

4: What to expect as the disease progresses

This section explores the progression of MND, end of life symptoms, and the options available for the introduction and withdrawal of treatment.

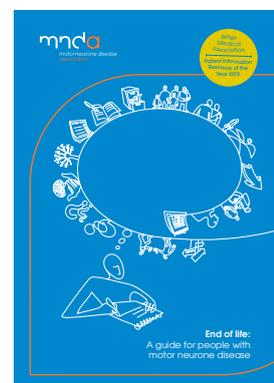
The following information is an extracted section from our full guide *End of life: A guide for people with motor neurone disease*.

All of the extracted sections, and the full guide, can be found online at: www.mndassociation.org/eolguide

The full guide can be ordered in hardcopy from our helpline, MND Connect:

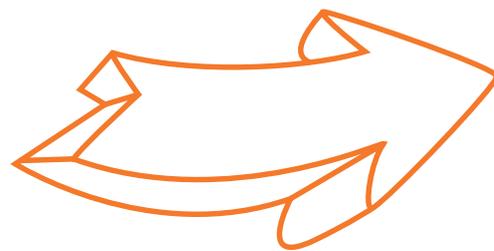
Telephone: **0808 802 6262**

Email: mndconnect@mndassociation.org



End of life:
A guide for people with
motor neurone disease

4: What to expect as the disease progresses



This section explores the progression of MND, end of life symptoms, and the options available for the introduction and withdrawal of treatment.

Please be aware that the following pages contain sensitive information. Read this information when you feel ready to do so. You can always return to this section later.

“ I think it’s important for me to understand, broadly speaking, what will happen and what the main decisions, options and changes will be.”

MND affects your motor neurones, which carry messages from your brain to your muscles to tell them what to do. Muscles that no longer receive these messages stop working. How you walk, talk, grip, eat, drink, swallow, breathe and think can all be affected.

Any disability that you experience is likely to get worse over time.

Although MND is incurable and cannot be reversed, there is much that can be done to manage symptoms and make your life as comfortable as possible.

See *Further information at the end of Part 2: What is likely to happen? for other publications on general symptoms and facts about the disease.*

How will the disease progress?

Everyone experiences MND in a different way. The range of symptoms and speed at which they progress varies from person to person, but can be rapid.

You may have serious problems at the start of your illness or these could take time to develop.

The way symptoms start with MND is unpredictable and it is unlikely that you will have the same symptoms in the same order as someone else with the disease.

Early management of symptoms (as soon as you notice any changes), will help you to cope with the impact and maintain the best possible quality of life.

“ I would like information on how the disease progresses... and what the future loss of bodily use is likely to be. Or shall I say, ‘what the future may hold for me!’”

The symptoms described in this section can happen throughout the course of the disease, not just at end of life. However, we have focused on the main concerns expected in the later stages of MND:

- mobility and movement
- speech and communication
- breathing
- swallowing
- changes to thinking and reasoning (known as cognitive change)

This information will help you understand what may happen and what can be done to reduce discomfort as the disease progresses. However, we recommend discussing your individual circumstances with specialist palliative care professionals for guidance.

“ I was told in full what to expect from life with MND. This has helped me to decide how my life is lived and has included end of life decisions.”

Mobility and movement

How will this affect me at end of life?

Some people remain mobile during the course of the disease (particularly if the disease starts in the face, throat or tongue). However, you may find your movement is severely affected in the later stages.

If you have increasing muscle weakness, it is likely to affect the use of your arms, hands and your ability to grip, as well as your mobility. For example, you may find it increasingly difficult to feed yourself, perform daily routines or transfer your body weight, such as from bed to chair.

“ I can see that I’m changing. I can’t hold things with my right hand... I can’t do all the things I want to do.”

What can be done to help?

If your mobility and movement are affected, you will need help with your care. As your needs increase, it may not be possible for your main carer to manage this alone. In the later stages, you are likely to need someone with you at all times.

Your care may be complex depending on the medical help needed.

You may feel great frustration as your independence reduces. Some people tell us that this can feel humiliating and can impact on confidence and self-esteem. If this affects you emotionally, ask your GP and health and social care team for advice. Again, planning ahead can help you manage some of the difficulties and feel more prepared.

“ I have found it enormously beneficial for me to consider the losses before they arrive.”

You may need to adapt your approach to daily activities. Accepting that you need equipment and assistive aids may feel difficult at first. However, this can help you maintain independence and your interests for as long as possible.

A physiotherapist or occupational therapist can assess your physical needs and advise on equipment and aids. Mobility aids such as wheelchairs, head supports, arm supports and hoists can make a big difference to your independence and what you can do. You may also wish to consider a riser-recliner chair and a powered bed for ease of positioning when sitting or sleeping.

