



# Motor Neurone Disease

## A Problem Solving Approach



THE ROYAL AUSTRALIAN  
COLLEGE OF  
GENERAL PRACTITIONERS

Endorsed by the Royal Australian College  
of General Practitioners

*The RACGP recognises that while it would be ideal to have published trial data for the management of all the troublesome symptoms of motor neurone disease, in many situations this is not available. Accordingly, clinical practice is guided by the opinions of experienced clinicians.*

**Motor neurone disease (MND)** is an uncommon degenerative disorder of motor neurones which leads to progressive paralysis of cranial and skeletal muscles. The onset is insidious. First symptoms can include stumbling, weakened grip, hoarse voice, cramp and muscle wasting. The sensory nerves and intellect are usually unaffected. The condition is incurable and leads to death within a few years of diagnosis, generally 1-5 years. Death is most commonly due to respiratory muscle weakness and ventilatory failure.

<b>Incidence:</b>	approx. 2 per 100 000
<b>Prevalence:</b>	6/7:100 000
<b>Male/female ratio:</b>	3:2
<b>Distribution:</b>	worldwide
<b>Average age of onset:</b>	most common in middle years

Motor neurone disease is characterised by progressive degeneration of motor neurones:

**Anterior horn cells in the spinal cord** - resulting in lower motor neurone (LMN) lesions:

- skeletal muscle wasting and fasciculation
- reduced muscle tone and stretch reflexes
- weakness of limb, trunk and respiratory muscles

**Cranial motor nuclei in the brain stem** - resulting in LMN weakness of the facial, jaw and bulbar muscles [bulbar palsy]

**Upper motor neurone (UMN) lesions in the motor cortex** - resulting in degeneration in corticobulbar and corticospinal pathways:

- spastic weakness of cranial and bulbar muscles [pseudobulbar palsy]
- spastic weakness of limb and truncal muscles
- exaggerated jaw, gag and cough reflexes
- pseudobulbar affect - emotional lability
- exaggerated limb stretch reflexes and extensor plantar responses

It is typical for patients to present with symptoms in one muscle group, for example, weakness and wasting of one hand or a unilateral foot drop. LMN or UMN signs may be present elsewhere on examination. It is also typical to find evidence of LMN and UMN features in the same limb, for example, leg muscle wasting and fasciculation in combination with increased muscle tone, exaggerated reflexes and an extensor plantar response. As the disease progresses other motor segments become involved.

Some patients may present with acute respiratory symptoms.

Sensory symptoms or signs are rare and should lead to review of the diagnosis.

Fronto-temporal cognitive changes have been associated with MND. Fronto-temporal dementia is prominent in 5-10% of cases. Recent neuropsychological studies also suggest that approximately one third may suffer from mild changes in cognitive skills and processes.

The motor nuclei controlling eye movements and the voluntary pelvic sphincter muscles remain intact until very late in the disease.

## Clinical Categories

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The disease can be categorised on the basis of sites of involvement at presentation and the balance between LMN and UMN features.

### **Amyotrophic Lateral Sclerosis - ALS**

- Most common form affecting about 65% of patients
- Mixed LMN and UMN signs usually beginning in the limb(s)
- Bulbar involvement later
- More common in men than women

### **Progressive Bulbar / Pseudobulbar Palsy**

- About 25% of cases at onset
- More common in women
- Progressive dysarthria and dysphagia
- Limb, neck and trunk involvement later
- Usually show mixed UMN and LMN features although one type may predominate

### **Progressive Muscular Atrophy**

- Progressive LMN muscle wasting and weakness, without convincing UMN features
- Relatively uncommon form of MND – less than 10% of cases
- Other causes of motor neuropathy need to be carefully excluded in these patients

### **Primary Lateral Sclerosis**

- Least common pattern of disease, about 1% of cases
- Progressive UMN syndrome with limb spasticity and weakness accompanied in time by a pseudobulbar palsy

(Other rare syndromes include; flail arm syndrome, man in barrel.)

The demarcation between the different clinical groups is frequently blurred. As the disease progresses there may be considerable overlap resulting in more generalised muscle wasting and weakness.

## **Sporadic MND**

In about 90-95% of cases MND develops for no apparent reason. It was first described in the medical literature in the mid-1860s. The incidence rate is around 2 per 100,000 people per year.

## **Familial MND**

Familial MND accounts for about 5-10% of all MND cases. A specific gene defect has been detected on chromosome 21 in 20% of familial cases, i.e. in 2% of all cases of MND. The gene is responsible for the production of superoxide dismutase or SOD1, an enzyme involved in the removal of toxic free radicals. Clinically the sporadic and familial forms are indistinguishable.

## **Mechanisms of motor neurone degeneration**

Polygenetic influences may predispose to the development of MND by affecting the levels of essential neurone survival factors increasing vulnerability to free radical damage, or predisposing to deleterious effects of a low level environmental toxin, or through the immune system, or by an infective agent. There is every possibility that a cascade of mechanisms may culminate in motor neurone fallout and that future effective therapies may involve a combination of several different strategies to interrupt different limbs of a complex patho-aetiological process.

Researchers increasingly understand the mechanisms involved in the production of MND. There is a strong theoretical implication from the recognition of superoxide dismutase mutations in some cases of familial MND that free radical damage may contribute to motor neurone disease. This provides a theoretical rationale (as yet unproven in clinical trials) for antioxidant therapy. The additional theoretical postulate that glutamate excitotoxicity may accelerate neuronal loss (with some support from clinical trials) provides a rationale for agents such as riluzole.

