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N393 Case Study: Communication

Amyotrophic Lateral Sclerosis (ALS), also well known as Lou Gehrig disease in the United States, is a selectively degenerative neurological disorder that affects both upper and lower motor neurons leading to motor dysfunction and eventually death (Porth, 2011). Primarily a disease of middle adulthood, with the onset of symptoms between the ages of 55-65, the incidence is higher in men than women, manifesting in about 2 cases in a population of 100,000 per year (Gordon, 2011). With a mean survival of 2 to 5 years from the onset of symptoms, ALS is considered a complex genetic and neurological disorder which generally follows a progressive course (Porth, 2011). Despite ALS being incurable, supportive therapies such as nutrition and respiratory management in addition to palliative care can help manage ALS symptoms and improve the patient's quality of life (Gordon, 2011).

The etiology of ALS has proven elusive. Five to 10% of cases are identified as familial, generally inherited as an autosomal dominant trait related to a mutation of the Cu-Zn superoxide dismutase type 1 (SOD1) gene on chromosome 21. Identified risks include age (55-65), sex (male), and smoking (Gordon, 2011).

Once manifested, ALS affects three distinct motor neuron locations: the upper motor neurons (UMN) of the cerebral cortex, the lower motor neurons (LMN) of the spinal cord – meaning the anterior horn cells, and the motor nuclei of the brain stem – principally the hypoglossal nuclei. As a progressive disease, manifestations are generally observed first in the distal portions of the spinal cord, as opposed to the proximal parts, leading researchers to believe that distal terminals degenerate with the disease progression moving in a centripetal direction ending in the death of the parent nerve cell. The resulting denervation leads to the shrinkage and atrophy of the musculature. This process is a direct result of the death of LMNs, a process also known as *amyotrophy*, and as a result is incorporated into the disease name. Alternately, the

term *lateral sclerosis* describes the loss of nerve fibers along the lateral columns of white matter in the spinal column in conjunction with the fibrillary gliosis (damage to glial cells due to CNS damage) that takes place (Porth, 2011).

Diagnosing ALS can be difficult, particularly since many of the early signs and symptoms are referable to various UMN and LMN degenerative diseases. Criteria, named the El Escorial criteria, were established in 1994 and revised in 1998 to standardize the diagnosis of ALS. Generally the diagnosis will be based on a history of progressive, and widespread, degeneration of both UMNs and LMNs. The findings are corroborated through electromyogram testing of at least three limbs, which is meant to rule out mimicking diseases and confirms the widespread degeneration of LMNs, a defining characteristic of ALS. Similarly, the absence of sensory involvement is also a defining characteristic of ALS. If an experienced ALS specialist and electromyographer diagnoses ALS based on the criteria above, the diagnosis is 95% likely to be correct (Gordon, 2011).

Manifestations of ALS are related to the death of UMNs and LMNs. In 65% of cases the most common manifestation of ALS is muscle weakness. Typical early symptoms include shoulder weakness, loss of hand dexterity, foot drop, and difficulty walking. Generally the symptoms associated with LMNs degeneration, such as cramps, weakness, fasciculation, and atrophy are more noticeable than UMN symptoms. Falls become common as limb function is lost leading to inability to walk, stand, or bear weight for transfers. Dependence on caregivers is unavoidable as a part of the disease progression (Gordon, 2011).

Further disease progression leads to axial musculature weakness, usually manifesting as kyphosis and head drop, both of which can cause pain and further imbalance. Some patients suffer from anarthria complicating patient communication. Similarly, swallowing problems are

common leaving the patient at risk for aspiration, malnutrition, dehydration, and possibly weight loss. Cognitive abnormalities are common, with 15-25% of patients suffering from impaired cognition, with particularly high incidences of frontal lobe dysfunction and dementia. Common abnormalities include changes in judgment, personality, language, and decision making abilities. Emotional lability, which is connected to the upper neural pathways associated with respiration, vocalization, emotion, and facial movements, is not uncommon due to the overall degeneration of UMNs. Similarly, depression and anxiety can develop as a symptom of disease progression complicating decision making (Gordon, 2011).

