Motor Neurone Disease

Aspects of care
for home and community care teams
Motor Neurone Disease Aspects of Care: for home and community care teams

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Publication feedback
We welcome suggestions from people using this document so that it can be improved over time.

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The information in this booklet is provided on the basis that all people accessing this resource undertake responsibility for assessing the relevance and accuracy of its content for their own purposes. In regard to symptom control: this publication is not an exhaustive source of information on symptom control. The medication suggested is not guaranteed to be effective or appropriate in all cases. Naturally, the decision rests with the prescribing doctor and/or nurse, taking into consideration the needs, wishes, and susceptibility of the patient.

If you have health concerns that may need immediate attention, you should seek medical advice.

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Introduction

Motor Neurone Disease Aspects of Care: for home and community care teams has been developed by the Motor Neurone Disease Association to provide home and community care providers with information about living with MND at home.

The decision to accept assistance from home and community care providers can be extremely difficult for both the person with MND and their family. It may represent a loss of independence and an acknowledgement of increasing levels of disability. This may be the first time that the family has accepted assistance in the home and they may feel a loss of personal privacy at having people from the community come into their home. They may also be concerned that their individual and complex needs will not be understood.

MND is a progressive disease and the person’s needs will change as they become more disabled. This change in the level of function can be quite rapid. Regular review and assessment of a person with MND is essential.

MND can affect the muscles used for speech and a person with MND may have a communication aid or special means of communicating. It is important for home and community care workers to take time to learn the means of communication used by the person with MND.

To assist with orientating each community care worker to the needs of an individual it may help to invite the person with MND and their family carer to write down specific care needs and preferences. Information about the person’s particular care needs obtained from family carers and appropriate allied health staff can also be used to establish care plans.

Case conferences can be very helpful if there are particular issues to discuss or for the review of care plans as the person’s needs and abilities change. Case conferences can include family members, the general practitioner, allied health professionals and the key worker as well as home and community care staff.

Publications in the MND Aspects of Care series

- Aspects of care: for home and community care teams
- Aspects of care: for staff of residential care facilities
- Aspect of care: for the primary health care team

For more information

Contact the MND Association. See back cover for full contact details.
About MND NSW

The Motor Neurone Disease Association of New South Wales (MND NSW) is a registered charitable not for profit organisation.

MND NSW offers a range of information and support services for people living with MND, family, friends and health and community care professionals. These include an information line, print and online information resources, education programs, MND Advisors, MND Coordinators of Support and the FlexEquip assistive technology service.

Information line

• The MND Info Line is available to answer questions from anyone who needs to know more about MND. This is a freecall number available weekdays between 9am and 4.30pm. Ph. 1800 777 175.
• MND NSW provides a range of print information and online information at mndnsw.asn.au.

Education

• MND education programs for people with MND, their family and carers are conducted by MND NSW with the assistance of local health and community professionals with experience in MND. Education programs are held in metropolitan and regional areas and include specific education programs for carers. Education programs are also available online.

MND Advisors and MND Coordinators of Support

• MND Advisors are qualified health and community professionals with specific knowledge and experience related to MND. They are a key point of contact for people with MND and their family. They provide information and support for people with MND, their families and carers and maintain contact through home visits, telephone and email.
• MND Coordinators of Support are qualified health and community professionals with specific knowledge and experience related to MND. They assist people with MND who are National Disability Insurance Scheme (NDIS) participants with the implementation of their NDIS plan.

FlexEquip

• The MND NSW FlexEquip Service provides assistive technology (aids and equipment) for people living with rapidly progressive neurological conditions, including MND. More information at flexequip.com.au.

Donations, fundraising and bequests fund 85% of MND NSW services. Just 15% of the Association’s income is received from government.

MND NSW advocates for change through:

• raising community awareness about the needs of people living with MND
• assisting people with MND, their families and carers self-advocate to service providers and funders
• proactively seeking systemic change in community, state and federal government agencies and services so they better meet the needs of people living with MND.

Membership of MND NSW is free for people with MND who live in New South Wales, Australian Capital Territory, Queensland Gold Coast and Northern Territory. Contact the MND Info Line ph. 1800 777 175 or 02 8877 0999 or visit mndnsw.asn.au for information.
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What is motor neurone disease?

Motor neurone disease is a progressive neurological disease. The motor neurones degenerate and die.

With no motor neurones to activate them, the voluntary muscles gradually weaken and waste. The voluntary muscles are the muscles we can control and use for movement, speech, breathing and swallowing.

In some countries motor neurone disease is referred to as amyotrophic lateral sclerosis, or ALS, or Lou Gehrig’s Disease.

We will be using the term motor neurone disease or MND.

Neurones

Neurones are nerve cells. Motor neurones take messages from the brain to the voluntary muscles and to some glands.

There are two types of motor neurones. Upper motor neurones take the messages from the brain through the spinal cord. Lower motor neurones take the message from the spinal cord to the muscles. MND leads to the degeneration of these motor neurones.

MND does not affect the sensory neurones – the nerves that receive messages from things we see, touch, smell, hear and taste.

Effect of MND

MND causes a person to become increasingly disabled. This is because the person’s voluntary muscles: those used for movement, speech, breathing and swallowing, are not being activated by the motor neurones.

There is no cure for MND.

The average life expectancy is two to three years from diagnosis, or three to five years from onset of first symptoms. However, about 5 to 10 per cent of people with MND will have a slowly progressive form of the disease and survive for more than ten years.
WHAT IS MOTOR NEURONE DISEASE?

Incidence and prevalence

MND facts

- MND is an uncommon, but by no means rare, disorder
- Each day in Australia and New Zealand two people are diagnosed with MND
- MND occurs at similar rates in most countries of the world
- Worldwide, about 6 to 7 people per 100,000 live with MND
- In Australia, about 2000 people have MND. In New Zealand, over 300 people have MND
- Each day in Australia and New Zealand two people die from MND
- Slightly more men than women are diagnosed with MND
- MND occurs most commonly in the 50 to 60 year age group. However, MND may be diagnosed in adults at any age

How do people get MND?

There is no evidence that MND is transmissible from person to person.

MND occurs sporadically, meaning occurring in scattered or isolated instances without a clearly identifiable cause, for about 90% of people with the disease. However, about 10% of people with MND have familial or hereditary MND. Of this 10% of people with MND, one-fifth have a genetic mutation on the SOD1 gene. Other mutations linked to MND include TDP 43 and FUS (Kiernan and others 2011).

Clinically the sporadic and familial forms of MND are indistinguishable

First signs and symptoms

For most people, MND usually begins by affecting a single limb or aspect of motor function. The diagnosis of MND is often clinically difficult. There is no single test for the disease and sometimes it is necessary to review a person for some time before the diagnosis becomes reasonably certain.

Some of the first signs and symptoms of MND

- weak hands
- fatigue
- muscle pain and cramp
- muscle twitches
- difficulty speaking or swallowing
- weakness in the legs
- loss of muscle tone
- difficulty getting comfortable
- immobility, discomfort and pain
- breathing difficulties
- drooling or thick saliva

The effects of MND become more generalised as the disease progresses. However, for some people, the symptoms are widespread from onset.
WHAT IS MOTOR NEURONE DISEASE?

Classification into types
MND can be classified into four main types depending on the pattern of motor neurone involvement and the part of the body where the symptoms begin.

Amyotrophic lateral sclerosis (ALS)
- Both upper and lower motor neurones affected
- Limb muscle weakness and wasting
- Amyotrophic lateral sclerosis is the most common type of MND. People with ALS have muscle weakness and stiffness, over-active reflexes and, in some cases, rapidly changing emotions. Initially muscles in the arm and/or leg are affected. The muscles of speech, swallowing and breathing are usually also affected later in the disease. ALS is the term commonly applied to MND in many parts of the world

Progressive bulbar palsy (PBP)
- Both upper and lower motor neurones affected
- Speech and swallowing muscle weakness and wasting
- Progressive bulbar palsy, mixed bulbar palsy and pseudobulbar palsy involve the muscles of speech and swallowing. The nerves that control these functions are located in the bulb (the lower part of the brain), hence the term bulbar palsy (paralysis). The arm and leg muscles may also later be affected

Progressive muscular atrophy (PMA)
- Lower motor neurones are affected
- Slower rates of progression and significantly longer survival compared to ALS and PBP
- Progressive muscular atrophy 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 type of MND to diagnose accurately. 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)

Primary lateral sclerosis (PLS)
- Upper motor neurones are affected
- Slower rates of progression and significantly longer survival compared to ALS and PBP
- Primary lateral sclerosis is very rare and diagnosis is often provisional. Initially muscles in the arm and/or leg are affected. The muscles of speech, swallowing and breathing are usually also affected later in the disease

Kennedy’s disease
Kennedy’s disease is a disorder of motor neurones which is not motor neurone disease. It is an inherited disorder affecting adult males caused by a mutation of a gene which plays a role in moderating the action of the male sex hormones. It causes slowly progressive weakness and wasting of muscles with only lower motor neurone involvement and other features.
WHAT IS MOTOR NEURONE DISEASE?

**MND progression**

As MND progresses more motor neurones die and when the motor neurones stop activating the voluntary muscles, the muscles weaken and waste. However, all people with MND will not be affected in the same way.

For example, although the first symptom of MND for some people is leg muscle weakness, the first symptoms for others may be arm muscle weakness or a weakness of the speech and swallowing muscles.

<table>
<thead>
<tr>
<th>People may experience any one or a combination of</th>
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<tbody>
<tr>
<td>• speech difficulties</td>
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<td>• swallowing problems</td>
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<tr>
<td>• immobility, discomfort and pain</td>
</tr>
<tr>
<td>• breathing difficulties</td>
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<tr>
<td>• drooling or thick saliva</td>
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**Cognition and MND**

Approximately 50% of people with MND may experience some change in cognition, language, behaviour and personality. When cognitive and behaviour changes occur in MND, it is because there have been changes in specific areas of the brain called the frontal and temporal lobes. 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 MND/FTD. Often the symptoms of dementia precede the motor symptoms, sometimes by a number of years.