**Antioxidants** - Vitamins E and C, beta carotene. As yet no scientifically performed clinical trial has been performed to test the effectiveness of these compounds.

**Viral infections and autoimmune mechanisms** - Continue to be areas of active analysis but have never indicated a likely aetiology or treatment alternative.

**Glutamate blockers** - The data supporting the use of riluzole in motor neurone disease has recently been subject to a rigorous Cochrane analysis. The conclusion was that a dose of 100mg a day is modestly effective in prolonging survival for people with ALS. In an Irish study riluzole was found to be probably more effective when started early in the disease course. In May 2002 riluzole was approved for prescription in Australia. It was made available on the Pharmaceutical Benefits Scheme from July 2003 for patients who meet defined selection criteria. Riluzole is marketed in Australia by sanofi aventis as Rilutek (see '*Further Reading*').

**Neurotrophic factors** - IGF-1 (myotrophin). A North American study indicated a very small survival benefit from IGF-1 but this was not supported by a European study. Trials are continuing.

Inhibition of p75NTR death signalling is being tested as a means of preventing motor neurone degeneration.

**Inflammation theory** - Inflammatory processes are being examined with trials of drugs such as Celebrex. Celebrex is ineffective in altering the rate of progression of MND. Many other experimental agents are currently under trial.

**Other developing areas of research** - the use of stem cells, axon guidance molecules and large-scale drug screening for potential value as treatments.

## **The Australian MND Registry (AMNDR)**

AMNDR provides a means to facilitate the collection and analysis of MND patient data in Australia. The goal of the registry is to improve patient care through continuous evaluation of patient management and associated outcomes. Following registration of the patient by a neurologist, other providers such as the GP or nurse can update patient details.

**More information on current research see '*Further Reading*'.**

MND is a life limiting disease characterised by a series of losses with the accompanying issues of grief and bereavement which affect the patient, the carers and the family from diagnosis.

Establishing links at an early stage can provide the primary health care team with a useful source of advice and support. It is important that people living with MND are clear about the role of palliative care services and the benefits that they can provide.

## **Palliative care services**

Each palliative care service provides its own program of services to the patient and their family. This may include: occupational therapy, social worker services, speech pathology, counselling, respite, dietary advice, loan of equipment, physiotherapy, opportunities for monitoring and review of symptom management, nursing, medical consultancy to the patient's GP, bereavement support, spiritual support, pastoral care, music therapy and a wide range of support from trained volunteers. These services may be provided in the home, hospice, residential care facility or hospital.

## **Community support services**

Community support services are subsidised by government. A range of services can be provided and may include access to: respite, counselling, home help, personal care, case management and leisure and recreational activities.

The Motor Neurone Disease Association in each state/territory is able to advise GPs and other health professionals regarding palliative care and local community support services.

## **Further Reading**

Clayton JM, Hancock K, Butow PN, Tattersall MHN, Currow D. 'Clinical practice guidelines for communicating prognosis and end-of-life issues with adults in the advanced stages of a life-limiting illness, and their caregivers'. In Press *Medical Journal of Australia*, accepted February 2007.

## **Motor Neurone Disease Associations**

Motor Neurone Disease Associations in each state can provide information on a range of services available to assist people with MND and their carer, ph. 1800 777 175. Several states have neurological clinics or MND clinics which provide a range of services to assist people living with MND, especially those newly diagnosed. Calvary Health Care Bethlehem Neurological Outreach Service in Victoria provides a national telephone consultancy service for health professionals on the management of degenerative neurological disorders, including MND, ph. 03 9595 3355.

**For end of life care see *'End of Life Care'*.**

The Motor Neurone Disease Association in each state/territory is able to advise GPs and other health professionals regarding local sources of supply of equipment which will enable people living with MND to enjoy the best possible quality of life. Most state MND Associations have a range of aids and equipment available for loan following a referral from the relevant health professional.

A range of aids and equipment can be viewed at Independent Living Centres (ILCs) which operate in every state/territory see <[www.ilcaustralia.org](http://www.ilcaustralia.org)>.

Before decisions are made regarding specific equipment, it is essential that accurate assessment be carried out by an **occupational therapist, physiotherapist, speech pathologist or nurse** depending on the area of expertise and the availability of professionals in any particular locality.

## Bathroom

- grab rails for bathroom and toilet
- chairs - shower/over toilet/commode

## Bedroom

- bed blocks
- bed sticks
- electrical high/low beds
- slide sheets

## Communication

- hands-free telephone
- call bells
- personal alarms
- magic slates/Etch A Sketch/Magna Doodle boards
- computerised communication aids including light touch keyboards and voice synthesisers
- eye-gaze boards
- computer programs
- voice amplifiers

## Daily living

- book rest
- page turner
- talking books

## Environmental controls

- personal alarms
- remote operation of lights/electrical equipment

## Furniture

- reclining chairs
- adjustable height chairs
- electric riser chairs
- mobile arm supports - wheelchair attached
- mobile arm supports - free standing or table attached

## **Grooming**

- velcro for assistance with clothing including shoes
- long handled aids for washing and hair grooming
- toothbrush holders and toothpaste squeezers
- wash mitts

## **Meals and feeding**

- modified cups, plates and cutlery
- non-slip mats
- cup holders
- collars
- splints

