Motor Neurone Disease

Aspects of Care

for the primary health care team

mnd Australia

Motor Neurone Disease: Aspects of Care for the primary health care team

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Publication history

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MND Australia is indebted to the MND Association of England, Wales and Northern Ireland for permission to amend and reproduce their publication for distribution in Australia.

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Disclaimer

Publication feedback

MND Australia welcomes suggestions from people using this document so that it can be improved over time.

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Please be advised

The following information should be considered, and extreme caution taken, for potential use of atropine drops (p. 17).

A number of papers suggest the use of atropine drops orally. A Therapeutic Goods Administration (TGA) safety update in 2018 advised health professionals to exercise extreme caution if considering off-label prescribing of atropine eye drops to treat hypersalivation. This follows the death of an adult patient in 2014 (N.B. not a person with MND) that has been attributed to accidental oral ingestion of a toxic quantity of atropine eye-drop, which had been prescribed off-label for sublingual administration. Hypersalivation is not an approved indication for atropine eye-drops in Australia.

Getting Started

The primary health care team will have a great deal of experience in caring for people with terminal disease. This booklet:

- outlines some of the particular problems encountered by people with motor neurone disease (MND).
- provides signposts to sources of expert help
- is complemented by and expanded upon at MNDcare, a web-based tool developed to get evidence based and best practice MND care research into practice

MND Australia and State MND Associations

As the national body for people living with motor neurone disease, MND Australia's Mission is to work collaboratively with our members, the State MND Associations, to promote, influence and advance MND care and research to improve the lives of all Australians impacted by motor neurone disease.

State MND Association objectives are:

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

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

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

State MND Associations offer services in all states and territories. The MND Research Institute of Australia (MNDRIA) is the research arm of MND Australia.

Information about MND, support services and research can be found on the websites of MND Australia and State MND Associations (see back cover for contact details). A limited amount of information in languages other than English is available from State MND Associations.

www.mndcare.net.au

MNDcare is the MND Australia website for health and community care professionals involved in MND care and support. MNDcare:

- provides up-to-date evidence based and best practice MND information
- provides links to further related resources, for both the professional and the person living with MND
- provides referral pathways for each identified need to assist with referral to the appropriate provider or service as soon as the need is identified
- promotes a process of regular, timely review and assessment of needs
- assists Australian health professionals and community care workers to help people living with MND to live better for longer
- provides access to MND Aware online training sessions for front line staff, working with people living with MND

www.alsmndalliance.org

MND Australia is an active member of the International Alliance of ALS/MND Associations:

- the International Alliance of ALS/MND Associations is 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

Neurological clinics and MND clinics

- State MND Associations have links with the neurological clinics or MND multidisciplinary clinics which operate in most capital cities. Freecall to contact your State MND Association: 1800 777 175

- In Victoria, Calvary Health Care Bethlehem – Statewide Progressive Neurological Disease (PND) Service provides a free national telephone service for health professionals on the management of MND: Phone (03) 9595 3355 Fax (03) 9595 3301 or email: statewidePND@calvarycare.org.au

Contents

Motor Neurone Disease

Miller et al. 2009a

Amyotrophic lateral sclerosis (ALS) is a neurodegenerative disease characterized by loss of motor neurons in the spinal cord, brainstem, and motor cortex. The cause of the disease is still not known. ALS is not curable, but a number of important therapies are available.

Chio et al. 2009

Amyotrophic lateral sclerosis (ALS) is a fatal neurodegenerative disorder characterized by a progressive degeneration of upper and lower motor neurons leading to limb paralysis, dysphagia, dysarthria, and respiratory failure.

The cause of the disease is unknown and there is no effective cure.

Although it is generally reported that the mean survival of patients from symptom onset is 3–5 years, ALS has a considerable variability in outcome and its prognostic factors are not satisfactorily defined.

The median survival from onset to death in ALS is reported to vary from 20 to 48 months with ALS referral centres reporting longer survival times. This wide range narrows when considering population-based studies, which are more likely to reflect the experience of the general ALS population (20–36 months). All studies report that 5 to 10% of ALS patients survive for more than 10 years.

Incidence and worldwide prevalence of MND

- incidence: approx. 2.65 per 100,000
- prevalence: 6/7:100,000
- male/female ratio: 3:2
- distribution: worldwide

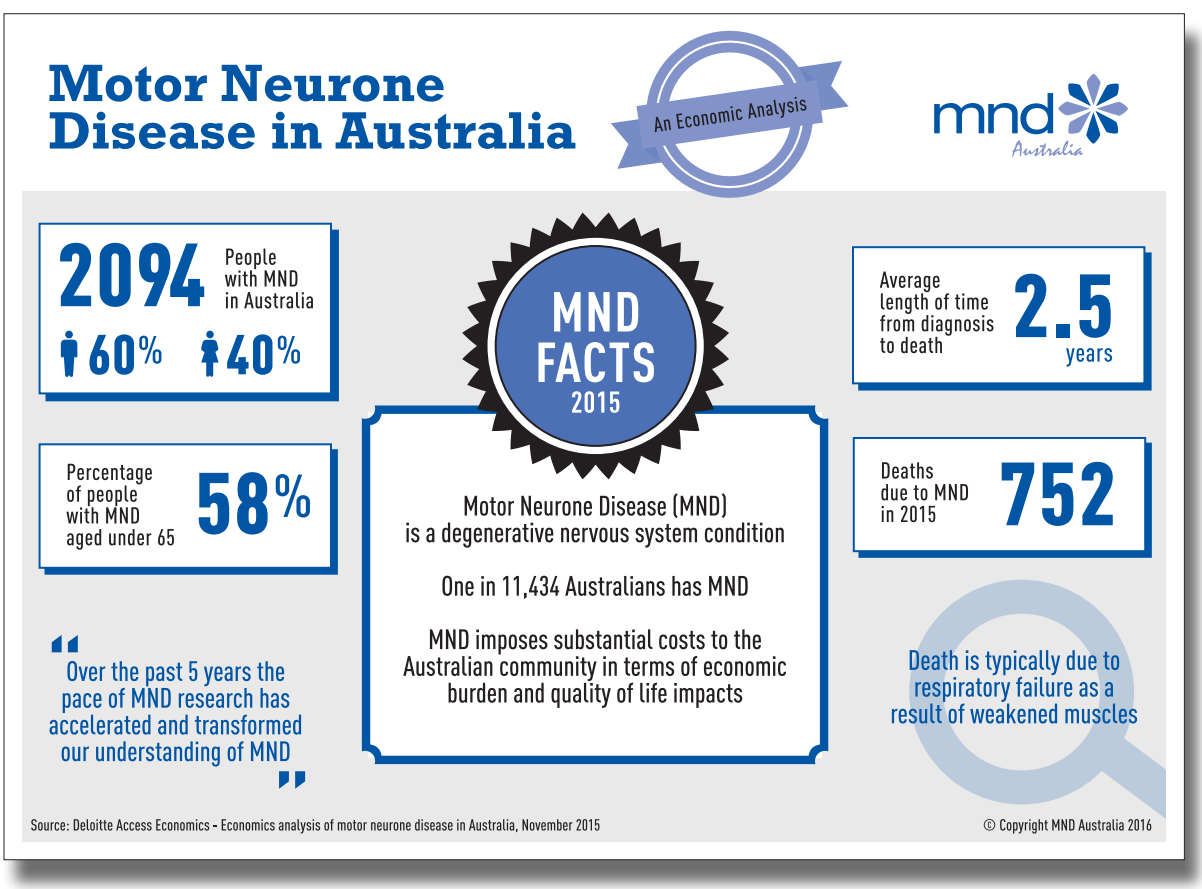


Figure 1: Infographic showing MND facts in Australia 2015. Deloitte Access Economics report available at www.mndaustralia.org.au/MNDcosts

Onset

MND onset is insidious.

Initial symptoms can include:

- stumbling
- weakened grip
- distorted speech
- cramp
- muscle wasting
- swallowing difficulties
- shortness of breath
- frontotemporal cognitive changes

Average age of onset is most commonly the middle years and onwards.

Presentation

Typically patients present with symptoms in one muscle group, for example, weakness and wasting of one hand or a unilateral foot drop.

- lower motor neurone (LMN) or upper motor neurone (UMN) signs may be present elsewhere on examination
- also typical to find evidence of LMN and UMN features in the same limb, for example, leg muscle wasting and fasciculation in combination with reduced muscle tone, exaggerated reflexes and an extensor plantar response
- frontotemporal dementia may also be present
- some patients may present with acute respiratory symptoms
- as the disease progresses other muscle groups become involved
- death is most commonly due to respiratory muscle weakness and ventilatory failure

MND can be categorised on the basis of sites of involvement at presentation and the balance between lower motor neurone and upper motor neurone features.

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

Clinical signs of motor neurone involvement	
Upper motor neurone (UMN)	**Lower motor neurone (LMN)**
Muscle spasticity	Muscle wasting
Hyperreflexia	Muscle weakness
Extensor plantar response (Babinski's sign)	Fasciculations
Preserved reflexes in wasted muscles	Absent or suppressed reflexes
Ankle clonus	
Pseudobulbar affect ('emotional lability')	

MND 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

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

Frontotemporal cognitive changes have been associated with MND. For more information see Cognition and behavioural change on page 24 and the MNDcare website at www.mndcare.net.au

The motor nuclei controlling eye movements and the voluntary pelvic sphincter muscles usually remain intact.

Phenotypes

Turner et al. 2013

With profound clinical, prognostic, neuropathological, and now genetic heterogeneity, the concept of ALS as one disease appears increasingly untenable. This background calls for the development of a more sophisticated taxonomy, and an appreciation of ALS as the breakdown of a wider network rather than a discrete vulnerable population of specialised motor neurons.