**What remains unaffected by MND?**

Sensory symptoms, that is, disturbances of touch, taste, smell, sight and hearing are rare. It is also important to understand that although the person with MND may have difficulty in speaking, his/her understanding and intellectual function are not usually affected.

Eye muscles, bowel and bladder control are usually not affected, however, constipation is often caused by lack of mobility, dietary changes and inadequate fluid intake.

**Cause of death in MND**

The usual cause of death in MND is breathing failure.

For some people, breathing difficulties may occur early in the course of the disease and the person may die before their level of general disability becomes very severe.

Others will lose the use of their arms and legs, communication and swallowing before breathing muscles become involved.

Distressing symptoms such as breathlessness, discomfort and pain are best managed in consultation with palliative care teams. See *MNDcare Approach - a person-centred approach* and *End of life care*, for more detail on palliative and terminal care of a person with MND.
Supportive interventions

Although there is no cure for motor neurone disease yet, research has shown some interventions can help people living with motor neurone disease to live better for longer.

<table>
<thead>
<tr>
<th>Interventions to live better for longer with MND</th>
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<tbody>
<tr>
<td>• multidisciplinary care</td>
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<tr>
<td>• good nutrition</td>
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<tr>
<td>• non-invasive ventilation</td>
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<tr>
<td>• riluzole (sold as Rilutek or APO-Riluzole)</td>
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</table>

There is one medication approved for treatment of the Global Type of motor neurone disease in Australia and New Zealand - riluzole (sold as Rilutek or APO-Riluzole). Research has shown that riluzole probably prolongs median survival by two to three months.

How people manage their earlier symptoms of motor neurone disease can affect how symptoms that may arise later can be managed. Early discussion about symptom management helps people with MND to plan ahead.

In brief

- MND is very different for every person diagnosed
- In any two people, MND may affect different areas of the body, progress in different patterns and at different rates
- The average life expectancy is two to three years from diagnosis, or three to five years from onset of first symptoms
- About 1 in 10 people with MND will have a slowly progressive form of the disease and survive for more than ten years
- There is no 'recipe' for managing MND. It is a matter of looking at the particular symptoms and needs of each person with MND and trying to reduce the difficulties caused by these symptoms
- The needs of people living with MND are complex
- Symptoms can progress rapidly leading to an escalation in the level of care needed
- It is important that a range of health and community care professionals are involved, enabling regular assessment and review of changing needs
Managing MND

There is no 'recipe' for managing MND. This is because the needs of people with MND vary considerably from person to person.

The MNDcare Approach

The MNDcare Approach is a person-centred approach. It is a model for

- talking with the person with MND about their unique symptom management and wellbeing and support needs
- developing strategies for meeting these needs
Multidisciplinary care

Multidisciplinary care is delivered by professionals from a range of disciplines who work together to address as many of a person’s health and other needs as possible (Mitchell and others 2008).

Step 1 Review and assess the needs of the person living with MND and the needs of the carer and family
- the needs of people living with MND can be broadly grouped into symptom management and wellbeing and support needs

Step 2 Provide information and the opportunity for discussion about the need
- the person with MND, their family, case manager and other service providers will require information about MND and its impact and management to assist them to make decisions about care
- people have varying informational needs and you will need to tailor information given to the requirements of the individual
- MND Associations in Australia and New Zealand provide written literature and web-based literature, telephone information lines, home and residential care visits and education sessions for people living with MND and health and community/residential care professionals

Step 3 Refer to the appropriate health professional, community care worker, agency or other service for
- rapid response
- coordinated action
- regular review

Multidisciplinary care can provide people living with motor neurone disease
- flexible, coordinated professional support
- referral to professionals in a coordinated way, so that any difficulties can be dealt with promptly
- regular review of symptoms
- opportunities to get specialised advice from other health and community care professionals and providers

The General Practitioner, community care team, allied health professionals and palliative care staff can work together to develop the best quality care management.

Occupational therapists can investigate the person’s best means of using communication aids including buzzers or call bells to let staff know they need help. Occupational therapists can also advise about comfort, positioning, pressure area care, splinting, mobility aids and ways to assist with personal care activities such as showering and dressing.

Physiotherapists have an important role in the comfort and care of the person with MND. They can advise about positioning. Physiotherapists can give exercises for muscles groups that are not affected by MND and exercises to help keep affected joints flexible.
Physiotherapists can also demonstrate the assisted cough technique for people who have a weak cough reflex and swallowing difficulty. They can provide advice on neck collars, foot splints and mobility aids to prevent falls.

Speech pathologists/therapists can assess the person’s swallowing and communication needs. They can recommend the best food and fluid consistency for the person and suggest ways to assist with safe swallowing and maintaining food and fluid intake. They can assess and advise about communication needs. Speech pathologists/therapists work with occupational therapists to find the best ‘set-up’ and positioning if a communication device is being used.

Dietitians assess the person’s nutritional needs and make recommendations to achieve the best level of nutrition and fluid intake.

Social workers refer people to appropriate services and benefits. They may also provide short term counselling and emotional support for people with MND and their families.

Palliative care specialist and clinical nurse consultants provide symptom control and medication advice for managing breathing difficulties and pain.

**Care 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 case discussions and team meetings ensure

- an optimal coordinated multidisciplinary team approach
- the continuation of regular assessment and review throughout the course of the disease

As a progressive disease, MND will cause the person’s condition to change over time. “The composition of the team may change to reflect the changing clinical and psychosocial needs” (Mitchell and others 2008) of the person with MND and their carer and family.

**Networking, managing and coordinating**

The needs of people living with MND are complex and vary from person to person. One agency/worker cannot meet all the needs of a person with MND. It is not uncommon for five to ten health and community care professionals to be involved in the care and support of one person with MND.

Networking, managing and coordinating are central to getting services and support up and running quickly for a person living with MND, their carer and their family.

Professionals providing multidisciplinary care can be from the same organisation, a range of organisations or from private practice from community, hospital, clinic, residential and other care settings.

Each discipline-specific team member enriches the knowledge-base of the team as a whole (Mitchell et al 2008).

Over time, the multidisciplinary team composition can change to reflect changes in the person’s needs (Mitchell et al 2008).
Key worker

One person, usually referred to as a key worker, case manager, care coordinator or team coordinator initiates effective and timely response when needs change.

Who the key worker is depends on

- local health and community care service availability
- the professional interests of individual health and community care professionals

The key worker may be a

- case manager
- general practitioner
- local occupational therapist, physiotherapist, speech pathologist/therapist or social worker
- MND clinic nurse
- MND Association advisor
- MND Association coordinator of support
- MND shared-care worker
- neurologist
- palliative care professional
- other health or community care professional with particular expertise in MND symptom management

A key worker

- maintains regular contact with the person with MND
- reviews needs
- initiates effective and timely response when needs change
- liaises with other team members, services and the person with MND and their family
- organises regular case conferences and team meetings
- maintains contact list of health and community care professionals involved, and contact details of family
- assists with team debrief

Multidisciplinary MND Clinics and Services

There are now several established MND specific clinics and programs of care throughout Australia and New Zealand. These specialised clinics provide an integrated approach to the management and clinical care of an individual with MND.

MND specific clinics and programs give the person with MND access to a range of health professionals who work together to provide a coordinated response to care. Team members may include the neurologist, rehabilitation specialist, palliative care specialist, respiratory specialist, physiotherapist, dietitian, social worker, occupational therapist, speech pathologist/therapist and registered nurse. In some areas, MND Association advisors also attend these clinics to provide information and support. MND specific clinics and programs often provide telephone consultancy for care providers within their designated health area. For more information contact the MND Association.
MND Association

The MND Association is an important part of the care coordination team.

The MND Association provides

- information and ongoing support
- home visits
- residential care visits
- assistance in accessing motor neurone disease clinics and local services
- equipment or assistance in accessing equipment from government agencies
- peer support and education programs
- carer workshops and programs
- support groups
- education sessions and information for health and community and residential care providers
- advocacy and influencing
- volunteers

For contact details see back cover of this publication.

Palliative approach

MND is a life limiting disease characterised by a series of losses with the accompanying issues of grief and bereavement.

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 and wellbeing needs 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.

Establishing links with palliative care at an early stage can provide the multidisciplinary team members 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. These services may be provided in the home, hospice, residential care facility or hospital.

Proactive planning

Ongoing review and discussion provides the person with MND and their family, and the community care staff and other members of the multidisciplinary team, with the opportunity to gently anticipate and talk about future needs. For example, a person developing speech muscle weakness may have no apparent need for communication aids now, but may need to become confident about using a communication aid before the aid becomes their primary means of communication.

Ongoing review and discussion also provides the opportunity for the team to understand the aspects of life that are most important for the person with MND.

Proactively planning for future needs can avert the need for future crisis management.
In brief

The needs of people with MND are complex and varied

The MNDcare Approach is a person-centred approach. It is a model for talking with the person with MND about their unique symptom management and wellbeing and support needs, and developing strategies for meeting these needs

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 and wellbeing needs management for the person with MND and their family is achieved

Be guided by the needs and wishes of the person with MND and their family carer

A team of health professionals is needed to provide regular assessment and to review changing needs

The team needs to share knowledge and experience

Home and community care staff are part of the team. Other team members include the general practitioner, occupational therapist, physiotherapist, speech pathologist/therapist, dietitian, social worker, palliative care consultant/nurse and orthotist

MND Association advisors can assist with contacting other health professionals

You can find more information about the MNDcare Approach online at www.mndcare.net.au
Breathing

Motor neurone disease often causes the muscles involved in breathing, the respiratory muscles, to weaken.

Respiratory muscle weakness usually develops gradually, but can occur suddenly.

Sometimes, respiratory muscle weakness may even be the first sign of motor neurone disease.

Signs and symptoms of respiratory muscle weakness

<table>
<thead>
<tr>
<th>Disturbed sleep</th>
<th>Daytime sleepiness</th>
<th>Increased fatigue</th>
<th>Morning headaches</th>
<th>Quieter voice</th>
</tr>
</thead>
<tbody>
<tr>
<td>Fewer words per breath</td>
<td>Shallow, faster breathing</td>
<td>Reduced movement of the rib cage</td>
<td>Excessive use of the muscles in the upper chest and neck</td>
<td>Weakened cough and sneeze</td>
</tr>
<tr>
<td>Breathlessness (dyspnoea) even at rest</td>
<td>Breathlessness lying flat (orthopnoea)</td>
<td>Impaired concentration or confusion</td>
<td>Irritability and anxiety</td>
<td>Decreased appetite</td>
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</tbody>
</table>

Shortness of breath, or dyspnoea, is one of the most frightening symptoms of MND. People with dyspnoea experience the feeling of not being able to get enough air, rather like the sensation of being in an overcrowded room.