## **Mobility**

- walking sticks
- walking frame
- rails/ramps
- splints
- wheeled trolley
- wheelchairs - carer or patient operated, manual/electric

## **Pressure care**

- cushions
- mattress overlays

## **Transfers**

- turntables
- hoists
- lifting belts

## Cause

- Weakness and paralysis of the lips, facial muscles, tongue, larynx, and pharynx resulting from affected trigeminal, facial, glossopharyngeal, vagus, accessory and hypoglossal cranial nerves
- Weakness of the muscles of respiration will impact on speech volume

## Effect

- Impairment of speech production (dysarthria) may begin with slurring, hoarseness and weak voice. This may progress to total loss of speech (anarthria)

## Treatment

Early referral to a **speech pathologist** for:

- advice on strategies for communication
- assessment for and provision of communication aids and training in their use
- the **speech pathologist** will work with an **occupational therapist** who can advise on seating, positioning, wrist supports, switches, pointers, mobile arm supports and tables, access to communication aids, computers and environmental controls

## Useful strategies

- Take time to create a relaxed atmosphere
- Encourage the person with MND to slow down speech and carefully articulate words
- Positioning - face-to-face, watch lips, eyes, gestures, reduce background noise
- Avoid interruptions or trying to finish sentences
- Encourage writing of key words to augment verbal communication
- Ascertain the individual's own preferred means of communication
- Establish gestures or signals for 'yes' and 'no'
- Ask questions which only need a 'yes/no' answer

## Communication aids

- Writing
- Alphabet board
- Communication charts
- Perspex eye-pointing frame (ETRAN board)
- Hands-free telephone
- Call bells
- Personal alarms
- Augmentative and assistive communication (AAC) devices e.g. computers, LightWRITER
- Telephone typewriter (TTY)
- Laser head pointer

## Emotional response

Poor communication ability can lead to:

- **isolation** - communication inadequate or avoided
- **exclusion from social interaction** - as speech becomes difficult/impossible to understand
- **frustration** - for both communication partner and person with MND - need time which may not be available
- **fear and anxiety** - unable to discuss their fears and concerns
- **low self-esteem** - others assume deafness and shout or assume intellectual impairment
- **loss of control and increasing vulnerability** - because misunderstood or opinion ignored or not sought
- **increased sadness** - isolation and frustration felt by the person living with MND, carer and family

**Depression may be present but can be masked by the progression of the disease and physical changes, and exacerbated by communication difficulties.**

Pain and discomfort in MND arise as complications of muscle weakness, stiffness and immobility. Pain from pre-existing conditions such as arthritis may be exacerbated by muscle wasting.

## Causes

- Loss of muscular control to stabilise large joints and maintain spinal posture
- Passive injury to joints when controlling muscles are weak e.g. shoulder joint damage during assisted transfers
- Muscle cramps
- Spasticity
- Skin pressure
- Constipation
- Dependent oedema
- Impaired circulation

## Management of postural discomfort and joint pain

- Early referral to a physiotherapist and an occupational therapist
- Careful positioning to support head, trunk and weight of limbs
- Regular repositioning for patients unable to reposition themselves
- Passive limb movements to relieve muscle and joint stiffness
- Allied health professional advice on most appropriate aids, positioning and transferring techniques and pressure relieving equipment
- Complementary therapies such as massage may be helpful

## Analgesia

Simple analgesics may be effective initially. Paracetamol 1g qid is an appropriate first line analgesic.

As a next step, it may be necessary to trial one of the following:

- add a non-steroidal anti-inflammatory drug if there is an arthritic or inflammatory component to pain
- small initial doses of an oral opioid are often helpful. Morphine is the most versatile drug being available in a number of immediate release forms - liquid, tablet and capsule. Start low e.g. 5mg orally and increase gradually if necessary. The dose may be repeated on a 2 hourly prn basis
- if regular analgesia is required slow release preparations are the simplest options
- where oral dosing is not possible and a PEG is available a slow release morphine suspension is available (MS Contin)
- where oral dosing is not possible and a PEG is not in place but regular analgesia is required consider a fentanyl patch (Durogesic)

- where oral dosing is not possible and a patch is ineffective or not indicated for some reason subcutaneous injection may be required
- where the patient is unable to tolerate morphine alternative opioid analgesics are available, oxycodone and hydromorphone, but dosing options are limited compared to morphine
- if you are unsure about opioid dosing options contact a palliative care specialist for further information and advice
- almost all side effects of opioids improve with time except constipation. Regular aperients are essential particularly in a population already at risk from constipation. For management of constipation see '*Dysphagia, Secretions and Nutrition*'

### Muscle cramps

These are more common early in the disease course.

Drinking tonic water may ease symptoms - it contains a small amount of quinine.

Drug treatment:

- diazepam, baclofen. Start with very small doses e.g. baclofen 5-10mg bd or diazepam 1-2mg bd. Only bedtime doses may be required. Note - observe for respiratory depression associated with sedatives/opioids/diazepam

### Spasticity

Anti-spasm drugs are well worth considering in the treatment of MND. However, it must be stressed that careful assessment of the effect of spasm medication is essential. Coordination with a **physiotherapist** is essential to determine the relative role of medication and regular physiotherapy e.g. passive stretching of limbs. Advice should be sought from the physiotherapist regarding optimal positioning in bed and chair.

