Review

Neurology® Clinical Practice

Neurologists as primary palliative care providers

Communication and practice approaches

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Abstract

Purpose of review: To present current knowledge and recommendations regarding communication tasks and practice approaches for neurologists as they practice primary palliative care, including discussing serious news, managing symptoms, aligning treatment with patient preferences, introducing hospice/terminal care, and using the multiprofessional approach. **Recent findings:** Neurologists receive little formal palliative care training yet often need to discuss prognosis in serious illness, manage intractable symptoms in chronic progressive disease, and alleviate suffering for patients and their families. Because patients with neurologic disorders often have major cognitive impairment, physical impairment, or both, with an uncertain prognosis, their palliative care needs are particularly challenging and they remain largely uncharacterized and often unmanaged. **Summary:** We provide an overview of neuropalliative care as a fundamental skill set for all neurologists. ***Neurol Clin Pract* 2016;6:40–48**

Palliative care is specialized medical care for people with serious illness. With few curative options in neurology, most patients need to learn to live with their disease for a long time before they die with—or from—that disease. The newly proposed concept of distinguishing primary palliative care (skills that all clinicians should have) from specialist palliative care (palliative care specialty skills)[1,2] is particularly relevant to neurology, in which palliative and neurologic care needs coexist.

In this review, we will follow 2 patients and their families through their illness trajectory (1 with amyotrophic lateral sclerosis (ALS) in the outpatient setting and 1 with severe stroke in the inpatient setting) and discuss proposed communication tasks. Rather than providing a comprehensive review of neuropalliative care, this article highlights some common issues faced by neurologists and aims to alert both neurologists and non-neurologists to the variety of palliative care issues in daily neurologic practice. For readers wanting a more in-depth and disease-specific review of palliative care, we refer them to several recent reviews.[3,4]

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Discussing serious news

In the early patient–physician encounters, the physician and the patient (with or without a family member) establish a diagnosis and review treatment options, but they also build a relationship.

Mr. Smith is in clinic to discuss his recent EMG results. He is a 47-year-old father of 2 teenagers who presented with progressive dysphagia over weeks. EMG is consistent with ALS.

Patients with ALS typically present when they are cognitively intact and functionally impaired but independent. Often they have seen a primary care physician, perhaps another neurologist, and read about ALS on the Internet. Patient experiences with the extent of testing, knowledge, and the time from symptom onset to diagnosis vary widely, so the conversation regarding the diagnosis should be specifically tailored to individual patient circumstances. The conversation might start with a question such as "Can you tell me what you know so far about your illness?" When delivering the diagnosis, it may be helpful to first ask for permission (e.g., "Would it be okay if we talked about…?") If permission is granted, the conversation may start by firing a warning shot (e.g., "I have difficult news for you") to give the patient a moment to brace for unpleasant news. Concise statements that confirm the diagnosis should follow the warning shot: "Based on your symptoms and the testing we performed, you have ALS." The delivery should be empathetic and the wording clear; this includes active listening with open-ended questions, reflective statements such as "I wish things were different,"[5] and empathetic responding (e.g., using the NURSE mnemonic[6] [table 1]). Time should be offered for patients and families to process the information before the physician inquires about how much additional information they would like to receive at that time.[7]

Given the poor prognosis associated with ALS, immediate questions from the patient and family may focus on life expectancy ("How long?") and the expected quality of life ("What will my life be like?").[8] These questions provide the clinician with an opportunity to discuss the prognosis and the typical disease trajectory honestly and in detail. Given the gravity of the news, conversations about options for life-sustaining therapies are typically best addressed at a scheduled follow-up appointment (table 2). It is important to broach prognosis early so patients can begin to discuss their preferences with their family members and their physicians. Knowing patients' goals and values will help family members and clinicians align their treatment plans accordingly.

Ms. Brown is 58 years old and was just wheeled into the emergency department after her daughter found her aphasic with right hemiplegia. She is intubated and drowsy and moves her left side but does not follow commands. Head imaging reveals an ischemic infarct involving the entire territory of the left middle cerebral artery (MCA).

Severe acute brain injury is a sudden neurologic catastrophe that catches both patient and family off guard.[9] Treatment decisions need to be made rapidly, including intubation, mechanical ventilation, and brain surgery (placement of ventricular drains or decompressive hemicraniectomy). Most often, the first meeting is between the (neuro-)hospitalist on call and the patient's family. The physician's duty is to tell the family about their loved one's medical situation and to ascertain patient preferences regarding life-sustaining therapies and goals of care. The top priority for the physician going into the meeting, however, is to build a partnership, establish trust, and express empathy, setting the stage for future decision-making.