Ravits et al. 2013

Amyotrophic lateral sclerosis (ALS) is characterized phenotypically by progressive weakness and neuropathologically by loss of motor neurons. Phenotypically, there is marked heterogeneity. Typical ALS has mixed upper motor neuron (UMN) and lower motor neuron (LMN) involvement. Primary lateral sclerosis has predominant UMN involvement. Progressive muscular atrophy has predominant LMN involvement. Bulbar and limb ALS have predominant regional involvement. Frontotemporal dementia has significant cognitive and behavioral involvement. These phenotypes can be so distinctive that they would seem to have differing biology. However, they cannot be distinguished, at least neuropathologically or genetically.

The four main phenotypes of MND are:

1. Amyotrophic lateral sclerosis (ALS)

- both upper and lower motor neurones are affected
- limb muscle weakness and wasting

ALS is the most common type, characterised by muscle weakness and stiffness, over-active reflexes and, in some cases, rapidly changing emotions. Initially the limbs cease to work properly. The muscles of speech, swallowing and breathing are usually also later affected. ALS is the term commonly applied to MND in many parts of the world.

2. Progressive bulbar palsy (PBP)

- both upper and lower motor neurones are affected
- speech and swallowing muscle weakness and wasting

When ALS begins in the muscles of speech and swallowing it is designated progressive bulbar palsy. Progressive bulbar palsy (PBP), also called progressive bulbar atrophy, involves the brain stem – the bulb-shaped region containing lower motor neurones needed for swallowing, speaking and chewing.

Symptoms include pharyngeal muscle weakness (involved with swallowing), weak jaw and facial muscles, progressive loss of speech and tongue muscle atrophy. Limb weakness with both lower and upper motor neurone signs is almost always evident but less prominent.

Pseudobulbar palsy shares many of the symptoms of progressive bulbar palsy (PBP) and is characterised by degeneration of upper motor neurones that transmit signals to the lower motor neurones in the brain stem. Affected individuals have progressive loss of the ability to speak, chew and swallow. Progressive weakness in facial muscles leads to an expressionless face. Individuals may develop a gravelly voice and an increased gag reflex. People with PBP or *pseudobulbar palsy* may have outbursts of emotional lability eg; laughing or crying uncontrollably.

3. Progressive muscular atrophy (PMA)

- lower motor neurones are affected
- slower rates of progression and significantly longer survival compared to ALS and PBP

PMA is characterised initially by lower motor neurone signs resulting in more generalised muscle wasting and weakness, absent reflexes, loss of weight and muscle twitching. PMA can be the hardest form of MND to diagnose accurately. Weakness is typically seen first in the hands and then spreads into the lower body where it can be severe. Other symptoms may include muscle wasting, clumsy hand movements, fasciculations and muscle cramps. The trunk muscles and respiration may be affected. Recent studies indicate that many people diagnosed with PMA subsequently develop upper motor neurone signs. This would lead to a reclassification to ALS. PMA may begin in the arms (flail arm type) or the legs (flail leg type).

4. Primary lateral sclerosis (PLS)

- upper motor neurones are affected
- very rare and diagnosis is often provisional

PLS affects the upper motor neurones of the arms, legs and face. It occurs when specific nerve cells in the motor regions of the cerebral cortex gradually degenerate, causing the movements to be slow and effortful. The disorder often affects the legs first, followed by the body, trunk, arms and hands and, finally, the bulbar muscles. Speech may become slowed and slurred. When affected, the legs and arms become stiff, clumsy, slow and weak, leading to an inability to walk or carry out tasks requiring fine hand coordination. Difficulty with balance may lead to falls. Affected individuals commonly experience pseudobulbar affect and an overactive startle response.

Sporadic and familial MND

Clinically the sporadic and familial forms of MND are indistinguishable.

- familial MND accounts for about 5–10% of all MND cases
- MND is sporadic in about 90–95% of cases, developing with no clearly identifiable causes

Turner et al. 2013

Two decades after the discovery that 20% of familial amyotrophic lateral sclerosis (ALS) cases were linked to mutations in the superoxide dismutase-1 (SOD1) gene, a substantial proportion of the remainder of cases of familial ALS have now been traced to an expansion of the intronic hexanucleotide repeat sequence in C9orf72.

Ravits et al. 2013

Approximately 60–70% of genes involved in familial type ALS have now been identified, the main ones being SOD1, TARDBP, FUS, C9ORF72, OPTN, VCP, UBQLN, and PFN1.

Orrell 2010

Several genes are linked to classical ALS, in particular SOD1 (copper/zinc superoxide dismutase), TARDBP (TAR DNA-binding protein 43) and FUS (fused in sarcoma/translated in liposarcoma), together with genes linked to other motor neuron disorders.

In clinical practice, it is assumed that all ALS/MND patients will respond similarly to potential treatment, but it is important that in future, patients with specific genetic mutations are identified and analysed as subgroups. It may be that some gene-directed therapies will be targeted on specific genetic conditions.

Genetic factors

Our understanding of the genetic basis of MND is increasing rapidly, but the relationship between genetic risk, environmental exposure and phenotype remains largely unknown particularly in sporadic MND.

Approximately 60–70% of genes involved in familial type ALS have now been identified. As genetic factors are identified and metabolic pathways become clearer, the potential environmental factors to test may become more obvious.

Al-Chalabi and Hardiman 2013

Current heritability studies suggest that about 60% of the risk of ALS is genetically determined, and the remaining 40% is environmentally determined. An understanding of the environmental contribution to ALS is essential since this is the only easily modifiable component of risk.

Turner et al. 2013

It is widely accepted that sporadic MND likely arises from the interplay of genetic mutations and developmental, environmental and age-related factors and events. The interplay between these factors is less well understood.

Kennedy's disease

Kennedy's disease (KD), also known as X-linked spinobulbar muscular atrophy (SBMA), is an inherited disorder of the lower motor neurones. Due to similar symptoms, people with KD are sometimes misdiagnosed as having MND. Kennedy's disease is characterised by slowly progressive weakness and wasting of muscles with only lower motor neurone involvement and other features.

- Kennedy's disease is an inherited disorder affecting adult males
- Females who have a father with KD are inevitably KD gene carriers and their sons (and daughters) have a 50% chance of inheriting the KD gene
- Female gene carriers do not usually develop any KD symptoms
- Males who have inherited the gene will develop KD symptoms as adults
- Kennedy's disease cannot be passed from father to son

Disease process

Disease process mechanisms

Turner et al. 2013
The enormous advances in scientific research during the past two decades have revealed the complexity of ALS pathogenesis which might have been underestimated at the time of the discovery of SOD1. Developments in molecular biology, particularly the discovery of C9orf72, when combined with new approaches in clinical neurophysiology and neuro-imaging have accentuated rather than lessened the importance of detailed clinical assessment. The mulitple pathogenic processes support the view of multiple routes to a common endpoint of progressive upper motor neuron and lower motor neuron loss, which might mean that successful treatment will ultimately involve multiple targets.

There are many theories about the mechanisms involved in the development of MND. These include:

- genetic factors
- physical trauma
- protein aggregation
- glutamate toxicity
- mitochondrial dysfunction
- dysfunctional signalling pathways
- exposure to environmental toxins and chemicals
- free radical damage
- infection by viral agents
- immune mediated damage
- loss of growth factors required to maintain motor neurone survival

It is thought that they may act individually or in combination to cause the disease.

Research throughout the world is ongoing. This has been underpinned by the development of a number of animal models (e.g. mice, zebra fish) and by DNA and tissue banks (storing DNA, neural tissue and blood samples from people with MND and control subjects).

The use of stem cells and large-scale drug screening for potential value as treatments are developing areas of research.

Disease process interventions

Riluzole

Miller et al. 2012
Riluzole 100 mg daily is reasonably safe and probably prolongs median survival by about two to three months in patients with amyotrophic lateral sclerosis.

Orrell 2010
Riluzole is currently the only medication approved by regulatory authorities for the treatment of ALS/MND, including Europe, the USA, and Australia.

There are two brands of riluzole available in Australia. Rilutek™ is manufactured by Sanofi and the generic version APO-Riluzole is made by Apotex. Both brands are available for eligible people at a subsidised price on the Pharmaceutical Benefits Scheme (PBS) under an authority prescription.

Riluzole appears to block the release of glutamate from neurones.

Miller et al. 2012
Adverse effects from riluzole are relatively minor and, for the most part, reversible after stopping the drug. The most frequent side effects are nausea and asthenia. Liver function becomes altered and requires monitoring. Riluzole affects liver function and should be prescribed with care for people who have pre-existing problems with liver function. Regular liver function testing is recommended.

Other disease process interventions

Huynh and Kiernan 2016

In the absence of a cure, the management of MND is primarily centred around symptom control and maintaining quality of life.

Other disease process interventions have been investigated but none are clinically significant to date. For more information see the MNDcare website at www.mndcare.net.au/disease-process-interventions

Clinical trials

The Australian New Zealand Clinical Trials Registry lists clinical trials including ALS/MND studies in Australia and New Zealand, as well as trials from across the globe that have been completed, are currently recruiting or are pending: www.anzctr.org.au

Special Access Scheme

The Special Access Scheme (SAS) is managed by the Therapeutic Goods Administration (TGA). SAS refers to arrangements which provide for the import and/or supply of an unapproved therapeutic good for a single patient, on a case by case basis. SAS applications are made to the TGA by registered medical practitioners, preferably the treating doctor.

Off-label prescribing

Sometimes a medicine may be licensed for one condition, but could have the potential to be used to treat other conditions or illnesses. This is referred to as 'off-label' use of an 'unlicensed medicine'. Off-label prescribing is when a registered medicine is prescribed for use that is not included in the approved product information. An unlicensed or off-label medication may be prescribed by doctors if they think it is likely to be effective for their patient and any benefits outweigh potential side effects or risks.

