

# Living with motor neurone disease

**Aspects of care**  
for people with MND,  
their family and  
friends



**Living with motor neurone disease: aspects of care for people with MND, their family and friends**

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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

*Living with motor neurone disease: aspects of care for people with MND, their family and friends* has been developed by the Motor Neurone Disease Association to provide you with information about living with MND at home.

MND is a very individual disease and not everyone gets the same symptoms. The patterns of weakness in MND vary from person to person. Therefore not all the information in this publication may be relevant to your particular situation. Just read those parts that you think will help you now. You can come back to the other parts if or when you need to.

### Publications in the Living with Motor Neurone Disease series

#### **Living with motor neurone disease: aspects of care**

- provides an overview of MND and strategies for symptom management

#### **Living with motor neurone disease: day-to-day**

- provides strategies for living at home with MND, more in-depth information and day-to-day tips

#### **Living with motor neurone disease: services and resources**

- provides contact details for organisations that provide services and information that may be of interest to people living with MND
- searchable online directory at [mnd.mndnsw.asn.au](http://mnd.mndnsw.asn.au)

#### **Living with motor neurone disease: for carers**

- for family and friends caring for a person with MND - provides information about the importance of looking after yourself

### For more information

Contact your MND Association. See back cover for full contact details.

## About MND New Zealand

The New Zealand Motor Neurone Disease Association (MND New Zealand) is a non-profit, registered charity established over 30 years ago for the purpose of supporting those living with MND.

We offer information and support to people living with a diagnosis of MND along with their whanau/carers and the health professionals and service providers involved.

We work to increase awareness and understanding of MND in New Zealand and to be a reliable link to information related to MND, its management, and ongoing research.

### Information

- MND New Zealand provides reliable information for people with MND and their whanau, through our support service and at [www.mnd.org.nz](http://www.mnd.org.nz)
- We provide education about the needs of people with MND to the broad range of health professionals and service providers involved in their care.
- We share research news and stories about living with MND from around the world on our Facebook page at [www.facebook.com/mndanz](http://www.facebook.com/mndanz)

### Support

- Our MND support advisors are qualified health sector professionals with knowledge and experience related to MND. They are a key point of contact for people with MND and their whanau and carers.
- Our support advisors offer free, personalised support and advocacy for those living with MND through home visits, phone, text and email.
- Our support advisors are well connected with local service providers and can liaise on behalf of those living with MND to facilitate access to services and equipment as appropriate to individual needs.

### Research

- MND New Zealand works to advance opportunities for New Zealanders with MND to participate in research if they wish, and grow the field of MND research in New Zealand.
- We fund the NZ MND Registry ([www.mnd.org.nz/registry](http://www.mnd.org.nz/registry)) and the NZ MND Research Network ([www.mndresearch.auckland.ac.nz](http://www.mndresearch.auckland.ac.nz)).

### Advocacy

- MND New Zealand advocates for change through raising community awareness about the needs of people living with MND.
- Our MND support service assists people with MND, their whanau and carers to self-advocate to service providers and funders.
- We advocate for the needs of people with MND at a political level and encourage statutory agencies to fulfill their obligations.

Donations, fundraising and bequests fund 90% of MND New Zealand services. Just 10% of our income is received from the Ministry of Health under a Disability Information & Advisory Services (DIAS) contract.

Membership of MND New Zealand is free for people with MND who live in New Zealand. Contact National Office on (09) 624 2148 or visit [mnd.org.nz](http://mnd.org.nz) for more 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.

### 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.

### First signs and symptoms

Some of the first signs and symptoms of MND may include weak hands, fatigue, muscle pain and cramp, muscle twitches, difficulty speaking or swallowing, weakness in the legs, loss of muscle tone, immobility, discomfort and pain, breathing difficulties and drooling or thick saliva.

Not all people with MND will be affected in the same way. For example, although the first symptom of MND for some people is leg muscle weakness, the first symptom for others may be arm muscle weakness or a weakness of the speech and swallowing muscles.

The diagnosis of MND is often clinically difficult. There is no single test for the disease and sometimes it is necessary for a doctor or specialist to review a person for some time before the diagnosis becomes reasonably certain.

The effects of MND become more generalised as the disease progresses. However, for some people, the symptoms are widespread from onset.

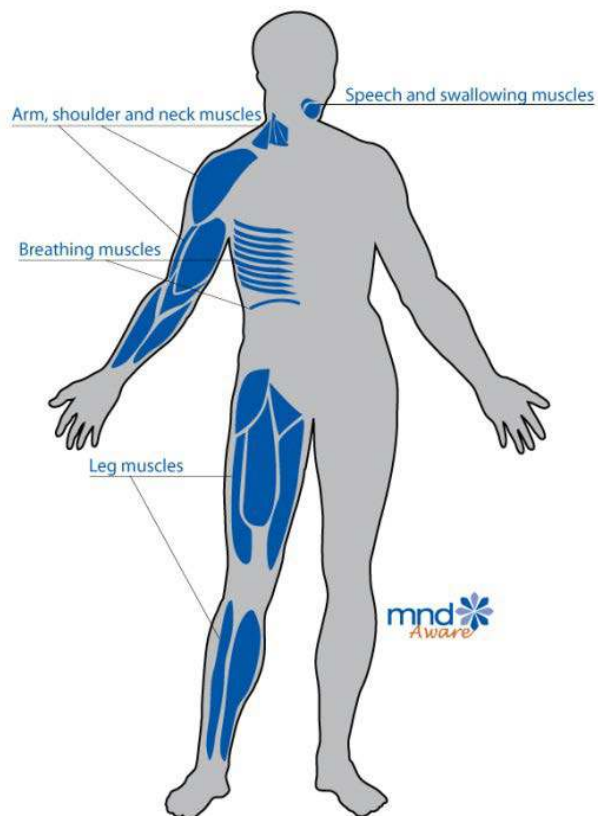


Figure 1 Voluntary muscles

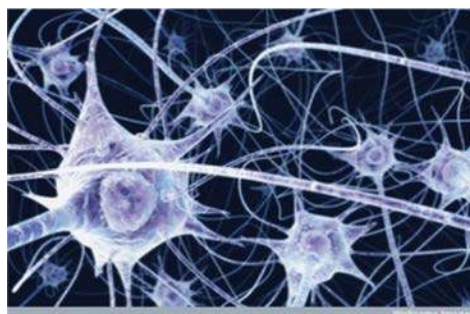


Figure 2 Illustration of a network of nerve cells in the brain

Source: Benedict Campbell, Wellcome Images

## 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.



## 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.

The patterns of weakness and rates of progression in MND vary from person to person and there is currently no cure for MND.