While pain is not generally associated with ALS, pain can be a feature of the disease process, possibly due to the degeneration of sensory tracts or nuclei. Certainly, the patient will be at risk for pain related to bedsores or atrophy, which is directly related to LMN and UMN degeneration, both of which lead to a loss of mobility. Interestingly, sensory and sphincter function are generally not affected from the disease process (Gordon, 2011).

Death is generally the result of respiratory complications late in the disease progression. Symptoms such as weakened cough, morning headaches, orthopnea, and dyspnea on exertion are common respiratory complications. Leading causes of death in ALS patients are pneumonia due to aspiration, respiratory failure, and pulmonary complications due to bulbar weakness (Gordon, 2011).

Once diagnosed, management of ALS focuses on symptom management and palliative care with the goal of providing a longer duration and better quality of life. Current research indicates that a multidisciplinary approach to ALS is vital to achieving the goal of longer duration and better quality of life. A comparison of survival curves indicates a 29.7% decreased rate of death and a 7.5 month increased median survival rate for patients whose ALS was

managed through interdisciplinary clinics and general neurology clinics. With such astounding results, it is important that the patient be aware of the necessity for close management of the ALS disease progression in order that any complications that arise can be immediately addressed by the multidisciplinary team. With that in mind, the multidisciplinary approach endeavors to manage ALS disease progression through drug, physical, and speech therapy, as well as nutritional and respiratory management (Corcia & Meininger, 2008).

Pharmacological management of the ALS etiological disease process is limited, though symptoms that are not disease specific, such as asthenia, fatigue, depression, anxiety, drooling, bronchial secretions, pain, pathological laughing, constipation, gastro-esophageal reflux, sleep disturbances, bedsores, venous thrombosis, pulmonary embolism, and cramps can all be managed through various modalities, including drug therapy. At this time riluzole, a glutamate-modulating drug, remains the only FDA approved drug to treat ALS. Suspected mechanisms of action include the stimulation of neurotrophic production factors and cell signaling actions through ion channels. In two clinical trials a daily dose of 100 mg per day over 18 months of treatment was shown to have a 7% difference between placebo and treated groups before adjustment, and a 35% difference after adjustment, effectively decreasing the incidence of death to 1 patient in 13 over a 12 month period of treatment. Rare adverse effects include raised hepatic enzymes, asthenia, and nausea, which generally appear within the first three months of use and disappear if the drug is stopped (Corcia & Meininger, 2008).

Physical and speech therapy are also vital components of ALS management. Physical therapy management focuses on the prevention of joint contractures and secondary pain related to joint stiffness which plagues disease progression and contributes to disability. Exertion is generally avoided since it increases the denervation process, with a focus on passive mobility

instead. Occupational therapy is used as a compliment to physical therapy with a focus on the improvement of performance in skilled tasks. Occupational therapists may also recommend equipment to improve a patient's ability to perform daily activities. Speech complications are common, with 80% of ALS patients suffering speech disturbances. Therefore, the focus of speech therapy is to maintain communication, prevent aspiration of food, improve the utilization of diaphragmatic and intercostal muscles for both speech and breathing, and suggest communication aids for advanced stages of ALS (Corcia & Meininger, 2008).

In conjunction with physical and speech therapy, nutritional and respiratory care are essential in the management of ALS. Denutrition leading to weight loss is a major prognostic indicator of death, with a 7 fold increased risk of death expected when the body mass index is $<18.5 \text{ kg/m}^2$. Causes for denutrition can be depression, upper limb disability, hypermetabolism, and/or bulbar involvement. Furthermore, aspiration is also a major concern for ALS patients sometimes requiring food texture modification in order to control weight loss and prevent aspiration pneumonia. New research also indicates that hyperlipaemia contributes to prolonged survival in ALS indicating that it is beneficial for patients to increase lipid rich nutrients or lipid intake. Some specialists recommend that gastrostomy (PEG tube) placement be utilized to maintain adequate nutrition; however, there is no evidence indicating that PEG tube placement prolongs survival, though some argue that it will improve QOL (Corcia & Meininger, 2008).