ALS Untangled provides reviews by clinicians and scientists of alternative and off-label MND drugs: www.alsuntangled.com

MND register and MND research

The Australian Motor Neurone Disease Registry

Clinical research is being undertaken in Australia through the Australian MND Registry, which 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, and by contributing to research.

Following registration of the patient by a neurologist, other providers such as the GP or nurse can update patient details. For more information see www.mndregistry.org.au

The Motor Neurone Disease Research Institute of Australia

The MND Research Institute of Australia (MNDRIA) is the research arm of MND Australia. MNDRIA promotes, supports and funds MND research with the greatest chance of making an impact on the future of MND.

The MND Australia Research Committee reviews grant applications and determines the distribution of funds within set policies, and according to stringent scientific assessment criteria.

Information on current research projects funded by the MNDRIA is available at www.mndresearch.org.au

The MND Australia Research Meeting is an annual meeting where researchers are invited to present their work.

Past presentations and research meeting summaries can be found at www.mndresearch.org.au

Current research reports and information

- MND care: www.mndcare.net.au
- MND Australia and State MND Associations: (see back page for contact details)
- MND Research Institute of Australia: www.mndresearch.org.au

MND care

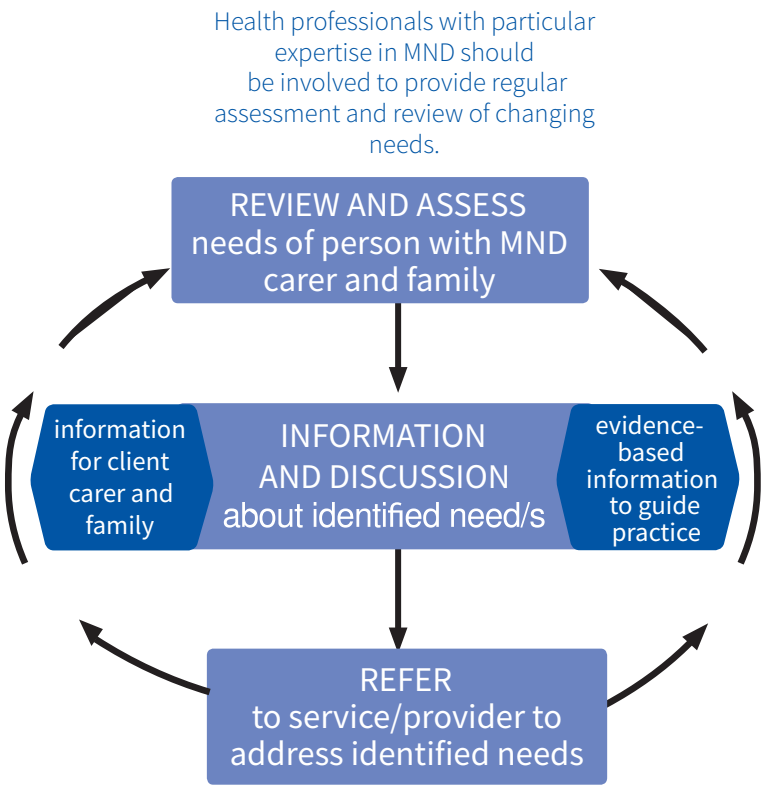
- MND may start in different areas of the body and progress in different patterns and at different rates
- care is very different for every person diagnosed
- there is no 'recipe' for managing MND but the MNDcare approach provides a framework for management
- the needs of people with MND are complex and vary from person to person.

MNDcare approach

www.mndcare.net.au

A person living with MND needs:

- information and support:
  - from the onset of symptoms
  - during the often protracted period of uncertainty prior to diagnosis
  - ongoing in response to diagnosis, 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, disability and community support services for:
  - assessment of needs (See Quality Measures in MND Care on page 34)
  - rapid response
  - coordinated action
  - regular review
  - equipment



Multidisciplinary care

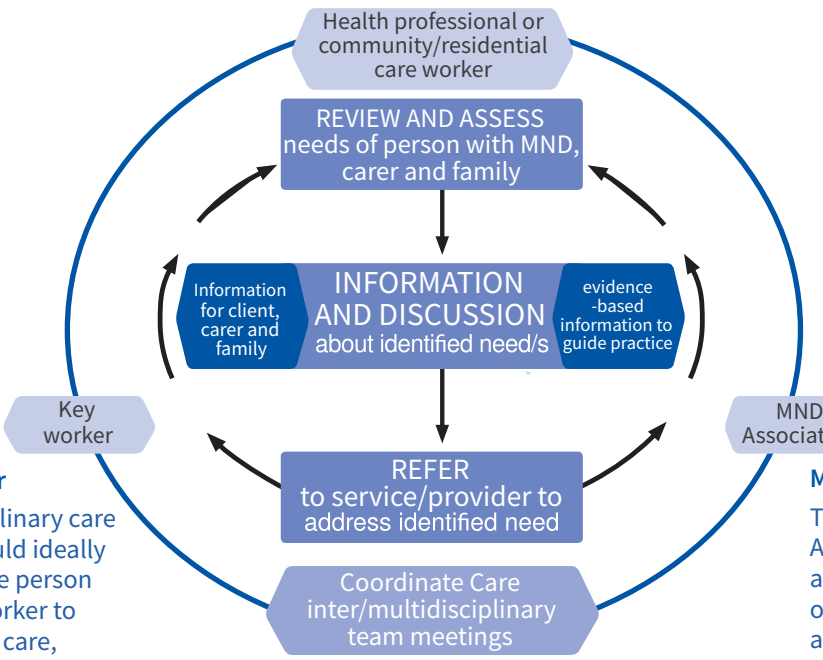
Mitchell et al. 2008

Multidisciplinary care occurs when professionals from a range of disciplines work together to deliver comprehensive care that addresses as many of the patient's health and other needs as possible. This can be delivered by a range of professionals functioning as a team under one organisational umbrella or by professionals from a range of organisations, including private practice, brought together as a unique team. As a patient's condition changes over time, the composition of the team may change to reflect the changing clinical and psychosocial needs of the patient. From the individual patient's point of view, this distinction is not critical — patients 'see' care delivered in similar aliquots, and by a similar range of disciplines.

The key worker, health professionals and community/residential care workers, and the MND Association coordinate care at inter/multidisciplinary team meetings.

Health professionals and community/residential care workers

There may be numerous (more than ten) health professionals and community/residential care workers from a range of services and organisations each reviewing and assessing the needs of the person with MND, their carer and family.



Key worker

Multidisciplinary care teams should ideally identify one person as a key worker to coordinate care, maintain all contacts, to facilitate referrals and to organise regular meetings.

Coordination

Coordination of care is vital as many professionals and services are likely to be involved in the care of a person living with MND during the disease trajectory.

Regular team meetings ensure an optimal coordinated multidisciplinary team approach and the continuation of regular assessment and review throughout the course of the disease.

MND Association

The State MND Association MND advisors provide ongoing support and assist people living with MND and their families to navigate the complex service system and to access services based on their needs.

Palliative approach

A palliative approach is required following a diagnosis of MND to ensure that early discussions around future care management decisions and advance care planning are held and optimal symptom management for the person with MND and their family is achieved.

- the aim is to assist people with MND to maintain quality of life and also to support MND carers to maintain their own health and well-being through a coordinated inter/multidisciplinary team approach
- this palliative approach is conducted within a neuro-rehabilitative framework in order to achieve holistic care and support based on the needs and wishes of the person with MND and their family

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 carer and the family from diagnosis.

- establishing links with palliative care professionals 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

Palliative care services provide their own program of services to the patient and their family in the home, hospice, residential care facility or hospital.

Services may include:

- counselling
- dietary advice
- loan of equipment
- medical consultancy to the patient's GP
- music therapy
- nursing
- occupational therapy
- opportunities for monitoring and review of symptom management
- physiotherapy
- social worker services
- speech pathology
- respite
- support – pastoral, spiritual, bereavement
- support from trained volunteers

MND clinics

Specialist MND clinics are available in Adelaide, Brisbane, Canberra, Fremantle, Perth, three locations in Victoria including Melbourne, and various locations in New South Wales. Referral to a MND clinic is recommended, where feasible, to provide confirmation of diagnosis. Teams at MND clinics are knowledgeable about MND and provide a coordinated multidisciplinary approach to care and ongoing expert review. The specialist MND clinics have links with other specialists, local primary health and palliative health professionals and the State MND Associations in each state.

Community support services

Community support services may be 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.

NDIS and My Aged Care

There are two portals of entry to be eligible to receive subsidised services:

- National Disability Insurance Scheme (NDIS) – for people aged under 65 years
- My Aged Care – for people aged 65 years or older

The State MND Association in each state/territory is able to advise GPs and other health professionals about local community support, subsidised services, palliative services and MND clinics. They also offer assistance to people with MND and their families to access these services.

Diagnosis

Anderson et al. 2012

Our objective is to present guidelines for making the correct diagnosis as early as possible. As no single investigation is specific to ALS, and there is no sensitive and disease specific biomarker, diagnosis is based on symptoms, clinical examination findings and the results of electrodiagnostic, neuroimaging and laboratory studies.

The diagnosis of MND is often clinically difficult, and sometimes it is necessary to review a person and observe symptom progression for some time before the diagnosis becomes reasonably certain. A general practitioner may suspect a neurological problem and organise referral to a neurologist. For MND signs and symptoms refer to Onset and Presentation on page 2.

MND Australia has developed a 'red flags' diagnostic tool for Australian GPs and health professionals. The tool (Figure 2) lists common MND signs and symptoms, prompts GPs to query MND and highlights the importance of initiating early referral to a neurologist.

