




End-of-life care of people with long-term neurological conditions

Krishnan Nair ,¹ Mark Lee,² Esther Hobson,³ David Oliver,⁴ Emma Husbands⁵

¹Management, Sheffield Teaching Hospitals NHS Foundation Trust, Sheffield, UK
²Palliative Medicine, South Tyneside NHS Foundation Trust, South Shields, UK
³Neurosciences, The University of Sheffield, Sheffield, UK
⁴Tizard Centre, University of Kent, Canterbury, UK
⁵Palliative Medicine, GHNHSFT, Gloucester, UK

Correspondence to

Dr Krishnan Nair, Neurology, Sheffield Teaching Hospitals NHS Foundation Trust, Sheffield, UK; siva.nair@nhs.net

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ABSTRACT

Guidance and protocols of end-of-life care have been directed towards the care of patients with cancer. It is possible to extrapolate some of these to people with long-term neurological conditions, but there are obvious differences. Neurological conditions have widely different time courses of progression, making the timing of these discussions challenging. The common issues around end-of-life care include knowing when to start discussions, approaching advance planning, managing common symptoms, diagnosing the dying phase, withdrawing life-sustaining treatments, providing support for family and carers and judging how to involve specialist palliative care teams. End-of-life care needs close collaboration between neurology, specialist palliative care and general practice.

BACKGROUND

Between 2001 and 2014, the number of deaths from neurological conditions in UK increased by 39%.¹ Palliative care is defined as ‘an approach that improves the quality of life of patients and their families facing problems associated with life-limiting illness, through the prevention and relief of suffering by means of early identification and impeccable assessment and treatment of pain and other problems, physical, psychosocial and spiritual’.²

- ▶ General palliative care would be provided by all those caring for patients with life-threatening illness, ensuring good communication with patients and families, shared decision-making, goal setting and symptom management.³
- ▶ Specialist palliative care would be provided by teams that would have this as their main activity and have received specialist training.³
- ▶ Neuro-palliative care is a holistic approach to the care of neurological patients with significant disability, complex needs and a potentially shortened lifespan.⁴

The UK National Service Framework defines a long-term neurological condition as ‘disease of injury or damage to the body’s nervous system (ie, the brain, spinal cord and/or their peripheral nerve connections) which will affect the individual and their family in one way or another for the rest of their life’.⁵ Neurologists address diagnosis, modification of disease course and management of symptoms for people with long-term neurological conditions such as Parkinson’s disease, multiple sclerosis and motor neurone disease. During the end-of-life phase of conditions, which in the UK is regarded as the expected last year of life, patients and their families, as well as other care providers, often turn to the neurologists for leadership and guidance.^{6,7} In this article, we discuss supporting people with long-term neurological conditions during the end-of-life phase.

IDENTIFYING THE END-OF-LIFE PHASE

The time course of progressive neurological conditions varies widely between and within disorders. While the average survival of patients with motor neurone disease is often 2–4 years from diagnosis, those with conditions such as Parkinson’s disease and multiple sclerosis may live for decades with significant symptoms.⁸ The unpredictable trajectory of neurological conditions presents a special challenge, especially when choosing the appropriate time to start discussions about the end of life. While patients, their family and carers can have an intimate knowledge of changes in a patient’s condition, they expect healthcare professionals to initiate these conversations. Although ‘end of life’ is defined as the expected last year of life, planning and discussions about it should not be restricted to this specific time frame, especially in long-term neurological conditions, as it is impossible to be accurate about when the last 12 months of life begins. Indeed, conversations around



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Box 1 Indicators of end of life in neurological conditions^{10–12}

- ▶ Rapid deterioration in mobility.
- ▶ Increasing dependence for personal care.
- ▶ Rapid decline in cognitive function.
- ▶ Loss of speech.
- ▶ Difficulty in swallowing.
- ▶ Loss of appetite and weight loss.
- ▶ Increasing fatigue.
- ▶ Recurrent aspiration pneumonia.
- ▶ Respiratory failure.

future and end-of-life care can be appropriate from the time of the diagnosis, and so professionals should be prepared to have these conversations; a prime example would be conditions where cognitive decline can be expected.