Approach to management

The person with respiratory muscle weakness needs to have a management plan that includes ongoing review.

Respiratory muscle weakness management plan

- Early assessment by a respiratory specialist
- Advice about managing respiratory muscle weakness
- Advice about non-invasive ventilation. To use non-invasive ventilation the person wears a mask connected to a small pump. This creates just the right pressure to keep the airways open so that room air can easily come in and out of the lungs.
- Strategies for ongoing review

The way a person with MND manages their respiratory symptoms may affect how other symptoms of MND can be managed. For example if, in the future, a person with MND needs liquid feed and fluids through a percutaneous endoscopic gastrostomy (PEG) tube their respiratory function needs to be at or above a certain level to ensure safe insertion of the PEG tube.
Avoid people with coughs and colds
People with respiratory muscle weakness should avoid people with coughs and colds. Referral to a doctor or respiratory physician for discussion about the influenza and pneumonia vaccines is often recommended.

If you have a cough or cold it is important that you do not provide care for the person with MND.

Well balanced diet of the right consistency
In undernourished people all muscles, including the respiratory muscles, are weaker. People with respiratory muscle weakness are less likely to have coughs and colds if they eat a healthy diet with the right mix of proteins, carbohydrates and other nutrients.

A weak cough can make it more difficult for the person with MND to clear thin liquids or thick and chunky foods that may accidentally enter their airway when they are eating and drinking. The speech pathologist/therapist and dietitian can advise about liquids and foods of the right consistency.

Positioning, room airflow and temperature
The way a person's body is positioned while sitting or lying down can assist their breathing.

Electric recliner chairs and adjustable wheelchairs enable experimentation with a number of different positions. Some people find it more comfortable to sit in a slightly reclined, or not so upright, position. Others prefer a fully upright position. When in bed, a person may find maximum comfort in a semi-reclined position. Regular pillows, boomerang pillows, foam wedges and bed adaptations, such as an electric bed with head-raiser, can be used to support the person’s upper body and head.

Having an open window in the room and using a fan to circulate air can also reduce feelings of breathlessness. It can also be helpful to use a humidifier to increase the moisture in the room air. Room temperatures that are too hot or too cold can also make the person with MND feel uncomfortable. A physiotherapist or occupational therapist can advise about positioning and where to get equipment.

Breathing exercises
Breathing exercises help the lungs to expand more fully and this reduces pooled air in the lungs. One simple exercise is to take five to ten deep breaths, with short rests in between, several times a day. The physiotherapist, palliative care team, specialist respiratory nurse and respiratory physician can advise about the right breathing exercises for a person with MND.

Fatigue management
Fatigue can be a major problem for people with MND. There is no advantage in them ‘pushing themselves’. Rather, the key is to encourage the person to save their energy for what they really want to do.

You can assist the person with MND to:
- respect the body’s limitations and to pace themselves
- find short-cuts for things they have to do
- use gadgets and labour-saving devices
• use equipment which helps save their energy. For example, they can take a wheelchair that they may not need to use all the time, when they go out.

Health professionals, such as respiratory nurses, physiotherapists, occupational therapists and rehabilitation specialist staff can provide information about energy saving tips, equipment and labour-saving devices.

**Relaxation techniques**

Anxiety or worry about breathlessness can affect a person’s breathing efficiency. Controlled breathing and other techniques can help the person relax and make breathing easier. The physiotherapist, palliative care team, specialist respiratory nurse and respiratory physician can advise.

**Non-invasive ventilation (NIV)**

Increasingly, many people with MND are choosing to use non-invasive breathing support, known as non-invasive ventilation or NIV.

NIV provides relief from symptoms such as fatigue, breathlessness and disturbed sleep patterns, but does not prevent progressive weakening of the respiratory muscles (Andersen and others 2007).

People with MND most commonly use variable positive airway pressure (VPAP) or bi-level positive airway pressure (BIPAP) machines. This is because these types of NIV machines can provide a lower level of pressure when the person breathes out. In addition, these machines can be adjusted to provide increased respiratory support if needed.

Over time, NIV will be less effective in helping the person with MND control their respiratory symptoms, because their MND will continue to progress.

To use non-invasive ventilation the person wears a mask connected to a small pump. This creates just the right pressure to keep the persons airways open so that room air can easily come in and out of their lungs when they breathe.

The NIV machine is usually used at night but as the respiratory muscles weaken the person with MND might use it at times during the day as well. When a person with MND starts using NIV they may need to try several different types of masks to find the right mask for them.

Regular mouth and facial skin care is important for people using NIV. This reduces the risk of infection and increases mask comfort.

**Medication**

There are a number of medications that reduce the symptoms of breathlessness. Small doses of morphine, or similar medications, may be effective in reducing the sensation of breathlessness and help make the person with MND feel more comfortable.

Anti-anxiety medication may be prescribed to reduce feelings of anxiety. The neurologist, general practitioner, palliative care or respiratory physician can advise about medication.
Swallowing and nutrition

Motor neurone disease causes the muscles involved in chewing and swallowing to weaken, although not all people with MND will have this symptom.

When a person has weakness in these muscles they may eat and drink less than they usually would and cough during mealtimes. They may take longer to eat or be worried and anxious about eating and drinking. This can result in weight loss, malnutrition, dehydration and a loss of enjoyment of life.

People with MND who are malnourished and dehydrated have poorer outcomes than people with MND who are well-nourished (Chio and others 2009).

Muscle weakness and swallowing

Muscle weakness caused by MND interferes with the swallowing process and reduces the person’s ability to swallow. This difficulty with swallowing is known as dysphagia.

If the swallowing process breaks down, control of the food or liquid is reduced. The effect of a break-down in the swallowing process depends on which of the swallowing muscles are affected.

For example, when the lips cannot close and the tongue is weak the person may be unable to gather the food together in their mouth ready to swallow.

If control of the throat muscles is reduced food may be left behind in the throat.

If the swallow reflex is delayed, the food passes over the airway opening before the larynx has had time to close.

If saliva is thickened and ropey, it becomes uncomfortable to swallow making it hard to clear the throat. This increases the risk of food and drink going down into the lungs.

Swallowing difficulties in MND

A person with MND who has swallowing difficulties will often have increasing levels of difficulty with swallowing as the MND progresses.

Swallowing difficulties can lead to dehydration, malnutrition and constipation. Usually, a person who has swallowing difficulties also has difficulties with saliva management and may experience dry mouth.

Weakness in the swallowing muscles may allow food or liquid to enter the person’s airway. This can result in chest infections or aspiration pneumonia.

People who have difficulty clearing food or liquid from their mouth, throat or airway may experience worry and anxiety about coughing. They may also have sensations of choking. This can be caused by muscle spasms, difficulty dislodging food in the throat, an inefficient cough or reduced airway protection from the swallow.
Signs and symptoms of weakness in the muscles involved in chewing and swallowing

- needing extra effort to chew
- coughing whilst eating or drinking
- taking several swallows per mouthful
- having a muffled or 'wet' sounding voice after eating
- finding eating or drinking tiring
- getting breathless after a meal
- taking more time than usual to eat and drink
- having frequent chest infections
- having slurred or indistinct speech
- finding it difficult to clear saliva

Approach to management

The person with swallowing difficulties needs to have a management plan that includes ongoing review. The aim of the management plan is for the person with MND to have as much pleasure as possible from food and eating, while consuming enough liquids and nutrients to remain well-nourished.

Swallowing difficulties management plan

- Assessment and advice from a speech pathologist/therapist 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
- Advice about gastrostomy. This is a medical procedure during which a short, permanent tube is placed into the stomach through the abdominal wall. The person can have liquid feeds and fluids through the tube, directly into the stomach, bypassing the mouth and throat
- Strategies for ongoing review

Positioning and swallowing

Correct positioning for eating and drinking is very important for airway protection. Swallowing is generally easier when the person is sitting as straight as possible with the head upright: avoid tilting the head back. Some people find swallowing easier if the head is positioned slightly forward, as if sniffing the air. Carefully arranged pillows, special chairs and tilt-in-space wheelchairs can be helpful in achieving the best position. After eating, the person with MND should remain upright for 30 minutes.

A physiotherapist can advise on positioning and head and neck support.
Fatigue
To minimise the effects of fatigue the person with MND can:
- rest before a meal
- make sure breathing has settled before starting the meal
- have smaller, more frequent meals, with the main meal during the part of the day the person is generally less fatigued
- eat and drink foods and liquids that are easier to chew and swallow
- time meals so that eating and drinking are not hurried or pressured.

Techniques in food and drink preparation, presentation and delivery

Liquids
When we drink or eat thin fluids such as water or clear soups, the liquid flows very quickly. We need strong lip, mouth and throat muscles to control and swallow the liquid so it arrives in the stomach as intended.

It is very important that a person takes an adequate amount of liquid every day for rehydration. Recommended fluid intake is at least 2100-2600ml per day (8-10 cups) for men and women, including plain water, milk and other drinks. This can include thick soups, yoghurt and custard. Have a drink after every meal and between meals, for example, mid-morning and mid-afternoon. A variety of fluids help tempt the taste buds. Always have drinks available. Regular small sips help reduce dehydration.

People with weakened lip, mouth and throat muscles often find it easier to swallow thickened liquids.

Commercially available liquid thickeners can be used to thicken the consistency of water and other liquids. Fruit nectars and milk shakes have a natural thickness. Liquids can also be thickened in other ways. For example, vitamised fruit can be added to fruit juice and pureed vegetables can be added to clear soups.

