]

One challenge with long-term mechanical ventilation is to maintain a seal at the tracheal tube cuff without damage to the trachea. When cuff pressure exceeds tissue perfusion pressure, ischemia leads to complications such as tracheal malacia, tracheal stenosis, tracheal-esophageal fistula, and erosion into the innominate artery. [16] The cuff monitoring techniques customarily used in the home are minimal leak technique, minimally occlusive volume, and/or squeezing of the pilot balloon, though these techniques are subjective and unreliable, and alone do not safeguard against hyperinflation. [17] The only way to assure pressures exerted against the tracheal wall do not exceed capillary perfusion pressure is to measure the pressure with a gauge or manometer every shift and as needed to assure it is below 25 cm H_2O.

Manufacturers generally recommend tracheostomy tube changes every 30 days. Nurses demonstrating competency can do this at the bedside with a healed stoma, though assistance is required -- changing the tracheostomy tube is a 2-person procedure. [18]

Attention to oral care and assessment is an important component of care for the chronically ill. Studies have demonstrated that bacteria that colonize in dental plaque contribute to ventilator-associated pneumonias. [19,20] Sponge cleaning of the teeth was found to have little effect on dental plaque, and is unlikely to reduce the risk of ventilator-associated pneumonias. [20,21] Though studies are ongoing, the current recommendation is to brush the teeth with a toothbrush 3 times a day. [21]

While prevention is the best treatment, dental problems can also occur: some dentists and hygienists do make house calls. Also of note, loss of muscle tone over time often causes the dental architecture to shift.

Enteral access can include gastric buttons, percutaneous endoscopic gastrostomy (PEG) tubes, and gastrostomy tubes changed at the bedside. For example, the Ponsky ^{TM} G-Tube is replaced by the nurse every month in a healed

stoma; the anchor device is a water balloon.

Nutritional options are not limited to supplements in a can. People with ALS are encouraged to avoid processed foods in favor of fresh, natural foods. [22] Even after the person is unable to take food by mouth, nearly all foods can be blended, strained, and given as a bolus enteral feeding. Some patients literally have what the family is having -- their meal is put in the blender, strained, and administered.

Menus can also be created based on nutrient-dense food choices ( ). It is a JCAHO requirement that clients receiving home nutrition care be monitored regularly to determine the effectiveness and appropriateness of therapy in meeting nutritional needs, [23] and this is particularly important to people with ALS as their clinical condition is in constant flux. Without verbal and nonverbal communication, assessment findings such as abdominal distension, high residual in the stomach before the next feeding, and bowel function are essential to assess gastrointestinal tolerance.

Table 3. Nutrient-Dense Menu Options for Enteral Feedings

Macronutrient	**Protein 20% to 25%** **4 kcal/g**	**Carbohydrates 55% to 60%** **3.3 kcal/g**	**Fats 20% to 25%** **9 kcal/g**
Nutrient dense menu options	Fish	Fruits	Avocado*
	Salmon+	Cantaloupe*	Coconut
	Tuna+	Grapefruit*	Tofu
	Mackerel+	Orange*	Nuts
	Herring+	Strawberries*	Olive oil
	Meat	Tomato*	Flax seed oil
	Chicken	Watermelon*	Fish oil products
	Eggs (not raw)	Vegetables	
	Yogurt	Acorn squash*	
	Goat milk	Asparagus*	
	Tofu	Cauliflower*	
	Soft or silken	Broccoli*	
	Protein powder	Potato*	
	Whey	Grains	
		Quinoa	
		Brown rice	
		Millet	

		Barley	

Assuring energy needs are met requires a baseline assessment and ongoing collaboration with the dietitian. Traditionally, caloric requirements are subtracted for each paralyzed limb, but studies that indicate patients with ALS are hypermetabolic [24] should be taken into consideration. Usual resting energy expenditure (REE) is between 20-25 kcal/kg, but the need can increase 50% with injury or illness. [25]

In the clinical setting, use of indirect calorimetry during mechanical ventilation provides the most precise measurement of daily caloric expenditure. [25] The technique measures oxygen consumption and carbon dioxide production to calculate REE. [25] Predictive equations, coupled with knowledge of an individual ALS patient and the clinical course, provide an estimate of energy requirements ( ).