There are no accurate ways to ‘diagnose’ the point at which someone has only 1 year left to live but various clinical indicators and assessment tools can be incorporated into practice. The Gold Standards Framework suggests using the simple ‘surprise’ question devised by Marie Curie: in this, healthcare providers ask themselves, ‘Would you be surprised if this patient died in the coming 6–12 months?’⁹ If the answer is ‘No’, this is an opportune time to start to plan for end-of-life care. The Supportive and Palliative Care Indicators Tool, the Gold Standards Framework and the UK End-of-Life Programme give overarching and specific clinical indicators relating to identifying people who are entering into end-of-life phase. These could be used to trigger discussions over end of life (box 1).^{10–12}

Awareness about these indicators will promote identification of the end-of-life phase that in turn enables a proactive approach to begin discussions about the patient’s priorities and planning for their care and support during end of life.

HOW TO TALK ABOUT PROGRESSING CONDITION/END-OF-LIFE PHASE

Identifying triggers (from the list above) gives an opportunity to open discussions with the patient and those important to them about their condition. The nature of many neurodegenerative conditions is such that capacity to make decisions can become impaired, and so early discussions give the patient a better chance to be included in decisions about their end-of-life care. The prognosis may guide the urgency of discussions but not everyone wants to engage in considering the future or wants to know about their expected prognosis. We must therefore explore the patient’s readiness for these discussions sensitively.

There are many techniques for talking about these challenging issues. Open questions allow clinicians to assess the person’s current understanding and to try to gauge whether they are ready for further conversations. Structuring consultations may help and there are many communication tools to support this: one example is the REMAP model (table 1).¹³

These conversations can be difficult, and patients and carers need support during the process. It may be necessary to arrange further reviews, maybe with a nurse specialist or members of the wider multidisciplinary team. No matter how thorough the conversations are, the patient needs a point of contact to call for further information and additional support.

Although these discussions should be individualised, there are some key things to explore: milestones in disease progression, the shifting focus from disease-modifying treatments towards management of symptoms and advance care planning.

HOW TO BEGIN PLANNING END-OF-LIFE CARE

Having recognised that the patient may be in the end-of-life phase (ie, the last year of life), healthcare professionals should sensitively offer opportunities to consider planning for future care, including at the end of life. For some patients, it may be more effective and

Table 1 REMAP model for end-of-life communication¹³

Reframe	<ul style="list-style-type: none"> ▶ Given your diagnosis, it is important we talk about things now, while you can express your priorities. ▶ We are in a different place because of what’s happened, but we know that for all of us, thinking about the future is important.
Expect emotion and empathise	<ul style="list-style-type: none"> ▶ It is hard to deal with all of this. ▶ I know it will be upsetting to think about situations before they are happening. ▶ I can imagine there are things that worry you about the future?
Map the future	<ul style="list-style-type: none"> ▶ Given the situation, are there things that you want to do? ▶ When you think about the future, what concerns you? ▶ Are there situations that you fear? Some people worry about being kept alive when they are at a point where they can’t communicate, for example.
Align with the patient’s values	<ul style="list-style-type: none"> ▶ As I listen to you, it sounds like the most important things for you are...
Plan next steps	<ul style="list-style-type: none"> ▶ It has been really helpful to hear what matters to you. These are the things that I think we can document to make sure that we manage situations in the future to try and ensure your priorities are achieved.

Table 2 Advance care plans/processes

Advance care plan	Legal status	Description
Advance decision to refuse treatment—England and Wales*	Legally binding if valid and applicable	State clearly, in specific circumstances, what treatments the person would not wish to receive, such as cardiopulmonary resuscitation, ventilatory support, antibiotics, gastrostomy/enteral feeding, hospitalisation.
Advance directive—Scotland*	Not legally binding	This allows someone to record treatments that they would want to decline in the event they lose capacity. While not legally binding, it should be referred to where a decision needs to be made.
Lasting power of attorney for health and welfare decisions—England and Wales† (referred to as welfare power of attorney in Scotland†)	Legally appointed role	The person appoints another person/s to support decision-making around their health/welfare needs if they no longer have capacity to do so. The appointed attorney/attorneys can accept or decline treatments offered. Note this role is distinct from lasting power of attorney for finance.
Independent mental capacity advocate	Independent role	An independent mental capacity advocate is an advocate appointed to act on someone's behalf if they lack capacity to make decisions and they have no family/friends to take part in best interests discussions.
Emergency healthcare plans	Advisory Not legally binding	Personalised plans in case of a future ANTICIPATED emergency when the patient may be unable to make an informed decision. ReSPECT process recommended by the Resuscitation Council UK. ¹⁴ Many areas also used treatment escalation plans.
Advance statement	Advisory Not legally binding	Sets out in general terms how the patient would like to be cared for. It is a record the person's wishes during the end of their life, including who they would like to be with, where they would like to be cared for, music, dietary needs, cultural or religious wishes and funeral plans. Options for donation of tissue—brain bank. These should still be taken into account when making decisions.