## How do people get MND?

There is no evidence that MND is transmissible from person to person. For about 90% of people with the disease, MND occurs sporadically. This means the person has the disease, but there is no clearly identifiable cause.

However, about 10% of people with MND have familial or hereditary MND. Of this small group, one-fifth has a genetic mutation on the SOD1 gene. Other mutations linked to MND include TDP 43, FUS and C9ORF72.

Clinically, the sporadic and familial forms of MND are indistinguishable.

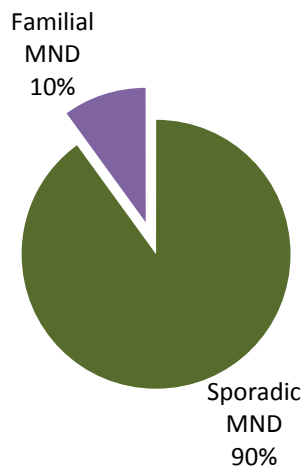


Figure 3 Incidence of familial and sporadic MND in people with the disease

## Incidence and prevalence

### Motor neurone disease

- 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

## MND progression

As MND progresses more motor neurones degenerate and die. When these motor neurones stop activating the voluntary muscles, the muscles weaken and waste.

Because MND affects the voluntary muscles, people with MND may experience any one or a combination of speech difficulties, swallowing problems, immobility, discomfort and pain, breathing difficulties, drooling or thick saliva.

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

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

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%) of all people diagnosed 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.

## Living with MND

People often describe the months before being diagnosed with MND as a time of uncertainty, frustration and worry.

*"We knew there was something wrong and Keith had many tests. Motor neurone disease had sort of been mentioned, but so were many other things. When it finally got to the stage of the doctor saying that it had to be MND, because everything else had been ruled out, we were relieved in a way. At last we had a name. But then that was sort of the end of the appointment and we drove home. I guess we were in a really slow form of shock. Not knowing anything at all really about MND but also sort of knowing that life as we knew it had changed."*

Simone, partner of Keith

A follow-up appointment with your doctor will give you the opportunity to ask questions about MND, discuss symptom management and plan regular review (see *Regular medical review*, page 13).

### Living with MND

- 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
- 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

## 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 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. You can get more information when you need it from:

- your neurologist, general practitioner and other health professionals
- other publications in the MND Association *Living with motor neurone disease* series
- MND Association (see page i).

#### Using the Internet to access health information

- The Internet can be a valuable source of health information that can help you understand your health issues as well as those of your friends or family
- Health information on web sites should not take the place of your health provider/patient relationship. There are many factors that need to be considered in relation to your health and the diagnosis of any condition. You should use the Internet as an information resource and ask a health professional about any issues raised by the information or anything that you don't understand, such as medical terms
- You should also avoid any online health practitioner who proposes to diagnose or treat you without a proper physical examination and a full consultation regarding your medical history

From Health Insite, the Australian Government gateway to reliable health information at [healthinsite.gov.au](http://healthinsite.gov.au)

### Psychological and spiritual needs

During the days, weeks and months following the diagnosis, people often start thinking about how having MND will affect their plans and hopes for the future.

*"We told the children. Well, they are not children anymore because they are all grown up now. And it was a bit of a relief when we'd done that.*

*Keith was still going to work, but we didn't know how long he could keep doing that for and when he should tell the boss. We were just sort of going on.*

*We didn't know how the disease would progress in him and the things he would be able to do. How much time did we have? We didn't know and it was really hard not knowing.*

*Some days I felt pretty angry. Other days I felt a bit numb. There were plenty of days I just tried not to think about it. There was one day we were watching a show on TV and I just started crying I was so sad. Then Keith started crying too. Then I said 'look at us' and we sort of laughed. We were all over the place."*

Simone, partner of Keith

Each person is individual and will react in his/her own way depending on personality, coping skills and relationships within the family. You may find that you and other family members and friends might experience one or more emotions at different times from each other. Suddenly the world you all knew has changed. Time is needed to take in what is happening and to work through the many emotions being felt. There may be times when you just want to run away, times when you become very angry or very sad. At other times you will be able to get on with living your life.

It can be helpful to talk to people who are not involved directly such as your doctor, MND Association advisor, counsellor, social worker, clergy or others living with MND. Additionally, many MND Associations may provide:

- information evenings for those newly diagnosed with MND
- local MND support groups or coffee mornings
- education about MND, for people with MND and their family members.

Sometimes you may not want to talk about what is happening, while others would like to talk through everything or vice versa. This can be very frustrating and if this is the case it is important to seek out and share your feelings with other trusted people. Just as there is no 'formula' for managing the physical aspects of MND, each person's way of coping emotionally will be different. There are times when people are simply unable to share their thoughts and feelings and this needs to be recognised.

### Your family

Telling family about the diagnosis may be difficult and traumatic. Some people choose not to tell others for some time, while others prefer that everyone knows. How and when you choose to tell family members is a very individual decision.

MND often brings about changes in family roles and relationships. The needs of the person with MND need to be balanced with those of other family members.

### Your children

The time after diagnosis is particularly stressful for parents. At this time parents are themselves emotionally vulnerable and trying to sort things out in their own minds. It may be difficult to explain things that you do not clearly understand and where so many uncertainties exist about the future course of the illness and the effects on the family. Thinking about the future may be emotionally painful and talking to children about the illness can be a realistic acknowledgement of what is happening.

Children will usually know when something is wrong by the changes in parental behaviour and attitude. They are likely to be more adversely affected by being excluded from what is happening. They need simple, truthful information from someone they love and trust. This is generally a parent. Parents may need encouragement, help and support in doing this. Children need to be given an explanation in words and ways that they understand and be included in family discussions wherever possible and appropriate.

A child's ability to understand the illness and its implications depends on age and intellectual development. If young children are involved it may help to talk to your MND Association advisor. Alternatively talk with a counsellor, social worker or your spiritual advisor to get their advice before you talk with your children.

An information pack of six booklets, *Talking with Young People about MND*, is available from the MND Association.

## Your friends

The way your friends will react to your diagnosis is very wide ranging and many people living with MND comment on the fact that sometimes their relationships with friends change. Some people begin to isolate themselves to avoid having to deal with their friends' reactions. Some friends may avoid you as they find it too difficult to cope with MND, however, many may become a lifeline for you and your family in the months and years ahead and your friendship may even become stronger due to MND. Very likely, among the people in your wider circle, new people will become close as they respond to your needs.

