

Ethical issues in neurological palliative care

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Summary. Palliative care for people with neurological disease may be appropriate at any stage in the disease progression, depending on their needs – physical, psychosocial and spiritual. The care for this patient group is often complex, with many issues faced by patients, families and professionals. There are often ethical issues to be faced – communication of the diagnosis, discussion of the genetic risks, decision making throughout disease progression, often when there may be communication or cognitive loss, advance care planning, care at the end of life, when decisions may be needed as to the most appropriate treatment and whether to withhold or withdraw treatment and discussions about assisted dying. At all these times there is a need for careful communication, listening to the patient and family and clear discussion of the benefits and risks of any plan or intervention.

Key words. Palliative care, neurodegenerative conditions, ethics, advance care planning, communication.

People with neurological disease, and their families/carers, face many different issues throughout the progression of the disease. All professionals involved in their care should provide generalist palliative care – attention to communication, decision making and goal setting and the management of symptoms. Specialist palliative care services are increasingly involved for more complex issues¹ and often there is close collaboration with neurology services².

Throughout the disease progression there may be ethical issues, which will vary according to the disease and to the individual person. These may be faced, by patients, families and professionals, at any time during the disease progression and include communication issues, the consideration of specific interventions, coping with cognitive change and care at the end of life. Although these issues may be primarily seen with people with progressive neurological disease, there may also be ethical dilemmas for people facing acute changes in neurology – such as stroke or brain injury – with the similar principles of discussion, consideration and decision making.

Aspetti etici nelle cure palliative in neurologia.

Riassunto. Le cure palliative per le persone affette da patologie neurodegenerative possono essere appropriate in ogni fase della progressione delle malattie, in base ai bisogni fisici, psicosociali e spirituali. Il percorso di cura per questi pazienti è spesso complesso a causa dei problemi che coinvolgono i malati, i loro familiari e i professionisti che li hanno in carico. Spesso emergono dilemmi di tipo etico che possono riguardare la comunicazione della diagnosi, le discussioni rispetto ai rischi genetici correlati, le decisioni da prendere durante le fasi di progressione e deterioramento, la comparsa di difficoltà comunicative, espressive e i deficit cognitivi, la pianificazione condivisa delle cure, le cure di fine vita, quando spesso si devono prendere decisioni rispetto ai trattamenti più appropriati, per esempio se iniziare o sospendere trattamenti di supporto vitale, o si venga coinvolti in discussioni riguardanti la richiesta di anticipazione della morte. In ciascuno di questi momenti è fondamentale una grande attenzione agli aspetti comunicativi, all'ascolto attivo del paziente e dei suoi cari e una estrema chiarezza rispetto ai benefici attesi e ai rischi potenziali correlati ai piani di intervento proposti.

Parole chiave. Cure palliative, patologie neurodegenerative, etica, pianificazione condivisa delle cure, comunicazione.

The telling of the diagnosis

The telling of the diagnosis is a very significant point for most people with a progressive neurological illness. How this is undertaken and the response of the professional involved may set the tone for the future care of the person, and affect how they perceive both the disease and the professionals involved in their care.

There is evidence that the process of telling the diagnosis is often less than ideal. Guidelines stress the need for honesty, response to the patient and family's wishes and concerns and ensuring that the information is given accurately and empathetically. For amyotrophic lateral sclerosis (ALS, also known as motor neurone disease - MND), the UK guidelines issued by the National Institute for Health and Care Excellence³ recommended:

- the diagnosis should be given by a consultant neurologist with knowledge and expertise in ALS;
- the discussion should include details of the disease, the prognosis, treatment options, the possible progression of the disease over time and the likely symptoms that may be encountered.

These principles would apply to any neurological disease. There are guidelines and protocols which outline the principles in all care, such as SPIKES – which suggests care at all stages: **S**etting up the interview; assessing the patient's **P**erception; obtaining the patient's **I**nvitation; giving **K**nowledge and information; addressing the patient's **E**motions with empathic responses; **S**trategy and summary⁴. However, a survey of patients with ALS in Australia showed that 36% were dissatisfied with the delivery of the diagnosis and the skills of the neurologist⁵.

The overall aim should be to ensure that the person, and their family, receive the diagnosis in a way that they can understand and enabling them to discuss the future options, with a knowledge to support this. Honesty and clarity are essential.