*No equivalent to the above in Northern Ireland.

†No equivalent in Northern Ireland. Power of attorney for finance can be appointed.

ReSPECT, Recommended Summary Plan for Emergency Care and Treatment.

acceptable to start with planning for the short term (next week/next month), before leading on to the end-of-life discussions. These conversations are often evolving discussions, which therefore are unlikely to

take place in one sitting or one clinic appointment. Promptly communicating the content of any initial conversations to other people supporting that person can allow them also to progress discussions. When

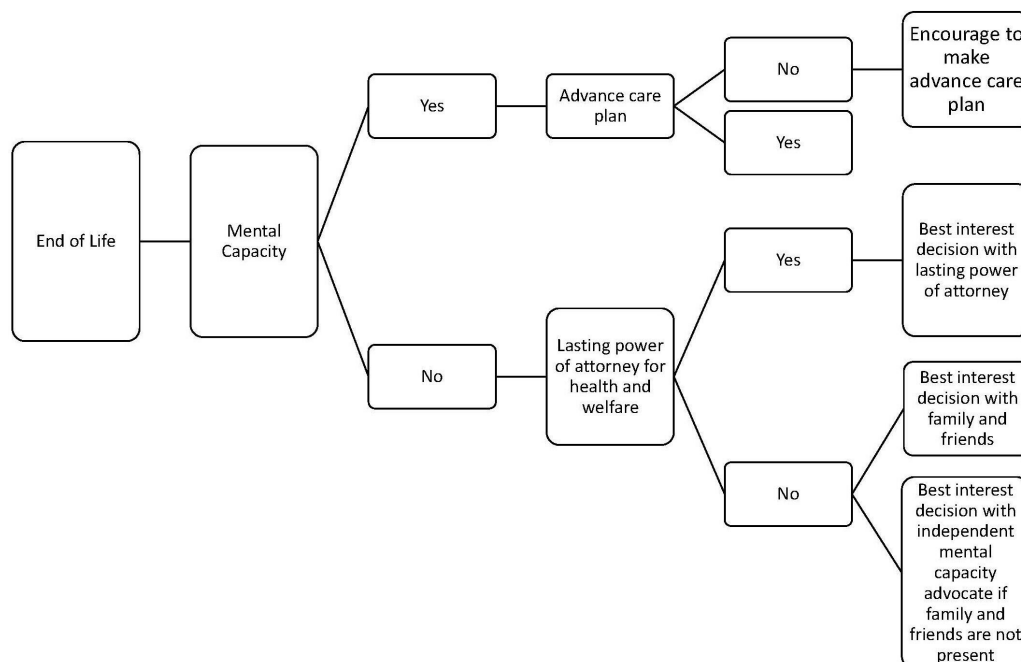
**Figure 1** Advance care planning: the process.

Table 3 Management of Parkinson's disease and seizures during end of life

Parkinson's disease	<ul style="list-style-type: none"> ▶ Prognosis thought to be weeks consider: <ul style="list-style-type: none"> – To minimise rigidity and /or prevent symptoms of dopamine withdrawal, consider a dopamine agonist patch (eg, rotigotine) or subcutaneous apomorphine if oral route is lost and no enteral route available. – If they have a percutaneous endoscopic gastrostomy or nasogastric tube, dispersible Madopar can be given. ▶ Prognosis likely days: the recommendation would be to focus on standard anticipatory medications, using midazolam as a muscle relaxant/opioid for pain. Even in last days of life, rotigotine is effective in preventing distress due to withdrawal symptoms. Common symptoms for relief during end of life include secretions, pain, agitation and fever.²⁷ ▶ Choice of anti-emetic may need to be considered with caution due to risk of increasing rigidity. Generally avoid metoclopramide/haloperidol and use levomepromazine/cyclizine with caution. Ondansetron and domperidone are safe.
Epilepsy	<ul style="list-style-type: none"> ▶ Cover risk of seizures if oral route is lost and no enteral route with recommendation for medication via a syringe pump. Midazolam remains the most common choice of anti-seizure medication in the dying phase with starting doses of 20–30 mg over 24 hours usually recommended. Other anti-seizure medications such as levetiracetam, phenobarbital and sodium valproate can be given via subcutaneous infusion but issues such as availability in community settings may focus the advice. ▶ An intravenous infusion may be appropriate during the dying phase for someone dying acutely but is less likely to be used in an expected/anticipated death.²⁸

someone is identified as likely being in their last year of life, the general practitioner needs to be informed to ensure that the patient is added to any local registers.