Having to repeat your story to many friends and colleagues may become tiresome or may upset you. Booklets are available from the MND Association that explain briefly about motor neurone disease. These may be useful to have on hand to give to people.

It may also be helpful to:

- ask one friend to communicate information and updates on your behalf to others
- use apps or a social networking site to set up a group so that you only need to communicate information to one 'place' (see the MND Association booklet *Living with motor neurone disease: day-to-day* for more information).

## Your partner

A life threatening disease has the power to strengthen healthy relationships, or shatter already weakened ones. It can bring out the best in some people and in others, awaken emotions they find difficult. As with all major life events, talking to each other and discussing feelings are very important.

Try to discuss emotional issues openly with your partner and allow plenty of time to listen. Sharing thoughts and emotions can help deepen respect and emotional bonds.

For some issues, you may find it helpful to speak with someone who is not your partner - your doctor, MND Association advisor, counsellor, social worker, clergy or others living with MND.

Physical intimacy can be a great way of expressing your feelings for your partner and bringing you pleasure in your life together. Continuing closeness between partners may help to reduce many of the fears and frustrations the disease produces.

Generally, MND does not prevent a man from getting an erection, or affect the physical side of sex for a woman. However the physical demands of illness, depression, fatigue, anxiety and stress can all affect the sexual desire of you and your partner.

As physical changes occur, such as muscle wasting and weakness, you may need to adjust your positioning or type of sexual activity. There are many ways to achieve intimacy and sexual satisfaction apart from intercourse - touching, kissing, stroking, cuddling can be immensely reassuring and satisfying. Playfulness and creativity are needed as the disease progresses and expressing affection can be incorporated into physical care.

## Spiritual needs

Spirituality can include both religious and non-religious elements. It can be a resource for coping and an arena to explore finding meaning.

### What is spirituality?

- “Spirituality has many definitions, but at its core spirituality helps to give our lives context. It's not necessarily connected to a specific belief system or even religious worship. Instead, it arises from your connection with yourself and with others, the development of your personal value system, and your search for meaning in life. For many, this takes the form of religious observance, prayer, meditation or a belief in a higher power. For others, it can be found in nature, music, art or a secular community. Spirituality is different for everyone.”

The Mayo Clinic (2010) ‘Spirituality and stress relief: Make the connection’

Talking to a spiritual advisor, religious leader, minister of religion may be helpful. A pastoral care worker may be available through the health care team.

## Planning needs

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.

Most people find that living with thoughts and fears about what lies ahead is often harder than talking about and setting in place arrangements that can allay some of these concerns.

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

These arrangements can be formal or informal, although some arrangements require specific documentation to be legally enforceable. This can become particularly important if you have specific wishes regarding health care management and interventions or if you expect to have future communication difficulties.

Sometimes, setting in place your own arrangements will avoid the need for a Tribunal or Court to appoint someone to act on your behalf.

Communicating with others about what is important to you is an ongoing process. Planning ahead reduces the possibility that your partner, family, friends or a court-appointed guardian will need to 'second-guess' what you would want to happen.

Often, people find that their wishes change over time. You can change your mind about your arrangements at any time as long as you still have the capacity to make decisions and you can communicate this to others.

Planning ahead involves:

- identifying what is important to you
- communicating this to others
- making arrangements to ensure that your wishes are carried out.

If you have made legally enforceable formal arrangements about decision-making on your behalf they are only accessed if:

- you no longer have the capacity to make decisions, or
- you cannot communicate your decisions to others, or
- (in some instances) you have determined that they will take effect immediately.

Arrangements you might consider setting in place include:

- a power of attorney or an enduring power of attorney, both of which allow you to give authority to someone you nominate to make decisions regarding financial issues on your behalf
- giving legal authority to someone you nominate to make decisions regarding personal, health and lifestyle matters on your behalf; and
- an advance health care directive which allows you to provide clear guidance to your doctors about your intentions for medical treatment in the future.

For more information see the MND Association booklet *Living with motor neurone disease: day-to-day*.

## Daily living and mobility needs

As MND progresses it causes weakness of the muscles in the legs, arms and neck; and weakness of the muscles used for speech and breathing. The pattern of weakness varies from person to person.

People with muscle weakness can use aids and items of equipment to help them live with MND in their home, participate in their work and leisure activities, and to continue to socialise with family, friends and the wider community. These often include walking sticks and frames, wheelchairs, grab rails and small ramps; and communication devices, such as alphabet or word charts, tablet computers and hands free computer pointers and mice.

Some aids and equipment items are designed to help maintain comfort for the person with MND. These include electric beds and armchairs, pressure care mattresses, over toilet aids and transfer slings.

*“Make sure you get advice from your occupational therapist or physiotherapist before making decisions about items of equipment and changes at home. We bought a wheelchair, but Joan only used it for couple of months before she needed a wheelchair that could tilt back to support her body better, and then we found out that we could have got the wheelchair through the MND Association.”*

Bill, partner of Joan

Ongoing assessment of your mobility, daily living and communication needs is important as these will change and you may need additional aids and equipment to manage at home. For more information see *Communication*, page 28 and *Movement and joints*, page 30.



## Personal care, home care and respite care needs

Initially, because MND has a progressive impact on activities of daily life, when assistance is needed it is usually provided by a person's partner, family member and/or close friend.

However, as MND progresses, people with the disease have increasing personal and home care needs. Home and community care services can help you and your family maintain your independence and quality of life. These services also provide some relief and respite for family members and friends who have stepped up to the carer role.

### Personal care, home care and respite care

- Personal care services are community based services that provide trained care workers who can assist you with bathing, dressing, eating and personal grooming
- Home care services are community based services that provide trained domestic care workers who can assist with cleaning, washing, ironing, shopping; and general household support, such as paying bills and accounts or assisting with telephone calls. Home care services can usually provide assistance with meal preparation and/or delivery
- Respite care services are community based services that provide in-home or centre based care for a person whose carer needs a break

### Saying yes to services

Personal care, home care and respite care services can make it easier for you and your family to live with MND. But it can be hard to say 'yes' to home and community care services when they are suggested. This can be especially true if you are currently managing quite well. Due to potential delays in getting services up and running, it is important that you:

- are linked into home and community care services as early as possible
- have regular review of your daily living needs.

Delays in accessing services can have a significant impact and can be very stressful for you and your family.

Your MND Association advisor can advise you about services available in your local area.