Recommendations for liquid consistency will usually be described as:
- Mildly Thick or Level 150 – the fluid runs freely off the spoon but leaves a mild coating on the spoon. Mildly thick fluids are thicker than fluids such as fruit nectars but not as thick as a thickshake
- Moderately Thick or Level 400 – the fluid slowly drips in dollops off the end of the spoon. Moderately Thick fluids are similar to the thickness of room temperature honey or a thickshake
- Extremely Thick or Level 900 - the fluid sits on the spoon and does not flow off it. Extremely Thick fluids are similar to the thickness of pudding or mousse

Food
Start with a balanced diet, including a variety of foods. A multivitamin supplement may be beneficial if it is not possible to eat the recommended amounts. A doctor or dietitian can advise.

People with weakened lip, mouth and throat muscles may also find it easier to swallow food of a particular consistency and the dietitian may suggest this for the person with MND. When weight loss is a problem, every mouthful needs to count.
Recommendations for food consistency in a modified diet are generally described as:

- **Texture A - Soft**: foods may be naturally soft, such as a ripe banana, or may be cooked or cut to alter texture. Foods are diced or sliced and require little, if any, cutting. This includes casseroles, pasta, fish and finely chopped meat moistened with gravy or a sauce.

- **Texture B - Minced and Moist**: foods that are naturally very soft or have been minced or mashed so that they are free from big lumps, skin, stalks or pips. The food keeps its shape when put on the plate. Additional sauce and gravy help in moisturising and mobilising the food.

- **Texture C - Smooth Pureed**: foods which have been puréed (vitamised or blended) so that they are smooth, moist and lump-free, but may have a grainy quality. They are not runny and do not need to be chewed.

### Preparation, presentation and delivery

- When preparing food it is often easier to prepare enough for three or four meals and freeze in serving sizes. This enables different combinations of meats and vegetables to be ‘mixed and matched’.

- Mixed consistencies can be very hard for the person with swallowing difficulty to manage.

- Make sure food is stored and served at the right temperature.

- Keep food types separate for serving. For example do not puree food together that is usually served separately.

- When puréeing or mincing meats, vary flavours with sauces, fruit juice, chutney and relishes.

### Assisting the person with MND eat or drink

If you are assisting a person with MND eat or drink, remember that people with weakened mouth muscles need more time to move the food into position for swallowing. Also:

- several swallows may be necessary to clear each mouthful

- the mouth needs to be clear before offering more food

- placing food towards the back of the tongue may be helpful.

### Utensils and aids for eating and drinking

There are many utensils and aids for eating and drinking that can be helpful for a person with swallowing difficulties and/or weak hand and arm muscles. These include cups that are lightweight with wide, flared tops; non-slip bowls and plates; one-way straws; shallow spoons; cutlery with easy to grip handles; spouted cups and syringes.

The occupational therapist and speech pathologist/therapist can advise about utensils and aids.

### Assisted cough technique

A cough is a very forceful breath out. When a person coughs, their abdominal muscles contract and push stomach contents up against their diaphragm. This causes the diaphragm to move up and force air out of the lungs. If these muscles are weakened, a cough will be less effectual.

A physiotherapist can advise on the assisted cough technique. This technique can help to keep the airway clear.
Severe coughing, choking and laryngospasm

Ongoing coughing and choking sensations can be caused by:
- difficulty dislodging food in the throat
- reduced airway protection during the swallow
- an inefficient cough
- muscle spasms

Coughing is the body’s way of moving food, drink or saliva away from the airway. Coughing while eating or drinking may be immediate, but it can also occur after a delay or at other times because food, liquid or saliva can collect in crevices in the throat and fall into the airway as a breath is drawn in.

Coughing may be severe and distressing but it is not in itself life-threatening.

Choking occurs when the airway is blocked partially or fully. Death from choking occurs very rarely in MND.

Laryngospasm is a sudden closure of the vocal cords resulting in wheezing and a short cessation of breathing. Triggering factors include smoke, alcohol, spicy foods and gastric reflux. The attacks usually resolve spontaneously in less than a minute, although they often create panic and anxiety.

People who have swallowing difficulties may be worried and anxious about coughing and sensations of choking.

Managing coughing, sensations of choking and laryngospasm

It is important for the person with MND to know that death caused by a choking attack is very rare in MND. The doctor, speech pathologist/therapist and physiotherapist can advise. The swallowing management plan usually includes strategies for managing coughing, sensations of choking and laryngospasm such as:
- encouraging the person with MND to stay calm and wait for the attack to pass
- using the assisted cough technique
- for laryngospasm, provide reassurance and encourage the person to cough or laugh as this will help to open the vocal cords
- the use of medications including morphine, amitriptyline, benzodiazepines and glycopyrrrolate
- when and how to seek medical advice.

Gastrostomy

Gastrostomy enables the person with MND to be fed through a small permanent tube that passes directly into their stomach, bypassing their mouth and throat.

Gastrostomy is a medical procedure during which a short, permanent tube is placed into the stomach through the abdominal wall. This procedure usually involves an injection of a local anaesthetic, a small incision and mild sedation.

Once the permanent tube has been positioned into the stomach it is prevented from moving by a widening of the tube internally and a small flange or collar externally. A cap is placed over the external end of the permanent tube. Fluids and liquid feed can be provided through the external opening of the tube. This type of feeding is called enteral feeding.

The person can tuck the external end of the tube under clothes when it is not in use. The permanent tube generally lasts for one to two years and can usually be replaced easily. Changing the tube does not usually require hospital admission.
Saliva and mouth care

MND does not cause more or less saliva to be produced. However, some symptoms of MND and their treatment can cause saliva management and saliva production difficulties. People with MND may have drooling saliva or thick saliva. They may also have dry mouth.

Mouth care, teeth care and MND

Mouth care is very important for people with MND who have tongue and throat muscle weakness. This is because 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.

Also, when a person has difficulty moving their tongue, it is very hard for them to move food particles around their mouth. Food can become trapped between teeth and between the teeth and the cheeks. This can cause mouth ulcers, gum disease and tooth decay, as well as bad breath.

It can be very difficult for a person with tongue and throat muscle weakness to manage toothpaste foam. Rinsing and spitting out or swallowing can sometimes be impossible.

Approach to management

Managing mouth and teeth care

- Use a low foaming toothpaste
- Use an electric toothbrush
- Swab the mouth with non-alcohol water-based solutions of bicarb soda (1/2 tsp to 1 cup of water), salt (1 tsp to 1 cup of water), peppermint oil (1 tsp to 1 cup water) or grapeseed oil (100%) can be an alternative to brushing the teeth. There are also commercially available mouth swabs such as Toothettes from Orthocare
- Avoid alcohol based mouthwashes and lemon and glycerine swabs as these can make the mouth dry
- Check the mouth daily for signs of caught food, oral thrush and ulcers

The doctor, nurse or speech pathologist/therapist can advise about dry mouth.

Drooling saliva

People with MND who have weakness of the tongue and throat muscles, or poor lip seal and head control, often have saliva that drools.

This is because weak tongue and throat muscles cannot do all the swallowing needed to clear saliva from the mouth. Together with a poor lip seal and weak neck muscles this may result in drooling. The excess saliva is unable to be kept in the mouth.

A person who has drooling saliva may feel embarrassed. They may withdraw socially and emotionally, causing a loss of independence.

Drooling saliva can contribute to dehydration and can affect a person’s speech and swallow.

Approach to management

Treatment for drooling saliva can result in other oral health problems, such as dry mouth or thick saliva.

The doctor, nurse, speech pathologist/therapist or physiotherapist can advise about drooling saliva.
Thick saliva

Thick, ropey and tenacious saliva in MND can be caused by mouth breathing, dehydration, weak cough, reflux, hot weather or air-conditioning. It can also be caused by medications used to manage drooling and other symptoms.

When saliva is thick, it becomes uncomfortable to swallow and hard to clear the throat. This increases the risk of food and drink going down into the lungs. Drinking can become more difficult and the risk of dehydration increases.

Thick saliva can make speech difficult.

Also, thick saliva is not very effective in protecting a person from microorganisms and toxins in food and in the air. Thick saliva in the mouth can catch bits of food that stay in the mouth after eating.

People who have thick saliva need to pay extra attention to mouth care.

Approach to management

The doctor, nurse or speech pathologist/therapist can advise about thick saliva.

Managing thick saliva

- Check fluid intake
- Offer soda water and suggest the person eliminate coffee
- Use mucolytic agents that break down the thick saliva, such as papaya enzyme lozenges and drinks or ice cubes of grape, apple, pineapple and papaya juice
- Try nebulised saline, which is a fine mist of water and salt, through a mask or mouthpiece
- Swab the mouth with plain water or one teaspoon bicarbonate of soda or salt dissolved in a glass of water
- Use commercially available products that moisten the mouth including specific gels and saliva spray such as Biotene
- Seek medical advice about medication such as bromhexine or Histidine
- Use the assisted cough technique to clear the thick saliva

Managing drooling saliva

- Get advice about head and neck support, and assist the person to position their body so their head is not tipped forward
- Use barrier creams and soft cloths to reduce skin irritation
- Use natural remedies such as horseradish tablets or sipping sage and hibiscus tea
- Remove excess saliva by swabbing the mouth or by using a syringe
- Seek medical advice about medication that reduces saliva production, for example, tricyclic anti-depressants and anticholinergics including atropine and hyoscine (Kwells) and/or injectable medications including low dose glycopyrrolate (Robinul) and botulinum toxin
- Following medical review, use a suction machine to remove the saliva
Dry mouth

Dry mouth is an uncomfortable condition experienced by some people with MND. People with dry mouth may also have saliva that drools or saliva that is very thick.

A poor lip seal, breathing through the mouth and not drinking enough fluids can all cause dry mouth. Dry mouth can also be caused by medications taken to reduce excess saliva.

People with dry mouth are more likely to get mouth irritations and infections such as thrush. Dry mouth can cause night waking, make chewing and swallowing harder and make it uncomfortable to speak and eat.

People with dry mouth may have difficulty in clearing their throat and may experience sensations of choking.

Approach to management

The doctor can advise about dry mouth.

Managing dry mouth

- Sip fluids frequently
- Use commercially available products that moisten the mouth including specific gels and saliva spray such as Biotene
- Seek a medication review
Communication

Speech difficulties in MND are caused by weakness and reduced coordination of the lips, facial muscles, tongue, larynx and pharynx.

