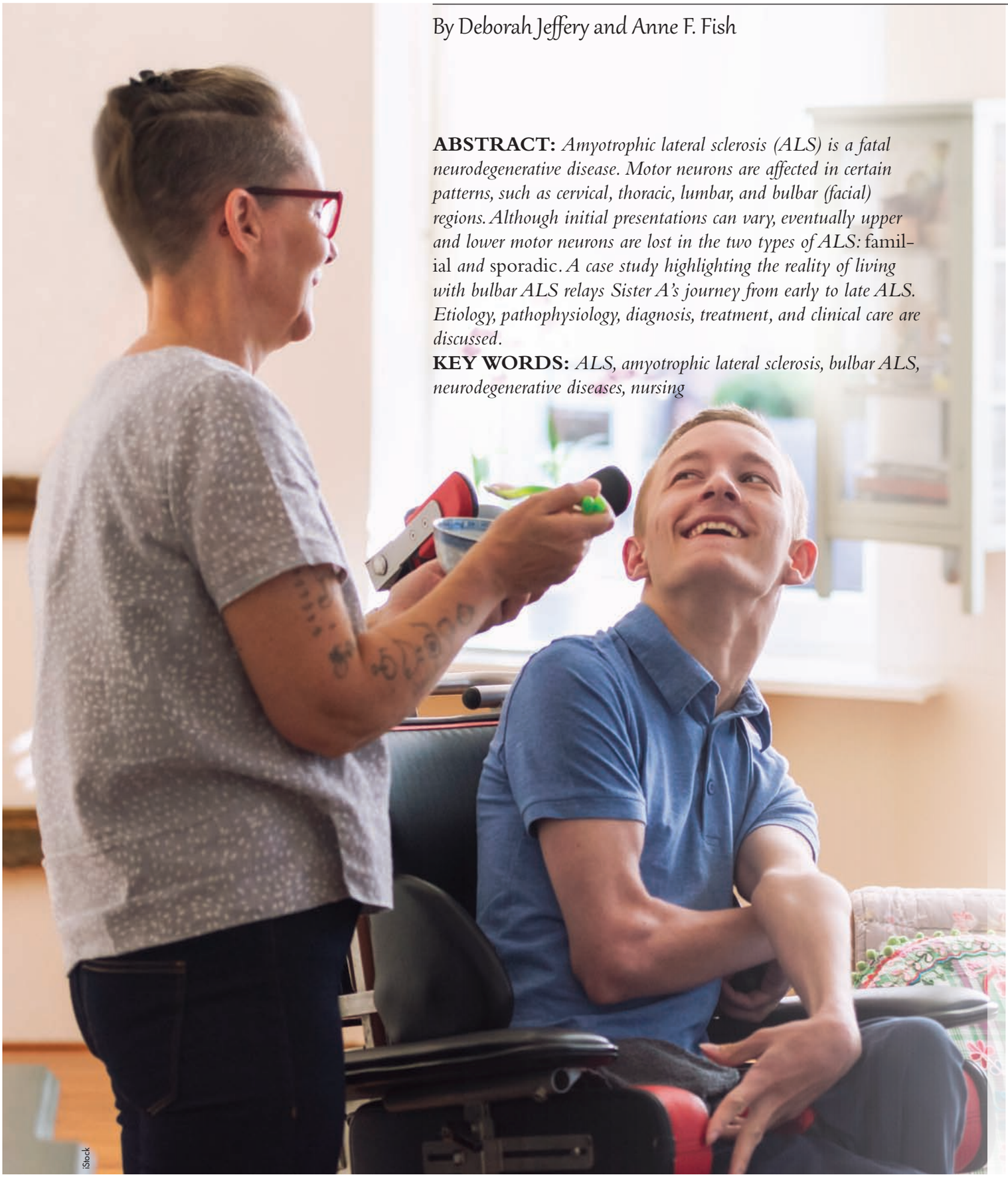
By Deborah Jeffery and Anne F. Fish

ABSTRACT: *Amyotrophic lateral sclerosis (ALS) is a fatal neurodegenerative disease. Motor neurons are affected in certain patterns, such as cervical, thoracic, lumbar, and bulbar (facial) regions. Although initial presentations can vary, eventually upper and lower motor neurons are lost in the two types of ALS:* familial *and* sporadic. *A case study highlighting the reality of living with bulbar ALS relays Sister A's journey from early to late ALS. Etiology, pathophysiology, diagnosis, treatment, and clinical care are discussed.*

