Symptomatic management of neurodegenerative disease in the elderly

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Abstract

The care of elderly people with progressive neurodegenerative disease is often complex and palliative care may be helpful from soon after diagnosis to enable the quality of life to be maintained as much as possible. Palliative care may be involved in an episodic way, as new problems and issues are faced, throughout the disease progression. There are many issues which require careful assessment, including symptoms, psychological and social aspects of care. This may include the assessment of cognitive change and ensuring that symptoms and issues are addressed correctly, even if the person cannot communicate or is cognitively compromised. This care will also include carers, spouses and families, professional carers and carers within residential and nursing homes. Patients and carers may need ongoing support, to cope with the multiple and continual losses faced within neurological disease progression, with the complex interaction with other comorbidities and the effects of ageing. As the progression continues the recognition of end of life, through the assessment of triggers, is important so that all can be prepared for dying and death.

Key words

Neurodegenerative disease

Palliative care

Elderly population

End of life care

Symptom management

The care of the patient with neurodegenerative disease is often complex and in the elderly patient may include the care of other co-morbidities that will influence the care provided. The common degenerative neurodegenerative disease in the elderly population are motor neurone disease / amyotrophic lateral sclerosis (MND/ALS), Parkinson's disease (PD), other Parkinson-plus disorders, multiple systems atrophy (MSA) and progressive supranuclear palsy (PSP) and the dementias. There will also be people with other diseases, such as multiple sclerosis (MS) or Huntington's disease (HD) which may not present initially in the elderly population but may continue to be a cause of disability with ageing.

The extent of the problem is not entirely clear for the elderly population and although the prevalence of these diseases may individually be low collectively, together, including dementia, neurological disease is a common cause of death – in the UK 4.4% of all deaths in people over 65 years old (1). The prevalences are MND/ALS 7/100,000, MSA 5/100,000, PSP 7/100,000, and PD 180/100,000, dementias 700 /100,000. (2,3).

There is no curative treatment for these diseases. However there is a great deal that can be provided to ensure quality of life is maintained to as high a level as possible and the patient and family are supported. Thus for people with neurodegenerative disease palliative care may be appropriate from diagnosis, particularly when there are other co-morbidities that may affect the prognosis (4). This approach has been suggested for neurological disease and an EAN/EAPC Consensus document on palliative care in neurology suggests a palliative care approach for this patient group (5). Moreover palliative care had been shown not only to improve quality of life but also length of life in lung cancer (6) and several studies have suggested that a multidisciplinary approach in MND/ALS may increase prognosis as well as improving quality of life (7). A specialist palliative care service has also been shown to improve the quality of life, and reduce symptoms such as pain, dyspnoea, sleep disturbance and bowel symptoms (8).

However the prognosis of these disease vary greatly – MND/ALS 2-3 years, PD has an average prognosis of years but patients may live for years, MSA 9 years, PSP 7 years and dementias vary but Alzheimer's disease has an average prognosis of 8 to 10 years. Thus end of life care may not be necessary for a long period of time after diagnosis but a palliative care approach, looking at all aspects of the patient and family, can be helpful throughout the disease progression. Moreover this may be on an episodic basis, when new symptoms or issues develop, such as at diagnosis, discussion and consideration of gastrostomy, development of respiratory problems and consideration of non-invasive ventilation and at the end of life for MND/ALS. (9). This may be a challenge for specialist palliative care services, which have traditionally been involved in a supportive way with patients and families. However the care of these patients may involve several different caring teams, such as elderly care services, primary care, social supportive care and rehabilitation services and there is need to be aware of the care collaboration and coordination of care between services, so that issues are not missed and there is no duplication (10).

Physical issues

As a group, older people have many unmet needs. They experience multiple problems and disabilities and require more complex packages of treatment and social care. The physical needs are common, and many are seen for all the main diagnoses. In particular problems in mobility and communication affect more than 80% of patients, weakness, spasms and bowels problems more than 50% and very frequent are also pain, psychiatric symptoms, sleep disturbance and bladder disorders (11, 12). The continuation of treatments may be important to minimise symptoms - for instance ensuring that dopaminergic medication continues for a patient with PD, and this may necessitate the use of a naso-gastric tube or transdermal preparations (13). There may also be treatments for patient with PD, such as deep brain stimulation or apomorphine infusions that may need to be continued as the patient deteriorates to reduce the risks of motor complications (14),

although consideration may also need to be given to withdrawal of treatment if there is no ongoing benefit.