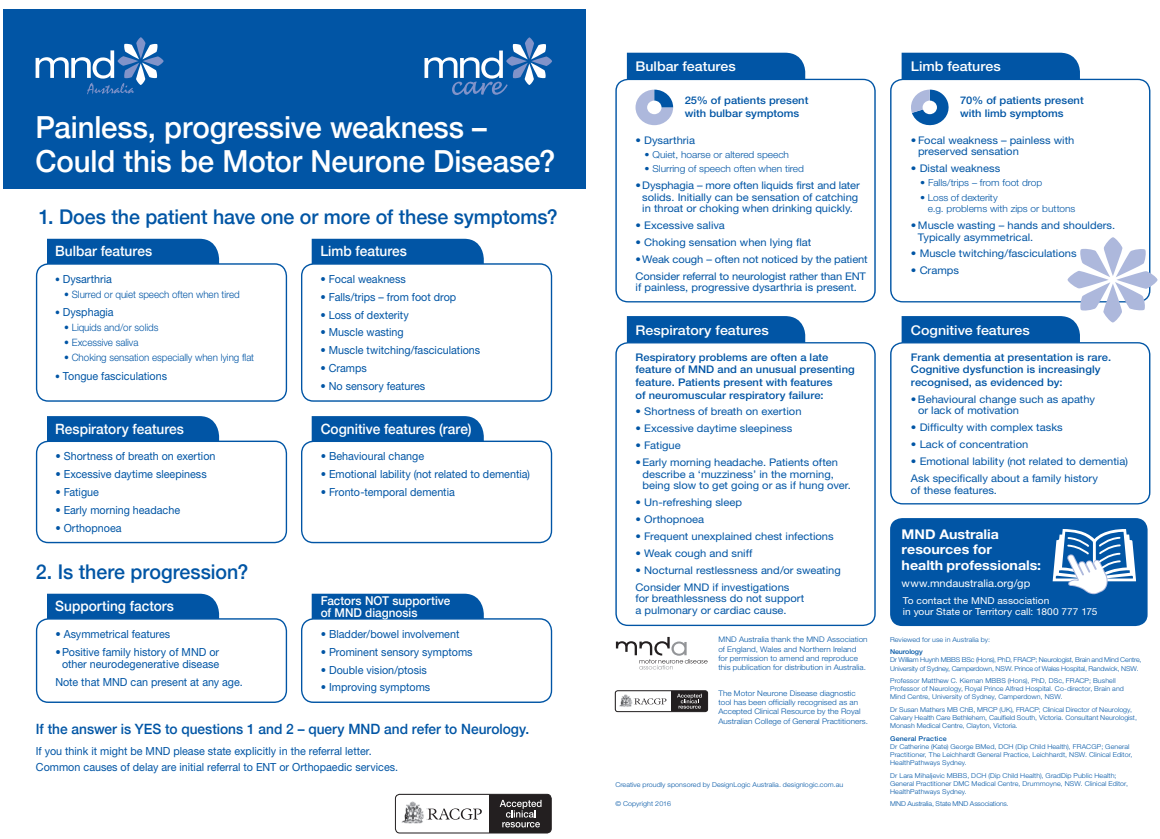


Figure 2: MND diagnostic tool illustrating 'red flags', respiratory and cognitive features of MND. Download available at www.mndcare.net.au/Diagnosis/Could this be MND?

Common diagnostic tests

There is no single diagnostic test for MND. Diagnosis is based on features in the clinical history and examination, usually accompanied by electrophysiological tests including electromyography (EMG) and nerve conduction studies.

Other tests may include:

- MRI scanning of the brain and spinal cord
- various blood tests
- lumbar puncture
- muscle biopsy

The purpose of these tests is to exclude the presence of other neurological conditions. In the early stages of MND, symptoms can be similar to those seen in other conditions, so people may spend months seeing various specialists before a diagnosis is reached.

Before the diagnosis

Anxiety can increase as a result of the onset of worrying symptoms, the difficulty in identifying a cause, the protracted period of investigation and the need for the doctor to be sure before giving the diagnosis.

National Institute for Health and Care Excellence (NICE) 2016

If you suspect MND, refer the person without delay and specify the possible diagnosis in your referral letter. Contact the consultant neurologist directly if you think the person needs to be seen urgently.

National Institute for Health and Care Excellence (NICE) 2016

Provide information and support for people and their family members and/or carers (as appropriate) throughout the diagnostic process, particularly during periods of diagnostic uncertainty or delay.

Giving the diagnosis

Communication of a diagnosis of MND is daunting for patients and neurologists. Results from a national survey showed that many health professionals lacked confidence imparting a diagnosis of MND.

Aoun et al. 2015

The communication of an MND diagnosis has implications for the way that patients and their families move from the news of their diagnosis to the actions required for support throughout the illness trajectory. This study reported on patients experience receiving a diagnosis of MND. Findings from the study centred around the SPIKES protocol – a widely accepted protocol for communicating bad news. 1) Setting; creating the right setting – preferably a private space with no interruptions. 2) Perception; determining what the patient/family already knows and how much detail they want. 3) Invitation; exploring what the patient/family are expecting or hoping for. 4) Knowledge; sharing information about how the diagnosis was reached, current state of understanding, current research and clinical trials and how to join the Australian MND Registry. Delivering the diagnosis accompanied by a consult letter, life expectancy, information and resources, contact details for the State MND Association and websites for further information. 5) Empathy; responding empathically to the feelings of the patient/family, delivering the diagnosis with warmth, care and empathy and allowing time to express their emotions. 6) Strategy; making a plan for immediate referral to State MND Association and community support networks and, if available, a multidisciplinary MND clinic referral. It was evident that the longer the patient spent with the neurologist during breaking such bad news the more satisfied they were with the experience.

Andersen et al. 2012

Imparting a diagnosis of ALS requires skill. If not performed appropriately, the effect can be devastating, leaving the patient with a sense of abandonment and destroying the patient-physician relationship. Patient/caregivers are more satisfied if effective communication strategies are used, and more time is spent in discussing the diagnosis.

Prognostic and end-of-life communication is a vital skill for health care professionals caring for patients with progressive life-limiting illnesses, and their families. The Australian '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' (Clayton et al. 2007) were developed to address these issues.

See the MNDcare website at www.mndcare.net.au/diagnosis/giving-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.

Aoun et al. 2017

Communicating the diagnosis of MND should always include family carers. Information on support provided by MND Associations should always be available at diagnosis. Results also showed that more attention should be given to family carers as a vital member of the MND care triad. And neurologists may require education and training in communicating an MND diagnosis.

Gallagher and Monroe 2006

Psychosocial care is the responsibility of everyone working with those affected by ALS.

Preparing to give the diagnosis

People with MND, their families and carers often suffer considerable psychological and emotional distress from diagnosis onwards.

- 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

Preparation

- take time to plan how the diagnosis will be communicated

Place, who is present and timing

- choose a quiet, relaxed, private space away from external distractions
- consider who will be present – partner, family members
- consider timing to ensure that the person can access other services if desired, for example, breaking the news on a Friday afternoon can greatly exacerbate anxiety

Telling the truth

- honesty is important
- avoid leaving the person feeling alone and unsupported
- prolonged uncertainty can exacerbate fear of the unknown, deny support and the opportunity to come to terms with mortality and to make decisions
- telling people in isolation can place unnecessary strain on relationships and families

How much information?

Initial shock often limits the ability to absorb information. At diagnosis people need:

- written information from the doctor e.g. MND – Some Facts, MND – More Facts. Copies are available from your State MND Association or for download at www.mndaustralia.org.au/information-resources
- opportunities to return for more information and to set own pace
- an identified source of on-going support and information for both the patient and the carer e.g. State MND Association.

Aoun et al. 2016

Best practice guidelines for MND stress the need for support for patients and family carers from diagnosis so that the individual's spiritual and emotional needs may be addressed alongside medical and practical needs. People living with MND require sensitive communication of the diagnosis combined with appropriate information about MND and support services, ensuring the provision of emotional/psychological support, a follow-up appointment within two weeks of diagnosis and direct referral to the relevant MND association.

Living with MND

Symptom management

The effects of MND – initial symptoms, rate and pattern of progression, and survival time after diagnosis – vary significantly from person to person.

> Miller et al. 2013
> The diagnosis of ALS has profound implications for the patient and his or her family. In recent analysis, too few patients received evidence-based treatment that can ease the disease burden. Although incurable at this time, advances in contemporary care options to the patient with ALS have been shown to prolong life and also to improve quality of that life.

> Miller et al. 2009b
> Effective management of symptoms is one of the primary goals of ALS patient care.

Breathing

> Berlowitz et al. 2016
> Respiratory failure is associated with significant morbidity and is the predominant cause of death in MND/ALS.

Respiratory weakness can develop at any stage of disease progression and may cause shortness of breath, fatigue, impaired quality of life and somnolence.

- referral to a specialist respiratory physician should take place early in the disease course and regular assessment initiated
- the way respiratory symptoms are managed may affect how other symptoms that may arise can be managed

Dyspnoea or shortness of breath

Dyspnoea is caused by weakened respiratory muscles – intercostals, diaphragm and abdominal muscles.

- dyspnoea is one of the most frightening symptoms of MND and there are a number of strategies that help to address this symptom and sensations of breathlessness
- early involvement of respiratory and palliative care specialists will be helpful in addressing this symptom and supporting the person with MND and their family

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 may include:
  - breathlessness on exertion, sometimes just the exertion of speaking or eating
  - sleep disturbance, anxiety and panic
  - orthopnoea – breathlessness lying flat
  - quiet voice and fewer words per breath
  - weakened cough and sneeze
  - hypoventilation is worse during sleep. Upper airway may also be partially obstructed due to bulbar and laryngeal muscle weakness
  - increasing blood CO_2 leads to restless sleep, inability to lie flat, headache on waking, daytime sleepiness, nausea, loss of appetite and rapid weight loss

Management

Regular respiratory assessments, including sleep studies, by a respiratory physician are indicated in most people 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 for the respiratory physician to initiate and prescribe non-invasive ventilation (NIV)

Non-invasive ventilation (NIV) and invasive ventilation

National Institute for Health and Care Excellence (NICE) 2016
Non-invasive ventilation can improve symptoms associated with respiratory impairment and can be life prolonging, but does not stop progression of the underlying disease.