Try to consider your future needs when looking at equipment, particularly when adapting your home. For example, a stairlift may not be the best choice if you are likely to need a wheelchair, as this would require two wheelchairs, one for each floor. You will also need to transfer between the wheelchair and stairlift, which may become difficult for you and your carer in the later stages.

A through-floor lift or a downstairs conversion may give easier access and there may be local authority or charitable funding to assist. Ask your occupational therapist for advice.

See also Section 15: Help from the MND Association for details about how we can support you with equipment and funding.

What should I consider when planning ahead?

Ask your health and social care team for advice about mobility and movement.

This may include guidance on:

- assessment of your needs, and those of your carer, by adult social care services, to help arrange appropriate care support at home and look at your future needs (your income and savings will be assessed to see if you need to make a contribution to any services you receive)
- assessment by a physiotherapist or an occupational therapist for access to aids to help you with mobility and movement
- assessment by an occupational therapist for an appropriate wheelchair
- home adaptations, which need planning early, as these can take a long time (particularly if you seek funding through a Disabled Facilities Grant)
- respite care arrangements to give your carer rest or time away from home
- residential or nursing home care if your needs are particularly complex or your main carer is unable to provide increasing levels of support at home.

See Section 6: *What care is available? for types of care and who can support.*

Speech and communication

How will this affect me at end of life?

Where speech is affected, this is likely to get worse over time. This may be due to breathing difficulties, which can reduce the volume of your speech. Your face, throat and tongue muscles may also weaken, which can change how you speak. If you find it more difficult to gesture or use facial expression, this will also make communication more challenging.

What can be done to help?

A notepad and pen can help you to communicate at first, but writing can become difficult if your grip is affected. Speech and communication aids can assist, from simple printed word or image cards, through to computer technology (which can be adapted for use with limited movement). A speech and language therapist with knowledge of MND should assess your needs, as not all items suit everyone. Some are costly and you may find that inexpensive and simple solutions can provide effective help.

What should I consider when planning ahead?

You may already use a computer and special applications to help you communicate or use the internet. Adapted controls can help you continue using this technology as physical disability increases. However, be prepared that in the later stages:

- you may find them more difficult to use
- some equipment may not be as easy to use in different locations, such as in hospital if you need treatment
- there is a possibility that these could fail to work, leaving you without a means to communicate.

If you already use computer technology, try to practise with basic aids, such as an alphabet board, as a back-up method. This way, you have an additional, simple way to communicate wherever you may be. Ask your speech and language therapist for guidance.

We recommend discussing your wishes about end of life with your main carer, family and health and social care team if you begin to experience any problems with speech or communication. As this gets worse, conversations are likely to become more difficult, particularly if some of the health and social care professionals you meet have little or no experience of MND.

You may wish to put advance care plans and advance decisions in place before your speech and communication get worse.

See Section 9: Advance care planning and advance decisions.

If you already have severe speech and communication problems, specialist palliative care professionals or other members of your health and social care team can help you with planning. Your main carer can also help ensure your wishes are understood.

Breathing

How will this affect me at end of life?

The muscles you use to breathe tend to weaken as the disease progresses. Symptoms and signs may include:

- disturbed or non-refreshing sleep
- shallow breathing, even when at rest
- difficulty breathing when lying flat
- repeated chest infections
- a weak cough or sniff
- morning headaches
- daytime sleepiness and feeling very tired
- confusion, poor concentration or memory problems
- poor speech volume
- excessive use of muscles in the upper chest and neck.

What can be done to help?

Ask your respiratory specialists for advice on breathing therapies and techniques, positioning to ease breathing, how to save energy and relax, and the types of mechanical breathing support available.

You may also be prescribed medication if suitable. Symptoms such as breathlessness can feel worse if anxiety increases and medication can help ease these feelings.

Pressure on your breathing may be relieved by using a riser-recliner chair, wheelchair, powered bed or other assistive equipment.

You can also try increasing the number of pillows you normally use, so that you don't lie flat at night.

If breathing problems increase, you may need to consider the following:

MND Just in Case Kit: a box you can keep at home, to contain medication for use in emergency situations. Having a kit nearby can be reassuring should you become breathless or have bouts of coughing or choking. We supply the box free of charge to your GP, at their request. Your GP then prescribes suitable medicine for you to keep in the box. This ensures medication is on hand, for a doctor or district nurse, if they are called out of hours.

It also provides guidance and (if your GP feels it is appropriate), medication for your carer to give to you while waiting for professional medical help to arrive.