An overarching goal of all discussions, no matter who has prompted them, is to understand better what matters to that individual; knowing what they most value and what they most fear forms a useful framework for care planning. This is the model used in Recommended Summary Plan for Emergency Care and Treatment approach.¹⁴

People do not always know what services and supports are available to them. They may have a preference over their desired place of death; where possible, it is important to share their views with those with whom they live. Hospital-based teams may not be familiar with community services, especially if a patient has come to a tertiary clinic, therefore it is important to involve primary care and other local services in these discussions.

OPTIONS FOR ADVANCE CARE PLANNING

The outputs that may arise from these discussions are often covered in the 'catch-all' term 'advance care plan', which may include both legally binding and legally non-binding elements (table 2). For those who lack capacity to discuss their treatment plans, it is necessary to involve a lasting power of attorney, where they have been appointed. Decisions that are made through a best interests process should involve any appropriate family/friends, although the decision ultimately lies with the lead clinician. In the absence of family or friends, an independent mental capacity advocate should be arranged (figure 1).

The treating team should explore whether any advance care plans are already in place, particularly those that are legally binding. These can be used to inform discussions and decision-making when the patient loses the capacity. By having a better understanding of a patient's preferences and goals, clinicians can ensure decision-making is guided by the patient's priorities, for example, how much value to place on quality of life versus life-sustaining treatment. Any

advance care plans should ideally be accessible to all care teams, including ambulance services, the local emergency department and general practitioners. There will be different ways to add such alerts in all areas, often through electronic patient record systems such as Summary care record or Electronic Palliative Care Co-ordination Systems, although there can still be a heavy reliance on people seeing the paper copy.¹⁵ Knowledge of systems being used locally can be sought from general practitioners and specialist palliative care teams.

REQUESTS FOR EUTHANASIA AND ASSISTED SUICIDE

People may wish to discuss an assisted death when addressing future care planning. Euthanasia (where a clinician administers medication with the intention of causing death) and physician-assisted suicide (where a clinician provides medication for self-administration by the patient that will result in death) are both currently illegal in the UK.¹⁶ These processes are legal in a small but growing number of countries. An expression of a desire to hasten death is often a cry for help and a desire to regain some control. Where patients make such requests, clinicians should always undertake a holistic assessment of any physical/emotional/psychological and emotional suffering and address these where possible, including assessing for and treating depression. A referral to specialist palliative care services for additional support may be indicated, allowing further discussion about an individual's needs and priorities. Patients need an honest and open discussion with someone who has capacity about the options that are open to them wherever they live. In the UK, this might include completion of an advance decision to refuse treatment (or regional equivalent) and the option to withdraw life-sustaining treatments. The British Medical Association has produced guidance on how to respond to a patient request for assisted dying.¹⁷ The House of Lords has been debating changes in the law around physician-assisted suicide and euthanasia, and so guidance may change in the coming years.



Figure 2 Brief targeted specialist palliative care intervention.

WITHDRAWING LIFE-SUSTAINING TREATMENTS

Withdrawal of treatments often poses complex ethical dilemmas. Ethically and legally withholding of treatments and withdrawal of treatments are usually seen as being the same. In these situations, it can be very helpful to involve the specialist palliative care team. In neurodegenerative conditions, the most common treatments that can be withdrawn are non-invasive ventilation or artificial nutrition/hydration.

Patients with capacity can request withdrawal of these treatments at any time. A patient may have documented in an advance decision to refuse treatment the point at which they would no longer wish for life-sustaining measures to be continued. With someone lacking capacity, if they have an appointed lasting power of attorney for health and welfare, the attorney(s) can request withdrawal on their behalf. Where someone lacks capacity but has no appointed lasting power of attorney, decisions would be made using a best interests approach.

