Ethics – Issues of care in progressive neurological disease

Introduction
Ethics can vary depending on culture and with language; however, the fundamental ethical principles of autonomy (respecting the patient’s view), beneficence (doing good), non-maleficence (not doing harm) and justice (rights and responsibilities of patients, families and professionals and the wider effects on others) underpin the care of all patients, including those with progressive neurological disease.

Disease progression in many neurological disorders is disabling. Disorders with variable prognoses include stroke, brain injury and multiple sclerosis (MS), for which disease-modifying treatments are available. For neurological disorders with no curative treatments (e.g. amyotrophic lateral sclerosis (ALS), Parkinson’s disease, multiple system atrophy (MSA), progressive supranuclear palsy (PSP), dementias), available treatments may slow progression and alleviate symptoms, but the patient will deteriorate over time and die from the disease. Planning for end of life is very important for the patient, in order for their views to be heard and for clear decision-making. This may be difficult and complex for all involved and can cause conflict.

KEY MESSAGES

- The fundamental ethical principles of autonomy, beneficence, non-maleficence and justice underpin the care of patients with progressive neurological disorders
- Planning for end of life is very important for the patient, so that their views can be heard and for clear decision-making
- Discussion of prognosis, treatment options, how the disease may progress over time and likely symptoms should be initiated when communicating the diagnosis
- It is preferable to make early decisions regarding interventions for potential future deterioration (difficulty swallowing, increased risk of aspiration, respiratory failure or cognitive change) rather than at the time of crisis
- Palliative care provides relief from pain and other distressing symptoms and affirms life, while viewing dying as a normal process; it intends neither to hasten nor postpone death, and integrates the psychological and spiritual aspects of patient care
- There is a role for palliative care across the trajectory of neurological disorders
- Recognition of deterioration over the last months and weeks, and diagnosis of the start of the dying phase, allow for appropriate end-of-life care
- In the context of public health priorities, rare diseases have a massive impact on life years lost with direct and indirect economic implications
- The majority of rare diseases are neurological, of which only 5% can currently be treated with approved therapies.

This report was made possible by an unrestricted educational grant from Cipla. The content of the report is independent of the sponsor. The experts participated voluntarily.

This report is from a joint congress of the Neurological Association of South Africa (NASA) and the African Academy of Neurology (AFAN), hosted in February 2019, furnished insights into many aspects of ethical care. Professor David Oliver (UK) shared his experiences on the needs, wishes and fears of the patient, family and healthcare team along the path from diagnosis to end-of-life care. The complex question of budget allocation for research into rare diseases was explored by Dr Johan Smuts (South Africa).
Communicating the diagnosis

The diagnosis should be given by someone who knows about the disease. Prognosis, treatment options, how the disease may progress over time and likely symptoms are all topics that should ideally be initiated in this conversation; it should also include the wishes, fears and concerns of the patient, their family and healthcare providers. This very difficult conversation requires time and empathy.

Decision-making

In the context of neurological disease, the patient is faced with many different decisions concerning treatment, interventions and end of life. These decisions need to be made as early as possible, according to the wishes of the patient and family. The benefits of early decision-making include increased discussion and involvement of the patient, family and healthcare providers so that options can be carefully considered; while cognitive impairment is less advanced, information can be given gradually with slower decision-making. At intervention or end of life, it may be too difficult to discuss options due to communication limitations and cognitive change, or the decision may have to be made in a crisis without time for consideration.

At diagnosis, discussion of treatment options, benefits vs risks, patient/family conflicts, costs and further care issues can be difficult. There is usually a general reluctance to discuss the future – patient fears, varying views within families and concerns of healthcare professionals make this a complex conversation. Often at diagnosis, knowledge and understanding of the disease is limited and modulated by fear/anger/delay/family pressure. The patient may have too little information or already have too much, such as with genetic disorders where families have experience of disease deterioration. Ideally, the person needs to be given information in balance with their ability to cope with that information.

The patient is facing multiple fears - anticipated physical (mobility, speech) and cognitive changes, fear of the disease from previous experience or from media, and fear of death and dying. Within families, there will be concerns regarding role changes, emotional changes, finances and their own fears of the disease, death and dying. Because these discussions can be complex and daunting, over-optimistic expectations on the part of the patient, family and professionals may lead to little discussion of options.