See Section 15: Help from the MND Association for details of how professionals can contact our helpline MND Connect for assistance.

Mechanical breathing support: is where a machine known as a ventilator supports your breathing. There are two types of assisted ventilation, as follows. With either, you are likely to become reliant on this support, as your breathing will continue to weaken.

- Non-invasive ventilation (NIV), where a portable machine provides extra air flow through a mask over your nose, or over your nose and mouth.
- Tracheostomy ventilation (also known as trache ventilation or invasive ventilation), where a machine supports your breathing through a tube inserted into the windpipe through the neck.

Ventilation is not suitable for everyone, but may help relieve respiratory distress, improve sleep and reduce fatigue.

Risk with use of oxygen: in most cases, ventilation for MND uses normal air to help you breathe. Extra oxygen is not usually recommended with MND, as it can upset the balance in your body between oxygen and carbon dioxide. However, if your oxygen levels are low, it may sometimes be used with caution. Any decisions regarding oxygen for home use should be discussed with your respiratory team, as high levels of oxygen in your blood can be harmful.

See Further information at the end of Part 2: What is likely to happen? for details about our MND alert resources and our range of information sheets 8A – 8D on breathing and ventilation with MND.

Weak cough: if your cough weakens, support can be given. This might include ways to increase the amount of air you take in, with a technique called breath stacking and the use of lung volume recruitment bags. You may also be offered a machine to assist your cough and help you clear mucus. Your health and social care team can advise.

Flu vaccination: chest infections can make breathing difficulties worse and increase mucus. Try to avoid people with colds or flu and request a flu vaccination from your GP. It can't stop colds, but does reduce the risk of flu.

The flu vaccination is usually available from October and you need it every year. You'll be checked to see if it's right for you. You may feel slightly unwell after the vaccine, but it won't give you flu. Ask your GP if your family can also have the vaccine to help protect you.

What should I consider when planning ahead?

You will be assessed to see if mechanical breathing support is suitable for you. You may or may not find the mask comfortable, but it is your choice to use this type of support.

Breathing support may reduce some of the symptoms of MND, but will not stop muscle weakness in your body from getting worse. This is likely to affect your quality of life.

There may come a time when you feel that breathing support is no longer helping or it has become a burden. You can ask for it to be withdrawn, even if this may shorten your life. If you have become reliant on ventilation and you are approaching end of life, death is likely to happen in a relatively short time following withdrawal of this support. This can vary depending on how weak your breathing has become. Your respiratory consultant or specialist palliative care professionals can advise. Medication before and during withdrawal can help you feel calm and relieve distress.

See Further information at the end of Part 2, for details about our information sheets on breathing, ventilation support and withdrawal. See also Section 5: How will I die?

You can record which treatments to withdraw, and precise circumstances in which you want withdrawal to take place, using an Advance Decision to Refuse Treatment (ADRT). A valid and applicable ADRT is legally binding, but will only be actioned if you become unable to make decisions or communicate. Tell everyone involved in your care about the ADRT. If they are unaware, they will not be able to use it.

See Section 9: Advance care planning and advance decisions for more detail on why an ADRT might be needed and how to complete the form.

It is also helpful to draw up advance care plans to tell health and social care professionals about your general needs and preferences. This is not legally binding like an ADRT. Not all professionals have experience of MND, so it can help guide how you are cared for, particularly in emergencies where you may not be able to communicate.

“ He was taken into hospital with a chest infection but they didn't really know what to do with him because the specialist nurses and the consultant were unavailable.”

For example, tracheostomy ventilation is often used temporarily to help someone recover if resuscitated in an emergency. The emergency team may not realise how difficult it can be to stop using this type of ventilation if your breathing muscles have weakened with MND. You may then have to accept this type of ventilation as ongoing support, which can affect plans for your future care.

If tracheostomy ventilation is something you definitely do not want in any circumstances, you need to make this clear to all those involved in your care. An ADRT may help you to do this.

Our *MND Alert Card* can also support you in emergency situations and has been included in the End of Life guide folder. It can be kept in a wallet, purse or pocket. If you are admitted to hospital for any reason during the course of the disease, it alerts staff that you have MND and directs them to key contacts for the specialist help you may need. Our *MND Alert Wristband* can also help flag that you have MND and that you may be at risk with use of oxygen.

“ Another thing that is really comforting is having a card with contact details...They told me that should I need to go into hospital I should show the card. The people concerned will then contact my consultant and the MND co-ordinator.”

We also produce an *Understanding My Needs* leaflet which you can use to write down a summary of your needs for use in hospital or during respite care.