KEY WORDS: *ALS, amyotrophic lateral sclerosis, bulbar ALS, neurodegenerative diseases, nursing*



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A Journey with Amyotrophic Lateral Sclerosis

A LIFE INTERRUPTED

Sister A had recently celebrated 50 years of religious life. She had spent her life serving others in various settings. Career highlights included social work and teaching English in Alaskan Native Eskimo villages. She also worked as a missionary, teaching English in South Korea for 2 years, and in settlements with underserved women and children.*

In 2015, Sister A was actively working in her religious community in St. Louis, Missouri, when, over several months, she noticed her ability to articulate speech had slowed, and swallowing had become an issue. Her voice weakness and speech difficulty began to interfere with her work in the contemplative community, where she helped bake and package altar bread for Catholic Masses. The increased need for transportation assistance and help with meals and other activities of daily life caused her to be temporarily assigned to the assisted living community nearby, where she received psychological counseling and medical testing. After 6 months, the diagnosis of amyotrophic lateral sclerosis was made. Sister A was 73 years old.

About 6,000 people in the U.S. are diagnosed with amyotrophic lateral sclerosis (ALS) yearly. A French physician, Jean Charcot, identified ALS in 1869; initially, the disease was called "Charcot Disease." However, in 1939, when the famous baseball player Lou Gehrig was diagnosed with ALS, it became known as "Lou Gehrig's Disease" (ALS Association, 2018a; Armon & Lorenzo, 2017). It is estimated that more than 20,000 Americans live with ALS at any given time (ALS Association, 2018b). Today, patients usually live 2 to 5 years after diagnosis, but progression of the disease is different in each person. It is common for patients to have remissions, lasting weeks to months, when there is little or no loss of function (ALS Association, 2016).

The etiology of ALS is unknown; however, research evidence suggests that genetics and environment play a role in development of the disease. ALS affects Caucasian and non-Hispanic populations and males at a higher rate. The U.S. Department of Veterans Affairs considers ALS a service-connected disease because military veterans are at higher risk to develop it. Veterans who served in the Gulf War are twice as likely to develop ALS than veterans of the same period who did not serve in the Gulf War (Mehta et al., 2017). Exposure to environmental toxins, such as lead and pesticides, appears to be a risk factor (National Institute of Neurological Disorders and Stroke [NINDS], 2018). Regarding age, sources offer varying ranges for highest risk of developing ALS, from 55 to 75 (Bellomo & Cichminski, 2015), 40 to 60 (NINDS), and those 60 to 69 (Mehta et al., 2017). This article relays the journey of Sister A after diagnosis of ALS.

PATHOPHYSIOLOGY

ALS is a motor neuron degenerative disease, involving the deterioration and death of lower and upper motor neurons, affecting the brain, brainstem, and spinal cord. *Lower motor neurons* that innervate voluntary muscles in the head, neck, and limbs, as well as *upper motor neurons* that descend through the pyramidal tract to synapse with the lower motor neurons, are affected. Initially there may be specific functional loss of only upper or only lower motor neurons, but eventually both are impacted. If ALS occurs concurrently with frontotemporal dementia, there is additional frontotemporal cortical neuron degeneration and cortical atrophy (Kasper et al., 2015).

When motor neurons die, denervation and atrophy of the innervated muscle fibers occur. As denervation progresses, the clinical exam and muscle biopsy reflect muscle atrophy changes. This is referred to as *amyotrophy.* When cortical motor neurons die, there is thinning in the corticospinal tracts that travel to the lateral and anterior white matter columns of the spinal cord. When this occurs, fibers and

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*Patient's name has been abbreviated to protect privacy.