Table 4. Determining Energy Needs

Step 1. Determine the basal metabolic rate (BMR). Also called resting energy expenditure (REE) and basal energy expenditure (BEE). Energy expenditure = BMR x stress factor x activity factor.	
Harris-Benedict formula	
Men	66 + (13.7 x wt in kg) + (5 x ht in cm) - (6.8 x age in years)
Women	655 + (9.6 x wt in kg.) + (1.8 x ht in cm) - (4.7 x age in years)
Step 2. Adjust the BMR for added stress (very general)	Surgery: 1.1-1.2 x Trauma: 1.1-1.4 x Infection: 1.1-1.5 x
Step 3. Determine level of physical activity	Ambulatory 1.3 x Confined to bed 1.2 x
Protein Requirements	
Healthy adult	0.8 g/kg per day
Stressed adult	1.5-2 g/kg per day

A food nutrient book allows the nurse to calculate nutritional information by hand for each general food item to create a menu template. Dietary software programs are a useful tool, but expensive to purchase. One free online program, The Interactive Healthy Eating Index, was identified, Though obtaining accurate weights may be difficult to impossible on an ongoing basis, body measurements, such as the neck and abdomen, can be taken to monitor weight status.

While whole foods provide a sound nutritional foundation, people with ALS may benefit from a good multivitamin liquid or powder and additional supplementation based on scientific evidence. Reference intake values are designed for populations, but can serve as a guide for individual needs. [26] Decisions on which supplements to choose may be determined from a combination of dietary software analysis; laboratory test results (CBC, SMA20, B12, red cell folate, prealbumin, and trace minerals); what the client tolerates; and what they can afford.

To minimize invasive devices, male and female urinals can be placed with care and utilizing padding. Suprapubic, in-and-out, and indwelling catheterizations are options, though chronic urinary tract infections can be common in people with ALS and difficult to manage. Cranberry-derived substances including glycoproteins, fructose, and condensed tannins (proanthocyanidins) prevent bacterial adherence, especially *Escherichia coli*, to urinary epithelial cells and

can minimize infection risk. [27-29] Whole cranberries can be included in blenderized feedings or administered in capsules of 300-400 mg twice daily. Free water should be adjusted to keep urine pale yellow to reflect adequate hydration. Sufficient body water is essential for healthy physiologic functioning. [30]

The maintenance of regular daily bowel movements is important for people with ALS to prevent obstruction. This complication is completely preventable with a bowel program scheduled at the same time each day, using a stimulant suppository such as bisacodyl for evacuation. Whole food should provide fiber that helps motility, and adequate hydration is important, particularly if psyllium is used. The use of acidophilus with active cultures in liquid, powder, or yogurt administered through the g-tube maintains healthy bacteria in the gut.

According to the literature, pain is generally not associated with ALS, but paralyzed limbs that are not moved can cause significant discomfort. JCAHO now includes pain standards in accreditation, [31] but because people with ALS in advanced stages may not have any objective communication, the nurse needs to be aware of subjective signs of pain, such as heart rate and blood pressure, and to document measures that minimize discomfort.

People with ALS need repositioning at least every 2 hours, with dependent bony prominences inspected. The nurse can instruct willing family members and friends in passive range-of-motion (PROM) exercises to assure there is adequate movement and sensory stimulation.

If the budget allows, weekly massages increase the blood supply to muscles, improve venous return to the heart, and help maintain flexibility to minimize pain. Massage also provides the touch that is a basic human need, resulting in a soothing, emotional release. Many practitioners will come to the home, and students work for a fraction of the cost. Caregivers, spouses, and significant others also benefit from massage. In a study of hospitalized cancer patients, mean scores for pain, sleep quality, symptom distress, and anxiety improved from baseline for subjects who received therapeutic massage. [32]

Therapeutic touch (TT) is another useful strategy. Mulloney and Wells-Federman [33] describe numerous studies citing the benefits of TT to reduce pain, promote sleep, reduce anxiety, and relieve depression. Also described was the nurse's experience while doing TT as one of expansion of consciousness as they assume and maintain a meditative state.

In ALS, the body withers while the mind stays intact. People with ALS have had to adjust from being healthy to, at this stage, being completely paralyzed. Remaining engaged in the world around them and feeling comfortable with their thoughts is a heightened challenge with ALS.

Family and friends may need to be educated that the person with ALS is not in a coma, though they might appear that way. They should be encouraged to share with the person with ALS what is going on in their lives, recognizing that it is important to still include laughter and fun in their conversation. Encourage family members to include the person's name in the conversation or use a touch of the hand to let them know they are included.