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.

Not all people with MND will experience difficulties with speech and non-verbal forms of communication. However, a person who does have communication difficulties will often have increased difficulty as the MND progresses.

When we can’t communicate

Difficulty with, or loss of speech can lead to a strong sense of isolation and feelings of vulnerability. Others may assume deafness or intellectual impairment. Opinions may be not sought or ignored. The person with MND may feel embarrassed and frustrated by their speech difficulties.

Managing communication difficulties

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

- **Speech pathologist/therapist**
  - early assessment

- **Occupational therapist**
  - seating and positioning
  - wrist supports
  - switches, pointers
  - mobile arm supports, tables

- **Speech pathologist/therapist, occupational therapist**
  - communication aids
  - computer
  - environmental controls

Effective communication

Establishing effective communication helps to build a sense of trust and will ultimately help you in caring for the person with MND.

The way you communicate with a person affects how easy or hard it is for them to communicate with you.
Call devices
A person can use a call device to let someone, who is within hearing distance of the activated device, know they need an item or assistance. Call devices are very important as they give the person with MND a greater sense of security. The person with MND may find it impossible to use their voice or a bell because of the effect of MND on the muscles used for speaking and hand and arm movement.

A modified door chime or intercom system could be used as a simple call device.

The switch adapted door chime is in two parts: the switch part and the chime part. The switch part remains with the person needing assistance and they press the switch when they need assistance. The chime part is positioned near the person responding to the chime. When the switch is pressed by the person needing assistance, the other person will hear the chime and know that assistance is needed.
Care alerters
Care alert systems differ from call devices. A care alert system is used to notify a person who is not within hearing distance or in the immediate area. The person needing assistance usually wears a special pendant or bracelet that has a button that can be pushed to activate the call for assistance. Care alert systems are provided by commercial providers. For more information on providers see the MND Association booklet *Living with motor neurone disease: services and resources*.

General communication strategies and aids
Low technology communication aids
Low technology communication aids include writing, flip-books, charts, boards and eye movements.

Writing is often preferred if hands are unaffected and speech is difficult to understand. Magna Doodle, white boards and note books can be used.

Several cards in a small flip wallet can be used for name, contact details, a short explanation of the communication difficulty and other frequently used phrases.

Communication charts and books can be made up according to each person’s need. These may include phrase, letter and picture charts. They can also be created in dual languages. Sample communication charts are usually available from a speech pathologist/therapist, the MND Association and online.

See-through boards, such as the ETRAN board, can be used with eye movements to point to letters or words. The board is positioned between the user and the person they are communicating with.

An agreed set of eye movements can also be used for simple communication. For example one blink or raising the eyebrows for ‘yes’ and two quick blinks or closing the eyes for ‘no’.

Signing with the hands to replace speech is not usually an effective communication strategy for most people with MND. This is because shoulder, arm and hand muscles may also be deteriorating, or may start deteriorating once the technique is established, resulting in additional communication issues as the disease progresses.
Battery operated communication aids
Small portable battery operated communication aids are commonly used by people with MND.

A voice amplifier includes a small microphone and speaker with adjustable volume. Is it useful if speech is very soft or if speaking loudly causes breathlessness.

The MessageMate stores and plays pre-recorded messages. It can be activated by touch or switch scanning*. Recordings can be made by the individual or a family member. It can be used as a language interpreting tool if set up with specific words/icons in one language and pre-recorded messages in a different language.

The Lightwriter outputs typed words and sentences as spoken words and/or text on two small screens positioned on the front and back of the device. It is not a computer. It is a text to speech device.

Portable devices such as the iPad and Android tablet computers can be used with specific apps that display or output spoken words from digital keyboards or communication charts. Commonly used communication apps include Verbally, SpeakIt, Proloquo2go (pictured) and Predictable. Some of these apps can be used with switch scanning* if the person’s hand function also deteriorates.

A laser head pointer, which is a laser light that can be fixed to a headband or cap, can be used with communication charts when mobility and hand functions are very weak, but head support and head movement is good. The laser light beam is directed by head movement to letters, words or phrases on a chart. It is used with a switch* for the user to turn the light on or off.

*See Switches that only need very small movements, page 30.

Use accessibility settings on everyday devices
Mobile phones, tablet computers, laptops and desktop computers often have standard features that include accessibility settings and ease of access settings. Many have features so that the device can be used without touch, a mouse or keyboard.
**Modified mouses**

Use of a standard computer mouse requires shoulder, arm, hand and finger dexterity and smooth movement. A modified mouse can be used to replace a standard computer mouse. Mouses described below use a USB connection, but may also be available in wireless styles.

Point-it! is a joystick USB mouse for a Windows computer. Knobs for the joystick aid comfort during use and include carrot, sponge ball and T bar styles. It can also be used with a switch* to select the left or right mouse click.

The switch adapted mouse is useful for people who can move a mouse but are not able to click the left or right mouse buttons. It is a standard USB mouse for a Windows computer with a socket that can be used to connect a switch* to select the left or right mouse click with one or more switches.

The SmartCat/EasyCat touch pad USB mouse for a Windows computer is much the same as a laptop mouse pad, minimising movement of the arm and hand for access to the computer. It has built-in left and right click buttons. The user taps, scrolls or moves across the touch pad with their fingers to move the cursor.

The Track Ball USB mouse for a Windows computer is available in various styles and designed for the user to move the ball with any part of the body, including the foot and toes. It has built-in left and right click buttons and can also be used with a switch* to select the left or right mouse click.

The Mini Track Ball USB mouse for a Windows computer is designed to be held in one hand while the user moves the ball with their thumb. It has built-in left and right click buttons.

The SmartNav USB mouse replacement for Windows or Mac computer includes an infrared camera and specialised reflective adhesive dots. One of the dots is placed on the user (usually on a cap worn by the user or the bridge of a pair of glasses). The infrared camera tracks the movement of the dot for computer cursor movement and operation of the onscreen keyboard.

*See *Switches that only need very small movements*, page 30.*
Switches that only need very small movements

Switches are often used by people who have reduced movement and/or communication difficulties. This is because they only require very small movements of the hand, finger, foot, elbow or chin to be activated. A person uses the switch to turn a device on or off, or to send a command such as a mouse click. Switches connect to switch compatible devices either directly through a 3.5mm switch input jack or indirectly through a switch interface. For more information on switch interfaces see Switch interfaces, page 31.

The following switches all connect through a 3.5mm switch input jack or a switch interface.

The Click switch is designed to be used with light finger control. The finger can be resting on top of the switch, until the user pushes it down to activate the switch. The force of the push needed can be adjusted.

The Cushion/Pillow switch is a soft switch particularly suitable for head or cheek activation. The switch is activated by pressing the top foam surface. The soft material allows it to be attached to a wheelchair or pillow and is removable and machine washable.

The Jelly Bean switch is the most commonly used switch and it is activated by pressing the surface. It has mounting sockets which allow for easy attachment to mounting systems for positioning the switch near where the person can activate it.

The Magic Wand switch is activated by the very smallest body movement and contact with bare skin. It is usually positioned near the head or hand, with the face or finger used to activate it with the slightest touch.

The Micro Light switch requires very little pressure, as little as 10g, to activate the switch. It is usually positioned near the user’s finger.

The Papoo switch is a hands-free movement activated switch. This device detects movement, such as a wave of a hand or finger over the sensor, up to 1cm away and registers this as activation.
Switch interfaces

Not all communication devices, including some tablet computers, have an input socket suitable for 3.5mm switch jack.

A Bluetooth switch interface can be used to connect a switch to a device that has Bluetooth capability but does not have an input socket suitable for 3.5mm switch jack. The 3.5mm switch jack can be plugged into the switch interface, which uses a wireless Bluetooth connection to the communication device, enabling the switch and the communication device to work together.
Explore mounting systems for communication devices
Mounting systems are used to temporarily or permanently position a communication device for use. Features include:

- how the mounting system is attached to a table, chair, wheelchair or bed
- how the device is attached to the mounting system and the length, breadth and weight of devices that can be attached
- how easy it is to position the device for user comfort and reach once the device is mounted
- how easy it is to move or remove the mounting system when the device is not in use.

The Latitude switch mount can be used to mount a switch to various surfaces including a wheelchair, table, bench and bed.

The RAM Ez Roll’r mounting system provides suction mounting for an iPad on suitable surfaces including a table, bench and bed.

The Universal switch mount is an adjustable mounting system for a switch. The base of the mount can either be free standing or attached by clamp.

Think about comfort
The Ergo Rest is a movable forearm support designed to provide support for the arm. The user can rest their arm in the support while using a device or switch.

Obtaining communication aids and devices
Some MND Associations and government services have a range of communication devices available following referral from a speech pathologist/therapist. Contact the MND Association for details.
Movement and joints

People use their neck, upper limb and lower limb muscles for movement and posture.

Although the first symptom of MND for some people is leg muscle weakness, the first symptoms for others may be hand or arm muscle weakness, or a weakness of the neck muscles. These symptoms may be experienced individually or in combination. As the disease progresses, the movement and postural support provided by the muscles can be greatly reduced. Joints and ligaments are unsupported and unprotected.

**Neck, upper limb and lower limb muscle weakness**

**Neck muscles**
Neck muscles are used to move and support the head. Weak neck muscles can cause the head to droop. This can cause injury to the neck and head. It can also cause neck muscle strain. Weak neck muscles can make it harder to breath, swallow and communicate.

**Upper limb muscles**
Upper limb muscles include the muscles of the shoulders, arms, elbows and hands. A person with shoulder and arm muscle weakness may have difficulty raising their arms or lifting things. Some people with weak shoulder and arm muscles may still have a strong hand grip. People with hand muscle weakness may drop things and have difficulty with tasks such as pinching, writing, typing, managing buttons or zippers and picking up small objects. A person with very weak upper limb muscle weakness may be more likely to have falls because they can no longer use their upper limb muscles for balance. People with very weak upper limb muscles cannot support the weight of their arms and hands.