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firmness are lost; this is *fibrillary gliosis*. This is the basis for the remainder of the disease name, termed *lateral sclerosis* (Kasper et al., 2015).

ALS is broadly categorized into two types, familial and sporadic. *Familial* ALS (FALS), representing 5% to 10% of all U.S. cases, is inherited, with each offspring having a 50% risk of inheriting the genetic mutation and expressing the disease. The genetic pattern is thought to be autosomal dominant inheritance, where only one parent carries the gene for ALS. The age of onset of FALS is approximately 10 years earlier than sporadic ALS, with a shorter life span (Bellomo & Cichminski, 2015). The most common type of ALS, occurring in 90% to 95% of cases, is *sporadic* ALS (ALS Association, 2018b; Mehta et al., 2017).

The World Federation of Neurology developed guidelines for ALS, known as the El Escorial Criteria, named for a city in Spain where professional groups reached consensus on this diagnostic model. To make a definitive diagnosis of ALS, it must be determined where there are upper and lower motor neuron deficits, as well as weakness (Agency for Toxic Substances and Disease Registry [ATSDR], 2018). On physical exam, the clinician assesses for diagnostic symptoms or manifestations of motor neuron dysfunction. A diagnosis of ALS can be made when motor neurons from three or four of these areas are affected:

- Bulbar (involving the face, jaw, larynx, palate, and tongue);
- Cervical (involving the hand, arm, and diaphragm);
- Thoracic (involving the abdomen and back);
- Lumbosacral (involving the abdomen, back, foot, and leg). (ATSDR, 2018, p. 4).

The sidebar, Clinical Diagnosis and Treatment of ALS, relays information about current diagnosis, treatment, and research into ALS.

JOURNEY THROUGH ALS

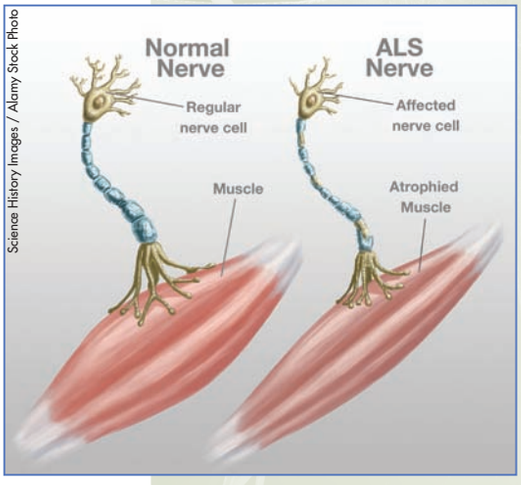
The diagnosis of ALS came as a shock to Sister A, who has lived an aesthetic life as part of her commitment to faith. She has not been exposed to risk factors predictive of ALS: cigarette smoking, alcohol consumption, occupational exposure to toxins, or being a military veteran. She had no family history of neurodegenerative diseases (Bryan et al., 2016; Mehta et al., 2016).

Yet, Sister A got the disease. She soon realized that she would be unable to return to her community and would need permanent placement at the assisted living community to begin intense management of ALS. When asked about progressing weakness in her hands, she noted that the inability to play the organ for Mass has been the most disappointing. In mid-2017, Sister A retained the ability to ambulate without assistive devices.

Sister A was referred to the local ALS Association after diagnosis. The ALS Association (2016) supports an initiative to "Create a World without ALS," which includes Team Challenge ALS and Walk to Defeat ALS. These nationwide community efforts raise awareness and generate revenue to fund research. The connection of ALS patients to the association is important, in order to help patients register with the National ALS Registry. The National ALS Registry was established in 2009 to record the incidence and prevalence of ALS, examine risk factors, and characterize the demographics of those living with ALS (Mehta et al., 2017). The services of the ALS Association are free of charge. The Association offers a community referral network

for additional support and resources within the community.