## Financial needs

There are several government pensions, allowances and benefits for people who are aged, disabled or a carer who meet eligibility criteria. Other subsidy, assistance and rebate schemes are available for various goods and services, for example, travel and taxi subsidies, leisure activity ticket discount, electricity rebates, medical supplies and equipment.

Some superannuation and life insurance schemes have provisions where, under certain circumstances including serious illness, benefits may be released early. Additionally, some superannuation funds include default life insurance, income protection and/or total and permanent disability insurance within their schemes.

Financial and legal advice, including advice about managing joint accounts and tax rules related to early release of superannuation benefits, is available from a range of non-government sources such as a solicitor, financial adviser, bank or retiree association.

For more information see the MND Association booklets *Living with motor neurone disease: day-to-day*, and *Living with motor neurone disease: services and resources*.



## Multidisciplinary care and MND

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. 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.

### Regular medical review

The effects of MND - initial symptoms, rate and pattern of progression, and survival time after diagnosis - vary significantly from person to person. Regular medical review ensures current evidence-based treatment and symptom management recommendations are fully utilised by the person with motor neurone disease.

Generally, regular medical review<sup>1</sup> will cover:

- |   |
|---|
| 1. Developing or updating a personalised MND multidisciplinary care plan<br>Frequency: at least once annually. <i>See page 14</i>   |
| 2. Discussing the drug riluzole which has been shown to slow disease progression<br>Frequency: at least once annually. <i>See page 14</i>   |
| 3. Screening for cognitive and behavioural changes<br>Frequency: at least once annually. <i>See page 38</i>   |
| 4. Providing symptom management for emotional lability, drooling or thick saliva and other MND-related symptoms. <i>See pages 40, 25, 33, 36</i>  |
| 5. Discussing signs of respiratory muscle weakness (awake or associated with sleep) and, if signs are showing, referral for respiratory function testing<br>Frequency: at least once every three months. <i>See page 17</i>   |
| 6. Discussing respiratory treatment options including non-invasive ventilation and assisted cough, if respiratory signs are showing<br>Frequency: at least once annually. <i>See page 17</i>  |
| 7. Screening for difficulty swallowing, weight loss, and impaired nutrition with results documented in the medical record<br>Frequency: at least once every three months. <i>See page 20</i>  |
| 8. Discussing nutritional support (dietary or enteral nutrition support via percutaneous endoscopic gastrostomy or radiographic inserted gastrostomy) for people with difficulty swallowing, weight loss or impaired nutrition<br>Frequency: at least once annually. <i>See page 24</i> |
| 9. Providing a referral for MND communication support (speech pathology) for an augmentative/alternative communication evaluation for people who have communication difficulties<br>Frequency: at least once annually. <i>See page 28</i>   |
| 10. Discussing MND advance care planning<br>Frequency: at least once annually. <i>See page 10</i>   |
| 11. Asking about any falls and trips<br>Frequency: all visits. <i>See page 30</i>   |

<sup>1</sup>Adapted from Quality improvement in neurology: Amyotrophic lateral sclerosis quality measures © 2013 American Academy of Neurology. Republished with permission.

## Living better for longer

### Interventions to live better for longer with MND

- Multidisciplinary care
- Good nutrition
- Non-invasive ventilation (NIV)
- Riluzole (branded as Rilutek or APO-Riluzole). Research has shown that riluzole probably prolongs median survival by two to three months. Regular blood testing to monitor liver function (every month for three months, then every three months for a further nine months and annually thereafter) is recommended for people taking riluzole

## Multidisciplinary care

You can live better for longer with motor neurone disease when health professionals have a coordinated, multidisciplinary approach to your care.

Multidisciplinary teams are also known as primary health care teams. Team members communicate with each other about your care and help you get care from other members of the team when you need it.

Professionals providing multidisciplinary care can be from the same organisation, a range of organisations or from private practice. They can work in the community, hospital, clinic, residential and other care settings. Each discipline-specific team member enriches the knowledge-base of the team as a whole and, over time, the multidisciplinary team composition can change to reflect changes in your needs.

For example, you may find you need to talk with a respiratory specialist about breathing, an occupational therapist about equipment, a speech pathologist/therapist about communication, a physiotherapist about joint stiffness, a palliative care team about support and your general practitioner and neurologist for regular symptom review.

## Getting multidisciplinary care

Multidisciplinary care is available at several MND-specific clinics in Adelaide, Brisbane, Canberra, Melbourne, Sydney and Perth. Teams at MND clinics are knowledgeable about motor neurone disease and provide a coordinated, multidisciplinary approach to the care of people with the disease.

Many people live too far from an MND clinic to attend or may live outside the MND clinic's area. If this is your situation, your multidisciplinary care can be provided by a local primary health care team. A primary health care team includes local health and community care professionals who provide a range of expertise, advice and support. Your primary health care team can liaise with your neurologist and your MND clinic, if you are attending one.

Members of a multidisciplinary care team	
<b>Community/aged care worker and case manager</b>	<ul style="list-style-type: none"> <li>Community/aged care workers provide general household assistance, emotional support, care and companionship to people in their homes. Case managers assist people to access health and community services.</li> </ul>
<b>Dietitian</b>	<ul style="list-style-type: none"> <li>Dietitians provide dietary and nutritional advice.</li> </ul>
<b>General practitioner</b>	<ul style="list-style-type: none"> <li>The general practitioner (GP) is a doctor providing general medical care. GPs are usually your first point of medical contact. The GP liaises with the neurologist and other health and community care providers.</li> </ul>
<b>MND Association advisor</b>	<ul style="list-style-type: none"> <li>The MND Association advisor assists people with motor neurone disease connect to the services they need. MND Association advisors also offer ongoing information to families and service providers as questions arise or needs change.</li> </ul>
<b>Neurologist</b>	<ul style="list-style-type: none"> <li>The neurologist is a doctor who specialises in disorders of the nervous system. The neurologist coordinates the tests you need for diagnosis. The neurologist also monitors the progress of the disease and management of your symptoms.</li> </ul>
<b>Occupational therapist</b>	<ul style="list-style-type: none"> <li>An occupational therapist (OT) helps you maintain mobility, function and independence. OTs provide advice about home modification, different ways of performing tasks and on selecting, acquiring and adapting specialised equipment.</li> </ul>
<b>Palliative care team</b>	<ul style="list-style-type: none"> <li>The palliative care team specialises in interventions that can improve quality of life for people with life limiting conditions. Palliative care services provide emotional support for people living with MND and can assist you to plan your future care.</li> </ul>
<b>Physiotherapist</b>	<ul style="list-style-type: none"> <li>A physiotherapist helps you maintain physical activity and mobility. Physiotherapists can also show your family or carer how to safely help you move from one position to another, for example, moving from a chair to a bed.</li> </ul>
<b>Respiratory specialist</b>	<ul style="list-style-type: none"> <li>The respiratory specialist is a doctor who specialises in disorders of the lungs and breathing. The respiratory specialist provides information and advice about breathing and motor neurone disease and has expertise in non-invasive ventilation.</li> </ul>
<b>Registered nurse, MND nurse, clinical nurse consultant or clinical nurse specialist</b>	<ul style="list-style-type: none"> <li>The role of the nurse is varied and can include ongoing care and care coordination, often for people in their own homes. Specialised MND nurses usually work in MND clinics and have particular expertise in motor neurone disease symptom management.</li> </ul>
<b>Social worker, psychologist, or accredited counsellor</b>	<ul style="list-style-type: none"> <li>A social worker, psychologist or accredited counsellor provides counselling on the psychological and emotional aspects of living with motor neurone disease. In addition, a social worker can provide information on community services that may assist you with accommodation, legal, financial and other issues.</li> </ul>
<b>Speech pathologist/therapist</b>	<ul style="list-style-type: none"> <li>A speech pathologist helps in the management of communication and swallowing. They can advise about communication aids and devices and also about swallowing techniques and food consistency.</li> </ul>