However, there may be issues with some families. There are some ethnic and cultural groups who may see these principles differently. Clinicians may face requests, from the patient and/or family, for non-disclosure of a diagnosis or the seriousness of the disease. This may lead to ethical dilemmas for the clinicians involved. There will need to be a balance between the ethical imperative to be honest and the right of the patient's right to knowledge and the cultural norms for a particular family⁶. This will need careful discussion and a response that allows the patient and family their specific wishes, but allows for open communication to continue and the relationship with the clinician to continue and be able to develop further⁶.

Another ethical dilemma that may be faced with some neurological disease when there is a genetic component to the disease. This will be seen with Huntington's disease – which shows an autosomal dominant inheritance of the gene expansion with 50% of children of an affected person developing the disease. This is increasingly seen in other neurological disease as the genetic influences in neurological disease is elucidated. For instance, gene mutations are seen in people with ALS – predominantly in those who have a family history of ALS – familial disease – but also in people with no clear family history⁷.

The genetics may be well understood in some families, particularly those with HD, when many generations may have been affected and the effects of the disease are only too well known, but there may be less understanding in some families. Ethically it is important that the affected person understands the significance of the genetic influences and the implications of these to both themselves and their families. However, there may be ethical issues raised if the patient does not want to share this information with their family, who are at risk of the disease, and may benefit from this knowledge – as they may be able to undergo testing for the genetic change, and reassure themselves that they have no risk or at least know what they

face, or they may wish to plan their own family using this genetic information.

The ethical dilemma may be faced, but the responsibility of the professional is to the patient and their view, if they have the capacity to make the decision, should be respected. However, this may place professionals in difficult circumstances when family ask them for information – information that they know but cannot tell due to the wishes of the patient. Careful consideration and support of all involved and essential advice may be required from relevant Ethics Committees, Professional licensing bodies or professional organisations.

Decision making

All decisions taken by a person should be according to their wish and respect their autonomy – that is ensuring that the person is able to state an informed preference or consent to whatever they do or is done to them. The process should ensure that the most appropriate and acceptable decision is made. If the person is not able to make the decision at the time, due to loss of capacity from cognitive change, loss of communication or loss of consciousness, it may be possible to take into account a decision that was stated when the person was able to do so.

In neurological care there may be many specific barriers to autonomous decision making, as cognitive and communication loss is common, particularly in certain diseases such as dementia or PD. There may be other circumstances that may affect decision making:

- insufficient time for decision making. Many decisions should be approached as a process – such as the discussion about gastrostomy which may start when there are mild swallowing problems, and further discussion as the issues increase with the options of meeting people with a gastrostomy, discussing with all the multidisciplinary team and family and carers, rather than a sudden decision when there are severe issues;
- unsupported information- any information should be clearly presented in language the person understands. Interpreters (for language or hearing or visual impairment) and communication aids and support from speech and language therapy may be necessary. There may be the need for different ways of providing information for some people – such as using pictures and less complex terminology for someone with intellectual disability;
- awareness of cognitive impairment – all people should be considered to have capacity to make decisions, and if this is in doubt full assessment is necessary. However, some people may be able to make some decisions if the options are presented clearly and without complex discussion.

However, many discussions are delayed until it is difficult for a person to be involved – due to cognitive or communication issues. This may be due to reluctance of the patient or family or, often, reluctance of the professionals to discuss complex issues. Clear explanation of any treatment intervention should occur, with full involvement of the family/carers if this is acceptable to the patient. Professionals may fear that such discussions remove “hope” from the patient. However, for a patient to make autonomous decision they should be involved and have the opportunity to discuss all the options and benefits and risks, and there may be a need to refocus the patient, and family, from hope of cure to hope of quality of life and death⁸.

There may be conflicts during these discussions. As it has been discussed above some families may consider that it is their role to make decisions on behalf of the patient, and this may be seen particularly in certain cultures⁷. Views may vary within families and may reflect existing family issues or tensions or fears/experiences of dying and death. It is important to listen to all involved and try to allow the patient the opportunity to be involved in decision making and the setting the goals of care. There may also be conflicts within the caring multidisciplinary team and again careful discussion is needed, perhaps with facilitation from outside the team itself, to resolve the problems. On occasions the issues may reflect other tensions and disagreements within the team^{9,10}.

