

Palliative and end-of-life care in advanced Parkinson's disease and multiple sclerosis

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Patients with advanced Parkinson's disease (PD) and multiple sclerosis (MS) can present with challenging clinical problems when admitted to acute hospitals. They are susceptible to life-threatening complications, most commonly sepsis, which arise against a backdrop of progressive decline in overall function. Both PD and MS may be complicated by cognitive impairment, swallowing or speech problems and refractory pain syndromes.^{1,2} Robust evidence supporting treatment decisions in advanced disease is lacking, but recent national guidelines recognise the importance of shifting goals towards a palliative approach as the disease becomes more advanced.^{3–6}

Importance of communication

Prompt and effective discussion between the patient's general practitioner and secondary care clinicians may both prevent inappropriate hospital admissions and achieve treatment decisions in the patient's best interests. Drawing on the strengths of multidisciplinary team working invariably ensures that patients receive better care. The key, however, to optimising care is taking time adequately to hear the patient's wishes before they might lose capacity.

Estimating prognosis

Both in advanced PD and MS it is difficult to anticipate life expectancy, but the

rate of overall deterioration is an important indicator of prognosis. Patients are particularly at risk of dying from aspiration pneumonia, urinary tract infections, complications from falls and fractures, and sepsis secondary to pressure ulcers. These are all commoner the more advanced the disease. Even so, some very disabled patients will live for many years while others may succumb quite suddenly from an episode of sepsis or, less commonly, from a pulmonary embolus. The unpredictable disease trajectory makes planning for the terminal event particularly difficult and most patients die in hospital rather than at home.⁷

Common themes and contrasts in advanced PD and MS

Cognitive/psychiatric

- *Cognitive changes* are common in MS, with loss of verbal fluency,

memory problems and difficulty with concentration.⁸

- *PD dementia* is common, characterised by illusions, visual hallucinations, visuospatial deficits, attentional and dysexecutive deficits.⁹ PD dementia can respond to acetylcholinesterase inhibitors.¹⁰ Hallucinations may respond to reduction in PD medication, particularly amantadine, monoamine oxidase type B inhibitors (MAOI-B), cholinergic medication and dopamine agonists.
- *Depression* is common in both illnesses. It may respond to selective serotonin reuptake inhibitors (SSRIs). In PD, optimisation of dopaminergic therapy and tricyclic antidepressants have also been shown to have benefit.¹¹ SSRIs should not be co-administered with MAOI-Bs because of the risk of serotonin syndrome.

Bulbar symptoms

- Hypophonia in PD and dysarthria in MS can make communication difficult and lead to immense frustration for patients and carers.¹²

Key Points

Palliative care in advanced Parkinson's disease (PD) and multiple sclerosis (MS) recognises the need to move from attempting to control function to affording comfort measures

Admission to an acute hospital can be highly disruptive to the nursing and drug routines for these patients

Advanced disease can lead to cognitive changes in MS, and in PD to dementia and hallucinations, each of which can rob patients of the opportunity to express their wishes and preferences as their disease progresses further

It can be empowering for those patients who still have capacity, sensitively to discuss whether they would wish to receive life-prolonging treatments as their disease progresses, such as intravenous antibiotics, percutaneous endoscopic gastrostomy feeding tubes and artificial ventilation

The terminal phase of PD and MS usually occurs against a backdrop of more rapid global decline and increasingly severe disability. The terminal event is often an acute episode of sepsis causing death over a few days

KEY WORDS: best interests, multiple sclerosis, palliative, Parkinson's disease, terminal

- *Swallowing* problems are associated with aspiration. Early expert assessment by a speech and language therapist is essential. Aspiration may initially be silent without apparent systemic effects, but the situation can change within a few hours as the patient develops systemic signs of sepsis.
- *Drizzling* of saliva is more common in PD. It can be managed by ultrasound guided salivary gland botulinum toxin injections, topical atropine 1% drops applied to the tongue or topical hyoscine patches.

Pain

Pain can be a disabling feature of PD² and may have a variety of causes including central pain and dystonia.

- *Neuropathic pain* in the limbs or face is frequently difficult to treat in MS.¹³ Amitriptyline and anticonvulsant drugs like gabapentin are often employed. However, both groups of drugs are less well tolerated in MS and there are often unacceptable anticholinergic side effects from tricyclics and sedation and ataxia from anticonvulsants.

- *Spasticity* in MS can cause painful spasms and limb jerking, both of which can be particularly troublesome during the night. Antispasticity drugs have a variable degree of success and tend to cause sedation and ataxia. Limb contractures are a sign of advanced disease.
- *Pressure ulcers*, usually over bony prominences, can be related to loss of mobility and inadequate nutrition.