What are the considerations when discussing interventions?

Swallowing

As disease progresses, the patient may have difficulty swallowing and an increased risk of aspiration. The issues of feeding and nutrition can be complex depending on the personalities involved, as the importance of food and its cultural aspects vary greatly depending on the patient and their family situation. Loved ones may nurture through feeding, the patient may ‘live to eat’ and may find it difficult not being able to enjoy the taste and social aspects of food. Complex decisions may be necessary when discussing possible interventions (use of a nasogastric tube, gastrostomy, intravenous fluids/feeding). These discussions will continue throughout the disease progression, and particularly towards the end of life, where there may be discussion of the continuation or withdrawal of feeding and/or hydration.

Respiratory failure

Respiratory support may include ventilatory support (non-invasive ventilation, tracheostomy) and use of opioids. Discussion may need to take place earlier in the disease progression so that all are aware of the patient’s wishes in the case of an emergency or sudden deterioration. These discussions about ventilation should include inevitable deterioration, even though breathing may improve.

Withdrawal of ventilation when the person does not want to continue with this intervention is ethical and legal in most countries, but this can be stressful for all concerned. Ethically, withdrawal of treatment is based on the patient’s best interests. The aim of all care is to ensure the comfort of the patient and respect their wishes. If the patient has capacity, ask them what they desire – they have

“…we need to have hope for cure, but we always have to have hope for care…”

Professor David Oliver
autonomy; but they cannot request a treatment which will not be helpful. If the patient has no capacity, it may be necessary to act in their ‘best interests’, taking into consideration the previously expressed wishes of the patient and the wishes and feelings of the family and team involved.

**Cognitive change**

Cognitive changes may occur in neurological disease and it is essential, when decisions are made, to ensure that the person does indeed have the capacity to make the decision. As communication and cognition deteriorate, it is important to plan ahead. This requires discussing all the options, benefits and risks and coping with conflicts between the patient, family and healthcare workers within a multidisciplinary team. Advance care planning can be helpful in resolving these complex issues and decisions and help to reduce conflicts within families. Options include an advance statement/directive, the agreement of a proxy decision-maker, will, funeral plans, and wishes for the place of care and death. The legality of advance care plans may vary from country to country.

**What are the considerations when discussing end of life?**

Patients and families may have concerns and fears of distress such as choking or pain at the end of life, loss of control or place of death. Ethical issues at end of life include decisions about life-prolonging medical treatments (including intravenous fluids, enteral or parenteral feeding, antibiotics, CPR, ventilation) and withdrawal of treatment, nutrition and hydration.

Assisted dying is increasingly discussed throughout the world. There is a distinction between euthanasia - a doctor intentionally killing a person by the administration of drugs, at that person’s voluntary and competent request - and physician-assisted suicide, whereby a doctor intentionally helps a person to commit suicide by providing drugs for self-administration, at the person’s voluntary and competent request. These actions are very different from the withholding or withdrawal of treatment, usually at the patient’s request. Communication skills are key in response to desire-to-die statements and it is important that there is an emphasis on understanding why such requests are made, rather than acting or resolving. If the patient is seeking assistance to end their life, always explore current feelings, suffering and distress (including considerations of suicide), rather than giving yes/no answers.

Terminal sedation for treatment of distress at the end of life is rarely used, usually only when distress cannot be managed by other means. Therapeutic/palliative sedation in the context of palliative medicine entails the monitored use of medications intended to induce a state of decreased or absent awareness (unconsciousness) in order to relieve the burden of otherwise intractable suffering in a manner that is ethically acceptable to the patient, family and healthcare providers.