Rather than "how long?," families of patients with severe acute brain injury typically ask "how likely" the chances are that their loved one will have a meaningful recovery. While helping the family develop realistic expectations, it is important to give them the chance to preserve hope. Anticipatory guidance (i.e., preparing the family step by step for what the future may hold) can help relieve some of the fear and anxiety of a stressful time. As family members make critical decisions in the immediate poststroke period, ideally these decisions

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Table 1 NURSE: A mnemonic to help with addressing patients' emotions with example statements

NAME the emotion	"I can see that you are sad (upset, angry) about…"
UNDERSTAND the emotion	"I understand that these results are not what you were expecting to hear" but also "I cannot imagine what it is like to (X)"
RESPECT the emotion	"I am impressed at how well you have cared for your father all this time"
SUPPORT the emotion	"I will be with you during the entire course of this illness"
EXPLORE the emotion	"Can you tell me a little more about your concerns regarding (X)?"

Adapted from Back et al.[6]

develop through a process of shared deliberation—a process of sharing information, knowledge, and emotions among patients, providers, and family members.[10]

Symptom management

Once a patient's diagnosis is established, clinicians turn their diagnostic expertise toward early detection and management of symptoms. Although the profile of symptoms for each neurologic disease may differ, each disease has symptoms that can be addressed and managed. Beginning with an open-ended statement such as "What symptoms are most concerning to you at the moment?" and following up with probing questions related to any of the palliative care domains of physical, psychological, spiritual, and social needs may allow for a better understanding of the sources of and contributions to an individual's suffering. The following section discusses 4 common symptoms that neurologists will encounter in the inpatient or outpatient setting.

Mr. Smith has lost weight. He admits that swallowing has become harder; he eats more slowly and chokes often.

Dysphagia is one of the most common symptoms in ALS, and it is present in more than half of patients at the time of diagnosis. Nutrition management is a central component of ALS care. Patients with bulbar dysfunction are referred to speech and language therapists for evaluation at the time of symptom onset. Altering dietary consistencies, adding nutritional supplements, and employing maneuvers to assist in swallowing are often recommended.[7,11] Alternative methods of nutrition need to be discussed, including the use of a percutaneous enteral gastrostomy (PEG) tube. Current evidence suggests that enteral nutrition in ALS is effective at stabilizing a patient's weight and confers a survival advantage.[11,12] It is important to discuss the option of a PEG tube early in the disease process so the procedure, if desired, can be performed nonemergently and while the patient has a vital capacity of >50% predicted in order to optimize the safety of the procedure.[11]

Muscle cramps are another common symptom in patients with ALS. They typically occur at night and may be triggered by exercise.[7] Painful cramps that interfere with a patient's quality of life can be managed with exercise and pharmacologic therapies. No clinical trials have demonstrated the efficacy of any specific therapy, but treatment with baclofen or gabapentin may offer symptomatic relief. The US Food and Drug Administration has restricted the use of quinine sulfate given safety concerns.[13]

Ms. Brown remains aphasic but awake and confused. She is attempting to remove lines and needs to be restrained. Her daughter asks what she can do to help.

Delirium develops in ~1 in 5 patients during their stroke hospitalization, with a higher risk in patients who are older and those with preexisting cognitive impairment, severe stroke, or a co-occurring infection. Delirium leads to a longer hospital stay, higher risk of

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Table 2 Suggested triggers for serious illness communication in neurology

General
Age >80 years and hospitalized
Patient or family asks to discuss these issues
Would you be surprised if patient died in next year?
Acute brain injury (stroke, hypoxic ischemic encephalopathy, traumatic brain injury)
Requiring mechanical ventilation
Predicted major functional and/or cognitive deficits requiring long-term assistance
Requiring long-term artificial nutrition
Amyotrophic lateral sclerosis
Routine
Periodic with scheduled clinic visits
Event-driven milestones
Dysphagia and considering artificial nutrition
Hypoventilation and considering noninvasive or long-term mechanical ventilation
Increased symptom burden and/or decreased quality of life
Major decrease in functional capacity
Malignant brain tumors
Routine
Periodic with scheduled clinic visits
Event-driven milestones
Second-line chemotherapy
Increased symptom burden and/or decreased quality of life
Major decrease in functional capacity
Parkinson disease
Routine
Periodic with scheduled clinic visits
Event-driven milestones
Presence of dementia or psychosis
Sleep disturbances, nocturia
Severe orthostasis with syncope
Considerable caregiver strain
Increased symptom burden and/or decreased quality of life
Major decrease in functional capacity
Decreased/insufficient nutritional intake
Other important comorbidity (stroke, cancer, congestive heart failure)
Dementia
Routine
Periodic with scheduled clinic visits
Event-driven milestones
Nighttime wandering
Any hospitalization
Concern for driving safety/giving up car keys
Considerable caregiver strain
Increased symptom burden and/or decreased quality of life
Major decrease in functional capacity
Decreased/insufficient nutritional intake
Other important comorbidity (stroke, cancer, congestive heart failure)