See Further information at the end of Part 2: *What is likely to happen?* for details about how to order additional publications, such as our *MND Alert Card* and the *Understanding My Needs* leaflet.

Swallowing

How will this affect me at end of life?

If you experience difficulty with swallowing, it may:

- affect how you eat and drink and you could lose weight
- make it more difficult to swallow your saliva (which can lead to drooling)
- affect how you take any medications you need (your GP can advise about different ways to receive these)
- increase the risk of food and drink entering the lungs (known as aspiration), which can lead to chest infections or aspiration pneumonia. Combined with a weakened cough, chest infections can be distressing and may happen repeatedly.

Swallowing difficulties can lead to mild choking, where the airway is partly blocked. Very rarely, severe choking might happen, where the airway is fully blocked.

What can be done to help?

You may wish to consider:

- using prescribed medication or over-the-counter remedies to relieve thin saliva, thick saliva or a dry mouth (your GP can advise on other ways to manage this)
- finding out how you and your carers can manage repeated coughing or choking, should this happen
- keeping a MND Just in Case Kit at home for emergency situations

See earlier heading, *Breathing for more detail on the MND Just in Case Kit.*

- asking your health and social care team about having a feeding tube inserted through the front of your abdomen directly into the stomach, to receive liquid food, medication and fluids (known as a gastrostomy).

See Further information at the end of Part 2: *What is likely to happen?* for a list of relevant publications on swallowing difficulties and alternative eating methods.

Tube feeding helps you to:

- receive the level of nutrients that you need
- reduce the risk of bits of food and drink entering your lungs, as you no longer have to eat and drink through your mouth (although it cannot prevent all risk of chest infection)
- receive medication in liquid form
- reduce the time needed to eat, which can become lengthy and tiring
- reduce stress for both you and your carer at mealtimes

“ Timing is crucial. My husband pushed for a feeding tube even before his health professionals thought it necessary...but he was right. A month later it would have been complicated, as his breathing was more affected.”

It is best to have a feeding tube fitted before you lose a significant amount of weight, but you do not have to use it immediately. You can eat and drink normally, for as long as you feel safe or comfortable. You can also taste small quantities of food and drink, while using the tube to receive the correct amounts.

At a later stage, you can use the tube for all of your food, fluids and liquid medicines.

If you live alone, it may be difficult to get the correct level of care required for a feeding tube.

What should I consider when planning ahead?

If you want to consider tube feeding, discuss the benefits and risks with your GP, dietitian and speech and language therapist in order to make the right choice for you.

See Further information at the end of Part 2 - What is likely to happen? for details about our resources on swallowing, tube feeding, and eating and drinking. Also, how to access the MyTube website for tube feeding videos.

Timing is important. Tube insertion requires a hospital procedure, which carries more risk if breathing problems get worse. There may come a point where it is no longer possible for a tube to be introduced. However, if you have one fitted and feel it is no longer helping, you can choose to stop receiving food by tube at any time, along with other decisions to refuse life-sustaining treatment.

When nearing the end of life, the body usually requires less food, whether taken orally or through tube feeding. This means that deliberately reducing or stopping the intake of food may not feel as uncomfortable as can be feared. Your family and carers may find it upsetting if they see you losing weight, but there is no need for them to encourage you to eat in the final stages. As you get closer to dying, you will be less likely to digest food properly and you will not feel the need to eat.

You can continue to receive water by tube to prevent dehydration if you wish, which may help you remain comfortable. If you have never had a tube fitted and can no longer swallow fluids, it can be helpful to keep the mouth and lips moist with a wet cloth or tiny amounts of water. Ask the professionals supporting you for advice.

If you are worried that you may not be able to express a choice for reducing or stopping intake of food in the future, you can record your wishes by using an Advance Decision to Refuse Treatment (ADRT), specifying the precise circumstances in which you would want withdrawal to take place.

See Section 9: Advance care planning and advance decisions.

Changes to thinking and behaviour

1 in 2 people with MND may have changes to thinking and behaviour (known as cognitive change). The number of people affected can increase in the later stages. Changes can be mild, but if more severe, making decisions may become more difficult.

The person affected may not be aware of changes, but family or a carer may notice different behaviour. However, changes may be due to other reasons, such as depression, poor sleep and emotional reactions to longterm illness.

Some people with MND also have unexpected emotional reactions, where they find themselves laughing or crying for no particular reason (known as emotional lability). This is more common if thinking and reasoning have been affected, but these emotional reactions can happen even when thinking processes are normal in MND.

See Section 2: Emotional and psychological support for more about emotional lability.