Lastly, respiratory management is likely the most fundamental and important aspect of ALS management, since respiratory complications are the leading cause of death in ALS patients. Complications arise from impairment of respiratory muscles, due to weakness, affecting the patient's ability to breathe, expectorate secretions, and cough leading, which in turn

leads to infection, hypoxia, and hypercapnia, any of which could lead to death. Due to the serious implications of dysfunction, pulmonary function must be evaluated every three months. Diagnostic exams which require frequent monitoring include blood gas analysis, spirometry, blood bicarbonate levels, transdiaphragmatic pressure (SNIP) test (measuring inspiratory muscle strength), maximum expiratory pressure (MEP – measuring expiratory muscle strength), and nocturnal oximetry and polysomnography (Corcia & Meininger, 2008). Regarding respiratory treatment options, nocturnal noninvasive positive-pressure ventilation (NIPPV) is now the standard treatment for respiratory insufficiency in ALS patients (Gordon, 2011). Should the patient require invasive ventilation due to an emergency, a ventilator is used, though the prognosis becomes very poor with 50% of patients dying within one month of invasive ventilation and only 20% surviving for more than two years (Corcia & Meininger, 2008).

Considering the life-limiting and terminal nature of ALS, a discussion of palliative care and hospice for ALS patients is necessary. Palliative care focuses on easing patient suffering, meaning an emphasis is placed on lessening pain and symptoms without curing the underlying disease or requiring that the patient give up life saving measures. Palliative care takes a multi-prong approach to care including the multiple goals of addressing physical, emotional, and spiritual care needs within the context of the patient's family and community. By comparison, hospice generally refers to a specific form of palliative care which is focused on easing patient suffering specifically at the end of life. While the hospice goals of managing the physical, emotional, and spiritual care needs of the patient are similar to the general palliative care movement, government Medicare hospice benefits require that enrolled patient no longer pursue life saving measures. In order to meet the various goals of palliative and hospice care, an interdisciplinary team including a physician, nurse, nursing assistant, chaplain, physical or

occupational therapist, social worker, and pharmacist all collaborate to meet patient specific goals and needs with a nurse mediating care and operating as the main contact with the patient and family (Zerwekh, 2006).

In particular, the patient must be made aware of how useful hospice can be in caring for the physical deterioration associated with ALS. It must be made clear that hospice care will offer a very tangible benefit to the management of the patient's pain, physical decline, all of which is in addition to the provisions available for psychosocial support according to the desires of the patient and family. Aside from the basic nursing functions of providing medication and teaching the family care techniques for the various forms of physical decline associated with ALS, the nurse must impress the importance of the interdisciplinary team as a resource for the care and management of ALS, which can make a huge difference in the QOL during the of end-of-life care. Interestingly, in an Australian study on living with ALS and the associated decision making that becomes necessary, the main concern identified by ALS patients was that information on how to live with ALS is not always forthcoming from health care providers (King, Duke, & O'Conner, 2009). With effective communication, this particular concern can easily be addressed by the interdisciplinary hospice care team. It is critical therefore that the patient and family be made aware of ways in which the hospice care team can be utilized as a source of information as well as its ability to ease the emotional and physical distress of coping with ALS as a life-limiting and eventually terminal disease.

Having discussed the pathophysiology and management of ALS, including what important information the patient should know about palliative and hospice care, a case study on ALS is analyzed and discussed as follows:

You have received a hospice referral for Mr. Jones who has ALS (amyotrophic lateral

sclerosis). He and his family, who are confused and anxious, listen to you as you describe what they should expect from the hospice experience. The family does not seem to understand why you are discussing end-of-life issues with them. You call Mr. Jones' family physician who tells you that the patient assured him that he talked to his family about his prognosis. You determine that Mr. Jones has in fact not told his family. You talk with Mr. Jones who admits that he has told his family he is stable and expected to have many years of life remaining. He asks that you help him break the reality of his poor prognosis to his family. ("Case Study: Communication," n.d.)

When critically reviewing the case study, the central theme that requires discussion is communication. As detailed and discussed previously, ALS is a severely life-limiting and eventually fatal disease with a complicated disease progression and constant patient decline. Therefore, communication within the interdisciplinary care team is fundamental to the successful management of ALS, while communication between the family and patient, as well as between the patient, family, and health care providers is even more important. Furthermore, being that the nurse's responsibility within the interdisciplinary team will be to coordinate care and make sure the patient's physical and psychosocial comfort needs are being met (Zerwekh, 2006), creating a working relationship with open communication between the nurse, patient, and family is a top priority of the nurse in this case study.