**What is the role of the neurologist in the terminal stages of disease?**

**Palliative care**

The World Health Organization describes palliative care as an approach that improves the quality of life of patients and their families facing problems associated with life-threatening illness, through the prevention and relief of suffering, early identification and impeccable assessment and treatment of pain and other distressing symptoms, affirm life and maintain the view that dying is a normal process, intending neither to hasten nor postpone death, while integrating the psychological and spiritual aspects of patient care. There is a role for palliative care across the disease trajectory of neurological disorders (Figure 1), from diagnoses such as ALS and severe stroke, at times throughout disease progression (Parkinson’s disease, MSA, PSP, corticobasal degeneration (CBD), MS) and at the end of life (any disease).
Assessment

Most patients with neurological disease will have many symptoms including pain, dyspnoea, dysphagia, incontinence, constipation, speech problems, delirium and anxiety/depression. It is important to ask the patient about specific symptoms, as many will not spontaneously talk about them otherwise. Pain is common in neurological diseases - ALS (76%), MS (82%) and Parkinson’s (62%). In dementia patients it is difficult to ask if there is pain and so proxy assessments are required. The causes of pain are variable - musculoskeletal, cramp, skin pressure, MS paroxysmal pain, constipation or urinary tract infection. Skin pressure pain can be managed with positioning and opioid medication.

Psychosocial and spiritual aspects of care for the patient and family are equally important. Spiritual aspects of care are not necessarily religious, but include the deeper meaning of life, fears of dying and fears of death. It is the role of multidisciplinary professionals to listen to the patient and family, co-ordinate the appropriate care and be open to all issues.

End-of-life care

When does end-of-life care start? Who is involved? How do we discuss this with patients? How do we improve care?

End-of-life care in neurological disease is a challenge, requiring collaboration between neurology, rehabilitation medicine, general medical care, primary care and specialist palliative care. The UK end-of-life care pathway (Figure 2) is helpful.

Recognition of deterioration over the last months and weeks, and diagnosis of the start of the dying phase, allow for appropriate management of interventions, medication, and carer and family support. Triggers for end-of-life care include patient/family request, dysphagia, cognitive decline, dyspnoea, repeated infections, weight loss and marked decline in the patient’s condition. End-of-life care triggers in specific neurological diseases are outlined in Table 1. The use of these triggers can be of help in recognising when a patient is deteriorating and allow the emphasis of care to be more on the support of the patient and family, the management of symptoms, such as pain and breathlessness, and the preparation for the end of life – including discussion of the concerns of patient, family and team and provision of medication for use if there is a sudden deterioration.
The ethics of budget allocations for rare diseases

In the context of public health priorities, rare diseases have a massive impact on life-years lost (double that of diabetes), economic implications of treatment and the indirect cost of family carers giving up employment. Rare diseases are both diagnostically difficult and often therapeutically problematic. Depending on the definition used, there are more than 7000 rare (orphan) diseases, of which approximately 5000 are neurological. Currently, only 5% of these can be treated with approved therapies.10

The US Orphan Drug Act (1983) was enacted to encourage the development of drugs for rare diseases through tax incentives, research subsidies and extended patient protection for companies that engaged in orphan drug research. The European Union (and Japan) have since enacted similar incentives and have included a greater number of conditions, including tropical diseases, that must be life-threatening or chronically debilitating. Arising from these incentives, at least 575 orphan drugs were approved by 2017. Also of consequence, the era of genomics has led to many common disorders being subdivided into genetically distinct ‘rare’ diseases that can then benefit from the incentives of orphan status. Many
of these drugs eventually make their way into the non-rare disease arena.11,12

Rare diseases, by the very nature of being rare, often have an unknown actiology, making therapeutic targeting difficult. The affected population is also very small, limiting clinical trials. The pharmaceutical industry has an obligation both to research and development, as well as to investors. Given the need to balance both research costs (including drugs that never get to market) and a reasonable margin of profit against a very small pool of patients, these therapies become extremely expensive and inaccessible to most. Where does the cost balance lie? How well does the drug work (quality-adjusted life years)? This threshold requires evaluation with each agent.

Spiralling costs of treatment for rare diseases have highlighted the need for a different approach to research and development. Pharmaceutical companies cannot be expected to pay for something which may potentially deliver no profit, while the public sector cannot pay for something that then becomes inaccessible due to price; so there needs to be a balance when it comes to risk-sharing. Strategies to address cost threshold include research and development being done by academics with public funding and not by drug companies; and transparent, collaborative patient registries being made available. Performance-related pricing is another option, so it is important that the use of these agents is monitored very carefully. Stakeholder investment can also provide patients for trial and assist financially (e.g. Cystic Fibrosis Foundation). A promising drug should be introduced into the market earlier, at a significantly reduced price under close supervision, until it has proven its worth.13,14

References

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