

CASE-BASED PALLIATIVE CARE THROUGHOUT THE CONTINUUM OF DISEASE

Maisha T. Robinson, MD, MS
Alan C. Carver, MD
Janis M. Miyasaki, MD, MEd, FRCPC

Introduction:

Neurological diseases are often chronic and progressive and the management is primarily palliative in nature. Palliative Medicine is appropriate for any patient who has a serious or advanced illness. The aim of this care is to improve overall quality of life by recognizing, preventing, and addressing physical, psychological, spiritual, and social suffering.¹ Palliative care can be delivered by any clinician who is caring for a patient with a serious illness, including a neurologist. Palliative care skills involve providing basic symptom management, addressing code status, and identifying the goals of care.² Specialty palliative care services are available for patients who have complex or unremitting symptoms, complicated social dynamics, or difficult decisions regarding treatment options.² Early, goal-directed conversations are essential as they can help set expectations and guide management plans.

Case-based examples:

Case #1: Mr. Johnson is a 64-year-old man with a two-year history of a right upper extremity tremor. Recently, his family noticed that his movements are slower, he is having more difficulty signing checks, and his handwriting has become smaller. His primary care physician was concerned that he had Parkinson Disease and he was referred to a local neurologist where the clinical diagnosis was confirmed. Mr. Johnson asks about the clinical course and the implications of this diagnosis on his current, very active lifestyle.

Mr. Johnson has provided his neurologist with an opportunity to discuss the typical symptom progression with Parkinson Disease and treatment options. Initial discussions may involve coping with his declining functional status and dependency on others for assistance with his activities of daily living or symptom management. Follow-up visits may incorporate advance care planning where dialogue can begin regarding life-prolonging care and treatment preferences, particularly as the disease progresses. Given the lengthy disease course and progressive debility, having an understanding of the trajectory of the disease may help Mr. Johnson and his family make plans for his care and for maintenance of some of the activities that he enjoys.

Discussing the neurological trajectories of illness³ may be useful to assist in framing conversations regarding symptom control and goals of care. The recognized trajectories to death include (1) a brief period of decline, which is commonly seen in patients with brain tumors or malignancies that are metastatic to the central nervous system. These patients have a relatively preserved functional status for the majority of their disease and then they have a precipitous decline; (2) catastrophic injury with subsequent decline, which is typically seen in patients who have strokes, intracranial hemorrhages, or other traumatic brain injuries; These patients have an acute deterioration at the time of the event; (3) prolonged decline, which is usually described in patients with neurodegenerative diseases. These patients have a slow, but steady, functional decline throughout the course of their disease; (4) relapsing and remitting, which is seen in patients with demyelinating diseases. These patients have periods of exacerbations, followed by some improvement, but their functional status does not improve back to their baseline.

Case #2: Ms. Jones is an 80-year-old woman with a history of congestive heart failure, poorly controlled hypertension, atrial fibrillation on anticoagulation, and an ischemic stroke with a residual right hemiparesis. She presents to the Emergency Department from a nursing home secondary to mental status changes. On examination, she is comatose, her pupils are anisocoric, and her respirations are labored. A non-contrast head CT reveals a large right fronto-temporo-parietal hemorrhage with 12 mm midline shift with uncal herniation. Her INR is 5.7. Her daughter, who is her health care surrogate, has arrived and she is requesting to discuss her mother's prognosis and treatment options.

Ms. Jones' presentation is consistent with the "sudden neurological impairment" clinical trajectory. From a palliative care standpoint, the discussion with her daughter would involve an update on her clinical status, a concise explanation of the options regarding neurosurgical intervention or comfort measures, and an attempt to understand Ms. Jones' preferences for life-sustaining care given her prior compromised functional status and her new deficits. Developing a plan for Ms. Jones' care relies on the ability of the clinician to communicate her poor prognosis to her daughter and to – in some instances – recommend a plan of action.

Case #3: Mr. Jefferson is a 43-year-old married father of two teenage children who has had a six-month history of progressive walking difficulty. Initially, he began tripping as he was climbing the stairs to his house and he attributed it to fatigue after long hours at work. Several weeks later, he noticed that his left leg began to feel weaker, to the point where he had to lift it with his hands when he got in to the car. He also noticed twitching of his muscles at random times throughout the day and mild changes to his speech. He has not had any difficulty with breathing or with swallowing. An extensive neurological, laboratory, radiological, and electrodiagnostic evaluation was performed and the diagnosis is amyotrophic lateral sclerosis (ALS). He and his wife return to the neurologist's office to hear the results of his testing and to discuss the diagnosis (see table 1 for tips on breaking bad news). He has researched ALS extensively online and he suspects that this is his diagnosis. After the visit, he calls and inquires about when hospice would be appropriate.