From the outset, the nurse immediately recognizes a very serious breakdown in communication within the family as well as between Mr. Jones and the primary care physician. Mr. Jones lied to his physician, stating that he had spoken to his family and informed them about his diagnosis and prognosis when he did not in fact do so, leaving his family very confused as to why they are attending a meeting with a hospice care nurse. Furthermore, the nurse can

extrapolate that there is likely a huge learning deficit, making it likely that Mr. Jones and his family will require teaching regarding the expected life-limiting and altering nature of ALS before an effective discussion on the benefits of hospice care for ALS patients can take place.

Difficulties in communication and its impact on end-of-life-care is a well-documented phenomenon. A study based in Norway identified that in order for patients to receive the highest quality end-of-life care, difficult conversations such as addressing the issues of life-prolonging treatment, hospitalization, and other patient and family preferences at end of life must be discussed, preferably before death is approaching (Gjerberg, Førde, & Bjørndal, 2011). While some patients and families find these conversations difficult, discussion can help the patient die a death of their choosing and avoid unwanted interventions. Therefore, while it is recognized that these conversations must take place, the best approach to increase communication is much discussed. In a literature review of 16 current articles addressing manners in which to enhance communication in end-of-life-care, three core themes were identified as being most effective: “utilizing education to enhance professional communication skills; using communication to improve patient understanding; and using communication skills to facilitate ACP (advanced care planning) (Barnes et al., 2012).”

Educating providers was a key theme of multiple articles in the literature review. Particularly helpful were workshops and training courses which helped improve the communications skills of the health care providers who participated. The goal of utilizing communication to improve patient understanding is more difficult to address. However, there are tools that improve communication and help the patient become more knowledgeable about their illness. The “Go Wish” game is meant to aid and focus conversations by using vocabulary to help focus and share topics related to the needs and concerns of the patient. Similarly

Norlander's ACP tool named "The Kitchen Table Discussion" is meant to aid a patient in discussing their illness, values and goals, their experience and perception of death, and empower them to speak with their physician. Furthermore, the use of a question prompt list can help reduce the patient's concern of what to expect in the future. Lastly, using a trained facilitator for ACP decisions can help increase patient knowledge, improve the quality of communication, lead to greater patient satisfaction with the decision making process, and lessen conflict during decision making (Barnes et al., 2012).

However, communication tools alone are not enough to ensure continuity of care. As mentioned previously continuous teamwork and collaboration within the interdisciplinary team is essential for to achieve continuity of care. Furthermore, pathways have been developed to clarify concepts, principles, responsibilities, roles, and enable decision making in end of life care. This is particularly important due to the ambiguity which many times surrounds role and prognosis-related communication which nurses sometimes face (Helft, Chamness, Terry, & Uhrich, 2011). The utilization of pathways effectively facilitates multidisciplinary communication and contributes to the continuity of care. Of particular interest are the palliative care pathway and urgent care pathway developed during a pilot study exploring a new model of collaborative working. These pathways were specifically developed to help direct the care of patients suffering from life-limiting diseases that are not cancer, encouraging providers to utilize multidisciplinary expertise when appropriate, in the process defining roles, concepts, and responsibilities regarding patient care along a continuum individualized to the patient's disease progress while addressing the end of life needs of the patient and family (Alsop, 2010).

Taking the above discussion about communication and its importance under consideration, Mr. Jones and his family would greatly benefit from a family discussion with the

primary care provider and the multidisciplinary end-of-life care team. An initial family discussion will allow for everyone to be informed of Mr. Jones's diagnosis, prognosis, and the services available to both the patient and family. This can then set the stage for continued open communication which is a fundamental necessity for palliative and hospice care to be successful. Furthermore, once the benefits of palliative care and hospice have been discussed, the foundation for further discussions regarding patient and family preferences for care at the end-of-life can then be approached.