The ALS Association gave Sister A information about the ALS clinic. At first, she was reluctant to use the clinic, as she felt that her neurologist should remain her primary connection to her care. She visited with the social worker from the ALS Association but felt that she did not want more tests. However, after her discussion with caregivers about being proactive and taking every opportunity to remain functional for as long as possible, she agreed to make an appointment at the ALS clinic at a university school of medicine.



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The ALS clinic offers patients an interdisciplinary team approach, including care by neurologists, pulmonologists, gastroenterologists, nurses; physical, speech, and respiratory therapists; dietitians; and social workers. Patients like Sister A typically see providers from all disciplines and receive testing each visit, according to an evidence-based clinical pathway. In addition, home care services are available from the Advanced Illness Management/Palliative Care Team. This team includes home visits by a physician, nurse practitioner, spiritual counselors, and volunteers who provide pet, art, and music therapy.

Sister A has all three pathophysiological aspects of bulbar ALS with dysarthria and dysphagia, as well as dominant arm involvement which, in her case, is complicated by prior carpal tunnel syndrome. For 4 months, she had episodes called *pseudobulbar symptoms* that included exaggerated, involuntary emotional responses, including laughing, sometimes noted as giddiness, by witnesses. Sister A also tested positive for mild cognitive impairment in short-term memory on her initial palliative care consultation.

Swallowing problems (dysphagia) cause difficulties with liquid and solid food, necessitating limited intake to avoid choking and aspiration. Sister A's decision to allow gastrostomy (G) tube placement for most of her nutrition and all medication administration was made 18 months after diagnosis. Evidence indicates that the decision to insert a G-tube is based on a person's

Clinical Diagnosis and Treatment of ALS

DIAGNOSIS: There is no specific diagnostic test for ALS, thus diagnosis may take 9 to 12 months and often is made by eliminating diagnoses. Estimates are that 50% to 70% of motor neurons are no longer functional when patients seek a diagnosis for symptoms (Mehta et al., 2017).

Overall diagnosis of ALS is based on clinical findings, but it is important to check diagnostic and laboratory tests for confirmation and differential diagnosis. Genetic testing may be done for familial ALS. Laboratory tests include urine and serum protein electrophoresis, 24-hour urine heavy metal panel, complete blood cell count and differential, vitamin B12 and folate levels, C-reactive protein and erythrocyte sedimentation rate, and thyroid and parathyroid function. Brain computerized tomography and magnetic resonance imaging are used to rule out other disorders, for example, brain tumor or multiple sclerosis (ALS Association, 2018a; NINDS, 2018). Electrodiagnostic studies for muscle wasting, sensory deficits, muscle weakness, and denervation include electromyography. Nerve conduction velocity studies and sensory studies may still be normal in ALS initially, but with severe muscle atrophy and denervation, conduction studies will be abnormal (Bellomo & Cichminski, 2015). Additional tests may include lumbar puncture, muscle and/or nerve biopsies, and myelogram of the cervical spine.

MEDICATIONS: There are two FDA-approved medications to treat ALS. A benzothiazole-class drug has been available for many years. Riluzole (Rilutek®) is believed to decrease the neurotransmitter, glutamate, that sends messages between neurons. Riluzole cannot reverse nerve damage, but it can prolong survival time by several months and can lengthen the time a patient can go without mechanical ventilation. The most common adverse effects to Riluzole are nausea, vomiting, diarrhea, abdominal pain, anorexia, dizziness, and asthenia. Liver function tests must be done periodically because there is a risk of liver damage in 10% of patients (Bellomo & Cichminski, 2015).

Edaravone (Radicava®), approved by the FDA in 2017, is the newest of the two primary drugs for ALS. It cannot restore lost function but slows the rate of loss of functioning. Edaravone is given by peripherally inserted central catheter infusion over 60 minutes daily for 14 days followed by 14 days with no infusion, then in cycles of 10 days on and 14 days off. Because it contains sodium bisulfite, there is increased risk of anaphylaxis and allergic reactions, especially in asthmatic patients. The mechanism of action is uncertain. Adverse effects include contusion, headache, and gait disturbances (Mitsubishi Tanabe Pharma America, Inc., 2018).