**Lower limb muscles**
Lower limb muscles include the muscles of the hips, buttocks, thighs, legs and feet. Early signs of hip, buttock and thigh muscle weakness include difficulties with climbing stairs, moving from a seated to standing position and difficulty getting in and out of bed. Early signs of leg and foot muscle weakness include foot drop, the dragging of feet and tripping. Some people with weak leg and foot muscles may still have strong hip, buttock and thigh muscles. A person with very weak lower limb muscles cannot move or support the weight of their lower body, legs and feet.

**Pain**
When a person has neck, upper limb or lower limb muscle weakness, pain and discomfort can occur because of muscle stiffness, immobility and unsupported head and limbs. Pain may also be caused by muscle cramps, spasticity, skin pressure and muscle twitching, known as muscle fasciculation. Getting good pain management strategies in place early can reduce the need for greater intervention later.
Approach to management
A person with neck, upper or lower limb muscle weakness requires a coordinated, multidisciplinary approach and regular review by their occupational therapist, physiotherapist and doctor.

Strategies for managing neck, upper and lower limb muscle weakness

<table>
<thead>
<tr>
<th>General strategies</th>
</tr>
</thead>
<tbody>
<tr>
<td>• See a doctor for cramp, pain, pressure sores or swelling. The medical review can often identify causes of uncomfortable symptoms. Short or longer-term medication for pain, swelling or cramp may be prescribed. For example, the prescription of baclofen for cramp</td>
</tr>
<tr>
<td>• Following medical review to eliminate other causes and problems, the physiotherapist and massage therapist can advise about gentle massage and range of motion exercises to reduce swelling and increase blood flow to immobile areas of the body</td>
</tr>
<tr>
<td>• gentle massage can assist with maintaining flexibility and reducing joint pain. For a person with MND, massage should be gentle because the ligaments holding the bones together may not have healthy muscular support. Gentle massage of the neck muscles is particularly important as these can become very stretched and painful</td>
</tr>
<tr>
<td>• range of motion exercises or passive stretching of the limbs, can reduce pain from spasticity caused by a tightening of the muscles in some people with MND</td>
</tr>
<tr>
<td>• it is not yet known if other forms of exercise, such as endurance and muscle strengthening exercises are beneficial or harmful for people with MND</td>
</tr>
<tr>
<td>• Relieve swelling by elevation of the affected limbs, with support that does not put direct pressure on the swollen area</td>
</tr>
<tr>
<td>• Have regular review by an occupational therapist to ensure comfort, especially if using a chair, wheelchair or bed for a prolonged period of time. The occupational therapist can assess the suitability of the chair or wheelchair and advise about pressure care cushions that can help with seating comfort. The occupational therapist can also advise about mattresses that can help with comfort in bed including foam mattresses and mattresses that provide alternating air pressure</td>
</tr>
<tr>
<td>• Use aids and equipment for postural support, movement and activities of daily living</td>
</tr>
<tr>
<td>• Get regular personal assistance for activities of daily living, to change positions for comfort and to avoid pressure sores</td>
</tr>
</tbody>
</table>
Positioning
The person with MND may need assistance to move weakened arms and legs into comfortable positions during the day and to turn in bed at night. Do not pull on a person’s arms or legs. The neck, shoulders, arms, hands, legs and feet may all need support for repositioning. It may take the person a minute or two to receive feedback from their body about the comfort of a new position and as a result it might seem that they make frequent requests to be repositioned. Very subtle adjustments of arms and legs may be necessary and the support of cushions and pillows often helps.
Cognition

Cognition is the mental process or action of acquiring knowledge and understanding. Approximately 50% of people with MND may experience some change in cognition, language, behaviour and personality. Most people experience relatively mild changes.

However, a small proportion (5-15%) of people with MND will show more significant changes and will receive a diagnosis of ‘motor neurone disease with frontotemporal dementia’ or MND/FTD. Often the symptoms of dementia happen before the motor symptoms, sometimes by a number of years.

When cognitive and behaviour changes occur in MND, it is because there have been changes in specific areas of the brain called the frontal and temporal lobes.

What do changes in cognition and behaviour look like?

When cognitive and behaviour changes are mild, they may not be noticeable or affect daily life. However, when changes are more pronounced, they can have a negative impact on a person’s lifestyle and relationships. Cognitive and behaviour changes can often be a source of confusion and misunderstanding for the person with MND, their family and community care team.

While the nature of changes in cognition and behaviour will vary from person to person, some of the most common symptoms are listed below.
Strategies for managing cognitive and behavioural change

If you or someone you know are experiencing these symptoms, it is important to consult with a general practitioner, neurologist or specialist MND clinic. Clinical neuropsychologists can also assist in diagnosing changes in cognition and behaviour, and can help the person, their family and the community care team to develop practical strategies to manage symptoms.

### Strategies often suggested for cognitive change

- Using simplified communication for example, giving simple directions, using short phrases and reducing distraction
- Keeping to a regular routine
- Using aids for remembering such as a diary and notes

### Strategies often suggested for behavioural change

- Using distraction and diversion
- Letting family and friends know about the behavioural change to increase their understanding of the situation
- Accessing support options such as talking about issues arising with understanding family and friends, and seeking counselling
- Getting a neuropsychologist cognitive assessment and advice if significant cognitive involvement is suspected
Emotional lability

A person’s emotions can also be affected by MND. Known as emotional lability or pseudobulbar affect, signs include inappropriate or exaggerated emotional responses. Emotional lability is different to cognitive and behavioural change.

Emotional lability and MND

Emotional lability is a common symptom of MND, although not all people with MND will experience it. Emotional lability is not a mood disorder.

A person with emotional lability may have unpredictable and uncontrollable episodes of laughing or crying. The involuntary laughing or crying response may not be reflective of their real feelings.

Effects of emotional lability

Sometimes, these involuntary episodes can be so disruptive that the person with emotional lability may avoid social situations. The person may feel embarrassed by these episodes.

I also noticed that I had a problem with laughing. I was afraid of going out socially in case I couldn’t control it.

Jane (who lives with MND)

Managing emotional lability

<table>
<thead>
<tr>
<th>Let others know</th>
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<tbody>
<tr>
<td>• that they have these unpredictable episodes — that their laughing or crying may not reflect what they are feeling inside</td>
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</table>

<table>
<thead>
<tr>
<th>During episode</th>
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</thead>
<tbody>
<tr>
<td>• distraction</td>
</tr>
<tr>
<td>• slow breaths in and out</td>
</tr>
<tr>
<td>• focusing on something unrelated may reduce symptoms</td>
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<thead>
<tr>
<th>Medication</th>
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<tbody>
<tr>
<td>• general practitioner</td>
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<td>• neurologist</td>
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<tr>
<th>Reassurance from others</th>
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<tr>
<td>• that they understand</td>
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</table>
Bladder and bowel

The bladder and bowel are not usually directly affected by MND. However, some symptoms of MND and their treatment can cause bladder and bowel management difficulties.

Bladder and bowel difficulties in MND

Some people with MND may have fasciculation, or muscle twitching, in the pelvic area. This may irritate the bladder and cause urinary urgency. People with MND who have limb muscle weakness may move more slowly to the toilet or need assistance to get to the toilet. They might be very concerned about getting ‘there’ in time.

People with MND who have reduced arm movement may find it difficult to arrange their clothing and manage toilet paper. Some people with MND will have very limited or no mobility and will use portable aids and equipment to assist them with passing urine and bowel movements. Others may find that their reduced buttock muscle mass may make it uncomfortable to sit on an unpadded toilet seat.

Although MND does not affect the anal sphincter muscles, if MND is affecting a person’s respiratory and abdominal muscles it can be difficult for the person to supply the push needed to have a bowel movement. This can result in constipation. Other people with MND may have constipation because of dietary changes, not having enough fluids and the effects of some medication.

Approach to management

The aim of the management plan is for the person with MND to feel comfortable, and less anxious, about going to the toilet. The Bristol stool chart is a useful reference and communication tool.

Bladder and bowel management plan

- Advice from a dietitian about fluid and dietary intake and preventing constipation
- Advice from an occupational therapist about helpful aids and equipment
- Advice from a physiotherapist about the most effective use of the abdominal and respiratory muscles
- Strategies for ongoing review
- Ongoing monitoring and advice from a community nurse

General toileting tips

An occupational therapist can advise about toileting aids and equipment including:

- increasing the height of the toilet
- using a toilet surround, raised toilet seat or over-toilet aid
- using a padded toilet seat
- using grab rails near the toilet
- using a mobile or bedside commode
- using a standard or portable bidet
- ensuring the toilet paper is within easy reach.
Bladder management
Incontinence is not common in MND however some people with MND may have increased urinary frequency and urgency, or difficulty in emptying the bladder. Sometimes pre-existing conditions may need to be taken into consideration.

If the person with MND is worried about when or how they are going to pass urine, they might drink less which can result in dehydration.

For people with MND who have swallowing difficulties, dehydration may lead to thickened saliva. Dehydration can also contribute to constipation.

Strategies for bladder management
It is important that the person with MND does not reduce their fluid intake because they are worrying about how and when they can pass urine. Regular toileting routines are important if the person is becoming less mobile. With a regular toileting routine, a person with limited mobility can feel it is okay to drink now, knowing that there will be the opportunity for them to pass urine at the scheduled time. Easy to manage clothing, continence pads and pants, and portable urinals for both men and women are often considered. Some people with MND may have a urinary catheter to pass urine.

Bowel management
Constipation is commonly experienced by people with MND. Causes include lack of mobility, weak respiratory and abdominal muscles, dietary changes, poor fluid intake and medications. Constipation can be very painful if left untreated. Early referral and intervention for the person with MND can reduce the need for more intrusive interventions later, such as suppositories and enemas.

Strategies for managing constipation
Weak abdominal and chest muscles and difficulty taking or holding a deep breath make it hard to bear down and supply the push needed for a bowel movement. Extra time may be needed, and if the person has reduced buttock muscle mass they may need a padded toilet seat for comfort. Optimum positioning, providing privacy and not rushing the person can be helpful. A physiotherapist can advise about positioning and the best use of muscles.

People who have swallowing difficulties often have dietary changes to accommodate fluid and food consistency needs. These dietary changes can also contribute to constipation, as can some pain medications. Constipation can also occur when people use tube feeding for their fluid and food intake.