As a hospice nurse, working towards the goal facilitating hospice care is very much in keeping with the nursing code of ethics which requires the nurse, amongst other responsibilities, to alleviate patient suffering (Izumi, Nagae, Sakurai, & Imamura, 2012). What is more, as a member of the end of life team, the nurse must mediate patient care; however, it is not appropriate for the nurse to "break the reality" of the prognosis to the family. It is the responsibility of the neurologist to inform the patient and family of the diagnosis and facilitate the commencement of a multidisciplinary approach to care, immediately (Corcia & Meininger, 2008). Therefore, while the nurse cannot personally resolve the initial communication gap present in this case study, she can alleviate the situation by facilitating a family meeting with the appropriate health care team members. The decision to facilitate communication between the clinician and patient/family is governed by the ethical duties of veracity, individual autonomy, and beneficence which require that the patient and family be told the true diagnosis, prognosis, and available end-of-life options, effectively respecting and facilitating the patient's right to make informed decisions for himself (McCluskey, 2007).

Having recognized the failure in communication, the nurse needs a research based approach to correct the problem. While initiating an initial meeting between the patient, family,

and primary physician is important, it is only a first step towards meeting the nurse's communication needs. Therefore, the nurse recognizes that there is a communication and learning deficit related to Mr. Jones not having communicated with his family and physician which is evidenced by the family's confusion regarding his diagnosis and prognosis, and Mr. Jones having lied about the situation to his primary care physician. In order to achieve the goal of communicating the benefits of hospice care to the patient and family, the nurse utilizes the most up to date research to compile a teaching plan to improve communication between the health care provider, family, and patient.

Based, on the research of Barnes et al. (2012), the nurse designs her teaching plan and nursing interventions based on knowledge that communication is most improved by the enhancement of professional communication skills, and utilizing communication skills to improve patient understanding of hospice care. Therefore, the nurse must first recognize whether or not she requires additional training or vocational courses to improve her professional communication skills. If she feels the need for additional training, she can attend professional courses or workshops (Barnes et al., 2012). Similarly, learning and utilizing skills that allow the nurse to evaluate patient learning and understanding will be crucial in assessing the Jones family after their family meeting with the primary care physician.

Due to the initial breakdown in communication, the family will certainly be shocked after learning that Mr. Jones has ALS, which is a terminal disease with an expected lifespan of two to five years. It is likely that the initial meeting will not be effective in communicating information about the progressive nature of the disease. What is more, the family may value the pursuit of medical intervention, even when it might not be medically indicated, a vivid example of medicine's technological optimism (Bishop, Brothers, Perry, & Ahmad, 2010). Furthermore, the

nurse needs to understand the emotional and physical needs of the family and patient. Therefore, the nurse must be able to assess where the family is holding in its emotional and cognitive understanding of Mr. Jones' disease progression.

Considering the multitudinous issues that require assessment, the nurse chooses to utilize the Ask-Tell-Ask model to assess the family and patient's readiness for learning about hospice care as well as to assess any knowledge deficits that need addressing. Using this tool the nurse is able to initiate communication that is centered on the patient and family, knowledge of the disease, and the expected physical changes. Based on the patient and family's perceptions and understanding, the nurse can target questions or issues that must be addressed (Shannon, Long-Sutell, & Coombs, 2011). In this manner the nurse will be able to assess the outcome of the family discussion with the physician as well as determine when the family will most benefit from a discussion about ALS and hospice care, since without a fundamental understanding of ALS disease progression, a discussion about hospice care will be ineffective.

During the process of using the Ask-Tell-Ask model, the nurse may recognize that the family and patient need help identifying topics which should be discussed, or concerns that need to be addressed. In order to facilitate further communication between the family, patient, and health care providers, the nurse may decide to use Norlander's ACP tool named "The Kitchen Table Discussion". This can effectively help the family and patient discuss the illness, their values and goals, their experience and perception of death, and empower them to speak with the health care providers with more clarity and focus (Barnes et al., 2012).

If the nurse utilizes the most recent research on communication properly, the desired outcome will be enhanced communication between the patient, family, and health care team. This improved communication will allow the multidisciplinary team to offer the highest quality

of care which not only meets the needs of the patient and family, but also fulfills the wishes of the family for end-of-life care. Indeed, once the foundation for open communication is set, the nurse will want to eventually discuss interventions at the end of life in order to avoid unwanted invasive interventions and allow the patient to experience a death of their choosing.