### Multidisciplinary care team key worker

The multidisciplinary care team key worker is the person on the team who can advise you about regular review of symptoms and coordinates your care.

Who your key worker is depends on where you live, local health and community care service availability and the professional interests of individual health and community care professionals in your area.

Your key worker may be a:

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

#### Your key worker:

- Maintains regular contact with you
- Initiates effective and timely response when your needs change
- Liaises with other team members and services
- Organises regular case conferences and team meetings

## 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.

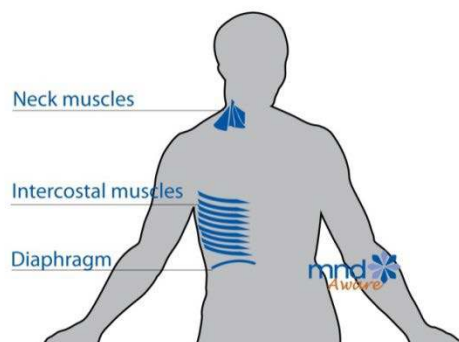


Figure 4 The respiratory muscles

### Signs and symptoms of respiratory muscle weakness

disturbed sleep	daytime sleepiness	increased fatigue	morning headaches	quieter voice
fewer words per breath	shallow, faster breathing	reduced movement of the rib cage	excessive use of the muscles in the upper chest and neck	weakened cough and sneeze
breathlessness (dyspnoea) even at rest	breathlessness lying flat (orthopnoea)	impaired concentration or confusion	irritability and anxiety	decreased appetite

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.

## Strategies for managing respiratory muscle weakness and sensations of breathlessness

### Avoiding people with coughs and colds

People with respiratory muscle weakness should avoid people with coughs and colds. Referral to a doctor or respiratory specialist for discussion about the influenza and pneumonia vaccines is often recommended. Let your family, friends and others visiting your home know they should stay away if they have a cough or cold.



Figure 5 Make sure people around you cover their mouths when coughing or sneezing

### Eating a 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 for you, see *Swallowing and nutrition*, page 20.

### Positioning, room airflow and temperature

The way a person's body is positioned while sitting or lying down can assist their breathing.



#### Forward lean standing

- Lean forward with forearms on a window ledge or other handy object, with shoulders and arms relaxed

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, aids and items of equipment.

#### Forward lean sitting

- Support the upper chest and chin or forehead on pillows. Remember to keep your back straight - extra pillows may be needed to help keep your back straight



#### Relaxed sitting

- Keep hands and shoulders relaxed and back straight

#### High side lying

- Make sure the pillows support your back well, keeping it straight, not slumped



Figure 6 Positioning to assist breathing

### 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 specialist can advise about the right breathing exercises for a person with MND.

### Managing fatigue

Tiredness can affect how you use your breathing muscles, see *Fatigue and insomnia*, page 33.

### Using 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 specialist can advise.

### Using 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.

NIV uses room air and is not oxygen therapy. To use NIV the person wears a mask connected to a small machine. This creates just the right pressure to keep the person's airways open so that room air can easily come in and out of their lungs when they breathe.

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

The NIV machine is usually used at night but, as the respiratory muscles continue to weaken, it is commonly used at times during the day as well. When a person starts using NIV they may need to try several different types of masks to find a mask that is comfortable, fits well and does not irritate the skin on the face and nose. Regular mouth and facial skin care is important when using NIV.

Over time, NIV will be less effective in controlling respiratory symptoms, because the disease continues to cause increasing muscle weakness. Usually, the doctor will discuss how NIV is withdrawn, before a person starts using NIV.

### Taking medication

There are a number of medications that help to 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 specialist can advise about medication.



Figure 7 NIV machine, hose and mask



## Swallowing and nutrition

Motor neurone disease may cause 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.

### 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 thick, ropery and tenacious, it is uncomfortable to swallow and makes it harder to clear the throat. This increases the risk of food and drink going down into the lungs.

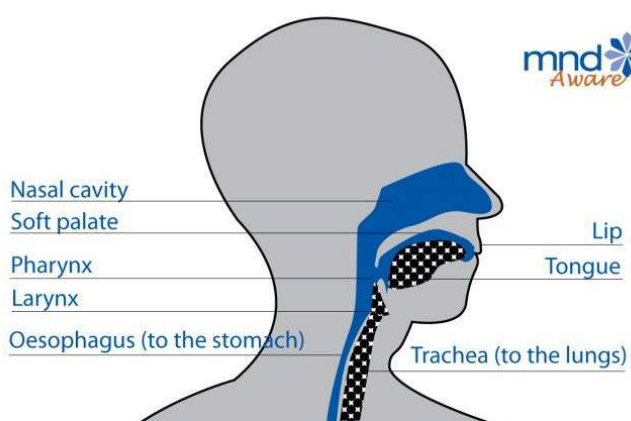


Figure 8 Normal swallowing

### Swallowing difficulties in MND

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

## Strategies for managing swallowing difficulties

### Positioning for eating and drinking

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 management

#### Minimise the effects of fatigue while eating

- 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 drinking or eating 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.

#### Food

Start with a balanced diet, including a variety of foods. Aim to eat the recommended daily amount of food from each food group. A multivitamin supplement may be beneficial if it is not possible to eat the recommended amounts. Discuss this with your doctor or dietitian.