If withdrawal is agreed to be the right approach, planning for the event should be undertaken promptly; this will probably be led by specialist palliative care and specialist respiratory teams. Neurology services may not be the lead but are likely to be part of multidisciplinary team discussions, particularly with a best interests decision.

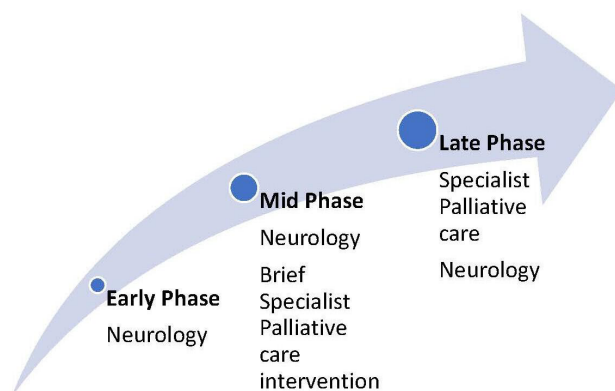


Figure 3 A model incorporating neurology and specialist palliative care in the management of long-term neurological conditions.

Guidelines on supporting withdrawal of ventilation and nutrition/hydration can provide information on the use of medications to ensure comfort.^{18–20}

It is important to plan support for the multidisciplinary team before, during and after any withdrawal of treatment, with an opportunity to discuss the ethical dilemmas and clarify the legalities of the process. If there is still uncertainty around a decision, local ethical committees may be able to provide an independent review.

HOW TO MANAGE SYMPTOMS IN A DYING PERSON

People with neurodegenerative conditions can deteriorate and die suddenly, sometimes in their sleep. The potential for sudden and therefore unpredictable, if not unexpected death, reinforces the importance of advance care planning to avoid inappropriate or unwanted interventions, such as attempted cardiopulmonary resuscitation. A more predictable decline is also possible. [Box 1](#) highlights some of the indicators to look for.

In the event of diagnosing dying as imminent, likely within days to weeks, enacting any agreed care guidance can support next steps. The neurologist's role here may be to confirm the assessment of primary care or other teams, and to provide medication advice, rather than seeing a patient if they are being cared for outside of the acute hospital. National Institute for Health and Care Excellence (NICE) guidance encourages open communication with a patient and those important to them; such communication is important in guiding people through changes and helping them to understand what will probably happen and what 'expected dying' might look like.²¹

Anticipatory prescribing for the dying phase is largely a standardised process for any health condition and local guidance will usually be available. This includes recommending medications by subcutaneous injection—recognising that the oral route is often lost—for common symptoms such as pain, breathlessness, agitation, secretions and nausea.²² If the patient can no longer take their usual medications for spasticity or pain, it is important to consider using other preparations to try to minimise their burden. For more complex medication regimens, it is best to liaise with specialist palliative care teams; there may also be disease-specific medication considerations ([table 3](#)).

Hydration and nutrition can be a hugely concerning issue for families. When someone who would usually take food/fluid orally is dying, their desire to eat and drink naturally diminishes as they deteriorate. It is important to reassure those important to the person that the patient is not starving or dehydrating to death. If there are concerns around symptoms of dry mouth or thirst, it can be worth considering a trial of

Table 4 Web-based resources on end-of-life care

Topics	Web-based resources
Advance care planning	https://www.resus.org.uk/respect https://www.gmc-uk.org/ethical-guidance/ethical-guidance-for-doctors/decision-making-and-consent https://www.mariecurie.org.uk/help/support/terminal-illness/planning-ahead/advance-care-planning?msclkid=31aa07774b7c143ebdd158ae9b0d9941 https://www.gov.uk/power-of-attorney https://www.nhs.uk/conditions/end-of-life-care/advance-decision-to-refuse-treatment/ https://compassionindying.org.uk/choose-a-way-to-make-an-advance-decision-living-will/
Withdrawal of hydration and nutrition, and withdrawal of ventilation	General Medical Council https://www.gmc-uk.org/ethical-guidance/ethical-guidance-for-doctors/treatment-and-care-towards-the-end-of-life https://apmonline.org/wp-content/uploads/2016/03/Guidance-with-logos-updated-210316.pdf Association for Palliative Medicine. Withdrawal of assisted ventilation at the request of a patient with motor neurone disease: guidance for professionals. Association for Palliative Medicine of Great Britain and Ireland, 2015. www.NIV4MND.co.uk —a website which provides evidence-based guidance to clinicians working with people with motor neurone disease on issues like withdrawal of non-invasive ventilation. Copyright NIV4MND 2020
Assisted dying	British Medical Association. Responding to patient requests for assisted dying: guidance for doctors. https://www.bma.org.uk/media/1424/bma-guidance-on-responding-to-patient-requests-for-assisted-dying-for-doctors.pdf . UK General Medical Council. https://www.gmc-uk.org/ethical-guidance/ethical-guidance-for-doctors/when-a-patient-seeks-advice-or-information-about-assistance-to-die