In summary, ALS is a devastating, life-limiting, disease with a general life expectancy of two to five years from the point of diagnosis. The QOL for ALS patients can be greatly enhanced by a multidisciplinary team approach to palliative and hospice care. The success of the multidisciplinary treatment is rooted in effective communication, which plays an essential role in the continuity of care and the patient's perception of high quality care at the end of life. In the case study presented, Mr. Jones, his family, and his primary care physician are all experiencing severe gaps in communication. In response, the nurse enhances her communication skills by reviewing evidence based research which enables her to improve communication by coordinating family meetings and utilizing new communication tools such as the Ask-Tell-Ask model and "The Kitchen Table Discussion" tool. As palliative and hospice care continues to evolve, the importance of effective communication within and between the multidisciplinary team, patient, and family will continue to be recognized as a fundamental necessity for the successful care and management of ALS.

References

- Alsop, A. (2010). Collaborative working in end-of-life care: Developing a guide for health and social care professionals. *International Journal of Palliative Nursing*, 16(3), 120-125.
Retrieved
from <http://search.ebscohost.com/login.aspx?direct=true&db=cmedm&AN=20357704&site=ehost-live>
- Barnes, S., Gardiner, C., Gott, M., Payne, S., Chady, B., Small, N., . . . Halpin, D. (2012). Enhancing patient-professional communication about end-of-life issues in life-limiting conditions: A critical review of the literature. *Journal of Pain and Symptom Management*, 44(6), 866-879. doi: 10.1016/j.jpainsymman.2011.11.009
- Bishop, J. P., Brothers, K. B., Perry, J. E., & Ahmad, A. (2010). Reviving the Conversation Around CPR/DNR. *The American Journal of Bioethics*, 10(1), 61-67. doi:
<http://dx.doi.org/10.1080/15265160903469328>
- Case Study: Communication* [Class Handout]. (n.d.). End of Life Care, Long Island University, Brooklyn, NY.
- Corcia, P., & Meininger, V. (2008). Management of amyotrophic lateral sclerosis. *Drugs*, 68(8), 1037-1048. Retrieved
from <http://search.ebscohost.com/login.aspx?direct=true&db=rzh&AN=2009965008&site=ehost-live>
- Gjerberg, E., Førde, R., & Bjørndal, A. (2011). Staff and family relationships in end-of-life

nursing home care. *Nursing Ethics*, 18(1), 42-53. doi: 10.1177/0969733010386160

Gordon, P. H. (2011). Amyotrophic lateral sclerosis: Pathophysiology, diagnosis and management. *CNS Drugs*, 25(1), 1-15. doi: 10.2165/11586000-000000000-00000

Helft, P. R., Chamness, A., Terry, C., & Uhrich, M. (2011). Oncology nurses' attitudes toward prognosis-related communication: A pilot mailed survey of oncology nursing society members. *Oncology Nursing Forum*, 38(4), 468-474. Retrieved from <http://search.ebscohost.com/login.aspx?direct=true&db=aph&AN=61890794&site=ehost-live>

Izumi, S. (., Nagae, H., Sakurai, C., & Imamura, E. (2012). Defining end-of-life care from perspectives of nursing ethics. *Nursing Ethics*, 19(5), 608-618. doi: 10.1177/0969733011436205

King, S. J., Duke, M. M., & O'Connor, B.,A. (2009). Living with amyotrophic lateral sclerosis/motor neurone disease (ALS/MND): Decision-making about 'ongoing change and adaptation'. *Journal of Clinical Nursing*, 18(5), 745-754. doi: 10.1111/j.1365-2702.2008.02671.x

McCluskey, L. (2007). Amyotrophic lateral sclerosis: Ethical issues from diagnosis to end of life. *Neurorehabilitation*, 22(6), 463-472. Retrieved from <http://search.ebscohost.com/login.aspx?direct=true&db=aph&AN=28117361&site=ehost-live>

Porth, C. M. (2011). *Essentials of pathophysiology: Concepts of altered health states*(3rd ed., pp.

913-914). Philadelphia, PA: Wolters Kluwer/Lippincott Williams & Wilkins.

Shannon, S., E., Long-Sutehall, T., & Coombs, M. (2011). Conversations in end-of-life care:

Communication tools for critical care practitioners. *Nursing in Critical Care*, 16(3), 124-130. doi: <http://dx.doi.org/10.1111/j.1478-5153.2011.00456.x>

Zerwekh, J. V. (2006). *Nursing care at the end of life: Palliative care for patients and families*.

Philadelphia, PA: F.A. Davis.