The timing of discussions about treatment options is complex as well. It may be possible to discuss future care at the time of diagnosis, although many people are shocked and unable to consider these wider issues, as they are coming to terms with the diagnosis itself. Moreover, initially there may be improvement in symptoms if there is appropriate and helpful treatment – for instance the patient with PD may experience a great improvement when started on dopa therapy. However, for many this may be the main opportunity to discuss the implications of the disease with a specialist team, as appointments may be limited or access to the neurological centre difficult, due to mobility or other issues. It may, thus, be necessary to try to discuss the complexity of disease progression at this early stage and the UK NICE Guideline on MND/ALS has recommended full discussion at these initial appointments³. In some centres a further appointment is set two weeks after the diagnosis is given, to allow for the wider discussion and for patients and families to be able to have discussion, having had the opportunity to think about what had been discussed initially¹¹.

Discussion of possible interventions may be possible throughout the disease progression. It may be easier, as discussed above, to have several discussions, particularly if there is deterioration. The UK NICE Guidelines on MND/ALS recommended discussion of options as part of routine monitoring – for

instance measuring respiratory function provides an opportunity to discuss future care, such a non-invasive ventilation, and provide information to facilitate later discussion³. Other guidelines have also suggested this approach for the discussion of both treatment options and end of life care¹².

The role of Ethics Committees remains controversial in supporting clinicians in making difficult decisions, as the majority of Committees are more involved in evaluation of research protocols, assessment of sponsored drug trials and allocation of resources. They often decline to be involved in clinical decision-making processes, particularly in some countries like Italy, and they feel that they are not competent to discuss issues around care at the end of life. This can lead to a sense of impotence and lack of support for those clinicians involved in the care of patients with neurological diseases facing dilemmas and sensitive decisions.

Advance care planning

As people with neurological disease often face loss of capacity to make decisions due to cognitive change, loss of communication or consciousness, advance care planning (ACP) is increasingly discussed. However, there may be complex ethical issues to be faced, from the assessment of the capacity of the person who completes the ACP to whether the circumstances were really appreciated by the person when they made their plan. ACP is defined as “the ability to enable individuals to define goals and preferences for future medical treatment and care, to discuss these goals and preferences with family and health-care providers, and to record and review these preferences if appropriate”¹³.

For a person to be considered to be competent to make a decision there is general agreement in the literature that a patient should be able to:

- understand the risks and benefits,
- appreciate the personal consequences of their choice,
- make a rational choice concerning treatment or research,
- express a choice^{14,15},
- act on a decision – in some countries¹⁶.

There are issues in how ACP is discussed and often people do not want to think ahead, even though they may have limited opportunity to do so before they lose cognitive or communication ability. There is increasing consideration of discussion of a person’s overall wishes of care and consideration of:

- What matters most to you in life when you are well?
- Which of these will become priorities when you become less well?

- How can you gain access to support your social network at a time when you become less well, so that you can ensure these priorities?
- How to cope with future periods of incapacitation¹⁷.

Thus, this does not focus just on planning for end of life but looks at person's values, considering what sort of person they are and whether they would wish to be cared for if there is deterioration, such as do they want life prolonged at for as long as possible or would prefer quality of life to be maintained even at the risk of a reduction in prognosis¹⁸. This enables people to think ahead, perhaps only for a few weeks initially but then allowing a wider consideration of end-of-life issues. This has been shown in multiple sclerosis care, where a realist review of the literature showed that it was important to enable people with MS to see that they had a life limiting illness, for ACP to be seen as relevant. Moreover, the importance of very careful communication by professionals, with training, was essential¹⁹.

Advance care plans may specify the circumstances when certain interventions or actions would be withheld – for example a person with ALS stating that they did not want tracheostomy and ventilation at the end of life, even if this led to their death. Such as request is often legally binding in many legislations, if completed in the correct way, although this does vary across countries. A person may also be able to define a proxy who would make decisions on their behalf, if they are unable to do so, such as defining a spouse or child to make decisions. The professionals would then ask this proxy for their opinion as to the person's wishes and act accordingly. In all cases ACP only becomes appropriate of the person has lost decision making capacity, as if they are able to make a decision, they should be asked directly and their decision respected.

ACP may also include wider issues such as ensuring a will and last testament is in place, so that the person's wishes about the future of their property and assets is clear; funeral plans; place of care and death – these may be different and it is important to ensure both are considered²⁰; how they feel about the family coping with the burden of care; particular wishes they may have such as particular foods they like or music they wish to listen to when they are not able to make the choice themselves.

End of life care

There are many issues with ethical dimensions as someone approaches the end of life.