Berlowitz et al. 2016
Non-invasive ventilation (NIV) improves survival in MND/ALS. The overall magnitude of benefit is 13 months and was largest in those with ALS-bulbar disease.

Rafiq et al. 2012
Respiratory muscle function significantly predicts survival and quality of life in patients with MND. The management of respiratory symptoms and maintenance of lung compliance are particularly important. NIV prolongs life and maintains quality of life in patients with MND. NIV requires a specialist service with ability to monitor and detect hypoventilation, to start NIV with the optimal machine and interface and optimised settings, to monitor the effectiveness of NIV and to manage complications. It is essential to have effective communication with the patient and family throughout the service. It is particularly important that the patient understands the role of NIV, and that the clinician records their wishes early on regarding management of intercurrent illness, increasing reliance of NIV and end-of-life care. The recent NICE guidelines concluded that such a service is a cost-effective use of NHS resources.

Evidence supports the use of non-invasive ventilation (NIV) to improve quality of life and to address the symptoms of CO_2 retention, poor sleep and dyspnoea.

Symptom control using NIV is often sustained for many months or years. A respiratory physician will need to assess the person in order to prescribe the appropriate settings for NIV.

- NIV is delivered via a mask usually at night initially and then as required during the day as MND progresses
- people living with MND are usually prescribed variable positive airway pressure (VPAP) or bi-level positive airway pressure (BIPAP) machines due to the lower level of pressure applied when exhaling. These machines can be adjusted to provide increased respiratory support if needed.

The person living with MND, together with their family, should be guided through a decision-making process related to management of respiratory symptoms and their future care. Detailed discussions should be held with regard to options early in the progression of the disease, considering quality of life, cost, support issues and the timing and process of withdrawal of NIV.

Life support via continuous ventilation with a tracheostomy (invasive ventilation) is not commonly used in Australia for a person with MND as their respiratory function will not recover and their disease will continue to progress. Some people may wish to be informed about and consider invasive ventilation.

Implications to consider for non-invasive and invasive ventilation:

- quality of life issues
- impact of other symptoms such as cognitive impairment and drooling
- cost of and access to assisted ventilation
- increased dependency on family and carers
- the impact on the carer who should be included in discussions and decision making
- power of attorney/guardianship, financial affairs, medical treatment/advance medical directives and end of life care

Oxygen

Respiratory muscle weakness occurs eventually in everyone with MND. Oxygen should be used with caution in the presence of respiratory muscle weakness because this weakness leads to the retention of carbon dioxide (hypercapnia) not hypoxia (inadequate levels of oxygen in the body).

In this situation, non-invasive ventilation may be the most appropriate treatment, as oxygen therapy can lead to further respiratory depression.

However, when oxygen levels are low, oxygen may sometimes be used with caution and appropriate monitoring – preferably under guidance of the person's specialist team to relieve symptoms/sensation of breathlessness. Oxygen can be used with NIV.

Additional management strategies and medications for dyspnoea

Rafiq et al. 2012

Practice Points:

- Domiciliary NIV therapy has been an important advance for patients with MND.
- Cough strength may be impaired in MND; a suitable cough augmentation technique can help such patients.
- Clinicians should pay careful attention to nutritional support in patients with MND.
- Pharmacological management of symptoms may include anticholinergics for excess saliva and mucolytics, for example, carbocisteine, to reduce secretion viscosity.
- Tracheostomy ventilation is an option for patients who wish to continue to live but who cannot use NIV.
- Palliative care needs, end-of-life issues and stopping respiratory support should be discussed regularly with the patient and the family.

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

- consult with a palliative care physician
- relieve anxiety – reassurance, meditation and/or medication
- correct posture in chair and bed
  - recliner chairs and beds with adjustable back and leg supports may be helpful
  - a semi-recumbent position may be most comfortable - this position allows the intercostal muscles and the diaphragm to work to greatest advantage
  - consult physiotherapist and occupational therapist
- improve secretion control
  - retained secretions in the mouth and pharynx accompanied with weakened cough further compromise the airway and add to the person's discomfort and panic, see Swallowing on page 16
  - the physiotherapist may also be able to reduce anxiety by teaching breath stacking and/or manual assisted cough techniques
  - for people with bulbar dysfunction, or whose cough is ineffective with unassisted breath stacking, consider assisted breath stacking (for example, using a lung volume recruitment bag)
  - a mechanical cough assist device can be helpful if assisted breath stacking is not effective, and/or during a respiratory tract infection.
- Atrovent via a nebuliser may be helpful in the early stages of difficulty with airway secretions

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

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

- there is no evidence that oxygen therapy could be advantageous to people with neuromuscular disease
- until more research is done, caution is recommended in the administration of oxygen to people with MND

Medications to reduce secretions

- drooling and pooling of saliva and weakened cough may compromise the airway
- for medications to reduce secretions see Swallowing on page 16 – these agents should be used with caution as they can produce a confusional state

Swallowing

Swallowing difficulties should be assessed and regularly reviewed by a speech pathologist. The goal of managing swallowing difficulties is to maintain optimal levels of nutrition and hydration and to reduce choking episodes. Speech pathologists work with dietitians to provide advice concerning food and fluid consistency, modification of diet and gastrostomy (see Gastrostomy on page 18).

Dysphagia

> Miller et al. 2009a
> Dysphagia is a symptom experienced by the patient and is prima facie evidence of swallowing dysfunction. The functional consequences are choking, aspiration, weight loss, and dehydration.

Dysphagia is difficulty in swallowing.

- caused by weakness and paralysis of the lips, facial muscles, tongue, larynx and pharynx resulting from affected trigeminal, facial, glossopharyngeal, vagus, accessory and hypoglossal nerves
- eventually affects about two thirds of people with MND

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

Dysphagia management

Dysphagia 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, gastrostomy – 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 gastrostomy – consult dietitian
- assessment and monitoring of respiratory function to assist with timing of gastrostomy – consult respiratory specialist
- head and neck support and positioning – consult physiotherapist
- training carers to perform assisted cough – consult physiotherapist
- information concerning managing swallowing difficulties, nutrition and gastrostomy – consult your State MND Association

Saliva management

Manage in consultation with speech pathologist.

Sialorrhea or drooling and pooled secretions

- position body and provide head and neck support (physiotherapist)
- reduce skin irritation
- oral swabbing and syringing
- 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: **extreme caution advised (see disclaimer section)**. Anecdotal experience in community settings suggests that 1% strength atropine eye dropscan 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
  - Atrovent Forte: anecdotal experience suggests that 2 sprays administered 3-4 times a day under the tongue, half an hour before meals or as required (dependent on timing of saliva issues) can help to manage saliva
- in the aged, anticholinergic drugs are associated with a significant incidence of cognitive and behavioural decline

Thick tenacious saliva

- check fluid intake
- mucolytic agents: papaya enzyme, papaya lozenges
- 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

Xerostomia or dry mouth

- often related to mouth breathing during sleep
- sip fluids frequently
- oral lubricants: Oralbalance gel, Biotene, 100 parts grapeseed oil to 1 part peppermint oil
- anticholinergic medication, especially at night, can exacerbate the problem

Oral hygiene

Mouth care is very important for people with MND who have tongue and throat muscle weakness. The anti-bacteria, anti-viral and anti-fungal properties of saliva are not as effective when a person has drooling saliva, thick saliva or dry mouth.

- immobility of the tongue decreases the natural ability to move food particles around the mouth – food can become trapped in the cheek pockets and this can cause mouth ulceration
- the person's mouth should be inspected each day for signs of oral thrush or ulcers
- teeth should be brushed carefully after meals
- make sure that excess secretions do not collect in the mouth, as this can cause problems swallowing
- it may be helpful to use an electric toothbrush
- low foaming toothpaste reduces the need to clear foam from the mouth when a person cannot spit due to muscle weakness
- swabbing the mouth with a cotton wool bud soaked in bicarbonate of soda and water (half a teaspoon to a glass of water) will help keep the mouth clean

Uncontrolled coughing and fear of choking

Swallowing will get more difficult over time and people with MND may experience episodes of severe uncontrolled coughing which increases their choking risk. Impaired respiration and swallow reflex, weak cough response, muscle spasm or stridor (due to acid reflux), and aspiration of food or fluids can increase choking risk. Severe uncontrolled coughing episodes are distressing for people with MND, their family and carers. It is important to stress to the patient and their carer that death caused by choking is rare.

Management

- encourage the person to stay calm and wait for the episode to pass
- advise the patient, carer and family on how to seek medical advice during or after an episode
- the physiotherapist may also be able to reduce anxiety by teaching breath stacking and/or manual assisted cough techniques
- for more information see Dysphagia on page 16

Treatment

- medications which help discomfort related to the experience of choking include morphine, amitriptyline, benzodiazepines and glycopyrrolate
- Clonazepam drops 2.5mg/ml. One drop = 0.1mg. Start with 5 drops prn
- oral morphine can reduce severe coughing episodes. Low doses such as 2–5mg as syrup, or subcutaneous injection

Maintaining nutrition

Chio et al. 2009
It is generally recognised that malnourishment is a relevant determinant of outcome in ALS

Some weight loss is an inevitable consequence of muscle wasting, however, as MND progresses eating may become difficult, exhausting and cause anxiety.