Hospice care is specialized medical care for people who have six months or less to live if their disease progresses in its normal fashion.⁴ It is important to note that all hospice care is palliative care but not all palliative care is hospice care. Mr. Jefferson is relatively early in his disease course and he likely would not qualify for hospice services at this time. Guidelines for hospice enrollment for patients with ALS include: severe respiratory dysfunction with a forced vital capacity of 30% of normal, dyspnea at rest, and plans not to pursue tracheostomy and mechanical ventilation, or rapid progression of disease AND malnutrition, or rapid progression of disease AND a life-threatening complication.⁵ Despite the fact that he is not ready for hospice, his question presents a chance to discuss his overall goals of care. Advance care planning can be addressed, specifically his preferences regarding life-prolonging care including percutaneous gastrostomy tube (PEG), non-invasive ventilation, and tracheostomy. Additionally, determining his wishes regarding cardiopulmonary resuscitation would also be appropriate. Palliative care for ALS patients may be provided by his primary neurologist, by an ALS Clinic team, or by a palliative care provider. Referral to hospice can be made when his disease progresses.

Case #4: Ms. Jackson is a 75-year-old woman with advanced dementia who lives at home with her daughter and she has a 24-hour caregiver who assists with all of her activities of daily living. Over the past year, her oral intake has slowly declined and she is barely eating 25% of her meals. Attempts at providing her with protein shakes for nutritional purposes have been unsuccessful. She refuses food and she is combative at times when feeding is attempted. Her daughter is concerned that she is not eating much and that she is losing weight. Her caregiver has had experiences caring for patients with feeding tubes in the past and she wonders whether Ms. Jackson could benefit from one.

Ms. Jackson has significant cognitive impairments that limit her desire to eat. It is understandable that her caregivers are worried about her weight loss despite reasonable efforts to increase her intake. A PEG tube may seem like a practical idea. However, in this patient population, PEG tubes have not been shown to reduce the risk of aspiration, to improve wound healing, to increase one's functional state, or to prolong survival.^{6,7} Negative impacts of feeding tubes include increasing agitation and the potential need for restraints and decreasing human touch. A feeding tube for Ms. Jackson is not recommended. Ms. Jackson's daughter and her caregiver should be encouraged to offer her multiple small meals a day, increasing her daily human interaction, and to offer comfort feeding.

Conclusion:

Caring for patients with neurological diseases requires a working knowledge of palliative care and competency surrounding breaking bad news, discussing prognosis, managing neurological and non-neurological symptoms, setting goals, and providing end-of-life care. There are opportunities to embed palliative care into the management plans for patients at the time of diagnosis and throughout the continuum of their disease. Families may also benefit from the conversations and preemptive planning that arise when the trajectory of disease and the goals of care are established early in the disease process.

Breaking bad news or relaying important medical information: SPIKES Protocol⁸	
S	SETTING the environment
P	Assessing the patient's PERCEPTION
I	INVITATION from patient to give information
K	Giving KNOWLEDGE and information
E	EXPLORE emotions and empathize
S	STRATEGIZE and SUMMARIZE

Table 1

References:

1. World Health Organization. *World Health Organization Definition of Palliative Care* <http://www.who.int/cancer/palliative/definition/en/>. Accessed November 30, 2016.
2. Quill TE, Abernethy AP. Generalist plus specialist palliative care--creating a more sustainable model. *The New England journal of medicine*. 2013;368(13):1173-1175.
3. Holloway RG, Gramling R, Kelly AG. Estimating and communicating prognosis in advanced neurologic disease. *Neurology*. 2013;80(8):764-772.
4. Centers for Medicare and Medicaid. *Medicare Hospice Benefits* <https://www.medicare.gov/pubs/pdf/02154.pdf>, November 22, 2016.
5. Mitsumoto H, Rabkin JG. Palliative care for patients with amyotrophic lateral sclerosis: "prepare for the worst and hope for the best". *Jama*. 2007;298(2):207-216.
6. Finucane TE, Christmas C, Travis K. Tube feeding in patients with advanced dementia: a review of the evidence. *Jama*. 1999;282(14):1365-1370.
7. McCann R. Lack of evidence about tube feeding--food for thought. *Jama*. 1999;282(14):1380-1381.
8. Baile WF, Buckman R, Lenzi R, Glober G, Beale EA, Kudelka AP. SPIKES-A six-step protocol for delivering bad news: application to the patient with cancer. *The oncologist*. 2000;5(4):302-311.