Drug treatment:

- baclofen, starting with 5-10mg bd. Gradually increase dose as required. It is seldom worth exceeding a total daily dose of 75mg. Possible side effects include drowsiness, increased muscle weakness and rash
- benzodiazepines e.g. diazepam, clonazepam. Patients seldom tolerate more than small doses during the day because of unwanted drowsiness. More helpful at night
- gabapentin (Neurontin) - 300mgs tds (renal function test)
- dantrolene sodium - 25mgs daily, increase slowly to 400mg per day (liver function test)

**Note:** Dosage of muscle relaxant should be carefully adjusted. The patient may experience increased weakness and decreased mobility.

People with MND, their families and carers often suffer considerable psychological and emotional distress. The major challenges are coping with loss and living with change. Much can be done to alleviate this distress, help people to adjust and make the most of their coping skills.

## **Before the diagnosis**

Anxiety can increase as a result of:

- onset of worrying symptoms
- difficulty in identifying cause
- a protracted period of investigation
- the need for the doctor to be sure before giving the diagnosis

## **Telling the diagnosis**

It is important to stress that the prognosis for people living with MND is quite variable and that some do a lot better than the average survival figures. Furthermore, some people living with MND will appear to enter a plateau where their symptoms do not change for some time.

## **Preparation**

Take time to convey the potential seriousness of diagnosis and prognosis.

## **Time and place**

Quiet, relaxed, private and away from external distractions. Choice of who will be present - partner/ family members.

## **Amount of information**

Initial shock often limits the ability to absorb information.

Need the doctor to give written information e.g. *Motor Neurone Disease - Some Facts*, *Motor Neurone Disease - More Facts*. Copies are available from MND Associations around Australia.

Need opportunities to return for more information and to set own pace.

Need an identified source of on-going support for both the patient and the carer e.g. State MND Association Care Advisory/Family Support Service.

## **Telling the truth**

Honesty is important.

Avoid leaving the person feeling alone and unsupported.

Prolonging uncertainty can exacerbate fear of the unknown, deny support and the opportunity to come to terms with mortality and to make decisions.

Telling relatives in isolation puts strains on relationships and families.

## **Emotional responses**

**Fear** - of increasing dependency, becoming a burden, of the unknown, of death and the process of dying. Allowing time and opportunities to discuss these concerns can alleviate these fears and produce practical solutions to some of them.

**Denial** - is a coping mechanism that sometimes operates alongside awareness of the condition and its implications. Unwillingness to discuss the eventual outcome of a life limiting disease, death, is often labelled as 'denial', a term which not infrequently carries with it pejorative overtones. It is important to distinguish between the individual who does not wish to talk about impending death and the one who denies the disease exists. Both denial and anger are normal grief reactions and can be associated with a refusal to accept help or to use equipment. Frustration felt by health professionals and carers confronted by these behaviours can be reduced by recognising the patient's right to retreat from the unbearable truth they face.

**Depression** - is not always easy to diagnose or differentiate from sadness and a recognition that many of life's expectations can no longer be realised. Diagnosis and treatment of depression, if present, is likely to have a positive effect on ability to cope.

**Acceptance/adjustment** - coming to terms with reality is constantly challenged by proliferating impairments and increasing severity of symptoms.

### **Emotional lability**

Upper motor neurone involvement is associated with pseudobulbar affect or emotional lability. This is a troubling symptom that occurs in as many as 50% of patients. Treatment with amitriptyline or drugs in the SSRI class may be helpful. Dextropethorphan (cough syrup) and modafinil have also been found to reduce this symptom.

### **Families and carers**

Motor neurone disease forces changes in roles and relationships. Children may need specialised support/counselling. Information on MND for children and parents is available from MND Associations. Consider ways of:

- balancing the needs of the person living with MND and other family members
- counteracting isolation of individuals and promoting awareness of each other's needs
- creating opportunities for expressing negative feelings without feeling guilty
- preventing carer burn out
- addressing the differing information and support needs of the patient and carer

The feelings of people with MND are often mirrored by their carers although not always at the same time. Additionally, responses may vary depending on cultural background.

Physical exhaustion from the caring role is coupled with powerlessness to prevent suffering. Contact the MND Association for information on carer specific resources and support.

### **Impact on professionals**

MND frequently arouses strong emotional and ethical challenges:

- attitudes to issues such as disability, quality of life, euthanasia and measures taken to prolong life
- frustration with the seeming inability of individuals or the system to provide solutions to problems
- subtle cognitive changes may impact on the individual's ability to accept advice

Multidisciplinary teamwork is necessary to provide support and encouragement. Regular case conferencing will assist with coordination, a seamless approach and support for the team.

If cognitive changes are present consult **neuropsychologist**.

## Dysphagia

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Dysphagia eventually affects about two thirds of patients.

### Causes

Weakness and paralysis of the lips, facial muscles, tongue, larynx and pharynx resulting from affected trigeminal, facial, glossopharyngeal, vagus, accessory and hypoglossal nerves.