As has been proposed for other medical illnesses,[32] we suggest triggers for serious illness communication in neurology.

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In addition to correctly diagnosing disease, it is equally important to correctly diagnose patient preferences for individual health outcomes and treatments.

institutionalization, and increased mortality.[14] Prevention is key and includes avoidance of sedative or neuroactive drugs, dehydration, and immobilization. General stimulation for thinking, concentration, and memory as well as frequent reorientation and a regular circadian rhythm can help maintain a calming, stable sensory environment. A family member at the patient's bedside may help with orientation and reduce anxiety. Once a patient develops delirium, the underlying cause needs to be explored by considering infectious, metabolic, or toxic etiologies. Treatment needs to include the above prevention measures. Antipsychotic agents may be used sparingly in the short term; benzodiazepines are not recommended. Dexmedetomidine is a new sedative agent used in anesthesia and intensive care that provides sedation without respiratory depression; it has been suggested as a promising agent for managing delirium in the intensive care unit,[15] but no studies have specifically addressed poststroke delirium.

Depression affects up to 1 in 3 stroke survivors, usually within the first year.[16] Poststroke depression (PSD) slows recovery, reduces quality of life, and is associated with increased mortality after stroke.[16] Every stroke survivor should be screened early and regularly for PSD. PSD may be prevented through early psychotherapy.[17] One small study suggested that early use of antidepressants may improve motor recovery as well as mood.[18] Tricyclic antidepressants have shown good effect in reducing depression, but their cholinergic side effects limit their clinical usefulness. The safety profile is more favorable for selective serotonin reuptake inhibitors[19] (e.g., citalopram or fluoxetine).

Aligning treatment with patient preferences

The overarching aim in palliative care (in all of medicine) is to assist the patient and family in establishing goals of current and future treatment (i.e., advance directives) in a process of shared decision-making. In addition to correctly diagnosing disease, it is equally important to correctly diagnose patient preferences for individual health outcomes and treatments. Knowing what the patient wants and values should allow the clinician to make appropriate treatment recommendations to achieve those outcomes.

In both chronic conditions such as ALS and acute conditions such as stroke, the use of a "best case, worst case, most likely case" scenario can offer patients and their families some insight into the range of possible outcomes, along with the most likely or expected outcome, and help weigh a treatment with its potential trade-offs.

These discussions occur at various time points along the disease trajectory. The need to renegotiate goals should also be anticipated when triggers of advancing disease suggest limited life expectancy or excessive suffering, either from the disease itself or from the high burdens associated with life-sustaining interventions. Delaying such discussions can have profound consequences on patients, families, and providers.

Here we provide examples of conversation "triggers" in both the outpatient and inpatient settings (table 2).

Mr. Smith comes back to clinic after a recent hospitalization for a mild pneumonia. He has recovered well but has been sleeping poorly since.

Poor, interrupted sleep may be a first sign of shallow breathing or apnea causing mini-arousals throughout an unrestful night. Respiratory symptoms typically occur when

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As patients live longer with neurologic illness, there is an increasing need to fully integrate palliative care with neurologic care.

the forced vital capacity is <50% predicted.[20] At this point, noninvasive positive pressure ventilation (NIPPV) at night is recommended. NIPPV improves quality of life, prolongs survival, and may slow the rate of respiratory decline in patients with ALS.[7,21–23]

Early discussions with patients and families about preferences and goals will help direct the conversation regarding the treatment plan when NIPPV is insufficient to manage impending respiratory failure. In the United States, fewer than 5% of patients opt for tracheostomy and mechanical ventilation.[24,25] Factors that contribute to this opting out include the financial burden associated with daily maintenance, quality of life concerns for patient and caregiver, and uncertainty about when to remove a patient from the ventilator.[7,20] If a patient elects to undergo tracheostomy and mechanical ventilation, it is critical for clear criteria to be established regarding ventilator withdrawal. Indications for withdrawal may include loss of all motor function, an unacceptable quality of life, or a specified date. Providers should not underestimate the emotional importance of helping patients achieve their goals as it relates to them personally. Being aware of and recognizing signs of mental and physical exhaustion and developing strategies to minimize their effects are part of optimal palliative care.