Regular fluid intake helps with managing constipation. A well-balanced diet will help avoid constipation, as can including particular foods and drinks that may have a laxative effect, such as prunes or prune juice. A dietitian can advise about this.

The dietitian or community nurse can also advise about commercially available medications that help maintain bowel activity. People taking fibre-based laxatives need to drink enough fluids to be able to pass bulky bowel movements. So a person who is not drinking enough fluids, has swallowing difficulties or who has abdominal and respiratory muscle weakness needs to take this into consideration before commencing treatment for constipation using fibre-based laxatives.

Other commercially available laxatives include stool softeners and stimulant laxatives. Stool softeners have a gentle effect while stimulant laxatives are usually used when other options have been exhausted.
Fatigue and insomnia

Fatigue and insomnia are often referred to as secondary symptoms of MND – not caused by MND, but often experienced by people with the disease.

Reasons for fatigue in MND

- fewer muscle cells getting messages from the brain
- cells getting the messages need to work at full capacity to perform the job or task
- reduced nutritional intake
- less air into the lungs
- side-effect of the medication riluzole

Fatigue management strategies

**Plan**

- plan activities in advance

**Rest**

- between activities
- before going out

**Energy conservation techniques**

- more intensive tasks for times of greatest energy
- aids or equipment for activities that are tiring
- conserve energy for important activities
- leave other tasks for others to complete
- domestic and personal care assistance

**Advice**

- respiratory specialist, nurse, physiotherapist, occupational therapist, rehabilitation specialist

Non-invasive ventilation, or NIV, provides relief from symptoms such as fatigue, breathlessness and disturbed sleep patterns, but does not prevent progressive weakening of the respiratory muscles. The NIV machine is usually used at night but as the respiratory muscles weaken the person with MND might use it at times during the day as well.
Reasons for insomnia
Insomnia, or sleeping difficulty, is common in the general population as well as in people with MND.

Reasons for insomnia
- ineffective breathing or nocturnal hypoventilation
- immobility
- pain due to stiffness of joints or muscles
- medication
- drooling saliva
- anxiety or other feelings associated with the emotional impact of MND

Managing general causes of insomnia
Different sleep strategies work for different people. They include:
- going to bed at the same time each night
- making sure the bedroom is quiet, cool, dark and comfortable
- avoiding over-exertion before going to bed
- avoiding caffeinated drinks
- avoiding heavy meals.

Breathing may be helped by elevating the head and chest with an extra pillow or raising the head of the bed on blocks. This relieves pressure on the diaphragm and improves lung expansion. The physiotherapist or occupational therapist can advise.

Sleep, positioning, muscle tightness and joint stiffness
While we are sleeping we often change position. However, because MND affects the muscles used for movement, the person with MND may need assistance during the night to change their sleeping position.

Inability to move and weight-loss can cause severe discomfort and skin breakdown. When lying in bed, the person with MND may need frequent turning. This can be as frequent as hourly repositioning. Very specific positioning of arms, legs and head may also be needed to maintain comfort. Light bedclothes and satin or silk sheets can allow for easier movement in bed.

An occupational therapist can advise about how side rails and extra pillows can provide added support. A segmented overlay mattress and specialised pillows can be used to distribute body pressure more evenly. An electric bed can be adjusted to a variety of positions.

Sleep, saliva and dry mouth
Drooling saliva, thick saliva and dry mouth can also cause sleep difficulties. Sleeping with the head elevated can prevent secretions from pooling in the upper airway.
Wellbeing and support needs

Wellbeing and support needs are general day-to-day needs that may arise from the impact MND is having on the person’s life, other family members and friends.

Information and planning

Information needs

Most people know very little about MND and seek information about the disease and any treatments that might stop or slow the disease progression. They, their family or friends will often continue searching for any information about a cure for MND. Others, however, may want to know very little about the disease. Some people may want lots of information, but only about specific topics. Sometimes, the person with MND may want very little information and the family and friends will read everything they can find. More information about MND is available from:

- the neurologist, general practitioner and other health professionals
- other publications of the MND Association
- the MND Association.

Planning

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

MND related planning topics

- symptom management
- accessing equipment
- daily living and mobility
- psychosocial and spiritual care
- family wellbeing and support
- support for children and families
- accessing superannuation, life insurance and financial advice
- financial assistance and benefits
- palliative care
- legal issues including setting in place arrangements for financial, lifestyle and health care decision making on the person's behalf
**Daily living and mobility**

Areas of daily living include personal care, domestic tasks, recreation, social interaction, community access and mobility.

People living with MND need individualised levels of support to help them manage their daily personal activities. Aids and equipment are used to assist the person to remain as independent as possible for as long as possible. They also help maintain comfort for the person with MND. Some aids and equipment items are used by a family carer or a community care worker to help them in their roles.

Ongoing assessment of daily living needs is important as these will change and the person may suddenly require additional aids and equipment to manage at home.

Domestic assistance may include assistance with house cleaning, washing, ironing, shopping and general household support, such as paying bills and accounts or assisting with telephone calls. Personal care needs may include assistance with performing essential self care tasks including bathing, dressing, eating and personal grooming. To maintain adequate nutritional intake the person with MND may require assistance with meal preparation and delivery.

Delays in accessing services, aids and equipment can have a significant impact on the person with MND. People with MND often have significant short-term, time-limited needs.

**Psychological and spiritual needs**

MND is a disease that has no cure and limited treatment beyond supportive care. Therefore the focus is on providing care that helps people with MND and their families cope with the progressive nature of MND and the ever-increasing disability it causes. Total care should include meeting physical, emotional and psychological needs.

Although often overlooked, psychosocial factors seem to play an important role in ALS/MND outcome (Chio and others 2009).

Psychosocial care is the responsibility of everyone working with those affected by ALS/MND (Gallagher and Monroe 2006).

**Motor neurone disease**

A diagnosis of MND turns a person’s whole world upside down and many assumptions, hopes, plans and expectations for the future have to be reviewed. The diagnosis does not only affect the person with the disease but also the people close to them. People with MND and their families often suffer considerable psychological and emotional distress.

Losses experienced in MND include loss of independence, role within the family, self-esteem, self-image and social interaction. Fears may include increasing disability, suffocation, choking and isolation, fear of the dying process and fear of the unknown.

Anxiety is emotional pain and worry. People with MND may have multiple causes for anxiety. These may include financial planning for their own and their family’s future, how the family will cope, as well as how they will be cared for as the disease progresses.

Depression is common (as it is in any devastating disease) and can be treated if persistent. Remember though, that a lack of facial expression may be due to weakness of the facial muscles, not depression. We rely so much on non-verbal responses that an expressionless person may appear angry, disinterested or depressed.

The major challenges are coping with loss and living with change. Much can be done to alleviate this distress, help people to adapt and make the most of their coping skills.
Psychosocial care
Just as there is no ‘formula’ for managing the physical aspects of MND, each family’s way of coping emotionally will be different. Psychosocial care is the responsibility of everyone working with those affected by MND (Gallagher and Monroe 2006). We need to think about “how we integrate these aspects of care into organisation of physical care” (McLeod and Clarke 2007). Spending time with the person with MND, talking through fears and worries can be very worthwhile. Services required may include psychological assessment, counselling and social support.

Spiritual needs
Spiritual care includes the health and community care team involved in care being aware of the individual’s spiritual and religious values. This can assist those working with the family to be respectful of the differences or similarities with their own belief systems. It is important for staff to always be appropriate and non-judgemental, to let the family know that they understand they have needs in this regard and that they are there to do the best for them. 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. Talking to a spiritual advisor may be helpful when these issues arise. A pastoral care worker may be available through the health, community palliative care or hospice care team.

Requests for voluntary assisted dying
Some people with MND may talk about wanting to hasten death through voluntary assisted dying. Currently, voluntary assisted dying is not legal in most states of Australia or New Zealand.

Discussions around assisted dying can be associated with a desire to keep control and a fear of how death will occur. People with MND need timely access to supports and services to maintain quality of life and independence and this is especially true in the later stages of the disease. It is important that they know that they are not alone and that there are people who have their interests at heart, on whom they can rely.

While many people with MND fear that they will choke to death, such a possibility is extremely rare. They can be reassured that death in MND is generally due to breathing failure which, for the majority of people, results in a gradual and peaceful lapse into unconsciousness prior to death.

Carer wellbeing and support
Usually, when a person with MND needs the assistance of a carer their partner, a family member or close friend steps up to the caring role. While caring can be a highly rewarding experience when appropriate support is available, without support, the responsibility of caring for a person with MND can have an adverse effect on a carer’s physical, mental and emotional health. It can also impact on the carer’s short and long-term financial security.

The demands of caring can make it harder to have a job, care for other family members and to take time to relax. Services provided to support the carer may include respite care, emotional support, MND education and assistance with accessing services.

Children and families
MND forces changes in family roles and relationships. The needs of the person with MND need to be balanced with those of other family members.

You can find more information about Wellbeing and support needs at www.mndcare.net.au
Advance care planning

It can be difficult for health and community care professionals to know when and how to have conversations about advance care planning with the person with MND and their family. Although there may be differences in how MND progresses, it is not uncommon for people living with the disease to become concerned about what lies ahead and how decisions about their finances, health and lifestyle will be made.

Planning discussions provide health and community care professionals with insight into how the person with MND and their family feel about the issues that may arise during the course of the disease.

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

This can be particularly important if the person with MND and their family have specific wishes regarding health care management and interventions.

For people with MND who expect to have future communication or cognition difficulties, advance care planning provides them with the opportunity to let others know about their wishes and preferences.

Getting started with advance care planning

Planning in MND has a two-fold purpose:
- plan for the future as much as possible
- plan for independence, with supports, for as long as possible.

Advance care planning might involve

Advance care planning is about having conversations and, if desired, drawing up formal documentation. Advance care planning might involve (Wilson 2010):

- organisational requirements
- advance health directive
- enduring power of attorney
Possible advance care planning steps

**Thinking about**
- future preferences
- who might make decisions on the person’s behalf

**Talking with**
- family and friends
- general practitioner or palliative specialist

**Recording wishes in legal documents**
- advance health directive
- enduring power of attorney

**Providing copies to family, doctor and/or hospital**

**Keeping copies available for emergencies**

(Wilson 2010)

It is important to note that even if the formal forms are not filled out the discussion, communication and awareness formulated throughout advance care planning processes may still be helpful (Wilson 2010).