People with weakened lip, mouth and throat muscles may also find it easier to swallow food of a particular consistency and your dietitian may suggest this for you. When weight loss is a problem, every mouthful needs to count.

Find out more about thickened fluids, food consistency, get sample meal plans and read about strategies for maintaining or gaining weight in the MND Association booklet *Living with motor neurone disease: day-to-day*.

### *Utensils and aids for eating and drinking*

Your occupational therapist and speech pathologist/therapist can advise about utensils and aids for eating and drinking that are suitable 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.

### **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.

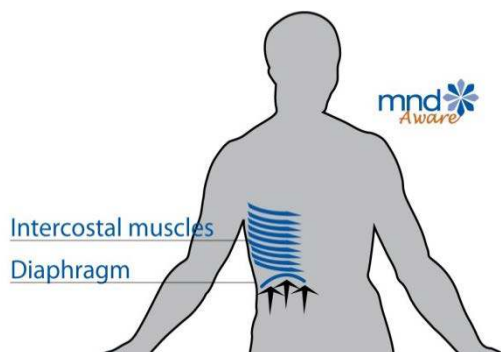


Figure 9 A cough is a very forceful breath out

### **Severe coughing, choking and laryngospasm**

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

Coughing is the body's way of moving food, drink or saliva away from the airway. It may 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. This can be caused by smoke, alcohol, spicy foods and gastric reflux. The attacks usually resolve spontaneously in less than a minute, but may create panic and anxiety. It can be helpful to reassure and to encourage the person to cough or laugh as this will help to open the vocal cords.

### *Managing coughing, sensations of choking and laryngospasm*

It is important for the person with MND and their carer to know that death caused by a choking attack is very rare in MND. If you have swallowing difficulties, your doctor, speech pathologist/therapist and physiotherapist usually develop a swallowing management plan for you. This can help reduce fears and anxiety.

#### Manage coughing, sensations of choking and laryngospasm

- Stay calm and wait for the attack to pass
- Use the assisted cough technique
- Use medications including morphine, amitriptyline, benzodiazepines and glycopyrrolate
- Know when and how to seek medical advice

## Gastrostomy (PEG or RIG)

If you have lost weight or have become dehydrated through inadequate nutrition, a gastrostomy may help you increase or maintain your weight, improve hydration and help to reduce tiredness and hunger.

Gastrostomy enables you to take fluids and liquid feeds, known as enteral feeds, through a permanent tube that passes directly into your stomach, bypassing your mouth and throat.

Two common methods are used to place the permanent feeding tube into the stomach through the abdominal wall. These are:

- percutaneous endoscopic gastrostomy, or PEG
- radiologically inserted gastrostomy, or RIG.

These medical procedures usually involve mild sedation for relaxation, an injection of a local anaesthetic and a small incision.

The permanent tube into the stomach 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. You can tuck this 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.

Often, people with motor neurone disease can continue to take some foods and fluids by mouth after a gastrostomy.

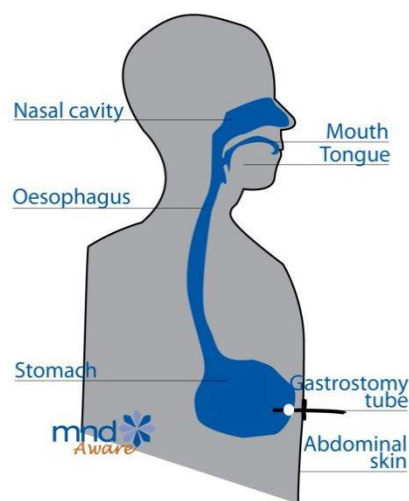


Figure 10 Gastrostomy (PEG or RIG)

### Getting advice about gastrostomy

- Your neurologist or medical specialist, and your general practitioner will be able to discuss the advantages and disadvantages of PEG for your individual circumstances
- Your respiratory physician should be involved to assess breathing function. It is safer to have the PEG done before significant breathing problems are evident
- Your speech pathologist/therapist will help determine the best time to have a PEG by assessing and reviewing your ability to swallow over time
- Your dietitian will assess if you have too much weight loss or poor intake of fluids. Dietitians can also advise about enteral feed products, suppliers, cost and subsidy programs

## 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 thick saliva or drooling 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

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

#### 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 (1tsp 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

### 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.

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

### Managing drooling saliva

- Get advice about head and neck support, and position your body so your 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

## 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 us 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

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

### Managing thick saliva

- Check that you are having enough fluids
- Drink soda water and 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
- Breathe in 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 (see page 23) to clear the thick 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

Your 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

We communicate with others many times a day by:

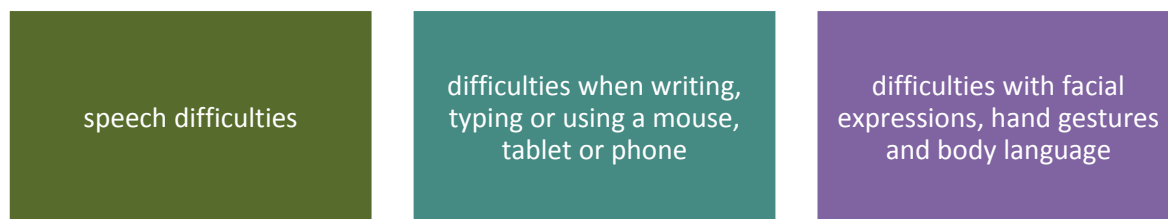
- talking in person or by phone
- using facial expressions and body language
- handwriting letters and notes
- sending text messages and emails
- using the internet
- using phone and computer applications such as Facebook, Twitter and Skype.

MND can cause muscles used for communication to weaken, although not all people with the disease will have these symptoms.

Being able to participate in conversations, whether in person, using handwriting, by phone or computer, or by speaking or typing, is important for well-being and quality of life.

Conversations provide us with the opportunity to be socially close to others. Conversations also enable us to share information and to communicate wants, needs and feelings. Being able to maintain communication with people important to you can provide you with invaluable support as you face the challenges of MND.

### Signs and symptoms of weakness in the muscles involved in communication



Speech difficulties in MND are caused by weakness and reduced coordination of the lips, facial muscles, tongue, larynx and pharynx. Vocal strengthening exercises to increase clarity of speech are not helpful for people with MND. 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. The person with MND may feel embarrassed and frustrated by their speech difficulties.

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, tablet or phone.



## Approach to management

A person with speech or other communication difficulty requires a coordinated, multidisciplinary approach and regular review. Communication needs often change over time.