A small minority of people develop a type of dementia called frontotemporal dementia (FTD), which is likely to involve more severe changes to reasoning and behaviour. These changes can be very difficult for the main carer to manage. Additional care support is likely to be needed.

See Further information at the end of Part 2: What is likely to happen? for detailed information about changes to thinking and emotions.

What can be done to help?

Ask your health and social care professionals, in particular your neurologist, for advice and ways to manage any symptoms relating to changes in thinking, reasoning or behaviour. Depending on the circumstances, you may be referred to other specialists, such as a clinical psychologist, usually with a specialist knowledge of neurological conditions, or a clinical neuropsychologist. They may be able to suggest ways to manage the impact for you and your family.

Make sure you are given plenty of time to understand any issues and ask for the professional to write down any important points or record the conversation (most mobile phones enable recording or you could take along another device). This will help you to remember what has been said.

Some symptoms may have other causes, for example you may feel very tired if your breathing has been affected and find it difficult to concentrate or retain information.



“ I need someone to help me to decide about choices at the moment...I can't remember things.”

If your main carer is likely to need more support, ask your local authority or adult social care services for an assessment of your needs and those of your carer. This will help determine what type of support you may need for personal care, including respite care to give your main carer time to rest. They can tell you what services are available and provide guidance. If you qualify for arranged services, these will be means tested to determine what contribution you may need to make.

Changes to thinking and reasoning may also affect the way you behave or react to others. You may wish to explore ways to feel calm and relaxed, which can alleviate some of the distress that may be felt. Medication can help or you can use other ways to relieve stress. For example, some people with MND find complementary therapies helpful, and activities such as music therapy.

What should I consider when planning ahead?

If you feel that it may become difficult to express your needs, wishes and preferences, try to have these discussions as early as possible.

Writing down how you would like to be cared for in the future will help to ensure your wishes are respected. Health and social care professionals can help with this.

See Section 9: Advance care planning and advance decisions.

You may also wish to get your personal affairs in order, such as making a will.

See Section 8: Putting affairs in order.

Other concerns

You may be worried about the following:

Pain: although MND does not directly affect the sensory nerves, immobility and weakness can lead to severe discomfort. Medication and other measures, such as physiotherapy, can help to control any resulting pain. The use of pressure relieving mattresses and cushions can also help, as can positioning. If you experience pain, ask your health and social care team or specialist palliative care professionals for assistance.

Incontinence and constipation: problems with the bladder and bowel are not usually direct symptoms of MND, but you may not be able to get to the toilet easily if your mobility is severely affected. With a type of MND called primary lateral sclerosis (PLS) you may feel a sense of urgency. Ask your health and social care professionals to advise about continence aids, a catheter or urinary sheath. Try not to cut down on fluids to avoid the need to go to the toilet, as this can lead to dehydration and constipation. They can also advise on bowel problems and constipation if needed.

“ How, where and when to go to the toilet has become more and more of an obsession as I go through my illness. The more immobile I have become, the more this question traps me...it is also much more difficult to open my bowels if I lack sufficient breath.”

Problems with the bowel can sometimes be managed through changes to diet and ensuring you receive enough fluids. Medication or other interventions may be advised in more severe cases, under medical supervision.

Key points

- One of the ways you can maintain control over your care is to ensure your wishes and preferences are written down. Ask your health and social care team about advance planning and advance decisions.

See Section 9: Advance care planning and advance decisions.

- Keep a log of any questions to ask health and social care professionals. This may help if you feel tired or overwhelmed at appointments. Note down answers or record the conversation to refer back to later. Ask for help to do this if needed.
- Think about how you would like your symptoms to be managed before they progress, particularly if your breathing or swallowing are affected. Share your thoughts about this with family and friends, as well as the professionals involved in your care.
- Before making decisions about equipment or aids, have your needs assessed by the relevant professional, eg an occupational therapist or speech and language therapist. Equipment may not always be suitable for everyone and mistakes can be costly. You may also be able to obtain certain items free from the NHS or adult social care services.
- You can be referred to specialists by your GP or other members of your health and social care team.
- Accepting appropriate care, equipment and adaptations can help you remain as independent as possible, for as long as possible. This can help you to save energy for the activities that are most important to you and take away some of the pressure from your main carers.
- If you feel home adaptations may be needed, think about this as early as possible, as both funding and practical arrangements can take time.

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For references and acknowledgements please
refer to the full guide, *End of life: A guide for
people with motor neurone disease*.

The full guide can be ordered in hardcopy
from our helpline, MND Connect:

Telephone: **0808 802 6262**

Email: mndconnect@mndassociation.org



This resource has been evidenced,
user tested and reviewed by experts.