Another newer medication, Nuedexta®, a combination of dextromethorphan and quinidine, can be utilized to treat pseudobulbar effect, along with other bulbar symptoms, such as difficult speech, facial muscle control, chewing, and swallowing (ALS Worldwide, 2017; Armon & Lorenzo, 2017).

An older antipsychotic drug used in schizophrenia and Tourette's syndrome, Pimozide (Orap®), has been shown in early research to help preserve neuromuscular function in ALS. Pimozide works by decreasing dopamine

in the brain but causes side effects such as drowsiness, dizziness, dry mouth, blurred vision, tiredness, weakness, extrapyramidal symptoms, and tardive dyskinesia. Phase II clinical trials are underway to document safety and efficacy in ALS treatment (Patten et al., 2017).

Ancillary medications are used in ALS for symptom management, including pain, depression, constipation, sleep disturbance, excess saliva and sputum, muscle cramps and stiffness, and pseudobulbar affect. These medicines include baclofen and diazepam for muscle spasm and glycopyrrolate to manage drooling saliva.

INTERDISCIPLINARY THERAPY: A multidisciplinary approach to care is important in ALS. Speech therapy and adaptive therapies can help preserve language; patients can respond to yes-no questions with eye movement or other nonverbal methods. Voice banking is a technique that stores the ALS patient's voice for use in computer-based speech synthesizers, when speech is no longer possible.

To make a definitive diagnosis of ALS, it must be determined where there are upper and lower motor neuron deficits, as well as weakness.

Physical therapy includes exercise that features low-impact and gentle techniques in walking, swimming, and stationary bicycling, which can help keep unaffected muscles strong. Stretching and range of motion exercises help prevent muscle contractures and painful muscle spasticity. Exercise also helps ameliorate fatigue and depression. Occupational therapy can assist with use of braces, walkers, wheelchairs, and ramps. Other healthcare professionals essential to optimal care include social workers, respiratory therapists, psychologists, nutritionists, acute care, home care, and hospice nurses (NINDS, 2018).

EXPERIMENTAL THERAPIES: Promising treatments for ALS are arising from cellular research focusing on understanding why motor neurons deteriorate and exploration of gene mutations. Autologous stem cell treatment is being investigated, along with research into biomarkers that can help identify the presence, rate of progression, or effectiveness of therapeutic interventions for ALS. Treatments using drug-like compounds, gene therapy, antibodies, and cell-based therapies are being explored. For example, researchers are exploring whether lowering levels of a certain brain and spinal cord enzyme in individuals with a specific gene mutation could slow the rate of ALS disease progression (Mehta et al., 2017; NINDS, 2018).

ALS is a non-notifiable condition with the Centers for Disease Control and Prevention. Thus, it is critical that ALS patients register with the National ALS Registry to further research on biomarkers, etiology, progression, and treatment effectiveness. Many barriers remain to identify prevention and causes of ALS, and most importantly, a cure. **—Mary Helming, Contributing Editor, *JCN***

definition of quality of life, acceptance of the disease, and consultation with caregivers. A patient's control over decisions, desirability of prolonging life, and progression of the disease are all considered when discussions about G-tube placement take place (Martin et al., 2016).

Sister A continued to eat small amounts of soft foods, including ice cream, for comfort feedings, but found it increasingly difficult to avoid coughing when swallowing food and liquids. Tube feedings were adjusted to maximize protein and fiber and to control diarrhea, as much as possible. Sister A remains continent of bowel and bladder.

As part of the home-based palliative care team assessment every 4 to 6 weeks, pain and function are evaluated, and adjustments are made to the medication regimen. Specialists started Sister A on Riluzole (Rilutek®), one of two medications with U.S. Food and Drug Administration (FDA) approval for use in ALS. In a Cochrane Systematic Review, Riluzole at 100 mg per day is reasonably safe and prolongs median survival by 2 to 3 months in ALS patients (Miller, Mitchell, & Moore, 2012). This increased median survival represents a 9% gain in the probability of surviving 1 year. Sister A is tolerating the medication, crushed in her G-tube daily. Riluzole resulted in a small beneficial effect on function but not on muscle strength.