Ms. Brown's head imaging on day 2 shows worsening of midline shift and impending uncal herniation. Her daughter consents for a decompressive hemicraniectomy because it would increase Ms. Brown's chance to see her first grandchild who is due that summer.

Patients with space-occupying ("malignant") MCA ischemic stroke have a 71% risk of dying, which is reduced to 22% by decompressive hemicraniectomy.[26] Survival, however, comes with a high risk of severe disability. Therefore, this surgery can be recommended only if the patient's individual preference is incorporated and there is clear understanding of how she values the possible trade-offs.

As illustrated in this case, treatment decisions after severe acute brain injury need to be made early, at a time when prognosis is uncertain and includes a wide range of potential outcomes, when the family members are actively grieving and may be uncertain about the patient's values and how they fit in with the proposed treatments and outcomes, and when a trusting relationship often has not been established with medical providers. It is critical that providers communicate honestly, acknowledge the burden of surrogate decision-making, and assure nonabandonment.

Introducing hospice for terminal care

Hospice is specialized care for patients who are in the terminal stages of their illness and likely to die within 6 months or less. Discussing hospice with patients and families is often viewed as a "bad news" discussion because it confronts the reality of death and the fact that curative treatments are no longer working.[27] This should not deter providers from talking about this underutilized service, which can provide optimal end-of-life care to terminally ill people, often in their homes. In a large cohort of "poor-prognosis cancer patients," hospice referral was determined by the physician's characteristics and referral history rather than patient prognosis or values.[28] When talking about hospice care with patients and families, it is helpful to use phrases such as "Hospice provides an extra layer of support for you and your family at home. They can provide medical care to you as your disease progresses and make sure your pain and other symptoms are treated at home." It is critical to prepare patients and caregivers for and guide them through the dying process. Knowing the typical progression of signs and symptoms and knowing how to handle palliative care "emergencies" are core competencies for practicing palliative medicine. A do-not-resuscitate order is not required for hospice enrollment.

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Take-home points

- *The serious news we discuss with our patients and their families should be tailored to the individuals and their disease; always start by exploring what they already know, what they have heard, and what they understand.*
- *Symptom management in neurologic disease requires particular diagnostic skill given patients' often-impaired cognition and should encompass questions related to any of the palliative care domains of physical, psychological, spiritual, and social needs.*
- *Aligning treatment with patient preferences is at the heart of clinical medicine. While helping the patient and family understand the disease from a medical perspective, the provider should explore who the patient is as a person, what the patient's values are, and what goals the patient wants to set.*
- *Hospice services provide palliative care for patients who are expected to live 6 months or less. This underutilized service can provide optimal end-of-life care to terminally ill people, often in their homes.*
- *A central component of palliative care is the team approach, which involves different professions as well as different disciplines.*

Mr. Smith admits to feeling anxious and lost. His breathing is more labored; he is on bilevel positive airway pressure 24 hours a day. He has declined tracheostomy and mechanical ventilation. He asks how long he will live.

Survival in patients with ALS is typically 3–5 years after symptom onset.[7] Although riluzole has been shown to slow disease progression and prolong survival by 2–3 months, the disease is incurable and the management is symptomatic and palliative.[13] For patients who decide to forgo tracheostomy, provide reassurance that their respiratory symptoms will be managed effectively and discuss hospice care.[20]

In the United States, hospice guidelines for ALS include (1) critically impaired respiratory function (forced vital capacity <30% of normal, dyspnea at rest, and a decision not to pursue mechanical ventilation), or (2) the combination of rapid progression of disease and critical nutritional impairment, or (3) the combination of rapid progression of disease and at least 1 life-threatening complication.[29] In discussing hospice with patients and families, it is often helpful to highlight the holistic care that hospice provides to both the patient and the family, including ongoing symptom assessment, support for the patient and family, and spiritual guidance; some hospice organizations also offer bereavement services.

Ms. Brown's family and her providers came to the conclusion that, although a trial of life-sustaining treatment seemed acceptable to her, she would not want prolonged dependence. Her hospitalization was complicated by continued delirium exacerbated by a urinary tract infection. At hospital day 14, she remains unable to communicate and is either confused and drowsy or confused and angry.