subcutaneous fluids with careful review of response and stopping after 48 hours if there is no benefit. Routine mouth care is widely recognised as a crucial part of care of the dying and should be encouraged, as well as offering sips of fluid as appropriate.

For people who are artificially fed/hydrated, it can be difficult to change regimens, even if dying is recognised. It is important to discuss the usual processes as above and to try to mirror these by reducing and sometimes stopping enteral regimens. It is important to reinforce that this is not causing that person to starve/dehydrate or even causing them to die, because the dying process is already happening.

It is important to address spiritual and religious needs appropriately; ideally these will already be known. Families/carers, including informal carers who may have known a patient for a long time, should be offered access to appropriate emotional support. The death of a person with a longstanding neurodegenerative condition can mean a loss of their caring role, possibly their income and even where they live, and so the process of bereavement can be complex.

HOW TO INVOLVE SPECIALIST PALLIATIVE CARE SERVICES

Most patients' end-of-life care can be managed by primary care teams and an important first step is to work in partnership with primary care services.

People often associate specialist palliative care only with death and dying but it is also about supporting and improving quality of life through planning and symptoms management. The involvement of specialist palliative care in neurodegenerative conditions will be

guided by the patient's needs; they do not need to be dying or even in the last year of their life to benefit from specialist palliative care input and timely discussions with local teams can be very helpful. They may already form part of multidisciplinary team clinics, where their introduction is seen as part of the routine care. Referrals to specialist palliative care could be considered in two circumstances.

Referrals for brief targeted palliative care interventions

Specialist palliative care teams can provide input for distressing physical symptoms such as pain, breathlessness, drooling and constipation that usual measures are not improving.^{7 23 24} They may offer assessment and advice; ongoing care can then continue with the neurology team with further referrals made as required (figure 2). They can help in managing challenging ethical dilemmas or complex needs of the patient and/or family. For example, in motor neurone disease, early referral to specialist palliative care may be useful to discuss interventions, such as gastrostomy or ventilatory support (figure 2).

Referrals for end-of-life care intervention

Patients who are approaching the end of life can benefit from palliative care interventions for the following^{23–26}:

1. To address complex/unmet physical, psychosocial and spiritual needs.
2. For referral to local hospice services that may have in-patient and day hospice/family support/bereavement services.
3. For withdrawal of life-sustaining interventions.

Key points

- ▶ Clinicians should support patients and family to explore advance care planning in a timely fashion, especially if the disease risks them losing mental capacity to make decisions.
- ▶ Clinicians should try to understand a person's priorities to focus care and ensure that unwanted or inappropriate treatments are not offered; this might include avoiding hospital admission if this is their preference.
- ▶ Brief, targeted involvement of specialist palliative care teams can help in managing refractory symptoms, even well before the end-of-life phase.
- ▶ Psychological and spiritual issues may greatly impact quality of life at the end of life.
- ▶ Families and carers should be offered support throughout their loved one's illness, including into bereavement.

Further reading

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Figure 3 shows a model incorporating neurology and specialist palliative care in managing long-term neurological conditions.

CONCLUSIONS

All the inherent uncertainties of progressive neurological conditions make management of end-of-life care complex. Neurologists play an important role in helping patients, families and other healthcare professionals to navigate the end-of-life phase. Collaboration with specialist palliative care teams can help with advance care planning, managing refractory symptoms, withdrawing life-sustaining treatments, dealing with psychological and spiritual issues, and supporting families who are struggling to cope.

Table 4 provides hyperlinks for web-based learning resources on this topic.

Twitter Esther Hobson @drestherhobson

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ORCID iD

Krishnan Nair <http://orcid.org/0000-0002-4004-2315>

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