Sister A's excessive oral secretions and drooling have been somewhat controlled with an anticholinergic, glycopyrrolate (Cuvposa®, Robinul®) and frequent oral swabbing with moist sponge swabs. Her pain associated with numbness and tingling in the upper extremities has been lessened with the use of gabapentin (Horizant®, Neurontin®), used in low doses to help alleviate pain assessed as neuropathic in origin.

Sister A's diagnosis of bulbar ALS causes a mixture of spastic and flaccid components that results in dysarthria, severe disintegration, and slow articulation. As her speech is no longer understood, she communicates with a writing board or through a text-to-speech computer application. She says the hardest thing will be when she loses the ability to laugh.

Motor neurons controlling extraocular movements are usually spared in ALS, allowing patients to use eye movements to control typing ability with special computers. To facilitate enhanced communication, Sister A learned to use an eye-tracking computer system (ETCS), the most advanced device for communication in patients close to a locked-in status, who due to a loss of motor ability, have lost speech. Patients in late-stage ALS often are left with only eye movements and blinking. ETCS allows patients to type text into a computer device that converts text to voice synthesis (Spataro, Ciriacono, Manno, & La Bella, 2014). This allows patients to access the Internet, webmail, and social networks, as well as communicate with caregivers. Studies suggest a relatively good rate of acceptance of the device. Its use requires cognitive function, ability to hold the head still, and accurate ocular movements (Spataro et al.). A device known as the *eyedrivomatic* allows patients to drive an electric motorized wheelchair with their gaze (Panjwani, 2017).

As motor neurons degenerate, muscle control associated with lung function causes decreased ability to move air in and out of the lungs. Cough assist techniques help clear large airways. Sister A wears an inflatable vest connected to an air pulse generator for 30 minutes daily, a treatment called High Frequency Chest Wall Oscillation. During therapy, the vest delivers a rapidly repeating pulse of air that tightens and releases chest muscles with an air flow system. Each squeeze stimulates a miniature cough that loosens, thins, and propels mucus toward major airways to reduce the possibility of infections and increase airway clearance. This therapy has been shown in clinical trials to be well tolerated by ALS patients and considered helpful in slowing the decline of forced vital capacity (Lange et al., 2006). Sister A refers to the treatment as the "jiggle vest."

The decision to use noninvasive ventilation (NIV) in the future is a decision that Sister A has not yet made. The determination of when Sister A will start NIV will be based on a combination of factors, including performance during pulmonary function testing, sleep quality, fatigue, orthopnea, dyspnea, and mental status changes. NIV is an important intervention in the later stages of ALS because death from ALS usually is due to respiratory failure.

NIV is an alternative to tracheostomy and invasive ventilator support. NIV can use a nasal, oro-nasal mask, or a mouthpiece. NIV is less secure than a ventilator, and the risk of aspiration is greater. The advantages are that tracheostomy is avoided, speech and swallowing are better preserved, and the possibility of home care is greater. The current standard of care strongly favors NIV, as it gives patients a stronger sense of control and avoids ventilator entrapment issues. Use of NIV with a face mask at various times during a 24-hour period may relieve the distressing symptoms of breathlessness and hypoventilation associated with ALS. Evidence shows that NIV improves survival, quality of life, sleep quality, cognitive function, and slows the rate of decline of lung function in ALS (Boentert, Brenscheidt, Glatz, & Young, 2015; Bourke & Gibson, 2004). An intervention involving surgically implanting a diaphragm pacing system, first used in spinal cord injury patients, has been shown to be effective in assisting ALS patients. However, the diaphragm must be thick enough to support implantation of the electrodes (Sanli et al., 2016).