- assessment and advice from a speech pathologist experienced in evaluating and treating swallowing difficulties, including management of coughing and sensations of choking
- advice from a dietitian about dietary intake and fluid consistency modifications
- advice from an occupational therapist about helpful utensils and aids

Gastrostomy (PEG and RIG)

National Institute of Health and Care Excellence (NICE) 2016
Discuss gastrostomy at an early stage, and at regular intervals as MND progresses, taking into account the person's preferences and issues, such as ability to swallow, weight loss, respiratory function, effort of feeding and drinking and risk of choking. Be aware that some people will not want to have a gastrostomy.

Best evidence to date supports the use of alternative feeding via a percutaneous endoscopic gastrostomy (PEG) or radiologically inserted gastrostomy (RIG) to improve nutrition and potentially to provide a survival advantage.

An important consideration with gastrostomy insertion is that respiratory muscle weakness and malnutrition can affect recovery from the procedure.

It is important that the person living with MND be aware of gastrostomy options in good time in order to obtain maximum benefit.

- assessment by a respiratory physician should be performed prior to the procedure
- for optimal safety and efficacy the procedure should be undertaken when the patient's FVC is more than 50% of predicted
- respiratory support during the procedure 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
- referring the person to a specialist and a hospital experienced in inserting a gastrostomy for people with MND may reduce the risk of complications

Constipation and urinary urgency

Rudnicki et al. 2015
Autonomic symptoms have been reported in up to 29% of patients with ALS. Common symptoms involve urinary and gastrointestinal dysfunction.

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, Durolax
- if the patient is able to swallow easily or a gastrostomy feeding is available - Movicol
- regular rectal evacuant to overcome weak muscles of defaecation e.g. suppositories
- microlax enemas, usually given three times per week
- note: diarrhoea may be due to constipation with overflow
- amitriptyline for urinary urgency – 12.5mg nocte

Communication

National Institute for Health and Care Excellence (NICE) 2016

When assessing speech and communication needs during multidisciplinary team assessments and other appointments, discuss face-to-face and remote communication, for example, using email internet and social media. Ensure that the assessment and review is carried out by a speech and language therapist without delay. Review the person's communication needs during multidisciplinary team assessment and liaise with or refer the person with MND to a therapist specialising in Assistive Augmentive Communication (AAC) equipment; alphabet word or picture boards, mobile devices or tablets and high-level technology such as voice output communication aids of eye gaze technology.

Andersen et al. 2007

Communication should be routinely assessed by a speech and language therapist (SLT). The goal of management of communication difficulties in ALS patients is to optimize the effectiveness of communication for as long as possible and to concentrate not only on the disabled person, but on personal partner-to-partner communication as well.

Dysarthria

Dysarthria is impairment of speech production.

- caused by 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 also impact on speech volume
- impairment of speech production may begin with slurring, hoarseness or weak voice
- this may progress to total loss of speech (anarthria)

Speech is not the only form of communication that can be affected by MND. Muscles used for non-verbal forms of communication can also be weakened by MND. These include muscles used for facial expressions, hand gestures and body language; and the muscles used for writing, typing and operating a mouse.

Management

Communication difficulties require a coordinated, multidisciplinary approach and regular review.

- early referral to a speech pathologist
- 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

Emotional response

Difficulties with communication can lead to decreased social interaction and feelings of isolation, loss of control, lowered self-esteem and increased vulnerability.

- speech may become difficult or impossible to understand
- opinions may be not sought or ignored
- others may assume deafness or intellectual impairment

Additionally, communication aids can cause frustration for both the communication partner and the person with MND.

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

Communication support 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

Care alerters are very important as they give the person with MND a greater sense of security. Switches are also often adapted onto modified door chimes and intercom system so they can be used as a care alert device. Other alert systems include commercial pendant-type back to base alerters and pressure buzzers.

An occupational therapist can help to find the person's best means of operating a call device or intercom.

Augmentative and assistive communication devices

- writing, alphabet board, communication chart, perspex eye-gaze frame (ETRAN board)
- hands-free phone, telephone typewriter (TTY), call bell, personal alarm
- laser head pointer, computer software, tablet computer, specialised apps, eye-tracking devices, Lightwriter

A person with speech or other communication difficulties requires a coordinated, multidisciplinary approach and regular review.

Refer to:

- Speech pathologist
  - early assessment
- Occupational therapist
  - seating and positioning
  - wrist supports
  - switches, pointers
  - mobile arm supports, tables
- Speech pathologist, occupational therapist
  - communication aids
  - using a phone, tablet or computer
  - environmental control units
  - voice banking

Movement and joints

Andersen et al. 2007

The degeneration of upper motor (UMN) and lower motor (LMN) neurons, leads to progressive weakness of bulbar, limb, thoracic and abdominal muscles causing muscle weakness, stiffness and immobility.

Assessment and continuing review by physiotherapist and occupational therapist for:

- management of exercise, positioning and pain management
- prescription of appropriate assistive technology to assist independence, mobility and comfort

Daily living and mobility

National Institute for Health and Care Excellence (NICE) 2016

Healthcare professionals and allied health practitioners, including physiotherapists and occupational therapists, should assess and anticipate changes in the person's daily living needs, taking into account the following; Activities of daily living, including personal care, dressing and bathing, housework, shopping, food preparation, eating and drinking, and ability to continue with current work and usual activities; Mobility and avoiding falls and problems from loss of dexterity; The home environment and the need for adaptations; The need for assistive technology, such as environmental control systems.

People with MND need different levels of support to help them manage their daily personal activities and to remain as independent as possible.

- assessment by an occupational therapist will help identify assistive technology and strategies that can assist the individual to achieve optimal performance in activities of daily living
- consultation by a physiotherapist is important in determining assistive technology needs to maintain optimal mobility

Exercise

National Institute for Health and Care Excellence (NICE) 2016

Consider an exercise programme for people with MND to; maintain joint range of movement, prevent contractures, reduce stiffness and discomfort, optimise function and quality of life. Choose a programme that is appropriate to the person's level of function and tailored to their needs, abilities and preferences. Take into account factors such as postural needs and fatigue. The programme might be a resistance programme, an active assisted programme or a passive programme.

Pain

Brettschneider et al. 2013

Pain in ALS is a frequent symptom especially in the later stages of disease and can have a pronounced influence on quality of life and suffering. Treatment of pain therefore should be recognised as an important aspect of palliative care in ALS.

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

Pain management

- 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

Initially, simple analgesics may be effective:

- paracetamol 1g qid is an appropriate first line analgesic

As a next step, consider:

- adding 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 gastrostomy tube is available a slow release morphine suspension is available (MS Contin)
- where oral dosing is not possible and a gastrostomy tube 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
- 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 Constipation on page 18.

If you are unsure about opioid dosing options contact a palliative care specialist for further information and advice.

Cramps

- common early in the disease course
- magnesium tablets may be helpful

Drug treatment

- start with very small doses e.g. baclofen 5-10mg bd or diazepam 1-2mg bd (or at least nocte prn)

Note: observe for respiratory depression associated with sedatives/opioids/diazepam

Spasticity

- anti-spasmodic drugs are well worth considering in the treatment of MND. However, it must be stressed that careful assessment of the effect of anti-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.

Assistive technology

Your State MND Association is able to advise GPs and other health professionals regarding local sources of supply of assistive technology which will enable people living with MND to enjoy the best possible quality of life. Early availability and adoption of assistive technology can prevent injury to the person with MND and provide safety for unpaid and paid carers.

- most State MND Associations have a range of assistive technology available, usually at no cost to the person with MND, following a referral from the relevant health professional
- a range of assistive technology can be viewed at Independent Living Centres (ILCs) which operate in every state/territory – see www.ilcaustralia.org.au
- people under the age of 65 may be able to access assistive technology through the National Disability Insurance Scheme
- before decisions are made regarding specific technology, 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
- assistive technologies that may be needed by people living with MND include:

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
- tablet computers
- voice amplifiers

Daily living

- book rest
- talking books
- tablet computers
- e-readers

Environmental controls

- personal alarms
- remote operation of lights/electrical equipment

Furniture

- reclining chairs
- adjustable height 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

Breathing

- non-invasive ventilation
- lung volume recruitment bag
- mechanical cough assist device

Meals and feeding

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

Pressure care

- cushions
- mattress overlays

Mobility and transfers

- walking sticks
- walking frames
- rails/ramps
- splints
- wheeled trolleys
- wheelchairs – carer or patient operated, manual/electric
- turntables
- hoists
- lifting belts

Cognition and behavioural change

Merrilees et al. 2010

The common pathological and anatomical overlap between frontotemporal dementia (FTD) and amyotrophic lateral sclerosis (ALS) suggests that the two disorders are strongly linked. Each patient displays a unique set of behavioural and cognitive changes. These may include:

- Executive deficits; reduced ability to plan, organise information, problem-solve, inhibit behaviour
- Apathy; interest and enthusiasm in physical activity may diminish
- Irritability; mood swings, bursts of anger, often over trivial details
- Poor judgement and impulsivity; may affect judgement when it comes to treatment options
- Loss of insight; may affect decision-making and adherence to treatment
- Emotional changes; lack of empathy, blunted emotional reactions or unpredictable emotional expressions such as crying or laughing without warning
- Aggression; moodiness, frustration or anger, aggressive behaviour during caregiving tasks

Strong et al. 2009

There is considerable evidence supporting the existence of cognitive and behavioural dysfunction in ALS, including a spectrum of frontotemporal syndromes and more classically defined dementias.

These include:

- ALS in association with cognitive impairment (ALSci)
- ALS with behavioural impairment (ALSbi)
- ALS with a concurrent dementia that meets the criteria for a FTD (ALS-FTD). The core aspect of this diagnostic categorization relates to the presence or absence of a frontotemporal lobar degeneration.