All efforts should be made to identify means of objective communication as long as any controlled movement is possible by the person with ALS. Though a new science, using the brain as a signal for communication is under research, and is already a means for some people locked-in to communicate basic needs. [34]

Other nontraditional strategies to stimulate the mind include use of a therapeutic light-sound machine. This device delivers precise frequencies of flickering lights from glasses coordinated with audio pulses from headphones, which moves the brain through different stages of frequency to induce specific states of consciousness. When the brain mimics the frequency of the stimulation, a synchronization, termed entrainment, occurs. [35] The device is not recommended if there is a history of a seizure disorder. Brainwave CDs alone can be used to modulate brainwave frequencies through music, meditations, and exercises. [36]

Maslow's Hierarchy of Needs [37] is relevant to people with ALS, not only in prioritizing care, but in recognizing the

potential for inner growth. In review, at the bottom of the hierarchy are physiological needs, such as food, water, and oxygen. The next level are safety needs, such as security; stability; dependency; protection; freedom from fear, anxiety, and chaos; and the need for structure, order, law, and limits. Next are belongingness and love needs, such as giving and receiving affection. Esteem needs are defined as the need for strength, achievement, adequacy, mastery, importance, dignity, or appreciation. Self-actualization is the highest level of accomplishment on the most personal, inner level of being: the peak of human achievement.

Supraquadriplegia does not preclude a person from becoming self-actualized, even if only for brief periods of time, though nurses and family provide a vital role in assuring lower level needs are met. Affirmations can be powerful tools in replacing negative self-talk and shifting awareness to facilitate expanded consciousness.

Spirituality promotes hope and optimism in the search for meaning through devastating illness. As a path to reduced anxiety, depression, and fear of dying, studies demonstrate the importance of spirituality on quality of life for people with chronic and terminal diseases. [38] Whether expressed through structured religion or intensely private beliefs, spirituality taps the best of us -- our core being. It is the basis of growth, stability, and feelings of wholeness. As Dennis Wholey [39] said, "The real journey of life is not upward, but inward." Inspirational works help guide the client and nurse to optimal awareness and growth.

To care fully for a person locked-in, the nurse must first believe that quality of life is possible. In the ALS Patient Profile Project, the conclusion was that anyone living with ALS has the potential to lead a life of quality, despite profound physical limitations from ALS. [40] One person with ALS at the age of 12 years called ALS "the loving disease," because it allowed her to be closer to the people she loved most. [3]

In order to enhance understanding of the individual's experience and use that awareness to facilitate the healing process, the American Holistic Nurses' Association requires nurses to integrate self-care, self-responsibility, spirituality, and reflection in their lives. [41] Through this process, the client and the nurse mutually participate in expanding consciousness. [9]

Florence Nightingale believed the quality of the patient's environment affected healing and recovery. [42] One aspect of that is the safety provided by consistent staff. Research demonstrated chronic critical care patients do not do well with a different nurse taking care of them every day, as continuity is crucial to the client's emotional and physical well being. [43]

Because only movement and vision (secondary to ophthalmoplegia) are affected, the other senses -- touch, sound, and smell -- should be enhanced to promote a healing environment. The concept of a healing environment is the concept of many therapeutic spas that can be applied in the home.

The nursing staff and family can create a healing place surrounding the bed or wherever the patient spends the majority of time. Consider the gentle flow of water in a fountain, quality incense or essential oils, soft clean sheets, and gentle music. Bathing requires an attitude shift to the spa environment as well. Rather than the assembly line approach, imagine a bath in a spa -- the fragrances and gentle touch. The bath is a wonderful opportunity to inspect and connect. That loving intent, accepting the person just as they are without judgment, opens the door that makes it easier for the patients to love and accept themselves as they are.

The most significant aspect of the healing environment is the healing that is taking place on the inside -- and the energy of love, appreciation, and caring is palpable, just as the energy of anger, fear, and confusion is palpable. What can be done to increase the amount of healing energy by those closest to the patient? Does the family have the support they need, physical and spiritual? Are there extended family, neighbors, or church members who could create a helping network?

Rather than have JA drift off in a dream state, the team sought to keep his mind stimulated, heart full, and life he valued so much worth living. Shift report was always given in his presence, and he was included in any decisions. In

the spring of 1996, JA was able to test one of the first mind-driven tools -- MCTOS or Mind Controlled Tool Operating Switch. Though he had tried other tools without success, after about an hour, he was able to make the machine alarm on command, and be still on command. This was tremendously empowering for JA, who after nearly 2 years of being completely locked-in was able to validate questions, such as, "Are we able to meet the majority of your needs the majority of the time," and "Is life still worth living?" His repeated, "yes" in answer to these questions was validating for the family and nurses. He understood now what was impossible to imagine in the beginning of his illness: that he could be loved, just as he was. See the Appendix for JA's plan of care during this time frame.

During 5 1/2 years at home, with a strong team and loving family, JA never went back to the hospital or even had an ED visit. Problems were acted on quickly, and interventions were consistent with a small staff. He was fortunate to have a physician who made house calls, while the majority of nurse-physician interaction took place by phone and fax. He received weekly acupuncture and therapeutic massage.