NURSING CONSIDERATIONS

Nursing management for ALS entails a high degree of compassion and presence, not only for the patient but for the family and caregiving community. Caregivers, including nurses, can pray for the ability to offer hope as ALS progresses. Sister A chose the following passage, John 10:14-17 (NIV), for strength and hope. This Scripture is inspirational to nurses who care for patients transitioning to end-of-life care.

> I am the good shepherd; I know my sheep and my sheep know me—just as the Father knows me and I know the Father—and I lay down my life for the sheep. … and there shall be one flock and one shepherd. The reason my Father loves me is that I lay down my life—only to take it up again.

Persons with ALS remain cognitively intact throughout the course of their illness. Caregivers must be especially sensitive to the patient's spiritual suffering, which may be particularly prominent for patients who remain fully aware of their worsening physical status. A spiritual assessment completed by the nurse, patient, family, social worker, and spiritual advisors, such as a chaplain, pastor, or priest, is invaluable. Not only are these patients facing death within an abbreviated time frame, but they watch their physical bodies becoming altered and incapacitated for years before dying.

Nurses can offer appropriate spiritual interventions, based on the spiritual assessment. Learn what is most meaningful to the patient. Prayer, readings from the Bible or other sources, music, inspirational recordings, and listening can be helpful interventions. Pastoral care can be offered and initiated by the nurse, whether the patient remains in the home, a long-term care facility, or an acute care facility. Importantly, nurses can be significant *active listeners* and engage in conversation as the patient and/or family and caregivers need.

The importance of preserving the dignity of the patient is of the highest priority. The nurse is a primary advocate for the rights and choices of the patient and family. Continually assess the patient's and family's support system and coping patterns throughout the ongoing loss of independence. Provide an accepting environment to have honest discussions about their fears and needs. This offers opportunities to relay problems to members of the provider team so that issues can be addressed proactively. Encourage realistic expectations, whenever possible. Refer to appropriate team members, such as counselors and ALS support groups.

Although loss of income and cost of care was not an issue for Sister A, financial concerns are a huge problem for ALS patients, as they lose the ability to work. As they deteriorate, accommodations must be made in the home, such as installing an entry ramp and widening doors for wheelchairs. Patients will eventually need a power

Web Resources

- ALS Association—http://www.alsa.org
- ALS Therapy Development Institute—https://www.als.net
- ALS Worldwide—http://alsworldwide.org
- Les Turner ALS Foundation—http://lesturnerals.org
- Agency for Toxic Substances and Disease Registry—https://www.atsdr.cdc.gov/emes/ALS/
- MedLine Plus—https://medlineplus.gov/amyotrophiclateralsclerosis.html
- Muscular Dystrophy Association—https://www.mda.org
- National Institute of Neurological Disorders and Stroke—https://www.ninds.nih.gov
- National ALS Registry—https://www.cdc.gov/als/Default.html

wheelchair and other assistive devices. For travel, a large van with a lift ramp will be needed. Mounting costs add stress to an already painful situation.

CLINICAL CARE

Nurses can facilitate better patient care through excellent communication about patients, as they work interprofessionally with the care team, including providers who write medical orders and those who offer support services. Nursing care related to activities of daily living is essential in ALS. Nurses can train and support caregivers in multiple aspects of care. Working with occupational therapists (OTs) and physical therapists (PTs), nurses can assist patient communication, using devices, such as the eye-tracking computer, magic slate drawing boards, and word boards.

Nutrition deficits related to swallowing difficulties may be addressed with OTs, as well as dietitians. It is recommended that ALS patients maintain a high-protein, high-carbohydrate diet, and fluid intake of 3,000 mL daily (ALS Association, 2017). By careful patient monitoring, nurses can recognize when the diet needs to change to soft or pureed foods, and eventually to G-tube feedings. The care of oral secretions is a common intervention.

Regarding the integumentary system, nurses should promote repositioning immobile patients every 2 hours to prevent skin breakdown. Maintaining intact skin is critical and is accomplished through careful cleansing of the skin, particularly after elimination, and maintaining skin moisture and hydration. A suitable mattress pad or air mattress that decreases skin breakdown risk can be recommended by the nurse.