### Effects

- Impaired ability to form a lip seal, chew, form a bolus and propel food/fluid with the tongue
- Impaired swallow reflex
- Impaired airway protection during the swallow

### Results

- Drooling
- Dehydration and malnutrition
- Aspiration and resultant chest infections which contribute to impaired respiratory function
- Severe coughing - choking risk

### Treatment

- Requires a rapid, coordinated, multidisciplinary approach and regular review
- Assessment and monitoring of swallow, advice concerning food and fluid consistency, modification of diet, modified eating and drinking utensils, percutaneous endoscopic gastrostomy (PEG) - consult **speech pathologist**
- Advice concerning modified plates, cups, cup holders, cutlery, and non-slip matting - consult **occupational therapist**
- Assessment and advice concerning nutritional intake and PEG - consult **dietitian**
- Head and neck support and positioning - consult **physiotherapist**
- Training carers to perform assisted cough - consult **physiotherapist**
- Information concerning managing swallowing difficulties, nutrition and PEG - consult **MND Association Family Support Service**

## Secretions

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Managed in consultation with **speech pathologist**.

**Drooling and pooled secretions** - anti-cholinergic medication will reduce and thicken saliva. Regular and excessive dosing may result in tenacious oropharyngeal secretions, consider prn.

Drug treatment - starting doses:

- tricyclic antidepressants - e.g. imipramine 10 mg/amitriptyline 25mgs at night
- clonidine - 0.1 mg at night
- glycopyrrolate - 0.4 mg subcutaneously up to three times a day or via a syringe driver. However, glycopyrrolate is not available on the PBS and may be difficult to obtain. It can usually be obtained through specialist palliative care services
- botulinum injections - into the parotid glands are used effectively to treat sialorrhea in some MND clinics in Australia
- atropine - anecdotal experience in community settings suggests that 1% strength atropine eye drops can be used orally to reduce saliva production. Drops can be diluted 1ml in 100ml of water and used as a mouth rinse up to three times a day. For patients who are physically unable to

rinse their mouth 2 or 3 drops can be given sublingually up to 3 times a day

In the elderly, anticholinergic drugs are associated with a significant incidence of cognitive and behavioural decline.

**Thick tenacious saliva** - check fluid intake. Mucolytic agents: papaya enzyme, juices and ice cubes - grape, apple, pineapple and papaya. Frequent swabbing of the mouth: using plain water or a mouth wash of one teaspoon bicarbonate of soda or one teaspoon salt in a glass of water, especially after meals (avoid harsh mouthwashes). Nebulised saline may also be helpful. Consider treatment with propranolol or metoprolol. Assisted cough technique can help, as can some extra water through the PEG (seems to have a diluting effect on the thick secretions for some patients).

**Dry mouth** - (often related to mouth breathing during sleep). Oral lubricants: Oralbalance gel, biotene products, 100 parts grapeseed oil to 1 part peppermint oil. Anticholinergic medication, especially at night, can exacerbate the problem.

**Choking** - attacks may be due to aspiration, impaired respiration or muscle spasm (see '*Dyspnoea and Choking*'). It should be stressed that death caused by choking attacks is rare and that the final stages of MND are usually peaceful and dignified (see '*End of Life Care*').

## Nutrition

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Advice concerning nutritional intake - consult **dietitian**. Some weight loss is an inevitable consequence of muscle wasting.

As the disease progresses eating may become difficult and exhausting, and cause anxiety. Evidence supports the use of alternative feeding via a percutaneous endoscopic gastrostomy (PEG) to improve quality of life soon after symptom onset. It is important that the person living with MND be aware of such options in good time in order to obtain maximum benefit.

Assessment by a **respiratory physician** should be performed prior to PEG placement.

For optimal safety and efficacy PEG should be placed when the patient's FVC is more than 50% of predicted. Respiratory support during PEG placement may be an option. A radiologically inserted gastrostomy (RIG) is a less invasive alternative procedure which may be an option for people with respiratory impairment.

Note: Not every patient will choose this type of intervention.

## Constipation

The sphincter muscles themselves are not weakened by MND. Incontinence is therefore not a common problem. Constipation is, however, a frequent problem and can be caused by:

- immobility
- modified diet and poor fluid intake
- medications - especially analgesics and anticholinergic drugs
- inability to strain due to weak abdominal, diaphragmatic and glottic muscles

## Management

- Optimise fluid intake if possible
- Faecal softeners if stool is hard e.g. Coloxyl tablets or lactulose syrup
- Bowel stimulants if bowel transit still slow e.g. Senna, Duroxal
- If the patient is able to swallow easily or a PEG is available - Movicol
- Regular rectal evacuant to overcome weak muscles of defecation e.g. suppositories
- Microlax enemas, usually given three times per week
- Note: Diarrhoea may be due to constipation with overflow

Respiratory weakness can develop at any stage of the disease progression and may cause shortness of breath, fatigue, impaired quality of life and somnolence. Dyspnoea is caused by weakened respiratory muscles – intercostals, diaphragm and abdominal muscles.

## Clinical features

Early in the course of the disease nocturnal hypoventilation may not be manifest by obvious shortness of breath but more by headaches or general tiredness. Later, more marked shortness of breath may appear but this tends to be at the time when the person living with MND has severe generalised weakness which may affect the throat and limb muscles. The symptoms at that time include:

- breathlessness on exertion, sometimes just the exertion of speaking or eating
- sleep disturbance, anxiety and panic
- orthopnoea - breathless lying flat
- hypoventilation is worse during sleep. Upper airway may also be partially obstructed due to bulbar and laryngeal muscle weakness
- increasing blood CO<sub>2</sub> levels result in headaches, nausea and somnolence, especially on waking
- hypoxia, especially if there is coexisting lung disease

## Management

Regular respiratory assessments including sleep studies are indicated in most persons living with MND. These should be initiated at a relatively early stage and their timing depends upon the level of abnormality detected. Where nocturnal hypoventilation is detected it may be appropriate to establish non-invasive positive pressure ventilation (NIPPV). Treatment of this type may prolong the life of a person living with MND however the primary aim of NIPPV is to improve the quality of life often at a time when limb function is still quite good.