Pain

Pain is considered an important symptom whose prevention and relief by means of early identification and impeccable assessment and treatment is a mainstream in palliative care being included in the WHO definition (15). Pain is highly prevalent in ALS/MND (16), in MS (17), in PD (18), HD (19) and dementia (20). In these conditions it can be caused by:

- Direct damage of the nerves, by demyelinization / nerve compression / deafferentation – presenting as neuropathic pain.
- Disease progression, for instance joint pain due to altered muscle tone or movement, or stiffness, cramps and spasticity.
- Secondary to other issues such as from bed sores, adverse effect of drugs or incidental pain form constipation to urinary tract infection, which may be difficult for a person with cognitive change to localise clearly.

For the effective control pain it has to be thoroughly assessed. This may be more complex for patients with communication impairment or cognitive decline. Many tools are available for this purpose, and consensus recommendations recently supported the The Pain Assessment in Advanced Dementia (PAINAD) and the The Pain Assessment Checklist for Seniors with Limited Ability to Communicate (PACSLAC) for use in assessing pain in nonverbal residents in nursing homes (21).

The treatment of pain will depend on the assessed cause. On occasions there may be an aggravating factor or medication contributing to the pain, and this should initially be stopped. In PD the careful titration of the medication may alleviate stiffness and pain. Physiotherapy and occupational therapy have an invaluable role in preventing and treating muscular stiffness, maintaining joint elasticity.

The choice of analgesia should follow the basic steps of the WHO ladder (22). Adjuvants like anti epileptics drugs, antidepressants, steroids can be added if appropriate. In the elderly it is important to titrate doses slowly and assess carefully for side effects. In this population it may also be important to assess the method of administration, for instance a transdermal patch, such as of buprenorphine or fentanyl, may be helpful if there are severe swallowing problems.

Many elderly people minimise their pain and the Panel on Persistent Pain in Older Persons of the American Geriatrics Society found that 25‐50% of older people living in the community have major pain problems, and 45‐80% of nursing home residents have substantial pain that is undertreated (23). Since older people commonly have multiple health problems (including arthritis and other bone joint and back problems), they often have several sources of pain (24). Many patients suffer from more than one pain, but overall are less likely to receive adequate painkillers if compared with cancer patients (19) even though it has been shown that opioids are effective and safe (25,26).

Dyspnoea

Shortness of breath is very common in some chronic conditions in the elderly, such as Chronic Obstructive Pulmonary Disease (COPD) and Chronic Heart Failure (CHF), but it is also very burdensome in neurological conditions like ALS/MND, advanced MS and Parkinsonisms. Dyspnoea is a symptom and has to be assessed in parallel with physical examination and respiratory testing. It can be caused by the disease, as in ALS/MND or MS where it is due to the insufficiency of the respiratory muscles, due to muscular rigidity in Parkinson's disease or PD, MSA and PSP, related to muscular exhaustion for continuous and relentless movements in HD or to neurological cachexia typical of advanced dementia.

Posture can play a determinant role in respiratory distress, such as if the person has restricted chest movement due to deformity or immobility. Rehabilitation, ensuring maintenance of physical functioning and mobility, play a fundamental role in prevention and treatment.

If there is evidence of respiratory failure, such as in MND/ALS, non-invasive ventilation may be considered, and, on occasions, invasive ventilation via tracheostomy (27). These two options have been shown to improve survival and quality of life in MND. It is not always well tolerated by old and frail patients and requires adaptation and collaboration from the patient.

Pharmacological treatment of dyspnoea is based on morphine and other opioids. It has been recommended in neurological guidelines (28) but its use is still too low, even though it has been shown to be safe when used carefully (26).

Dysphagia

Dysphagia and nutritional issues are very common in the elderly affected by neurodegenerative disorders. It is normally determined by the lack of coordination of the muscles of swallowing. This symptom can occur at different times during the various disease trajectories, but it may develop particularly at the end of life phase (29).