Because of muscle atrophy, spasticity, and weakness, nurses can help manage muscular issues by promoting active and passive range-of-motion exercises up to four times daily. For optimal respiratory care, keep the head of the bed elevated at 30 degrees or higher to improve respiratory exchange and digestion. Physical examination of the chest for abnormalities is vital. Encourage deep breathing and coughing every 2 hours while awake. Use of oxygen may be essential, as well as suctioning the airway. The nursing role may include teaching family members how to manage suctioning and encouraging incentive spirometry. Ventilator care may become a significant part of nursing care. To avoid pulmonary embolism, initiate the use of compression stockings for deep vein thrombosis prophylaxis.

Monitor the patient's urinary system, looking for signs of urinary retention and infections. Bowel regimens may also fall under the care of the nurse, to both prevent impaction from constipation and to manage diarrhea from liquid supplements. Many times, nurses may find themselves needing to teach a considerable number of skills to the patient's family and caregivers. The ALS Association (2017) offers excellent, in-depth information on nursing care of ALS patients.

A LIFE WITH ALS, A FACE OF FAITH

In January 2017, Sister A was approaching her second-year anniversary since diagnosis of ALS. She no longer drove a car and required assistance with nutrition and medication administration through a G-tube. Although her symptoms were not yet severe enough to completely limit her activities, she made the decision in early 2017 to move to skilled nursing care in her religious community. She takes a mild antidepressant and uses antianxiety medication, meditation, and prayer to deal with the emotional symptoms associated with a terminal illness. Sister A has appointed a Durable Power of Attorney for healthcare, who will act as an advocate and make decisions on her behalf, especially decisions about hospitalization. She signed an Advance Directive, stating she does not want life-prolonging (mechanical ventilation) or life-saving (cardiopulmonary resuscitation) treatments.

Through 2017, as communication with speech became difficult, Sister A learned how to use a speaking computer. With numbness in her fingers and feet, she was still able to use her writing board and ambulate short distances with the use of a cane. She attended Mass most days and enjoyed reading Scripture.

Most importantly, Sister A continues to do as much for herself as she can. She inspires the nursing staff each day; she smiles and greets everyone with dignity and courage. She has chosen the following Scripture, as she faces each day:

> Love is patient, love is kind. It does not envy, it does not boast, it is not proud. It does not dishonor others, it is not self-seeking, it is not easily angered, it keeps no record of wrongs. Love does not delight in evil but rejoices with the truth. It always protects, always trusts, always hopes, always perseveres. (1 Corinthians 13:4–7, NIV)

As of March 2018, Sister A had declined significantly. She is unable to stand or bear weight. She has had choking issues so we agreed to use a sport bottle for food that has been puréed, and for smoothies and shakes. Her nutrition is mainly G-tube feedings during the night. She was evaluated with a 2-night sleep study for a bi-positive airway pressure (BIPap) machine to assist with breathing. She can no longer speak, and her writing board ability is gone, due to weakness and tremors. We use sign language to communicate.

I (Deborah) took her to the St. Louis Zoo in fall 2017, using a borrowed power wheelchair. We rode the train and visited the bears, big cats, and Children's Zoo. Our favorite thing was the polar bears swimming toward her to touch through the glass. She still has her smile, and she is a gracious presence for all of us. She knows about this article and has promised to read it, unless Jesus calls her home first.

Currently, half of the people affected by ALS live 3 or more years after diagnosis; 20% live 5 or more years; and up to 10% live more than 10 years (ALS Association, 2016, 2018c). Sister A hopes to be in the 10%. She has accepted the fact that she has a limited future. When she learned she had ALS, she felt Jesus' arms holding her, as she prayed in chapel. She said the promise of this verse gave her strength at that time: "The LORD is close to the brokenhearted and saves those who are crushed in spirit" (Psalm 34:18, NIV). Sister A is

using the resources available to her and using her faith to accept each day as it comes. She presents a face of faith, an acceptance of a life with ALS.

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