RECOGNITION AND DISCUSSION OF END OF LIFE

It is important to ensure that, if possible, all are aware that the person may be entering the final stages of the

disease progression. This will allow planning for end-of-life care, ensuring that ACP is clear and gives the opportunities for all involved – patient, family and professionals, to be prepared. For patients this may allow their wishes to be known, if they have not been expressed before in ACP, the opportunity to undertake any particular wishes – such as seeing family, visiting important places, reliving and telling memories; for families to be prepared for their death and ensure all who would wish to know, and perhaps visit and say goodbye, are involved; for professionals to prepare for end of life care – stopping unnecessary medication, investigations or interventions, ensuring medication is available for any symptoms which may occur, such as pain, breathlessness or distress²¹. However, there may be resistance to consider these issues by anyone – patient, family or professionals.

It may be helpful for professionals to use triggers to help in this process of the recognition of the end of life. These may be: consideration of the “Surprise question” (Would you be surprised if this patient died in the coming year?)²² or the use of specific tools, such as SPIC²³, NECPAL or RADPAC. The use of these tools may help in identifying patients who are deteriorating and allow conversations with patients, if appropriate, families and the wider MDT about the future and allow the wishes of all to be recognised and then acted on, as appropriate and if possible. Although there may be resistance to these discussions it may be very important if the person, and their family, is to make appropriate decisions, that they know of the possible future deterioration and share their views. For an autonomous decision to be made the person does need to be able to discuss all the important information – benefits and risks – as discussed above.

WITHHOLDING AND WITHDRAWING TREATMENT

Many people with progressive neurological disease may benefit from interventions that may improve quality of life and/or increase length of life. However, there may be occasions when what is possible may not be appropriate – as the risks of harm are considerable, the benefits are very limited, there may be long term implications that may be unacceptable to the person. The discussion of these interventions is again complex – including consideration of gastrostomy, parenteral feeding / fluids, ventilatory support, such as non-invasive ventilation (NIV) or tracheostomy ventilation (TV), cardiopulmonary resuscitation (CPR), antibiotic treatment (orally or intravenously). All of these discussions may need complex discussion, of the benefits and risks and it is recommended, in many guidelines, that these discussions take place earlier in the disease progression, so that cognition

and communication may be easier and time can be taken in the process of decision making³.

Ethically it can be argued that withholding treatment – i.e., not giving antibiotics or parenteral fluids at the end of life – and withdrawing treatment – actually removing or stopping interventions – such as stopping an infusion or removing ventilatory support – are similar²⁴. The primary goal of any medical treatment is to benefit the patient by restoring or maintaining health, maximising benefit and minimising harm. Treatment that does not provide net benefit to the patient may, ethically and legally, be withheld or withdrawn and the goal should shift to the palliation of symptoms. These issues, however, feel complex when managing any individual patient and family, and good communication, discussion within the multidisciplinary team and involving others, as a second opinion, are often very helpful²⁵.

The most complex and difficult area for patients with progressive neurological disease, and in particular ALS, is the removal of ventilatory support at the end of life. Usually the withdrawal of the ventilation – whether NIV or TV – will lead to the death of the patient within a relatively short time, from minutes to hours. Moreover, the removal of ventilatory support will often cause severe distress, of breathlessness, and it may be necessary to ensure the patient has these symptoms managed effectively, in advance of the withdrawal^{26,27}. The discussions may be complex for everyone and may lead to much discussion amongst the MDT, with differing views expressed²⁷.

The ethical dilemmas may be relatively clear in some cases. If a person with decision making capacity does request the withdrawal of an intervention this should be respected. Careful explanation of the implications is necessary but the autonomous decision of a competent patient must be respected, in most jurisdictions. However, if the patient is not able to make the decision themselves it may be necessary to look at any ACP, as they may have specifically requested withdrawal at a certain stage of disease progression, such as loss of communication, or discuss with any proxy for decision making and with the wider family and MDT. Any decision should be taken in the patient's best interests, after careful consideration of the views of the family and MDT. This may vary across countries but the ethical approach will be the same.

