

section 2.6.2: Neurodegenerative conditions

Motor Neurone disease (MND)

General description of the disease and progression

An incurable degeneration of motor neurones leading to loss of voluntary muscular contraction and muscle wasting that ultimately leads to death due to respiratory failure:

- diagnosis may be delayed due to the non-specific nature of the presentation of this disease and patients therefore may be at an advanced stage by the time they are seen in a medical setting
- prognosis – less than 50% survive longer than 3 years post diagnosis therefore the aim of care is palliative from diagnosis – Riluzole is the only disease-modifying treatment currently approved for use but may only provide modest slowing of progression¹
- presentation can be varied: usually commences with weakness of the skeletal muscles causing clumsiness, progressing to involve the tongue, pharynx, larynx and muscles of respiration and relentless and progressive weakness and wasting
- difficulty with speech and inability to swallow food and fluids safely may occur with disease progression:
 - impaired cough, airway protection may be compromised with an increased risk of aspiration/pneumonia
 - reduced nutritional intake may not support nutritional requirements
 - excessive, tenacious secretions are problematic
 - access to suctioning equipment is often essential
 - decreasing saliva production, dietary modification
- touch, taste, sight, smell and hearing are not affected by the disease, nor directly bladder, bowel or sexual function – intellect is usually not affected, but in a small minority there will be some degree of cognitive impairment
- mobility management
 - in advanced stages patients are often dependent on someone else for their every need, as they are unable to walk, speak or swallow food safely
 - eventually the respiratory muscles are affected and this weakness ultimately leads to death
- treatment options other than directed to relief of symptoms are limited
 - discussions regarding nutritional or ventilatory support in those patients who have bulbar involvement are complex and take time and consideration – encouraging these discussions whilst the patient is able to participate and be understood is important

Identification of potential problems

- observe for changes in speech
- inquire about any difficulties swallowing or episodes of choking
- observe for dribbling and inquire about difficulties clearing saliva
- ask about difficulties breathing, disturbed sleep, nightmares, headaches on waking and day-time sleepiness
- assess mobility and test limb function and power
- ask specifically about pain and conduct a full pain assessment

Specific considerations for end stage disease

- exploring patient preferences regarding future care options including percutaneous endoscopic gastrostomy, mechanical ventilation
- using a multi-disciplinary approach is essential with speech pathology, dieticians, occupational therapists playing essential roles
- ensure advanced directives for end of life care have been discussed
- assess caregiver status
 - emotional and physical impact, including depression and back injuries
 - loss of own independence
 - access to information and support
 - provide training in hands-on care, eg bathing, gastrostomy management
- adaptation to disease – MND is characterised by continual loss of strength and function, thus allowing little stability or time to adapt or grieve for the loss of being with their family
- access to respite – this provides patients and caregivers with a break, which can help to deal with feelings of guilt or resentment as well as create time for the caregiver to recover physical stamina
- support groups – peer support groups encourage participants to share experiences and challenges with others in similar situations from a perspective of “one who has been or is there”
- dealing with uncertainty – MND is generally a steadily progressive disease over 2-5 years, but the rate of progression varies greatly from one person to another and this varied pattern of trajectory may make uncertainty more difficult
- bereavement risk
 - initial research data indicates that bereavement in relatives after an MND patient's death may be particularly severe and prolonged², and may be due to the huge burden of care in the months preceding death
 - there may be an extended bereavement period in which the patient, caregiver and family members can also mourn both the loss of the patient's physical function and their respective independence

references

¹ Kiernan M (2005) Riluzole: a glimmer of hope in the treatment of motor neurone disease *MJA* 182 (7): 319-320.

² Martin J, Turnbull J (2000) Lasting impact, and ongoing needs, in families months to years after death from ALS. *Amyotr Lat Scler* 1(suppl 3):514–15.