Barriers to the use of advance care planning and advance health directives

- don’t know how to
- don’t know enough about them
- prefer to leave decision to doctor
- don’t like to think about these end of life issues
- prefer to leave decision to family
- prefer to leave it until situation arises

While many people welcome discussions about end-of-life planning, some prefer to delegate decision-making authority to proxies, such as a family member rather than make rigid decisions now for complex decision making in the future (Wilson 2010).

In conclusion, advance care planning helps people with MND and their family to talk about concerns and fears about what lies ahead. It enables them to set in place arrangements for financial, health and lifestyle decision-making that may allay some of those concerns and fears.

You can find more information about Planning for future care needs - advance care planning at www.mndcare.net.au
End of life care

MND is a progressive neurological condition. Over time, muscles become weaker because there are fewer and fewer healthy motor neurones controlling them. The most common cause of death in MND is breathing failure.

As the needs of the person with MND and their family become more complex, increased involvement by a palliative care team can be integral to optimal symptom management towards the end of life.

During this time the person with MND and their family need practical and emotional support. Often, this is provided by the multidisciplinary team, including palliative care.

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 whether they will need to go to hospital or a hospice or whether they can stay at home for as long as possible, even until they die.

It is important for these fears and questions to be discussed. Palliative care professionals can help with this. The community palliative care team should be involved and can provide support to both the person with MND and the family.

One of the most common fears is that of the person choking to death. Although a person with MND may experience some choking episodes, it is very rare for this to be the cause of death.

Some people with MND do die suddenly due to a sudden weakening of the respiratory muscles or chest infection, but most people gradually deteriorate due to increasing respiratory weakness.

Palliative care

Palliative care teams provide expert symptom management and emotional and social support to people with life threatening illnesses and their families in their own home.

The palliative care team usually includes allied health professionals, as well as nursing and medical staff. Some palliative care teams provide diversional, music and alternative therapies and volunteer visits.

Establishing links for the person with MND and their family with palliative care provides the opportunity for relationships to be formed before the very final stages of the disease.

Additionally, the palliative care team can provide advice and support to other multidisciplinary team members.

A late referral to palliative care, although not ideal, can provide the person with MND and their family with practical and emotional support.

Palliative care may be available 24 hours a day and can be accessed through the local hospital, hospice or community health centre.
Features of optimal end of life care

End-of-life care that leads to a more peaceful terminal phase and then death, is more likely to occur when

- care plans and information are shared
- adequate nursing cover is available
- comprehensive symptom control is used
- psychosocial support is offered
- support for family and friends is offered
- the person with MND has the opportunity to say goodbye to those closest to them

The terminal phase

The terminal phase of MND is recognised as increased progressive weakness. The health of the person with MND may appear to suddenly deteriorate over a few days or, for some people, over a few hours.

This may be preceded by:
- reduction in chest expansion when breathing in
- quietening of the breath sounds
- use of accessory muscles for breathing
- morning headache from carbon dioxide retention overnight.

These signs may be noticed by the family or by a member of the home and community care team.

Preparing for the coming days

Recognising these signs provides the opportunity for the team to help prepare the family for the coming days and the person’s impending death. Although the person may have had MND for several years, this preparation may help prevent the family shock of an ‘unexpected’ death.

Preparing for the coming days involves talking about

- the signs of impending death
- what to expect
- if pain is experienced, pain management
- how and who to contact for immediate advice
- how to involve others
- additional comfort items
- managing the moments following death

Medications in the terminal phase

Morphine is frequently prescribed for breathlessness, anxiety and/or pain. Morphine may be given orally as a liquid, or tablet, or a continuous injection of small doses via a self managed syringe driver (a needle is placed under the skin and left in place for a couple of days attached to a small machine). The dose is carefully monitored by the general practitioner, palliative specialist, community nurse or palliative care nurse.

There are a number of other medications such as Lorazepam, Diazepam (valium) or Midazolam - which may be prescribed to treat anxiety and restlessness.
Oxygen may be prescribed to minimise the feeling of breathlessness in the end stages of MND.

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

### Medications in the terminal phase
- **Opioid analgesics** - reduce cough reflex, relieve dyspnoea, control pain and help to reduce fear and anxiety
- **Anti-cholinergics** - such as hyoscine hydrobromide and glycopyrrolate reduce saliva and lung secretions
- **Sedatives** - such as diazepam, midazolam, clonazepam and chlorpromazine reduce anxiety
- **Oxygen** - can be used to relieve the sensation of breathlessness
- **Haloperidol** - for terminal restlessness

### Bereavement
For someone bereaved by ALS/MND it may be difficult to accept that the illness journey has ended (McMurray and Harris 2006).

*For each person the bereavement experience is personal and unique. In ALS/MND, 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.*

### Strategies to assist in making real the loss
*(McMurray and Harris 2006)*
- provide an opportunity to view the body of the person who has died
- allow for the expression of feelings
- acknowledge the normality of grief
- review the loss experience
- provide information and support about grief
- assist with anniversaries and memories
- consider and acknowledge secondary losses
Dying and death
Dying and death is an intense time for families and for health and community care professionals. It is important to recognise and acknowledge the impact on yourself.

Providing support and care for people living with MND
Providing support and care for people living with MND is challenging because it can be both physically and emotionally demanding.

People living with MND often need very complex care. It often requires a lot of your time, resources, creativity and energy to ensure the person gets good quality care. Over time it is not unusual for health and community care professionals to develop a strong relationship with the person with MND and their family.

MND is often described as a devastating disease. No matter what your role is, watching someone lose their functional ability and being part of the support team can cause stress. It is important that we recognise this and take steps to manage our own stress and frustrations in these situations. It is also important to acknowledge our own grief and the grief of others.

Aspects of self-care when working with people with MND
- developing workplace preventative strategies
- looking after yourself outside of work
- recognising that death is a part of life
- looking out for signs of depression or burnout
- accessing resources to keep you informed about MND

You can find more information about End of life care and self-care for health and community care professionals at www.mndcare.net.au
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The signs and symptoms of MND

**Emotional lability**
- inappropriate or exaggerated emotional responses

**Cognition**
- mild changes in behaviour, cognitive skills and cognitive processing

**Swallowing and nutrition**
- extra effort to chew
- coughing whilst eating or drinking
- several swallows per mouthful
- eating or drinking is tiring
- breathlessness after a meal
- mealtimes take longer
- frequent chest infections
- slurred or indistinct speech
- muffled or ‘wet’ sounding voice after eating
- difficulty clearing saliva

**Neck weakness**
- head drops
- strain and injury
- harder to breathe, swallow, communicate

**Breathing difficulties**
- disturbed sleep
- daytime sleepiness
- morning headaches
- quieter voice
- fewer words per breath
- shallow, faster breathing
- weakened cough and sneeze

**Bowel and bladder**
- constipation
- move more slowly/need assistance

**Foot muscle weakness**
- foot drop
- dragging of feet
- tripping

**Fatigue and insomnia**
- muscle tiredness
- low energy
- general tiredness
- trouble sleeping
- restless sleeping

**Leg weakness**
- difficulty climbing stairs, standing up, getting out of bed
- no longer support own weight

**Communication and speech**
- weakness and reduced coordination of the muscles used for speech — lips, tongue, facial muscles, larynx, pharynx

**Saliva and mouth care**
- drooling saliva/thick saliva/dry mouth
- difficulty swallowing or coughing up saliva
- increased risk of aspiration
- dehydration
- sensation of choking

**Arm and shoulder weakness**
- difficulty raising arms
- difficulty lifting
- pain from being lifted under arms

**Hand weakness**
- drop things
- difficulty pinching
- writing and typing
- managing buttons or zippers
- picking up small objects

**Pain management**
- muscle spasms and cramps
- swelling and joint stiffness

**General comfort issues**
- muscle stiffness and immobility
- unsupported head and limbs
- muscle cramps
- spasticity
- skin pressure
Managing the signs and symptoms of MND

**Cognition**
- simplified communication
- regular routine
- aids for remembering
- education and support for carers/family
- professional consultation

**Swallowing and nutrition**
- soft/puree diet
- thickened foods/liquids
- correct positioning for meals
- consider peg

**Emotional lability**
- let others know
- medication

**Communication and speech**
- speech pathology review
- effective communication techniques
- low technology communication
- battery operated and electronic aids and equipment
- computers, i-pad/ tablets

**Saliva and mouth care**
- position body, provide head and neck support
- oral swabbing and syringing
- medication
- dark grape juice, papaya extract
- mouth care

**Neck weakness**
- supportive collar
- headrest
- reclining chair
- neck and limb muscle support during transfers

**Breathing difficulties**
- avoid people with coughs and colds
- well balanced diet
- positioning, room airflow and temperature
- breathing exercises
- fatigue management
- non invasive ventilation
- medication

**Arm and shoulder weakness**
- support arms, do not pull or lift under arms
- transfer belt or hoist
- investigate shoulder injury
- adaptive cutlery
- modified buzzer

**Bowel and bladder**
- maintain adequate fluid intake, dietary intake
- bowel management regime
- optimal positioning
- aids and equipment
- regular toileting
- ongoing monitoring and advice

**Hand weakness**
- passive exercises
- consider hand splints

**Foot muscle weakness**
- ankle and foot support
- walker

**Foot muscle weakness**

**Fatigue and insomnia**
- plan activities in advance
- rest
- energy conservation techniques
- nutritional review
- medication review
- respiratory review

**Leg weakness**
- physiotherapy and occupational therapy review
- mobility aids
- wheelchair
- raiser/recliner chair
- bed blocks, electric bed
- toilet raiser, shower stool/chair
- home modifications – ramps, grab rails

**Pain management**
- treat muscle spasms and cramps
- multidisciplinary care plan

**General comfort issues**
- physiotherapy and occupational therapy review
- frequent change of position
- frequent turning in bed
- pressure cushions and mattresses

*mnd Aware*
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