The majority of acute stroke deaths are associated with a decision to withdraw or withhold life-sustaining treatment. Having decided on 1 "heroic measure" (in this case decompressive hemicraniectomy) is not necessarily indicative of wanting continued aggressive life-sustaining treatment. Rather, the surgery can be seen as a time-limited trial (i.e., "an agreement between clinicians and the patient/family to use certain medical therapies over a defined period of time to see if the patient improves or deteriorates according to agreed-on clinical outcomes"[30]). If a trusting relationship has been built between the clinician and the family, they may explore alternatives to continued treatment, including no escalation of treatment or withdrawal of life-sustaining treatments. Hospice criteria for patients with stroke lean on functional and nutritional status, including major weight loss, severe dysphagia without artificial nutrition, or hydration and pulmonary aspiration.

The multiprofessional approach

Palliative care requires the concerted effort of multiple members of an interdisciplinary and multiprofessional care team including but not limited to physicians, nurses, spiritual care providers, and social workers. As neurologists cultivate their primary palliative care skill set, they will also learn to identify complex suffering, conflict, or other scenarios that need to be addressed by palliative care specialists and other consultants at an early stage.

For both Mr. Smith and Ms. Brown, palliative care specialists may have been involved in any one of the tasks described above. The neurologist may have wanted assistance with communicating the diagnosis and prognosis, meeting with the family, or helping them with treatment decisions; the family may have needed an additional layer of support with regard to symptom

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management and exploring their own values or those of their loved one in order to decide on the treatment option that would best fit their needs. As with most things in medicine, there is no clear line distinguishing primary from specialty palliative care. Although all clinicians who treat patients with serious illnesses should be capable of delivering primary palliative care,[2] a palliative care consult should be initiated for complex symptoms or existential distress, refractory pain and suffering, or mediation for a difficult family meeting. In addition, a hospital or clinic chaplain or the patient's community-based faith leader may be called on to provide spiritual, emotional, and bereavement support. Social workers assist with discharge planning and locating appropriate community resources while providing patient and family caregiver support.

As patients live longer with neurologic illness, there is an increasing need to fully integrate palliative care with neurologic care. The observation in cancer trials that early palliative care leads to improved quality of life and increased survival[31] has nurtured the idea that (1) neurologists need to develop a primary palliative care skill set,[2] (2) more palliative care specialists need to be trained to handle problems specific to neurologic patients, and (3) neurology needs "a triage system for calling on palliative care specialists when necessary."[1]

REFERENCES

1. Quill TE, Abernethy AP. Generalist plus specialist palliative care–creating a more sustainable model. N Engl J Med 2013;368:1173–1175.
2. Institute of Medicine. Dying in America: Improving Quality and Honoring Individual Preferences Near the End of Life. Washington, DC: National Academies Press; 2014.
3. Connolly S, Galvin M, Hardiman O. End-of-life management in patients with amyotrophic lateral sclerosis. Lancet Neurol 2015;14:435–442.
4. Oliver DJ, Borasio GD, Caraceni A, et al. A consensus review on the development of palliative care for patients with chronic and progressive neurological disease. Eur J Neurol Epub 2015 Oct 1. doi: 10.1111/ene.12889.
5. Quill TE, Arnold RM, Platt F. "I wish things were different": expressing wishes in response to loss, futility, and unrealistic hopes. Ann Intern Med 2001;135:551–555.
6. Back A, Arnold RM, Tulsky JA. Mastering Communication with Seriously Ill Patients: Balancing Honesty with Empathy and Hope. New York: Cambridge University Press; 2009.
7. Elman LB, Houghton DJ, Wu GF, Hurtig HI, Markowitz CE, McCluskey L. Palliative care in amyotrophic lateral sclerosis, Parkinson's disease, and multiple sclerosis. J Palliat Med 2007;10:433–457.
8. Holloway RG, Gramling R, Kelly AG. Estimating and communicating prognosis in advanced neurologic disease. Neurology 2013;80:764–772.
9. Geurts M, Macleod MR, van Thiel GJ, van Gijn J, Kappelle LJ, van der Worp HB. End-of-life decisions in patients with severe acute brain injury. Lancet Neurol 2014;13:515–524.
10. Epstein RM, Street RL Jr. Shared mind: communication, decision making, and autonomy in serious illness. Ann Family Med 2011;9:454–461.
11. Miller RG, Jackson CE, Kasarskis EJ, et al. Practice parameter update: the care of the patient with amyotrophic lateral sclerosis: drug, nutritional, and respiratory therapies (an evidence-based review): report of the Quality Standards Subcommittee of the American Academy of Neurology. Neurology 2009;73:1218–1226.
12. Katzberg HD, Benatar M. Enteral tube feeding for amyotrophic lateral sclerosis/motor neuron disease. Cochrane Database Syst Rev 2011:CD004030.
13. Miller RG, Jackson CE, Kasarskis EJ, et al. Practice parameter update: the care of the patient with amyotrophic lateral sclerosis: multidisciplinary care, symptom management, and cognitive/behavioral impairment (an evidence-based review): report of the Quality Standards Subcommittee of the American Academy of Neurology. Neurology 2009;73:1227–1233.
14. Shi Q, Presutti R, Selchen D, Saposnik G. Delirium in acute stroke: a systematic review and meta-analysis. Stroke 2012;43:645–649.
15. Mo Y, Zimmermann AE. Role of dexmedetomidine for the prevention and treatment of delirium in intensive care unit patients. Ann Pharmacother 2013;47:869–876.
16. Hackett ML, Anderson CS, House A, Xia J. Interventions for treating depression after stroke. Cochrane Database Syst Rev 2008:CD003437.
17. Hackett ML, Anderson CS, House A, Halteh C. Interventions for preventing depression after stroke. Cochrane Database Syst Rev 2008:CD003689.