Careful feeding is an appropriate and sensitive approach that can often be adopted until very advanced stages of the disease. The adoption of accurate postural hygiene techniques, mouth care, appropriate food consistency can allow to provide enough food and liquid to very ill and frail patients. In some neurological conditions, such as MND the placement of a PEG (Percutaneous Endoscopic Gastrostomy) has been suggested when swallowing impairment becomes more noticeable and weight is lost and may improve the patients' quality of life and prevent aspiration pneumonia (27). However in many elderly people with neurodegenerative disease PEG placement may not be helpful and there is no evidence of positive impacts in terms of survival, quality of life or prevention of collateral effects (30).

In the last days of life it is also important to explain to the family members that their beloved one is not starving to death, but losing the capability to eat is part of the normal process of dying. However as eating and feeding are often perceived as a normal life sustaining procedure there are often

ethical discussions as to whether to use enteral tube feeding or intravenous/ subcutaneous fluid replacement. Discussion with the family and, if possible, the patient may allow a clear plan to be made, accepting that death is now inevitable and the patient's best interests may be best met by comfort care alone.

Confusion

Decline in cognitive function can be a common feature of many neurological disorders in the advanced stages. Typically in Alzheimer's Disease and dementias the clinical picture is dominated by loss of memory and relentless cognitive deterioration that leads to loss of autonomy and total dependency in the ADL. This can appear in other conditions like the HD or in PD and PSP. MS often cause psychiatric symptoms and in later stages it is not easy to discern between severe communication impairment and cognitive decline. In MND/ALS Frontal Temporal Dementia (FTD) is seen in about 15% of patients and milder cognitive change, which may affect executive, decision making, in up to 50% of patients (31).

Confusion has to be assessed carefully to detect possible reversible causes. For example systemic infections, like the urinary tract infection, are very frequent in disabled elderly patients, above all if not well hydrated or with indwell bladder catheters, can cause acute confusion state. In this case a specific antibiotic treatment can reverse the confusion. It also important to ensure that other medication is not adding to confusion, or that medication is not being taken correctly. In PD medication needs to be taken regularly and at the correct dose and if this is not checked the patient can become confused and unwell.

Urinary and bowel problems

Urinary incontinence or retention may be due to direct neurological damage, or as a consequence of a general physical decline. Careful attention to any issues that may be exacerbating the problems is important but urinary catheterisation may become necessary.

Constipation is very common in PD, but can appear in all neurological conditions. It can be worsened by the use of symptomatic drugs like opioids, anticholinergics, and PD specific treatments. It is very important to provide adequate education to patients and carers on how to take liquids, manage residual mobility, posture and use correctly laxatives.

Psychological issues

A person with a neurological disease faces many issues which cause psychological and emotional distress. This may be compounded in an elderly person, who is also facing issues and problems associated with other co-morbidities. The particular issues that may be faced are:

- Concerns and fears of increasing disability
- Fears over coping at home – either being a burden on spouse/ family or ability to cope alone
- Fears over the process of dying – many neurological illness may be associated with a distressing death. For instance due to the publicity of some people with MND applying to the courts for assisted dying there has been discussion about the distress of choking to death with MND, even though the research has shown this to be very rare (25)
- Fears of dependency if they become more disabled and less mobile
- Concerns about wishing to end their life – although in most countries assisted dying is not legal many people may express the wish to die (32) – either as a result of actual or perceived risk of distress – and may feel isolated as they feel unable to express these concerns either or family and friends or professional carers (33)
- Feelings of depression or anxiety as they face physical symptoms or increasing disability or reduced abilities – in mobility, speech, swallowing and breathing.

- Fear of losing control – of bodily functions or of their cognition and ability to communicate and be themselves. This may be particularly seen for a person with a neurological disease which leads to cognitive change, dementia or PD or MND/ALS, as they are always aware that any small change may be a sign of mental deterioration and the onset of dementia.

There may be no easy answers to these concerns but professional carers should be open to discuss these issues, and be able to allow people with neurological disease to express their views and fears. It may also be important to facilitate discussions within families so that the fears and concerns can be shared (34).

Social issues

Most people are part of a wider social group – whether family or friends. However an elderly person may have particular issues as they may be bereaved, from their partner and/ or friends leading to less support and increasing loneliness or isolation.

