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Oliver, David J. and Turner, Martin (2010) Some difficult decisions in ALS/MND. Amyotrophic Lateral Sclerosis and Frontotemporal Degeneration, 11 (4). pp. 339-343. ISSN 2167-8421.

### DOI

<https://doi.org/10.3109/17482968.2010.487532>

### Link to record in KAR

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## Some difficult decisions in ALS/MND

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## Introduction

Those with amyotrophic lateral sclerosis (ALS)/ motor neuron disease (MND) currently face inevitable progression of their disease, with increasing immobility and variable problems with communication. As well as the personally devastating impact of a shortened life span felt by most patients, the effects of disability go far beyond the patient and inevitably alter relationships with partners, family and the wider social network. There is an impact on all the professionals involved in the care. The Oxford neurologist Bryan Matthews (1920–2001) recognized a balancing act between the needs of the patient, family and physician, commenting in his textbook 'Practical Neurology' that: "The best test of a physician's suitability for the specialized practice of neurology is not his ability to memorize improbable syndromes but whether he can continue to support a case of motor neuron disease..." (1).

There are many decisions to be faced during the individual ALS patient 'journey', and no rigid model to fit the wide range of human experience, emotion and behaviour (2). Management decisions often require conversations that go beyond simply obtaining informed consent for a procedure, and other issues may be difficult for patients, families and professionals and may expose aspects of personality or past experience. Such conversations can be a source of intense discomfort to the patient, families and professionals, and as a result such discussions may be overlooked or avoided.

Commonly challenging discussions include:

- The use of gastrostomy
- The use of assisted ventilation
- End-of-life planning
- Requests for assisted dying

There is now clear clinic-pathological overlap between ALS and frontotemporal dementia, and with rigorous neuropsychological testing it may be possible to demonstrate mild cognitive impairment in up to 50% of patients (3). Although frank dementia is rare (and often an early feature when present), mild frontal lobe dysfunction might in theory impair executive function and influence decision making (4). Thus there may be a need to consider some decisions earlier on in the disease process, to allow patients to be fully involved (5). However, earlier decisions may be difficult due to uncertainty about disease course, complicated by a desire to avoid considering future deterioration. The aim of all decision making is to empower the person's autonomy – "informed preference or consent to whatever we do or what is done to us". This will be helped by ensuring the person is fully involved in the process, and this requires that:

- They have sufficient background information to help in the decision.
- Information is provided in a sympathetic and empathic way, with both sides of the argument given.
- Time is spent explaining the various points and aims of any interventions or treatments.
- The person is supported, if they wish, by family or friends.
- Written information may be of help to allow further consideration.
- The decision making is seen as a process, taking place over a period of time, allowing careful consideration to be given to the decision (6).

These areas are never easy and often complex, with the need for team discussion and close involvement of both the person with ALS and their family. There may be multiple interactions, with conflicts between individuals, both within families and professional teams. These discussions may bring to light difficult areas for all, and professional carers may inevitably bring their own cultural experiences and prejudices to these discussions. In considering some of these issues we hope to promote more proactive discussion with patients and families, involving multidisciplinary teams and the wider global ALS care community.

Through all decisions and interventions it is essential to stress that supportive care will continue to be provided, with the aim of managing symptoms and minimizing any discomfort or distress. Stating "we will never ignore or neglect you" explicitly can be very helpful in assuaging fears that might otherwise lead to avoidance of palliative interventions.

## Gastrostomy

*“I’ll put that off for as long as possible. It’s permanent and feels like admitting defeat.”*

Failure to maintain adequate nutritional status is an established adverse prognostic factor in ALS, and gastrostomy is a well-established safe and practical alternative to oral feeding (7).

Bulbar symptoms are frequently the most feared by ALS patients. As the first symptom they are associated with poorer prognosis (although not invariably the case), and limb-onset patients often await their development as a marker of the ‘final stage’ of the disease (although there is surprisingly little known of the probability of a limb-onset patient needing gastrostomy at some point in their disease course). Thus, gastrostomy, above other palliative measures, may be perceived by patients as the chief surrogate marker of deterioration, or succumbing to the disease. Moreover, as an invasive procedure (unlike NIV, where a mask can be removed), gastrostomy may raise particularly strong feelings of loss of control or a perception of ‘giving in’ and accepting ‘artificial’ support. Pejorative terms in common media uses such as ‘tube feeding’ compound such views.