Up to 50% of people with MND can experience changes in cognition, language, behaviour and personality (ALSci and ALSbi). Most people experience relatively mild changes. However, a small proportion (5-15%) will show more significant changes and will receive a diagnosis of 'motor neurone disease with frontotemporal dementia' or ALS-FTD.

Management of cognitive and behaviour change (ALSci and ALSbi)

- help in simplification of environment and communication:
  - short phrases
  - stop/think reduce distractions
  - memos, aids
- carer and professional awareness about cognition and behavioural change in MND
- carer education
- discussion within family and within multidisciplinary team
- support for person and their family
- accept the issue
- pharmacological
  - SSRI antidepressant
- behavioural
  - distraction
  - diversion

Management of frontotemporal dementia (ALS-FTD)

Management and treatment of ALS-FTD is an emerging area. Consultation with a neuropsychologist for cognitive assessment and advice is beneficial if significant cognitive involvement is suspected.

Lillo and Hodges 2009

A fuller characterisation of the extent of cognitive and behavioural dysfunction in MND is not simply of academic interest but has important implications given that the burden and stress for carers of patients with FTD is very great. It also has relevance to effective communication, legal issues and end-of-life decision making by patients with MND.

Pseudobulbar affect (PBA)

Miller, Pratt & Schiffer 2011
Pseudobulbar affect (PBA) consists of uncontrollable outbursts of laughter or crying inappropriate to the patient's external circumstances and incongruent with the patient's internal emotional state.

Pseudobulbar affect – also known as 'involuntary emotional expression disorder' (but often incorrectly labelled 'emotional lability') – is a troubling symptom that occurs in as many as half of all people living with MND. PBA can cause severe distress, embarrassment, social disruption and isolation adding to the burden of communication and cognitive difficulties with which patients and their carers must cope.

Managing PBA

- clear explanation about the involuntary nature of emotional outbursts associated with PBA
- treatment with amitriptyline or drugs in the SSRI class may be helpful

Miller, Pratt & Schiffer 2011
Dextromethorphan/quinidine is a novel PBA therapy with antiglutamatergic actions. Its recent FDA approval for treating PBA makes it the first medication with this indication.

Fatigue

Fatigue is a common symptom of MND. It is caused by a number of factors:

- as MND attacks motor neurones, they become unable to send commands from the brain to the muscle cells that they control – movements must then be performed by a depleted number of nerve and muscle cells – this means that muscles tire quickly
- other metabolic changes
- weight loss and reduced food intake due to swallowing difficulties are likely to affect the person's energy levels
- when MND affects breathing muscles, less air is drawn into the lungs and when activity increases, it becomes more difficult for the lungs to supply enough oxygen to the body

Managing fatigue

- rest following physical activity and in the later stages of MND. Washing, dressing or using the hoist may exhaust the person and it may take some time for them to recover
- health professionals, such as respiratory specialists, nurses, physiotherapists, occupational therapists and rehabilitation specialist staff can advise on energy conservation techniques, labour-saving devices and respiratory support options

Insomnia

Causes of insomnia include:

- discomfort arising from immobility
- pain due to stiffness of joints or muscles
- muscle spasticity
- shortness of breath
- excessive saliva or dry mouth
- taking stimulants at bedtime

Additionally, because breathing cannot be controlled voluntarily when sleeping, weakened muscles may lead to ineffective breathing (nocturnal hypoventilation). Sleep apnoea is the temporary cessation of breathing while sleeping. This may cause wakefulness, as the disruption of the oxygen supply wakes the person to resume normal breathing.

See other sections of this booklet for information on addressing these causes.

Wellbeing and support needs

Planning

Although there may be differences in how MND progresses, it is not uncommon for people living with motor neurone disease to become concerned about what lies ahead and how decisions about their health, lifestyle and finances will be made.

Planning ahead provides the person with MND and their family with the opportunity to think about, discuss and set in place arrangements for health, lifestyle and financial decision-making.

This can become particularly important if they have specific wishes regarding health care management and interventions or if they expect to have future communication difficulties

State MND Associations provide information and support to assist with planning. Contact details for State MND Associations can be found on the back cover of this publication.

Planning issues can be addressed in the context of support and referral via the planning advisor and the multidisciplinary team.

Review and delays

Ongoing review of daily living needs is also essential as the needs of the person with MND can change quite quickly because MND is progressive.

Delays in accessing services can have a significant impact on the person and their family carer. People with MND require early access to the National Insurance Disability Scheme (NDIS) and community aged care services and fast-tracked plans for care, support and equipment 'bundles' including assistive technology, communication technology and non-invasive ventilation to support breathing and quality of life.

Advance care directives

National Institute for Health and Care Excellence (NICE) 2016

Think about discussing advance care planning with people at an earlier opportunity if you expect their communication ability, cognitive status or mental capacity to get worse.

Bloomer et al. 2010

Most people look to their GP to initiate end-of-life discussions. End of life care planning is a process which needs to take place over a number of discussions and to include nominated decision makers, family members or significant others.

Borasio and Voltz 2006

The consideration of advance directives is increasing in many parts of the world, allowing the wishes of the patients to be clear, even if they are not able to express them due to the progression of the disease.

Advance care directive legislation varies in each state and territory in Australia. Your State MND Association can provide information on advance care planning resources available.

Psychosocial factors

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 may alleviate these fears and produce practical solutions

Denial and anger

- are coping mechanisms 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 individuals who do not wish to talk about impending death and

those who do not wish to accept they have MND
- 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 issues

Depression

- 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
- may be present but can be masked by the progression of the disease and physical changes, and exacerbated by communication difficulties
- diagnosis and treatment of depression, if present, is likely to have a positive effect on ability to cope

Anxiety

- people living with MND may be anxious about:
  - financial planning for their own and their family's future
  - how the family will cope
  - how they will be cared for as the disease progresses
- fears may include increasing disability, suffocation, choking and isolation, fear of the dying process and fear of the unknown
- spending time with the person with MND, talking through fears and worries can be very worthwhile
- if at all possible, provide the opportunity for fears to be expressed and openly discussed
- social workers, pastoral care workers or members of the palliative care team can help in providing counselling

Intimacy and sexuality

Bromberg 2014

MND affects all aspects of intimacy and sexuality for the patient and caregiver. Despite its importance, little is written about the intimate interactions between people with MND and caregivers. Sexual activity occurs in people with amyotrophic lateral sclerosis (ALS) including those on long-term mechanical ventilation. The topic is not often brought up at clinic visits, leaving patients and caregivers unsure as to whom to discuss issues with.

Stausmire 2004, in Gallagher and Monroe 2006

Although most professionals cannot be expected to become specialist psychosexual counsellors, all have a responsibility to become comfortable with offering and responding to cues about sexuality, offering first line help and referring for specialist support.

Loss and bereavement

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

- coming to terms with reality is constantly challenged by proliferating impairments and increasing severity of symptoms
- establishing links with palliative care 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

Spiritual care

Being attuned to the spiritual care needs of the family and the person with MND can help in guiding the family to where these needs may be met.

Lambert 2006

Spirituality is defined to include both religious and non-religious elements. Spirituality can be considered as a resource for coping, as a potential source of conflict and suffering, and as an arena to explore finding meaning.

We need clinically based, non-denominational spiritual care and it is important for all clinicians to be aware of their patients' spiritual values.

Isolation and social support

Mockford et al. 2006

Future strain and distress is alleviated by the greater the number of friends and relatives seen at least once a month, and the less the patient's illness is affecting other areas of their lives.

State MND Associations provide a range of services to assist people with MND and their family and carers to remain engaged with the community. Services may include MND advisor, support groups, carer programs and home visits.

The online MND community is very active both nationally and internationally. Social media platforms can be easily accessed and allow people with MND to share their thoughts and exchange ideas with the broader community. For people with limited movement and/or communication difficulties social media can be accessed using communication devices.

Carer wellbeing and support

Carers Australia 2014

Providing end-of-life care in the home is particularly intense for family and friend carers. While managing their own grief and the grief of others, carers will be providing high level physical and emotional support that a patient needs at the end-of-life. They will need to liaise with a range of service providers, making clinical and other care decisions and making the necessary preparations for the time following the death. It follows that if the carer is well supported throughout this experience, they will have more control over end-of-life care for the patient and greater satisfaction from fulfilling the patient's wish to die at home. And with greater numbers of carers supported to provide end-of-life care in the home, there will be less dependence on institutions and hospitals.

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.

Aoun et al. 2012

Family members caring for people with motor neurone disease may experience exceptional strain due to the usually rapid and progressive nature of this terminal illness.

Gallagher and Monroe 2006

Carers require:

- adequate nursing support
- confident, committed family doctors
- good symptom control for the patient
- coordinated care that is individual and flexibly delivered
- access to specialist care
- practical help: household tasks, personal care, equipment
- respite care as an inpatient or a home sitting service
- knowledge about the illness and training in skills to enhance patient comfort
- financial support
- advice and information on services available and help to secure them
- emotional support directed specifically at the carer

Contact your State MND Association on free call 1800 777 175 for information on carer specific resources and support, local respite and community services.

Children and families

MND forces changes in family roles and relationships and children may need specialised support and/or counselling.

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 person with MND and the carer

Information on MND for children and parents is available from your State MND Association.

The MND Australia 'Talking with Young People about MND' information pack is designed for parents to assist them in communicating about MND with their children. There is an information pack available for teenagers and young people to help them identify and express feelings about a parent with MND.

Visit www.mndaustralia.org.au/get informed/information resources or contact your State MND Association.

The MND Association of England, Wales and Northern Ireland have resources for children and young people including young carers. Visit www.mndassociation.org

Impact on professionals

MND frequently arouses strong emotional and ethical challenges. These include:

- differences in 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 that may impact on the individual's ability to accept advice

Multidisciplinary teamwork is necessary to provide support and encouragement to team members.