Over the last 5 months of his life, JA seemed to be shutting down. He was thin, pale, and more prone to bouts of illness. He contracted a gastrointestinal virus that led to severe diarrhea and abdominal distention. One evening, his heart rate dropped suddenly into the 30s; his MD was consulted, and his acupuncturist came to the home on a Friday night for acupuncture and moxibustion, while his family rushed to his bedside from out of state. Unlike other times when JA had been sick, however, he looked "well" -- skin pink and his energy felt peaceful. With no resistance, it was the right time to say good-bye. It was sad, but it was joyful too -- JA had worked hard to come to a place of expanded consciousness through his illness and pain, and now he was free to let go.

ALS is a progressive motor neuron disease that can result in supraquadriplegia and years on a ventilator. A multidisciplinary approach to comprehensive care may involve an APN, general practitioner, neurologist, pulmonologist, physical therapist, occupational therapist, registered dietitian, social worker, home healthcare nurses and aides, and hospice services. Particular challenges occur with variations in insurance coverage and skill mix: RNs, LPNs, CNAs, and lay caregivers may all participate in the delivery of care, depending on the coverage available and financial resources.

Blending art and science, APNs can optimize the client's quality of life, improve outcomes, decrease healthcare costs, and ease the burden on caregivers through the fear and uncertainty of advanced ALS. Within a holistic atmosphere of love and connectedness that facilitates expanded consciousness, in the words of Alexander Pope, "Hope springs eternal."

- ALS Association

  Information, resources, research updates, and existing clinical studies.

- The ALS Clinical Assessment, Research, and Education (ALS C.A.R.E.) Program

  Provides a database and advisory board to develop new and effective ways to manage the care of people with ALS.

- ALS Digest

  bro@met.fsu.edu -- email to subscribe

  Email list for more than 4700 patients, researchers, and families in the worldwide ALS community.

- ALS-Therapy Development Foundation

  Research and development of ALS drugs.

- Discuss ALS

  ALS discussion group for patients, families, caregivers, and medical professionals.

- Family Caregiver Alliance

  Web site with information and support for families of patients with chronic illnesses.

- National Guidelines Clearinghouse

  An evidence-based review of the care of patients with ALS

- Muscular Dystrophy Association: ALS division

  ALS information, resources, research updates, and existing clinical studies.

- National Institutes of Health

  Patient link to clinical trials. Search by disease, location, treatment, or sponsor.

- National Library of Medicine: PubMed at Medscape

  Search by disease for scientific abstracts and full-text articles.

- American Association for Respiratory Care (AARC) clinical practice guideline: Long-term invasive mechanical ventilation in the home

- AARC clinical practice guideline: Suctioning of the patient in the home

- Quick reference guide for clinicians: Mechanical ventilation beyond the intensive care unit

- American Society for Parenteral and Enteral Nutrition (A.S.P.E.N.): Standards for Home Nutrition Support

- American Society for Parenteral and Enteral Nutrition (A.S.P.E.N): Standards of Practice for Nutrition Support Nurses

- Arbor Nutrition Guide

  Web site with nutritional information, calculations, and extensive links.

- Interactive Healthy Eating Index

  A free dietary assessment tool that calculates an entered diet compared with recommendations based on the Food Guide Pyramid.

- National Institutes of Health: Office of Dietary Supplements

  Health information, FDA guidelines, and research regarding dietary supplements.

- MJA Website

  Provides free software with nutrition calculations for Windows and Palm OS

- Anna Wise Center for Awakened Mind Training

  Provides information on consciousness development and related products with link for purchase.

- Learning Meditation

  Provides information and speech guided meditations.

- Self-actualization

  Web site describes Maslow's Hierarchy of Needs, characteristics of self-actualized people, and provides an email link for self-actualization affirmations.

- Brain Actuated Technologies, Inc.

  Provides descriptive and purchase information on Cyberlink, a device that senses and responds to electrical signals generated from muscle, eye movement, and brainwave activity.

- Communication Independence for the Neurologically Impaired, Inc. (CINI)

  An organization devoted to improving the quality of life of people with ALS by disseminating information about communication technology.

- NeuralSignals

  Provides descriptive information for the Brain Communicator, the first experimental brain implant approved by the FDA for study in humans. Also provides purchase information for the Muscle Communicator, a noninvasive device that detects imperceptible muscle activity.

- Technos America

  Provides descriptive and purchase information for MCTOS: Mind Control Tool Operating Switch. Device can be operated by muscle twitch, eye movement, or bioelectrical activity for people without movement.

- The Thought Translation Device

  Information about a device that facilitates communication, using self-control of slow cortical potentials.

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