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18. Chollet F, Tardy J, Albucher JF, et al. Fluoxetine for motor recovery after acute ischaemic stroke (FLAME): a randomised placebo-controlled trial. Lancet Neurol 2011;10:123–130.
19. Creutzfeldt CJ, Holloway RG, Walker M. Symptomatic and palliative care for stroke survivors. J Gen Intern Med 2012;27:853–860.
20. Miller RG, Rosenberg JA, Gelinas DF, et al. Practice parameter: the care of the patient with amyotrophic lateral sclerosis (an evidence-based review): report of the Quality Standards Subcommittee of the American Academy of Neurology: ALS Practice Parameters Task Force. Neurology 1999;52:1311–1323.
21. Bourke SC, Gibson GJ. Non-invasive ventilation in ALS: current practice and future role. Amyotroph Lateral Scler Other Mot Neuron Disord 2004;5:67–71.
22. Kleopa KA, Sherman M, Neal B, Romano GJ, Heiman-Patterson T. Bipap improves survival and rate of pulmonary function decline in patients with ALS. J Neurol Sci 1999;164:82–88.
23. Lyall RA, Donaldson N, Fleming T, et al. A prospective study of quality of life in ALS patients treated with noninvasive ventilation. Neurology 2001;57:153–156.
24. Albert SM, Murphy PL, Del Bene ML, Rowland LP. Prospective study of palliative care in ALS: choice, timing, outcomes. J Neurol Sci 1999;169:108–113.
25. Bradley WG, Anderson F, Bromberg M, et al. Current management of ALS: comparison of the ALS CARE Database and the AAN Practice Parameter. The American Academy of Neurology. Neurology 2001;57:500–504.
26. Vahedi K, Hofmeijer J, Juettler E, et al. Early decompressive surgery in malignant infarction of the middle cerebral artery: a pooled analysis of three randomised controlled trials. Lancet Neurol 2007;6:215–222.
27. Casarett DJ, Quill TE. "I'm not ready for hospice": strategies for timely and effective hospice discussions. Ann Intern Med 2007;146:443–449.
28. Obermeyer Z, Powers BW, Makar M, Keating NL, Cutler DM. Physician characteristics strongly predict patient enrollment in hospice. Health Aff (Millwood) 2015;34:993–1000.
29. Mitsumoto H, Rabkin JG. Palliative care for patients with amyotrophic lateral sclerosis: "prepare for the worst and hope for the best". JAMA 2007;298:207–216.
30. Quill TE, Holloway R. Time-limited trials near the end of life. JAMA 2011;306:1483–1484.
31. Temel JS, Greer JA, Muzikansky A, et al. Early palliative care for patients with metastatic non-small-cell lung cancer. New Engl J Med 2010;363:733–742.
32. Bernacki RE, Block SD; American College of Physicians High Value Care Task Force. Communication about serious illness care goals: a review and synthesis of best practices. JAMA Intern Med 2014;174:1994–2003.

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