**Palliative Care for People with Progressive Neurological Disease: What is the role?**

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Although patients with neurological disease do receive palliative care, particularly those with amyotrophic lateral sclerosis (ALS), the involvement of palliative care may only occur in the final stages of the illness trajectory, when communication and cognition may be affected and the influence of palliative care is limited. In the United Kingdom, neurological patients do not receive palliative care on a regular basis — less than 10 percent of people who die under hospice care have a neurological diagnosis (1).

Why is it that people with neurological disease do not receive the care they need and deserve? Although the numbers of people with any individual progressive neurological disease are low (the prevalence of individual neurological diseases are: ALS, 7/100,000; multiple sclerosis [MS], 110/100,000; progressive supranuclear palsy [PSP], 5/100,000; multiple systems atrophy [MSA], 5/100,000; and Parkinson’s disease [PD], 150/100,000), approximately 12 percent of all deaths are from progressive neurological disease (2). The populations of Western countries are ageing, so there will be increasing numbers of people with symptomatic neurological disease, and many will have comorbidities.

For patients with neurological disease, prognosis is uncertain. They generally survive longer than many patients with cancer: MS has an average prognosis of about 30 years; PD, 11 years, although this can vary, and some patients live for 20 years after diagnosis; PSP, 5 years; MSA, 5 to 8 years; and ALS, 3 to 5 years, although 10 percent may be alive after 10 years (3-5). These patients have many symptoms and issues — physical, psychological, social, and spiritual — and they would benefit from palliative care and a wider holistic and multidisciplinary approach (6, 7). Complex physical and cognitive changes can occur in these patients, and thus their care must be of a different order than that for cancer patients. These complex changes include: psychiatric changes and hallucinations in PD or PSP; behavioural changes in PSP, ALS, or MS; and dementia in PD or ALS (8).A palliative care approach could be helpful in dealing with these issues.

There is increasing use of interventions in the care of patients with neurological disease. These include: gastrostomy insertion for those with swallowing difficulties; noninvasive ventilation or, on occasion, invasive ventilation with tracheostomy for ALS patients with respiratory failure due to respiratory muscle and diaphragmatic weakness (9, 10); and invasive measures, such as deep-brain stimulation or the infusion of apomorphine for those with PD. These interventions are usually intended to palliate symptoms, but there may be limited discussion about their palliative intent, resulting in a lack of clarity in the minds of patients, patients’ families, and professional carers. This, in turn, can give rise to an overly optimistic view of the prognosis, limiting further discussion of the disease progression and impeding preparations for eventual death from the disease. Referral to palliative care may be postponed, because it is viewed negatively and associated with impending death.

Many patients with neurological disease live with progressive deterioration over a period of many years. They have episodes of severe illness followed by low-level recovery. In light of this, it is difficult to determine when initiation of palliative care would be appropriate, especially since many patients and patients’ families associate palliative care or hospice care with imminent death and so are reluctant to consult the multidisciplinary team (11). In addition, palliative care units and hospices fear that if they admit patients with neurological disease, then these patients will block beds and prove difficult to discharge home. However, a UK study demonstrated that the median stay in one UK hospice for someone with ALS was 15 days, a period similar to that for all of the hospice’s patients (12).

Key questions are how to recognize the end-of- life phase and when to refer a patient to palliative care. Some have suggested using triggers to deter- mine when a patient has entered the last year of life (8). The Gold Standards Framework suggests that certain criteria may help with specific clinical indicators for neurological disease and ALS, MS, and PD; it also suggests employing the “surprise question”: “Would you be surprised if this patient were to die in the next few months, weeks, or days?” (13). The Supportive and Palliative Care Indicators Tool **(**SPICT) recommends similar criteria: progressive deterioration in physical and/ or cognitive function, despite optimal therapy; speech problems, with increasing difficulty communicating and/or progressive dysphagia; recur- rent aspiration pneumonia; and breathlessness or respiratory failure (14). These have not been fully evaluated, but they may offer some guidance, and they may facilitate the appropriate involvement of the specialist palliative care multidisciplinary team. In the United States, there is specific pressure to look at this area, as funding for hospice care can depend on a prognosis of less than six months; due to such funding concerns, in 2011, the median length of stay in hospice was 19 days (15). Specific triggers have been identified for determining whether people with progressive neuro- logical disease have reached the end-of-life phase. These were developed through consensus by specialists in neurological care (8, 16). The generic triggers for all progressive neurological disease are: patient request, family request, dysphagia, cognitive decline, dyspnea, repeated infection (in particular, aspiration pneumonia), weight loss, marked decline in condition, and significant complex symptoms (such as pain, spasticity, nausea, psychosocial issues, or spiritual issues). In addition, there are specific triggers for each disease group: for ALS, respiratory failure or increased breathlessness, reduced mobility, and dysphagia; for MS, dysphagia, choking attacks, poor hydration and nutrition, frequent infections, cognitive decline, reduced communication, profound fatigue, and reduced response to the environment; for PD, rigidity, pain, agitation/confusion from sepsis, and neuropsychiatric decline; for PSP, dysphagia, speech problems, weight loss, severe pressure sores, and psychiatric symptoms, as well as medications losing their effectiveness (8, 16). These triggers have been tested in a retrospective study, which demonstrated that the number of triggers increases as death approaches (17).