Shortly after diagnosis, reassuring patients proactively that those with ALS do not ‘choke to death’ can help to allay a major concern, and set the scene for a better understanding of the purpose of gastrostomy if this becomes indicated later. Managing patient expectations when the time comes is also important. Although the maintenance of nutritional balance is intuitively likely to improve overall survival, gastrostomy is primarily a comfort measure, for example avoiding hunger pangs or dehydration, or reducing the anxiety and social isolation caused by prolonged mealtimes. Emphasising positive messages that the oral route may still be used after the insertion and for tasting foods, even if most nutrition is administered via the gastrostomy, or that a gastrostomy is not visible through normal clothing, can all be helpful, along with photographic examples of the external tube in situ.

Percutaneous endoscopic gastrostomy (PEG) requires patients to have mild sedation to allow passage of the endoscope, and a degree of recumbence. A lying vital capacity of at least 50% is a typical benchmark to assume both will be safe (8). This means that if the inevitable decline in respiratory function associated with ALS occurs to a significant degree before the bulbar need for gastrostomy, then the risks of a PEG may outweigh benefit. While a radiologically-inserted gastrostomy (RIG) offers an alternative sedation-free procedure in such circumstances, it may be better to have introduced the idea of gastrostomy soon after the development of any bulbar symptoms. This allows the patient to feel active in the decision rather than under pressure before further respiratory decline. Emphasizing a prophylactic role for PEG in preventing nutrition-related weight loss, perhaps not being used for some months after insertion, supports this.

## Assisted ventilation

*“Why can’t I just have a tracheostomy? Even if I can’t move then at least I can watch my children grow up.”*

Non-invasive ventilation (NIV) is now a routine part of care for many ALS patients, ideally managed in partnership with a respiratory team (9). By reducing hypercapnoea the most dramatic effect is in improving quality of life through reduced sleep fragmentation, morning headache, and daytime somnolence. NIV has a significant effect on overall survival (10), but this should not be the main indication, and is an important message in managing patient expectation, especially when it may not be suitable for, or tolerated by, all patients.

Patients with respiratory failure may develop incipient symptoms over a prolonged period of time, or very rarely present with catastrophic symptoms in an emergency situation. Where the latter happens outside a tertiary neurological centre, there needs to be prompt consideration of the diagnosis of ALS to prevent unwanted or futile intervention. Routine monitoring of respiratory function in the clinic setting is now standard, and there may be evidence of decline before significant symptoms have developed. Given that NIV may lengthen survival, it has the potential to be instigated before the person is ready to consider all the implications. The discussion of NIV should include end-of-life issues, for example increasing disability and dependency, and the inevitability of death despite NIV. Such a discussion should also include mention of the mode of death, explicitly addressing the common fear of ‘suffocating to death’ by outlining the nature of the drowsiness that usually occurs in the context of end-stage

hypercapnoea and hypoxia.

Ventilation via tracheostomy (tracheostomy with invasive ventilation (TIV)) is taken up by as many as 30% of ALS patients in some countries. The perception by many patients (and some physicians) of uniformly long-term survival benefit is less certain. The potential to be 'locked in' without even eye movements for communication and the possibility of dementia must form part of the discussion (11). TIV brings with it a requirement for high levels of nursing care. Clear plans in the event of acute complications relating to the entry site and tube need to be established. Instigation of TIV in an emergency setting is never advisable. The high expense of TIV means it is considered non-standard within many models of health care cover. However, it is unhelpful to raise this as an issue with patients as it can then become the focus of a perceived (possibly legal) battle with 'the system' that needlessly risks emotional harm and an erosion of the doctor-patient relationship, when outlining some of the practical issues above would lead many to an informed decision to decline TIV anyway (12).

As the person deteriorates they may wish to consider withdrawal of assisted ventilation, either themselves at the time or as part of an Advance Decision to Refuse Treatment (ADRT). It is important to explore the emotional impact as this decision will be difficult for both patient and family, with a high potential for feelings of guilt in the latter. With sensitive discussions it may be possible to establish a clear plan. There are guidelines for the withdrawal of NIV (13), and these need to be considered. Patient, family and professionals will all require support.

### **End-of-life planning**

*"All I ask is that I can die at home."*

For people with ALS to be able to make informed decisions about end-of-life issues these areas must be discussed openly. The early reassurance of both patients and carers that they will not 'choke to death' or 'suffocate' can be very powerful at allaying concerns. Key issues include:

- 1) Place of care. Where would they wish to receive care during the disease progression? This is not necessarily synonymous with place of death (14).
- 2) Place of death. Do they wish to die at home, in a hospice, residential home or hospital? It may not always be possible for them to die in their preferred place of care. Many hospices are unable to provide ongoing care over a long period of time, although they may be able to admit patients for a period of symptom assessment. However, active planning may enable people to remain at home. The provision of appropriate medication 'in case of need' may be appropriate (e.g. the Breathing Space Programme in the UK suggests the provision of morphine, midazolam and glycopyrronium bromide injections in case of a sudden deterioration or distressing symptoms) (15). Carers must feel comfortable with this, however, and it must be made clear that they are not hastening death.
- 3) Attempts at resuscitation in the event of cardio-pulmonary arrest are likely to be futile at the end of life for someone with ALS. The completion of a DNAR (Do Not Attempt Resuscitation) order can ensure that the person can be allowed to die naturally in this rare circumstance without confusion among ward staff and family. Exploration about where to deposit such orders, e.g. with the local out-of-hours medical and nursing services, ambulance base or hospital is useful.
- 4) The completion of advance care plans can be very helpful, ensuring that the person's wishes are understood if they are unable to communicate them or there is a loss of capacity. In the UK, options include:
  - An advance statement - this outlines the person's overall wishes but without specifying the actions wished.
  - An ADRT defines specific treatment refusals for circumstances of deterioration, understanding that refusal may lead to death. This is legally binding if it has been completed appropriately, and when a person has lost capacity the health-care professional can act accordingly.
  - Lasting Power of Attorney where a patient nominates someone to make financial and care decisions on their behalf if they lose capacity. Even if this formal measure is not invoked, suggesting to patients that they actively address financial matters, e.g. through the creation of a Will, is a very useful though often neglected suggestion.

### **Requests for assisted death**

*“I can’t bear being a burden to my partner and family. I want them to remember me when I was healthy.”*

There is increasing general awareness of assisted dying. In countries that permit assisted dying (The Netherlands, Belgium, Switzerland, Luxembourg and Oregon and Washington States in the USA), a significant proportion of those with ALS receive assisted death. In the Netherlands up to 20% of ALS patients receive euthanasia or physician-assisted suicide (PAS) and in some centres the figure may be as high as 50% (16). Of the 115 UK citizens to date who have received assisted suicide from Dignitas in Switzerland, 27 were ALS patients (17). Debbie Purdy, a person with multiple sclerosis in the UK, recently challenged the High Court to allow her husband to accompany her to Dignitas without the risk of prosecution for assisting suicide (a criminal offence in the UK). This was granted and the Director of Public Prosecutions has produced guidelines which clarify the legal position. However, coverage of this case, and the earlier case of an ALS patient, Diane Pretty, while increasing awareness of ALS has undoubtedly had an adverse effect in emphasising the idea of an otherwise distressing death when the evidence from specialist palliative care has shown this is the rare exception. A legitimate concern is that messages of hopelessness make patients vulnerable to the sense that they are a burden to society for whom assisted dying is an ‘honourable’ duty.

A person with ALS who is asking for an assisted dying may be doing so for one of many reasons and discussion of these issues, meeting the question head-on, may be very helpful:

- An expression of frustration. Raising the issue of suicide can be a way to vent anger at the perceived impotence of the physician, carer or ‘system’.
- Fear. Is the person fearful of a distressing death? There are often fears of ‘choking to death’, which can be justifiably allayed when studies have shown that this to be exceptional (18).
- Explanation and preparation for dying, including the provision of appropriate medication, advance care planning and support of the person and their family can alleviate these fears. Being ‘kept alive’ by interventions such as TV may be a particular concern in countries where subsequent withdrawal is not possible. This may be helped by careful discussion and completion of an advance care plan.
- Depression. Debatably uncommon in patients, although undoubtedly a major issue for carers. Both might be helped by psychological intervention or medication.
- Control. Some people with ALS wish to have control over the end of their lives, particularly if they feel their quality of life in the future may become intolerable for them (19), but also sometimes as a way to claw back some control in a disease they may perceive to have otherwise stripped them of it completely.

In a small minority of people there may be a profound and continuing request to die despite discussion. As professionals we hear the distress but in the majority of countries no action can be taken. There is a need to continue to support the patient, family and all professionals involved in the care of such patients. It may be important to stress that life would not be prolonged by intervention, and completion of an ADRT may provide reassurance despite appropriate care involving only comfort measures, rather than active intervention.

## **Summary**

The physical and emotional damage caused by ALS creates ripples that reach a wide group of individuals beyond the patient. Decisions about care and end of life are never straightforward; solutions are always bespoke and must involve the wider multidisciplinary team. There is a need for awareness of how these discussions will be affected by personal attitudes, experience, culture and issues in our own professional and personal lives. The aim should be to respond to patient and family and be honest, sensitive, empathic, understandable and direct, with a constant balance of hope with realism. Finally, as professionals we have our own need for support and to be aware of our own feelings to prevent the very real threat of compassion fatigue (20). A strong multidisciplinary team is greater than the sum of its parts, and can provide robust internal support to its members, using their pooled expertise to serve patients and their families more effectively as a result.

**Declaration of interest:** The authors report no conflicts of interest and are alone responsible for the content and writing of this paper.

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