The spouse / partner and other family may have similar fears and concern as the patient – about the disease, disability, dependency and dying and death. It is important that they have the opportunity to share these concerns and, if possible, share together with the patient (34). There may also be very practical issues providing care as the partner, and often the children, who may themselves be over 65 years old, may have their own physical issues or mental / emotional issues. If the partner has a serious co-morbidities themselves, or evidence of cognitive change or dementia it may be very difficult to provide care at home, as the patient may have been the primary carer within the relationship and the change of roles may cause great disruption within the household. The housing may also need to be reassessed if there are issues with mobility or fears of safety, and this may be a source of distress if an elderly couple have lived in their present housing for many years.

Spiritual issues

The spiritual issues faced by a person with progressive neurological disease may involve their own particular experiences, beliefs and values. This may be manifested in a religious belief and practice but many people may not espouse such beliefs, but still have spiritual values and may search for meaning in their illness and lives (35). There may be no easy answers to these questions – such as the meaning of life or of suffering – but it is important that they are heard and acknowledged. Counselling and involvement of faith leaders may be helpful for some.

Carer support

The support of carers – both family and other carers supporting the person and their family at home - is very important. They will have their own particular concerns, as outlined above, which need to be addressed (34). Time is needed to hear the family and carers concerns and wishes, which may need to be adjusted to take into consideration the issue that the person feels. However time supporting the family may allow a person to have their wishes more carefully respected and enable the care to continue.

Many elderly people will be within a residential care home or other facility. The carers there may also need their own support and attention to their concerns is important if the patient is to remain in the home setting and feel secure and well supported. Carers may have variable experience and have their own concerns, particularly of dying and death. A clear plan of care is important and many care homes now allow for extra training of their staff and a wider multidisciplinary discussion, including the patient if possible, the family and the general practitioner, to ensure that all know the plan of care if problems are encountered.

Professional support

The care of a person with a progressive neurological disease can be stressful and hard for the professional carers, particularly if there is cognitive change and confusion. Support of the professions is important and the EAN / EAPC Consensus recommends “Professionals involved in the

care of progressive disease should receive education, support and supervision to reduce the risks of emotional exhaustion and burnout" (5). Ongoing support and supervision is very helpful and should be encouraged at all levels of care.

End of life care

As the disease progresses there is the need to recognise and prepare for end of life. The recognition of this stage may be complex in many patients, particularly when there are co-morbidities. However there are certain triggers that have been suggested for recognition of end of life for people with neurological disease, which may be seen as relevant in the elderly:

- swallowing problems
- recurring infection
- marked decline in functional status
- first episode of aspiration pneumonia
- cognitive difficulties
- weight loss
- significant complex symptoms (36,37).

The recognition of this phase is important in allowing families and carers to be more prepared for the death and for all to make appropriate preparations. For instance for some patient and their families it may be the opportunity to ensure that the person's wishes for care at the end of life, including place of death, their Will, the funeral arrangements and wishes for people they wish to see can be ascertained and if possible acted on.

There is often a need to ensure that some of these preparations are made in advance of the final stages of life, as there may be a reduced ability to communicate, due to speech difficulties, or cognitive change. If these are likely discussion of advance care plans is important while the person

can make decisions for themselves, so that if they do lose the capacity to make these decisions, due to communication loss or cognitive change, their wishes area known and can be respected. The discussion of the advance care plan may be difficult but may allow a person to clearly define their wishes.

There is also the opportunity for carers and professionals to ensure that preparations for care at the end of life have been made, including discussion and completion of Do not attempt cardiopulmonary resuscitation orders, ensuring advance wishes are undertaken and appropriate medication for end of life care – such as injections of morphine for pain, midazolam for agitation and an anticholinergic medication, such as glycopyrronium bromide, to reduce respiratory secretions and an antiemetic , are available at home or in the ward situation so that they are readily accessible for administration if they become necessary. In the UK the MND Association have prepared a leaflet and a box for this medication, known as the "Just in Case" kit (38).

Conclusion

The careful assessment of the issues faced by a patient with progressive neurodegenerative disease, and their family, is essential by a wider multidisciplinary team. In this way the care of the patient can be co-ordinated and the quality of life maintained as long as possible and the quality of death maximised.

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