When more severe shortness of breath appears the following measures are indicated:

- relieve anxiety - reassurance, meditation and/or medication (see below)
- correct posture in chair and bed. Beds with adjustable back and leg supports and recliner chairs may be helpful - consult **physiotherapist** and **occupational therapist**. Most people living with MND find a semi-recumbent position most comfortable. This position allows the intercostal muscles and the diaphragm to work to greatest advantage
- improve secretion control. Retained secretions in the mouth and pharynx further compromise the airway and add to the person's discomfort and panic (see '*Dysphagia, Secretions and Nutrition*'). The **physiotherapist** may also be able to reduce anxiety by teaching controlled breathing exercises and assisted coughing techniques
- Atrovent via a nebuliser may be helpful in the early stages of difficulty with secretions

## Consider

- Ideally, referral to a specialist respiratory physician should take place early in the disease course and the person living with MND, together with their family, should be guided through a decision making process on whether they wish assisted ventilation to be instituted
- Intermittent NIPPV by mask at night initially. As required during day as MND progresses
- Life support via continuous ventilation with a tracheostomy is available but is not an easy decision to make. Detailed discussions should be held with regard to this early in the progression of the disease, considering the significant quality of life issues in the face of progressive weakness

- It is important to discuss the process and timing of withdrawal of NIPPV or invasive ventilation. Implications to consider:
  - quality of life issues
  - power of attorney/guardianship, financial affairs, medical treatment/advance medical directives, end of life care
  - cost of and access to assisted ventilation
  - increased dependency on family and carers
  - the carer should be included in discussions and decision making

## Symptom control

Breathlessness is one of the most frightening symptoms of MND. Few patients choose the option of invasive mechanical ventilation, although some achieve good palliation of symptoms of CO<sub>2</sub> retention and dyspnoea using NIPPV. These benefits are often sustained for many months or years. Ultimately, however, the breathing difficulties become life threatening.

## Medication

**Opiates** - Symptom control of dyspnoea and anxiety are best achieved using opiates.

Starting dose: morphine 2mg orally or subcutaneously (the subcutaneous dose is usually half the oral dose). Increase gradually as required.

Usually patients benefit from a bedtime dose, but may use it intermittently in the daytime to assist with anxiety. When anxious or distressed by breathlessness or retained secretions in the throat, a small dose 45 minutes before meals can improve symptoms of dyspnoea while eating.

Regular dosing should be repeated 4 hourly but can be given more often if needed.

**Benzodiazepines** - Small doses of diazepam, clonazepam or midazolam may be added to help control anxiety. Clonazepam has the advantage of coming in drops which can be given more easily orally (or sublingually if there is tongue control). Midazolam or clonazepam may be needed in small doses subcutaneously for respiratory distress. Lorazepam (Ativan) can be useful as an agent to help anxiety.

**Oxygen therapy** - Oxygen therapy to treat hypoxia, in the setting of CO<sub>2</sub> retention, is likely to lead to increasing hypercarbia, coma and death and should be used with caution.

**Medications to reduce secretions** - Drooling and pooling of saliva may compromise the airway. For medications to reduce secretions see '*Dysphagia, Secretions and Nutrition*'. These agents should be used with caution as they can produce a confusional state.

## Choking

Attacks may be due to aspiration, impaired respiration, muscle spasm or stridor (due to acid reflux).

## Management

It is important to stress to the patient and their carer that death caused by choking attacks is rare and that the final stages are usually peaceful (see '*End of Life Care*').

Encourage them to stay calm and wait for the attack to pass.

Consult a **physiotherapist** for advice on assisted cough technique.

## Treatment

There are medications available which help discomfort related to choking. Medications which may alleviate this symptom include morphine, amitriptyline, benzodiazepines and glycopyrrolate.

It is important to establish the patient's wishes in regard to end of life care while they are still able to communicate easily.

The terminal stage is recognised as progressive weakness and often a sudden deterioration over a few days or hours.

The most common cause of death is respiratory failure, usually following upper respiratory tract infection.

## **The carer**

The carer and the family will need practical and emotional support. Care plans and information must be shared by all members of the care team, and adequate nursing cover needs to be maintained at the home, hospice, aged care facility or hospital. Comprehensive symptom control and optimal psychosocial support is essential in the management of a peaceful and dignified death.

## **Communicating with the patient**

This may become extremely difficult but eye pointing or single response answers to closed end questions can be maintained.

## **Symptoms**

The terminal stage may be preceded by reduced chest expansion, a quietening of the breath sounds, use of accessory muscles (if any are left), and morning headache from CO<sub>2</sub> retention overnight. The signs may be noticed by a physiotherapist or other clinician on the team. It can be useful to know so the family can be prepared for the coming days and the patient's imminent death (after many months/years of disease). This may help prevent the shock of an 'unexpected' death (with the risk of more severe bereavement) or the family inadvertently ringing an ambulance after the patient has died at home.

Note: It is important to reassure patients and carers that death from choking is rare.

## **Action**

Check all symptom control and support needs. For management guidelines refer to other sections in this booklet:

- pain
- dyspnoea
- dysphagia, dry mouth and salivation
- equipment
- dysarthria
- psychological support

Reassess needs of carer and family.