Decision making

Broaching the topic of life-prolonging treatments is best done when the patient is relatively well and still has capacity for the particular decision. No one should be coerced into advance care planning but some patients will feel more in control having signed an advance decision (refusing specified treatment) (Table 1).^{14,15}

Decision making in the acute situation

For patients who lack capacity and are acutely unwell, there is a requirement to act in the patient's 'best interests' as prescribed by the Mental Capacity Act 2005 (Table 2).¹⁶ Optimum decision making in advanced PD and MS revolves around 'trying to get it right' for the individual patient. As far as possible, this should be what the patient wants or, if they have lost capacity, would have wanted. The patient's previous experience of hospital admissions may influence their preferences.

*Withdrawing/withholding treatment and practical symptom management*¹⁷

As the patient becomes increasingly bedbound, sensitive discussions need to take place with the patient and family about altering the focus of care.⁵ In both diseases, the aim of treatment shifts from maximising function to minimising suffering and aiming for the patient's comfort and dignity. Common goals are:

Table 1. Pertinent topics to consider in advance decisions (refusing treatment) in Parkinson's disease and multiple sclerosis.

Practical problem	Potential treatment
Difficulty taking adequate nourishment orally ¹⁵	Nasogastric feeding tube – short-term (up to 2 weeks) PEG feeding tube
Unable to take adequate liquids orally ¹⁵	Either via feeding tube or iv or sc fluids
Major infection, typically involving chest, urinary tract or via pressure ulcer	Antibiotic therapy, usually iv May require prolonged or repeated courses of antibiotics
Respiratory failure secondary to aspiration pneumonia	NIV May require ITU support and tracheostomy ventilation
Cardiorespiratory arrest	Cardiorespiratory resuscitation (to be effective in advanced PD and MS, this should include discussion about admission to ITU) Sudden unexpected cardiorespiratory arrest is uncommon in PD and MS If the patient is happy to discuss advance decisions, the more likely scenarios above should be addressed first

ITU = intensive therapy unit; iv = intravenous; MS = multiple sclerosis; NIV = non-invasive ventilation; PD = Parkinson's disease; PEG = percutaneous endoscopic gastrostomy; sc = subcutaneous.

Table 2. Making 'best interests' decisions, as described in the Mental Capacity Act 2005.¹⁶

1	Assess capacity for decision in question (eg a potentially life-prolonging treatment)
2	Act in patient's best interests:
	(i) Consider all relevant circumstances
	(ii) Capacity for matter in question?
	– help patient to participate if possible
	– must not be motivated to cause death of patient
	– consider patient's past/present wishes/feelings
	– patient's beliefs and values if had capacity
	– views of those close to patient

Table 3. Tips for the acute medical take.

Neuroleptic malignant syndrome (pyrexia, muscle rigidity, raised creatinine phosphokinase)	May follow acute levodopa withdrawal (eg in a patient who loses the ability to swallow medication)
Antiparkinsonian drug routine.	A potentially treatable cause of acute deterioration in PD patients that should not be overlooked Great psychological attachment is placed by patients and carers on this Concern often justified as PD patients who appear to be deteriorating may improve when the medication regimen is restored
Inability to swallow oral PD medication	Patients who usually take levodopa are at risk of NMS; nasogastric dispersible co-beneldopa is generally preferable in this situation The topical dopamine agonist rotigotine can be a useful substitute (unlicensed indication)
Inability to swallow antispasticity drugs in MS	Midazolam can be given subcutaneously via a syringe driver to relax skeletal muscle (unlicensed use) Typical starting dose 10 mg/24 hr, increasing to 20–30 mg/24 hr if required Sedation from midazolam is dose-dependent

MS = multiple sclerosis; PD = Parkinson's disease.

- freedom from pain
- relieving anxiety and agitation (particularly in PD)
- dealing with troublesome symptoms including:
 - drooling in PD
 - constipation
 - bladder spasms
 - insomnia.

One of the features of advanced PD is the inability to tolerate sufficient dopaminergic medication.¹⁸ In both PD and MS it is appropriate to minimise oral medication, particularly statins, antihypertensives and bulk-forming laxatives. Practical suggestions in managing complications for the acute physician are given in Table 3.

Managing the terminal phase

When the terminal phase can be anticipated by an acceleration in the patient's global deterioration, a decision may have been taken with the patient and family not to treat further episodes of infection. A careful check for specific symptoms should be sought from the patient directly or from observing the patient for any signs of distress. Close family members will often recognise signs of unspoken distress. The views of experienced clinical staff may determine if the

patient is frightened, in pain or has different nursing requirements.

For patients unable to swallow in the terminal stage, medication can be administered subcutaneously as needed or continuously using a syringe driver.¹⁹ Medication can be given, if necessary, to relieve specific symptoms as follows:

- midazolam for fear or agitation
- hyoscine butylbromide for drooling or chesty secretions
- morphine for pain

If pain is present, a sufficient dose of morphine should be used to relieve it but without causing undesirable opioid side effects.

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