It will be less stressful for you, your family and friends if you try out alternative ways of communication while you can use your usual means of communication such as speech and typing.

Assistive technology solutions can be very simple, and often just involve using everyday items, such as a pen, phone, tablet or computer, in a new or different way.

People often try several different strategies or systems before finding the solutions that are right for them. You may find a variety of tools or strategies useful depending on who you are communicating with and what you want to say.

There are health professionals who can assist you to decide on the best strategies and solutions for you.

The communication referral, assessment, trial and supply of equipment and installation/implementation process may take from three to nine months, and sometimes longer.

### Communication management plan

- Early assessment by a speech pathologist/therapist
- Advice from a speech pathologist/therapist about
  - communication aids
  - computer aids
  - environmental controls
- Advice from an occupational therapist about
  - seating and positioning
  - wrist supports
  - switches, pointers
  - mobile arm supports and tables
- Strategies for ongoing review

## Getting communication aids and devices

Your speech pathologist/therapist and occupational therapist can advise about communication aids and devices that may be available from a government subsidised equipment agency. Also, some Australian MND Associations have a range of communication devices available for people with MND following referral from a speech pathologist/therapist.

## 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. This can leave joints and ligaments 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 breathe, 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.

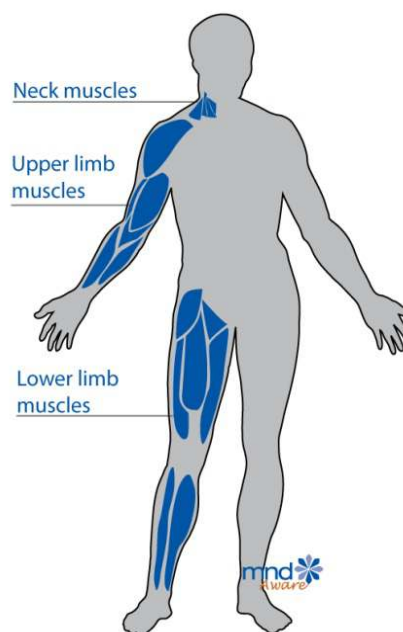


Figure 11 Neck, upper limb and lower limb muscles

## 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

### General strategies

- 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
- 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
  - 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
  - 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
  - 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
- Relieve swelling by elevation of the affected limbs, with support that does not put direct pressure on the swollen area
- 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
- Use aids and equipment for postural support, movement and activities of daily living
- Get regular personal assistance for activities of daily living, to change positions for comfort and to avoid pressure sores



Figure 12 A selection of collars



Figure 13 Pressure cushion



Figure 14 Transfer belt



Figure 15 Pivot hoist



Figure 16 Electric tilt in space wheelchair

### Particular strategies for neck muscle weakness

- A person with progressive neck muscle weakness usually starts using a foam collar for postural support
- As the weakness progresses a more structured collar or chin support is often prescribed by the physiotherapist
- Headrests and reclining chairs are also used to support the neck and head
- When transferring from one position to another, ensure that the neck and head are supported during the movement. For example, while transferring from a chair to a bed

### Particular strategies for upper limb muscle weakness

- The shoulder joint is a highly mobile but unstable ball and socket joint. As the shoulder muscles weaken, the shoulder is at risk of subluxation, a small dislocation. You may experience shoulder injury or strain if your arms are pulled by a person assisting you to reposition in a chair or bed, or assisting you to stand. The occupational therapist and physiotherapist can advise about aids and equipment for transfer and postural support. These often include the use of a transfer belt or hoist for movement
- Other equipment items often prescribed include reclining chairs and wheelchairs with headrests
- Slings and pillows to provide shoulder, arm and hand support may also be recommended
- Shoulder pain, frozen shoulder and rotator cuff injuries are not symptoms of MND. Shoulder pain may be the result of poor positioning. The physiotherapist and general practitioner can advise about treatments and positioning for shoulder pain and injury

### Particular strategies for lower limb muscle weakness

- The occupational therapist and physiotherapist can advise about aids and equipment for mobility, transfer and postural support
- These may include ankle and foot support and equipment items such as a walker, wheelchair, recliner chair, hoist, transfer belt, bed blocks, electric bed, toilet raiser, shower stool and shower chair
- Home modifications such as grab rails and ramps are also often considered

## Getting aids and equipment

Your speech pathologist/therapist, occupational therapist and physiotherapist can advise about aids and devices that may be available from a government subsidised equipment agency. Also, many Australian MND Associations have a range of aids and equipment available for people living with MND following referral from a health professional.

## Fatigue and insomnia

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

### Reasons for fatigue

People with MND get fatigued or tired easily. This is because:

- as the motor neurones in the body become less effective, they become unable to send commands from the brain to the muscle cells that they control. Fewer muscle cells get the commands. The muscle cells that do get the commands must then try to perform jobs usually done by the full number. The muscle cells that receive the commands are working at full capacity to perform the job or task.
- if MND is affecting the breathing muscles, the person draws less air into their lungs. When the person's activity level increases, it becomes more difficult for the lungs to supply enough oxygen to the body. This causes general fatigue.
- difficulties eating may reduce nutritional intake, affecting energy levels
- fatigue is a common adverse effect from the medication riluzole, which is often taken by people with MND.

### Strategies for managing fatigue

#### Getting advice about fatigue

- Health professionals, such as the respiratory specialist, nurse, physiotherapist, occupational therapist and rehabilitation specialist can advise on energy conservation techniques, breathing and MND
- An occupational therapist can advise about aids and equipment that can conserve energy. There is no advantage in 'pushing yourself'. Rather, the key is to save your energy for what you really want to do
- A dietitian or nutritionist can advise about the right balance of foods to maintain energy levels
- The general practitioner, neurologist or palliative care physician can advise about adverse effects of riluzole

#### General tips for fatigue

- Eat enough to maintain your energy levels
- Plan activities in advance
- Rest between activities, for example, after dressing in the morning have a rest before doing the next activity
- Keep more intensive tasks for times of greatest energy, for example, after a rest or sleep
- Use aids or equipment for activities that are particularly tiring, for example, use a walking frame or wheelchair just when going out
- Carry out tasks or activities that are the most important to you and leave other tasks for others to complete
- Get domestic assistance for cleaning and food preparation, and personal care assistance
- Get advice about fatigue from the multidisciplinary care team

## Reasons for insomnia

Insomnia, or sleeping difficulty, is common in the general population as well as in people with MND. Specific causes of insomnia in people living with MND include:

- ineffective shallow breathing (nocturnal hypoventilation) causing frequent waking because of weakened respiratory muscles
- discomfort because of immobility
- pain due to stiffness of joints or muscles
- medication
- drooling saliva or dry mouth
- worry or anxiety.