Liase with palliative care team.

## Medication

Refer to other sections of this booklet. Additionally:

- opioid analgesics - reduce cough reflex, relieve dyspnoea, control pain and help to reduce fear and anxiety
- anti-cholinergics - such as hyoscine hydrobromide and glycopyrrolate reduce saliva and lung secretions
- sedatives - such as diazepam, midazolam, clonazepam and chlorpromazine reduce anxiety
- oxygen - can be used to relieve breathlessness
- haloperidol - for terminal restlessness

**Dosage and modes of administration** of medications should be discussed with the **palliative care physician** to ensure optimal end of life care.

## Bereavement

Bereavement follow-up and support for the family may be available from the local palliative care service and the MND Association.

## Needs of the person living with MND

Information and support:

- from the onset of symptoms
- during the often protracted period of uncertainty prior to diagnosis
- ongoing in response to progressive deterioration and impact on family life

Access to the skills of experts from a variety of agencies including medical, nursing, allied health, palliative care, MND Associations and community support services for:

- assessment of needs
- rapid response
- coordinated action
- regular review
- equipment

## The multidisciplinary team

A breadth of expertise from a multidisciplinary team is necessary to manage MND and to ensure optimal care however accessing all these services is a daunting prospect.

The number of agencies, referral criteria and complexities of communication place additional burdens on the person with MND and their carer who are often still reeling from the shock of diagnosis.

**A key worker** can:

- develop a relationship as trusted family confidant
- coordinate the activities of other team members
- interface with other service providers
- organise regular case conferences
- alert team members and initiate effective and timely responses to changing needs

**Anticipating and avoiding crises** depends on:

- building and maintaining trust
- confidence in a rapid response to requests for help
- communication within the team and effective delegation of responsibility
- access to a responsive level of service

## Rarity of MND

GPs rarely see a case of MND but the primary health care team will have a great deal of experience of caring for people with terminal disease. This booklet aims to outline some of the particular problems encountered by people with MND and to provide signposts to sources of expert help.

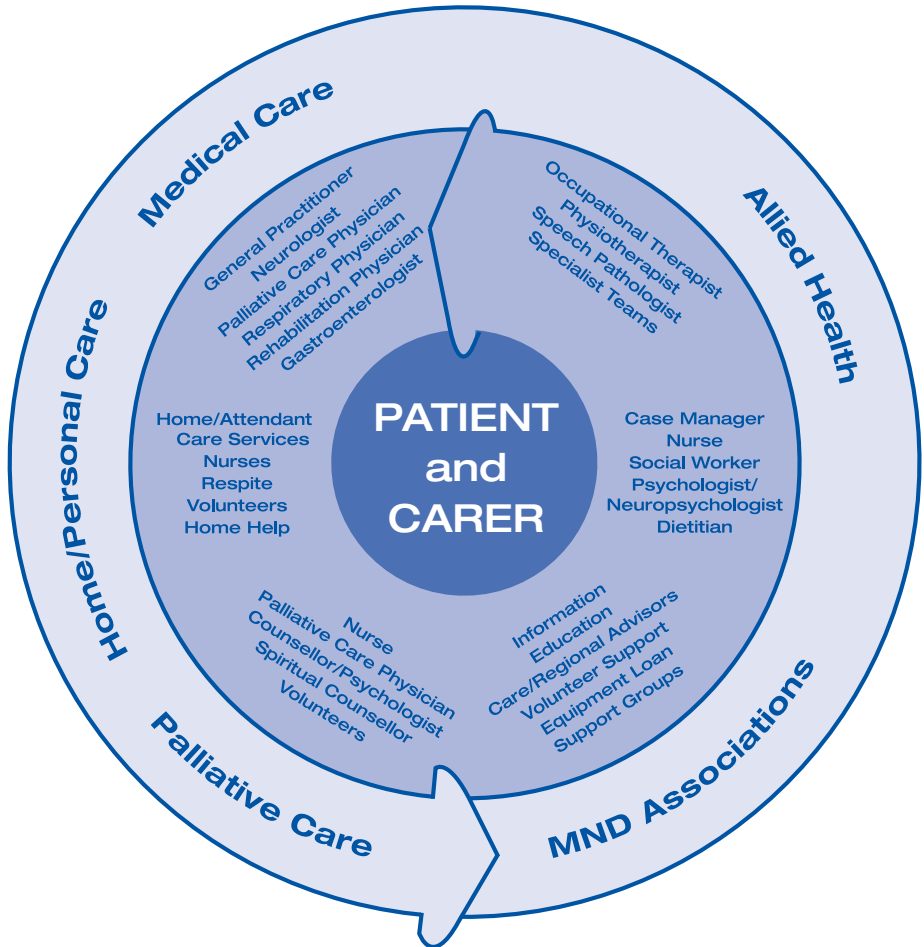
## The primary health care team

The anticipatory care approach in modern primary health care provides the ideal supportive framework to retain dignity and autonomy for the individual during the relentless progression of this devastating disease.

The primary health care team can consist of a number of health care professionals delivering a range of supports to optimise the quality of life for the person with MND. Depending on the individual situation these may include the GP (following diagnosis by a neurologist or physician), speech pathologist, occupational therapist, physiotherapist, dietitian, social worker, nurse and MND Association care/regional advisor.

Medicare funds GPs to coordinate or participate in case conferences and care planning through the Enhanced Primary Care (EPC) initiative. EPC also covers consultations with counsellors and private allied health professionals.