Regular case conferencing will assist with coordination and support for the team.

Personal care and community services

Search for state specific personal care and community services by using the Referral Pathways link on the MNDcare website at www.mndcare.net.au/referral-pathways.

Personal care

Personal care assistants can provide assistance with showering, toileting, feeding, respite care and housework. Personal care service availability varies from state to state.

Nursing services

Registered community nurses are accessed from the local hospital home nursing service or community health centre. Registered nurses are also part of the palliative care team, which may be accessed from the local palliative care hospital, hospice or community health centre.

Palliative care

Palliative care teams may be accessed through the local hospital, hospice or community health centre. Depending on local availability and the needs and wishes of people with MND and their families, members of the palliative care team may be involved as part of the multidisciplinary team in providing care and support from diagnosis onwards or only in the later stages of MND. See also Palliative approach on page 9.

MND Associations

Your State MND Association provides a range of services that may include all or some of the following – MND Info Line, MND Advisors, support groups, carer support, education and information sessions, information resources, assistive technology, membership and events. Contact details for State MND Associations are available on the back cover of this publication.

Case management

Specialised case management services are available in some states and territories to assist people to access a range of services to meet their individual needs. Your State MND Association is able to advise on these services.

Care packages

Care packages are government funded and are aimed at people who require more than one type of service to assist them to live at home.

- there are four levels of Commonwealth funded Home Care Packages aimed at people over 65 who have a variety of care needs and whose needs are changing. An Aged Care Assessment Team (ACAT) must first assess the person's needs. Contact My Aged Care 1800 200 422 or contact your State MND Association on free call 1800 777 175.
- people under the age of 65 may be eligible for the National Disability Insurance Scheme. For more information contact your State MND Association on free call 1800 777 175

Financial help and support

General financial advice is available from a range of non-government sources such as financial advisers, banks and self funded retiree associations. There may be a provision with some superannuation funds of total and permanent disability insurance. Appropriate advice should be sought before resigning from employment.

Fact sheets on financial matters are available from your State MND Association.

Centrelink

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

- Age Pension
- Disability Support Pension
- Mobility Allowance
- Health Concession Cards
- Working with an illness, injury or disability
- Carer Payment
- Carer Allowance
- Combining Caring and Working
- Essential Medical Equipment Payment

Financial help is also available for carers.

For more information visit www.humanservices.gov.au or call 13 27 17.

Veterans

Other benefits may be available to veterans and their widows.

- Contact Department of Veterans' Affairs on 13 32 54 or for regional callers dial 1800 555 254 or visit www.dva.gov.au

Getting assistive technology

The MND Association in each state/territory is able to advise GPs and other health professionals regarding local sources of supply of assistive technology, which will enable people living with MND to enjoy the best possible quality of life. See Assistive technology page 23.

Information resources

A range of online and printed information is available from State MND Associations: facts about MND, advice for living with MND and a series especially for families and young people.

End of Life

National Institute for Health and Care Excellence (NICE) 2016

Offer the person with MND the opportunity to discuss their preferences and concerns about care at the end of life trigger points such as: at diagnosis, if there is a significant change in respiratory function, or if interventions such as gastrostomy or non-invasive ventilation are needed. Be sensitive about the timing of discussions and take into account the person's current communication ability, cognitive status and mental capacity.

Clayton et al. 2007

Discussions about prognosis and end-of-life issues can be conceptualised as a process of ongoing conversation over time, rather than a single discussion.

Initiating end of life discussions

The acronym 'PREPARED' has been recommended in the Australian Clinical Practice Guidelines (Bloomer et al. 2010) as a guide to assist health professionals in facilitating end of life discussions.

Prepare for the discussion

- Ensure you have confirmed results of investigations and received relevant reports, ensure uninterrupted time, and arrange for those who need to be present

Relate to the person

- Ensure you have rapport and show appropriate empathy and compassion

Elicit patient and caregiver preferences

- Identify the reason for the consultation and elicit the patient's (and the caregiver's) expectations
- Clarify their understanding of the situation, and establish what and how much detail they want to know
- Consider cultural and contextual factors that may influence information preferences

Provide information

- Provide information that is specific to the patient's needs

Acknowledge emotions and concerns

- Check understanding of what has been discussed and if the information provided meets the patient's and caregiver's needs

Realistic hope

- Offer realistic hope and encourage questions

Document

- Document a summary of the discussion and communicate the decisions to appropriate other health professionals.

Fears and questions about death

The person with MND is likely to have fears and questions about death and about how they will be cared for in the very final phase of the disease.

The person with MND and their family may also have questions about when to go to hospital or a hospice or whether they can stay at home for as long as possible, even until they die.

Ideally consultation by a palliative care team or physician should be initiated early in the course of the disease with subsequent periodic review. See Palliative approach on page 9.

Palliative care involvement can assist the multidisciplinary team with the initiation and timing of end of life discussions.

Communicating with the patient

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

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

The terminal stage

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 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_2 retention overnight
- signs may be noticed by the carer or a member of the multidisciplinary team
- the family can be prepared for the coming days and the patient's imminent death (often after several 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.

Medications at end of life

A range of medications will need to be considered at end of life to address worsening symptoms related to breathing, pain, saliva management and anxiety. The palliative care team should be actively involved to ensure optimal end of life care.

Refer to other sections of this booklet and to the MNDcare website. 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 – to relieve the sensation of breathlessness
- haloperidol – for terminal restlessness

Dosage and modes of administration of medications should be discussed with the palliative care physician.

Action

- check all symptom control and support needs. For management guidelines refer to other sections in this booklet:
  - pain (page 21)
  - breathing (page 13)
  - swallowing (page 16)
  - equipment (page 23)
  - communication (page 19)
  - psychosocial factors (page 26)
- reassess needs of carer and family
- advise the carer and family on managing in the moments following death
- liaise with the palliative care team

Caring for 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
- ensuring the use of assistive technology e.g. lifting equipment provides safety for carers
- comprehensive symptom control and optimal psychosocial support is essential in the management of a peaceful and dignified death

Carer and family bereavement

McMurray and Harris 2006

For each person the bereavement experience is personal and unique. In ALS, there may be a lengthy period of anticipation of the death that may affect the process and outcome of the bereavement. A range of support should be offered according to the individual needs of the bereaved. There is a need for further research to determine whether there are significant differences in responses for people affected by ALS related bereavement.

Some State MND Associations and palliative care organisations offer individual or group bereavement support for carers.

Contacts of Interest

MND Australia

Your State/Territory MND Association (see list on back cover)

MNDCare

Australian Motor Neurone Disease Registry

Carers Australia

Palliative Care Australia

Advance Care Planning Australia

Independent Living Centre

Australian Government, myagedcare

National Disability Insurance Scheme

International Alliance of ALS/MND Associations

ALS Untangled

Stem Cells Australia

Quality Measures in MND Care

This information is based on the American Academy of Neurology ALS quality measures developed by Miller et al. 2013. © 2013 American Academy of Neurology. Republished with permission.

Amyotrophic lateral sclerosis quality measures
Measure title and description
1. ALS multidisciplinary care plan developed or updated Develop multidisciplinary care plan for patients diagnosed with ALS, if not done previously. Frequency: update plan according to disease progression and patient's changing needs, but as a minimum once annually.
2. Disease-modifying pharmacotherapy for ALS discussed Discuss with patients diagnosed with ALS the use of disease-modifying pharmacotherapy (riluzole) to slow ALS disease progression. Frequency: as required depending on disease progression and patient's changing needs, but as a minimum once annually.
3. ALS cognitive and behavioral impairment screening Screen patients diagnosed with ALS for cognitive impairment (e.g. frontotemporal dementia screening or ALS Cognitive Behavioral Screen [CBS]) and behavioral impairment (e.g. ALS CBS). Frequency: as required depending on disease progression and patient's changing needs, but as a minimum once annually.
4. ALS symptomatic therapy treatment offered Offer patients diagnosed with ALS treatment for pseudobulbar affect, sialorrhea, and ALS-related symptoms.
5. ALS respiratory insufficiency querying and referral for pulmonary function testing Discuss with patients diagnosed with ALS symptoms of respiratory insufficiency (awake or associated with sleep) and when appropriate refer for pulmonary function testing (e.g. vital capacity, maximum inspiratory pressure, sniff nasal pressure, or peak cough expiratory flow). Frequency: at least every three months.
6. ALS noninvasive ventilation treatment for respiratory insufficiency discussed Discuss with patients diagnosed with ALS respiratory insufficiency treatment options for noninvasive respiratory support (e.g. noninvasive ventilation, assisted cough). Frequency: as required depending on disease progression and patient's changing needs, but as a minimum once annually.
7. ALS screening for dysphagia, weight loss, and impaired nutrition Screen patients diagnosed with ALS for dysphagia, weight loss, or impaired nutrition and document the result(s) of the screening(s) in the medical record. Frequency: at least every three months.
8. ALS nutritional support offered Offer patients diagnosed with ALS who experience dysphagia, weight loss, or impaired nutrition, dietary or enteral nutrition support via percutaneous endoscopic gastrostomy or radiographic inserted gastrostomy. Frequency: as required depending on disease progression and patient's changing needs, but as a minimum once annually.
9. ALS communication support referral Offer patients diagnosed with ALS who are dysarthric referral to a speech-language pathologist for an augmentative/alternative communication evaluation. Frequency: as required depending on disease progression and patient's changing needs, but as a minimum once annually.
10. ALS end of life planning assistance Offer patients diagnosed with ALS assistance in planning for end of life issues (e.g. advance directives, invasive ventilation, hospice). Frequency: as required depending on disease progression and patient's changing needs, but as a minimum once annually.
11. ALS falls querying Ask patients diagnosed with ALS about falls. Frequency: all visits.

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