## Strategies for managing insomnia

### General tips for insomnia

- Go to bed at the same time each night
- Make sure the bedroom is quiet, cool, dark and comfortable
- Avoid over-exertion before going to bed
- Avoid caffeinated drinks and heavy meals

## Breathing and insomnia

Sleep apnoea is the temporary cessation of breathing while sleeping. This disruption to the breathing pattern wakes the person so that they resume breathing, but disrupts sleep.

Some people with MND experience morning fatigue. They may wake up feeling tired, sometimes with a headache and shortness of breath. This may be caused by a weakness in the diaphragm.

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. A physiotherapist or occupational therapist can advise on positional strategies for managing insomnia.

A person who wakes up regularly during the night and doesn't feel refreshed by sleep is usually referred to a respiratory specialist for a sleep study. They may be prescribed non-invasive ventilation. You can find out more about respiratory muscle weakness, see *Breathing*, page 17.

## Positioning, muscle tightness, joint stiffness and insomnia

While sleeping, we often change position. Light bedclothes and satin or silk sheets can allow for easier movement in bed.

However, because MND affects the muscles used for movement, a person with MND may need frequent assistance during the night to change their sleeping position. This can be as frequent as hourly repositioning. Very specific positioning of arms, legs and head may be needed to maintain comfort.

Inability to move and weight-loss can cause severe discomfort and skin breakdown. An occupational therapist can advise about how side rails, extra pillows, a pressure relief mattress overlay and specialised pillows can be used to increase comfort in bed and distribute body pressure more evenly. An electric bed can be adjusted to a variety of positions.

### **Saliva, dry mouth and insomnia**

Droling 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, see *Saliva and mouth care*, page 25.

### **Worry, anxiety and insomnia**

The person with MND may be experiencing difficulty sleeping because of worry or anxiety following the diagnosis of MND. Relaxation techniques, soft music, reassuring conversation and some medications can promote sleep. You may find it helpful to speak with your general practitioner, neurologist, palliative care team or the MND Association about ongoing support, through counselling and/or attendance at an MND support group.



## 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 person with bladder or bowel difficulties needs to have a management plan that includes ongoing review. 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.



Figure 17 Toilet raiser



## 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.

## 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.

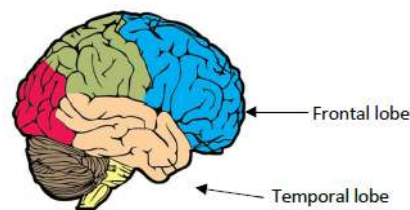


Figure 18 The frontal and temporal lobes of the brain

Many people with MND do not experience cognitive and behaviour change

Symptoms might be caused by factors other than changes in the brain (for example, depression or medication effects) so these need investigating

Clinical neuropsychologists and neurologists can assess changes in cognition and behaviour with a view to helping you to develop practical strategies

If you or others notice changes, supports are available to assist with management of these symptoms

### 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 and their family.

While the nature of changes in cognition and behaviour will vary from person to person, some of the most common symptoms are listed below.

Changes in cognition can include difficulties with

- Concentration
- Thinking quickly
- Learning new things
- Recalling information from memory
- Reasoning and problem solving
- Planning and organising
- Awareness and insight into MND symptoms

#### Changes in behaviour or personality can include

- Rigidity – resistance to changing routine or attempting new activities
- Impulsivity – doing or saying things without considering consequences
- Irritability – experiencing a ‘shorter fuse’ or reacting with disproportionate anger to events
- Disinhibition – difficulties in controlling behaviour or making social judgements, such as saying inappropriate things during social events
- Apathy – reduced motivation and less interest in previously enjoyed activities
- Difficulty understanding and expressing emotion. For example, difficulty in picking up on emotional cues and responding appropriately

#### Changes in language can include

- Unusual speech patterns, writing or spelling
- Difficulty finding the right word in conversation
- Taking longer to respond in conversations
- Problems in understanding the meaning of words
- Using odd words to get a message across
- Repeating particular words or phrases over and over

### 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 and their family 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
- (If significant cognitive involvement is suspected) getting a neuropsychologist cognitive assessment and advice

## 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 going out or being in other social situations. The person may also 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 has MND

### Strategies for emotional lability

Your general practitioner or neurologist can advise you about emotional lability.

#### Strategies often suggested for emotional lability

- Letting others know that the laughter or tears may not reflect what you are feeling
- Using distraction to think of something else
- Taking slow breaths in and out
- Focusing on something unrelated
- Taking medication as advised by the general practitioner or neurologist
- Getting reassurance that others understand

## Care in the later stages of MND

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 the disease and their family become more complex, it is not uncommon for the local palliative care team to become involved in care at home. Together with the multidisciplinary team, the palliative care team can provide the person with MND, their carer and the family with practical and emotional support.

### 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 community palliative care team can provide advice and support to both the person with MND and their family.

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 with palliative care at an early stage of the disease provides the opportunity for relationships to be formed before the very final stages of the disease. 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 your doctor, the local hospital, palliative care facility or community health centre.

### 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. There may also be questions about if or when to go to hospital, palliative care or when to remain at home.

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 muscle weakness.

### 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.

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.

Speaking with your family, general practitioner, neurologist or palliative care physician about plans for future care and preferences can assist with relieving anxiety. It may also be helpful to talk with a psychologist, social worker, counsellor or other member of the care team.

## The end phase

The end 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.

The general practitioner and the palliative care team can advise on comfort, medications and symptom control. Particularly, they can talk with the person with MND and their family about the signs of impending death, what to expect, how any pain might be managed, who to contact for immediate advice, how to involve others, where to get additional comfort items and managing the moments following death.

You may be advised to complete an adult palliative care plan. This plan details individually tailored treatment specified for you by your GP, including the administration of medications and other actions to relieve and manage symptoms in the home.

You can find out more about care in the later stages of MND in the booklet *Living with motor neurone disease: day to day*.

## Publications in the Living with Motor Neurone Disease series

### Living with motor neurone disease: aspects of care

- provides an overview of MND and strategies for symptom management

### Living with motor neurone disease: day-to-day

- provides strategies for living at home with MND, more in depth information and day-to-day tips

### Living with motor neurone disease: services and resources

- provides contact details for organisations that provide services and information that may be of interest to people living with MND

### Living with motor neurone disease: for carers

- for family and friends caring for a person with MND - provides information about looking after yourself

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