# Patient and Carer Centred Wheel



General financial advice is available from a range of non-government sources such as financial advisers, banks and self funded retiree associations. Fact sheets on financial matters are available from some state/territory MND Associations.

There are a number of pensions, allowances and benefits available from the Commonwealth Government for people who are aged or disabled. Financial help is also available for carers.

## Centrelink

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Some payments may be subject to an income and asset test.

### Age Pension

Must meet Australian residency requirements. Available to men 65 years and over and women aged from 60.5 years to 65 years depending on date of birth.

### Disability Support Pension

Available to people aged 16 or over and under Age Pension age who are unable to work for a prolonged period because of a disability.

### Health Concession Cards

Three health concession cards are currently available through Centrelink. These are a Commonwealth Senior Health Card, Pensioner Concession Card and a Health Care Card. All cards provide concessions on pharmaceutical prescriptions. Some state, territory and local government services will provide other discounts on services to holders of these cards.

### Carer Payment

An income support payment for people who cannot support themselves through participation in the workforce while caring for someone with MND. Contact Centrelink for information about conditions which may apply.

### Mobility Allowance

Mobility allowance helps meet the extra cost of travel for disabled workers or trainees. If the person is still working the mobility allowance can be used to purchase a modified vehicle or access subsidised mobility costs to remain employed.

### Carer Allowance

A person may be able to claim Carer Allowance if they live with and look after an adult who requires a lot of additional care because of their MND.

Further information on pensions and benefits - contact Centrelink ph. 13 2717.

Information is available in languages other than English - contact Centrelink ph. 13 1202.

Centrelink employs Indigenous Customer Service Officers to provide for the needs of people of Aboriginal or Torres Strait Islander descent.

A free Centrelink newsletter with information for carers and people with disabilities, *Disability and Carer Connections News for Carers* is available ph 13 2717.

For more information see <[www.centrelink.gov.au](http://www.centrelink.gov.au)>

## Veterans

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Other benefits may be available to veterans and their widows. Contact the Department of Veterans' Affairs ph. 133 254 or see <[www.dva.gov.au](http://www.dva.gov.au)>.

## Commonwealth Carelink Centres

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Commonwealth Carelink Centres are information centres for older people, people with disabilities and those who provide care and services. Centres provide free and confidential information on community aged care, disability and other support services available locally, interstate or anywhere within Australia ph. 1800 052 222 or see <[www.commcarelink.health.gov.au](http://www.commcarelink.health.gov.au)>.

## Commonwealth Carer Resource Centres

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Commonwealth Carer Resource Centres are located in each state and territory. These Centres provide carers with referral to services and practical written information to support them in their caring role ph. 1800 242 636.

## Commonwealth Carer Respite Centres

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Commonwealth Carer Respite Centres coordinate access to local respite services. They can give advice about respite services and also arrange 24 hour emergency respite care ph. 1800 059 059.

## The Motor Neurone Disease Associations

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These are not-for-profit organisations that aim to bring together people concerned with MND including those living with MND, their carers, volunteers and health professionals.

Their objectives are:

- to ensure that people affected by MND secure the care and support they need
- to promote research into causes and treatments

Their funds are used to support the care of people with MND by:

- providing ongoing information, education, advice and support to people with MND, their carers and health professionals
- providing equipment on loan - the range of equipment available from state associations varies
- supporting research into the disease and its management

The International Alliance of ALS/MND Associations continues to be the global organisation representing ALS/MND Associations around the world. Member organisations and associations provide direct support and services for people living with the disease. MND Australia is an active member of the International Alliance of ALS/MND Associations.

Additional information about motor neurone disease, support services and research can be found on the websites of MND Australia and State MND Associations (see *back cover*).

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## Websites of Interest

- MND Australia - [www.mndaust.asn.au](http://www.mndaust.asn.au)  
 MND Research Institute of Australia - [www.mndresearch.asn.au](http://www.mndresearch.asn.au)  
 Australian Motor Neurone Disease Registry - [www.amndr.org](http://www.amndr.org)  
 Carers Australia - [www.carersaustralia.com.au](http://www.carersaustralia.com.au)  
 Palliative Care Australia - [www.pallcare.org.au](http://www.pallcare.org.au)  
 Respecting Patient Choices - [www.respectingpatientchoices.org.au](http://www.respectingpatientchoices.org.au)  
 Independent Living Centre - [www.ilcaustralia.org](http://www.ilcaustralia.org)  
 International Alliance of ALS/MND Associations - [www.alsmndalliance.org](http://www.alsmndalliance.org)  
 MND Association of the UK - [www.mndassociation.org](http://www.mndassociation.org)  
 ALS Association of the USA - [www.alsa.org](http://www.alsa.org)  
 World Federation of Neurology - [www.wfnals.org](http://www.wfnals.org)  
 Rilutek Hotline sanofi-aventis Group - 1800 00 58 58

# Contacts

Service	Name	Phone
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MND Association .....

MND Care Advisor/Regional Advisor .....

Neurologist .....

Other Medical Specialist .....

Case Manager .....

Speech Pathologist .....

Occupational Therapist .....

Physiotherapist .....

Dietitian .....

Social Worker .....

Palliative Care .....

Psychologist .....

Home Care Coordinator .....

Nursing .....

Other .....

.....

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## Member Organisations

### MND ACT

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Freecall: 1800 642 479  
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