Neurological patients do not receive palliative care for a number of reasons, but what are the ways forward? There is increasing pressure within the fields of neurology and palliative care for change. International guidelines encourage the involvement of palliative care: the guideline of the European Federation of Neurological Societies on ALS recommends that palliative care be included in the care plan from the time of diagnosis (18), and the US practice parameters on ALS state that “hospice is a major provider of care in the final stages of ALS” (19). In the UK, national reports have recommended including palliative care as part of ongoing care. The National Service Frame- work for Long-term Conditions contains the quality standard, “people in the later stages of long-term neurological conditions are to receive a comprehensive range of palliative care services” (20), and the document *End of Life Care in Long Term Neurological Conditions: A Framework for Implementation* stresses the need to provide palliative care for people with progressive neurological disease (16).

The European Federation of Neurological Societies — now the European Academy of Neurology

— and the European Association for Palliative Care have created a joint task force to look at neurological palliative care. The task force has produced and submitted for publication a consensus review on palliative care for patients with progressive neurological disease. The review considers the need for neurologists to become more aware of palliative care and the need for palliative care services to improve their knowledge and aware- ness of the requirements of neurological patients. Several areas have been highlighted for particular consideration: the early integration of palliative care; the involvement of the full multidisciplinary team; communication with patients and their families, including discussion of advance care planning; symptom management; end-of-life care; carer support and training; and education for all professionals involved in the care of these patients and their families (21).

There is increasing evidence of the need for palliative care and the effectiveness of it as part of the ongoing care offered to patients with progressive neurological disease. Two studies have shown that palliative care improves symptoms and quality of life for patients with progressive neurological disease (7, 22). The involvement of palliative care services will vary according to the disease: ALS patients often require specialist support from diagnosis (11), whereas MS patients may need only limited support throughout the disease progression and perhaps also at the end of life.

There is a need for close collaboration among all the services involved — neurology, rehabilitation medicine, specialist palliative care, and primary care, as well as respiratory medicine or gastroenterology when specific expertise, such as gastrostomy feeding or ventilatory support, is required (18, 23). Such collaboration has been encouraged in several guidelines (18, 19) and by the national organizations. In 1996, the American Academy of Neurology declared that neurologists have to learn and apply the principles of palliative care because many patients affected by neurological disorders die after a long period of illness, and their neurologist is often the principal or the only consulting physician (24, 25). The importance of such collaboration and the need for increased awareness of palliative care within neurology services is also emphasized in the European Academy of Neurology/European Association for Palliative Care consensus review document (21). Palliative care for patients and their families does not have to be offered exclusively by a palliative care team. Any professional multidisciplinary team with the knowledge and skills to support patients can provide this care. The responsibility for its provision can remain with neurology or rehabilitation services, or it can be shared with specialist palliative care services. Sharing this responsibility could cause some tension or conflict, as the various teams may have very different attitudes and goals (26). These tensions must be recognized and addressed to ensure that care is coordinated and does not cause confusion for all involved, particularly patients and their families

(26).

Palliative care services should be made available throughout the disease trajectory according to the specific needs of individual patients and their families (27); for instance, ALS patients may need help coping with their diagnosis, and they may need to be informed about and discuss genetic testing, increasing symptoms, gastrostomy feeding, and ventilatory support at different points as they approach the end of life (18, 27). Thus, the involvement of palliative care may be episodic throughout the disease progression as new issues emerge and as various decisions have to be made. Patients with a number of conditions may have little need for palliative care for long periods, but there will be occasions, especially as the end of life approaches, when the wider multi- disciplinary approach is required (8, 27).

Palliative care is developing. More palliative care is being provided to patients in more disease groups. We must be able to respond to patients with progressive neurological disease, and their families, whether their needs are physical, psychosocial, or spiritual. We all need to take up this challenge if we are to help our patients effectively. This will involve increasing collaboration and interaction among teams and professionals and acknowledging the tensions that this may cause. Our efforts should result in improved care for patients with progressive neurological disease and their families.

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