The withdrawal of ventilation may require medication to be given, before the withdrawal, to ensure that there is no distress, there are guidelines to help in this²⁷ but it is often stressful for all concerned. Studies in the UK of palliative medicine doctors showed that they found the discussions and procedure particularly stressful, rating the challenges practically as 5.81 out of 10, challenges emotionally at 6.22/10 and challenges ethically as 5.08/10²⁸. Some stated that it felt to them as if they were un-

dertaking euthanasia, even though they knew that the ethical discussions were clear that removal of a treatment that was no longer appropriate and requested by a patient was completely ethical²⁸. The withdrawal of ventilation appears to be an emotionally charged situation with many conflicts both for the doctors themselves and within the wider MDT^{28,29}. There was a pronounced difference between the complex ethical discussions and the emotional reactions experienced – the conflict between the head and the heart²⁸. Support of all involved, the patient, family and the wider MDT is essential, often with the opportunity to discuss the issues afterwards in an open and non-judgemental way²⁷.

Thus, the use of ACP is very important when interventions are started, so that all may be clear later as the disease deteriorates. The UK NICE MND Guidelines³ recommend that discussion about the future care should be started before, if possible, and certainly when a new intervention is discussed and then started. In this way patients will be able to make clear decisions, based on the benefits, which may be profound, but also aware of the risks of disease deterioration and the implications of the intervention on future care. As discussed above, these discussions may be difficult but are necessary, if truly autonomous decision making is to be facilitated.

ASSISTED DYING

There is increased discussion of assisted dying throughout the world: physician assisted suicide (PAS) - a doctor intentionally helping a patient to commit suicide by providing drugs for the self-administration at the person's voluntary and competent request- and euthanasia - a doctor intentionally killing a person by the administration of drugs, at that person's voluntary and competent request³⁰. There is a clear distinction between assisted dying and the management of distressing symptoms, by the use of medication, such as opioids, or in rare occasions with palliative sedation, where medication is given to induce unconsciousness to relieve severe distress³¹.

In countries where assisted dying is permitted people with neurological disease are often over represented in the deaths, compared to the population. For instance, in Oregon, USA, where PAS is allowed 7.7% of the people who have died from ingesting a lethal overdose of medication between 1998-2020 had ALS, whereas the ALS represents only 0.08% of the population³². In the Netherlands, where euthanasia and PAS are permitted, one study showed that 22% died as a result of an assisted death³³.

Assisted dying often leads to controversy, within the health professionals, patients and families and society in general. There are many ethical issues, with

those advocating for assisted dying emphasizing the importance of patient autonomy, whereas others are concerned about the loss of trust in doctors, the fears of an escalation in the reasons for permitted assisted dying, the lack of compliance to the rules and the gradual erosion of the rules to include those who are not competent to make decisions, even to babies and children^{34,35}.

In all jurisdictions, including those where assisted dying is not permitted, there is a need to hear the concerns and fears of patients and families and why they may be asking for life to be ended. This may be due to the fear of the future, and many people fear a distressing death from progressive neurological disease, although there is increasing evidence that dying of a disease, such as ALS, may be no more distressing than any other disease, when good palliative care is provided^{36,37}. The aim of discussions may need to be to understand their request, rather than necessarily acting on it³⁸. Neurological disease has been shown to lead to existential distress both for patients, families and health care professionals and careful exploration of the real concerns may be helpful³⁹. Moreover, clear explanation of the likely symptoms and issues at the end of life, reassurance that these symptoms can be managed effectively, discussion that the patient can request that life prolonging treatment is withheld, and careful exploration and discussion of the deeper existential, emotional and spiritual issues may all be helpful.

Even though assisted dying remains outside the palliative care precinct, the EAPC recommends that all requests for euthanasia and PAS require respect and careful attention, together with open and sensitive communication in the clinical setting and that individuals requesting euthanasia or PAS should have access to palliative care expertise³⁰. This is due to evidence that often, although not always, requests for euthanasia and PAS may be altered by the provision of comprehensive palliative care. Suffering from physical symptoms can be alleviated, and psychosocial and spiritual care can improve well-being of patient and caregivers. Information about the available palliative care options may provide reassurance and diminish anxieties about the subsequent disease trajectory³⁰. The authors suggest how palliative sedation may offer an option for many conditions in which patients may request euthanasia or PAS, in case of refractory physical symptoms or untreatable psycho-existential suffering when the person is close to the end of life³¹.

The care of people with neurological disease, particularly when this is progressive, will include the consideration of many ethical issues. Careful discussion with patients and families is essential, together with discussion amongst all members of the multidisciplinary team. This will allow a considered discussion

and often a mutually acceptable management plan, which will allow the person to maintain as good a quality of life as possible, and maintain quality of life and allow the person to die peacefully.

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