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Progressive muscular atrophy (PMA)

Information for people with or affected by progressive muscular atrophy


If you have been diagnosed with a type of MND called progressive muscular atrophy (PMA), you may have concerns about how to manage the condition.

This information sheet includes details about this rare form of motor neurone disease (MND), ways to manage symptoms and how to access further support.

The information is split into the following sections:

- 1: **What is PMA?**
- 2: **What are the symptoms?**
- 3: **What help can I get?**
- 4: **How do I find out more?**

This information sheet includes details about life expectancy, and you may prefer not to read any further until you are ready.

 This symbol is used to highlight **our other publications**. To find out how to access these, see *Further information* at the end of this sheet.

 This symbol is used to highlight **quotes** from other people with or affected by MND.

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

What do the words mean?

Atrophy:	Where muscles waste and reduce in mass.
Electromyogram (EMG):	A test using fine needles to measure the activity in the muscles to detect abnormalities or damage.
Fasciculation:	Muscle twitching which can sometimes be seen as a rippling effect under the skin.
Flail arm syndrome:	Where the arm muscles waste and become limp and cannot be moved voluntarily.
Magnetic resonance imaging (MRI):	A scan which involves lying inside a large tube-shaped scanner which produces detailed images of the inside of the body. It is used to rule out certain abnormalities of the brain and spine.
Nerve conduction test:	This test uses small patches placed on the skin. Small electrical impulses are used to check the speed of electrical conduction in nerves.
Palliative care:	Specialist care area focused on improving quality of life for people with life-shortening conditions. This can include symptom control and support for physical, psychological, social, spiritual, religious or other needs.

1: What is PMA?

PMA is a rare form of MND that affects the nerves, which run from the spinal cord and control your muscles, known as lower motor neurones. PMA results in muscles gradually losing their mass, known as atrophy or wasting. This causes the muscles to become weak, and a twitching sensation that ripples under the skin (known as fasciculation).

It usually starts in the arms or legs, and may only affect one part of the body for a number of years before spreading to other areas.

Like other forms of MND, it typically affects people aged over 50 years but younger people can also be diagnosed with PMA. It is a little more common in men.

What causes PMA?

Like other forms of MND the causes of PMA are unknown, and research is ongoing. As in MND as a whole, it is thought that a combination of environmental and genetic factors play a part, but it is not currently possible to give a clear answer about the precise triggers.

How is PMA diagnosed?

There is no single test to diagnose PMA or any other form of MND. Your neurologist will first rule out any other conditions that may be causing your symptoms, such as multiple sclerosis, spinal damage or tumours in the brain or spine.

It can take time to be sure that you have PMA, and most people will have had symptoms for a few years before a definite diagnosis can be made by a neurologist.

If your doctor thinks you may have a form of MND or another neurological condition, they would usually refer you to a neurologist. After discussing your symptoms and examining you, the neurologist may arrange a series of tests including:

- a full assessment of your symptoms and family medical history
- scans to check for any structural problems in the brain and spinal cord
- tests on your nerves and muscles
- blood tests to exclude other conditions
- testing the fluid from around your spine, known as lumbar puncture.

How will PMA affect my life expectancy?

PMA usually progresses more slowly than some forms of MND, and many people with this form of MND live for five years or more.

However, it is important to note that some people with a diagnosis of PMA have their diagnosis changed to amyotrophic lateral sclerosis (ALS) as their symptoms become clearer. ALS is the most common form of motor neurone disease which can progress more quickly, and has additional symptoms.

2: What are the symptoms?

The early symptoms of PMA are very similar to those of the other forms of MND, so it can be difficult to tell them apart at first.

Not everyone will experience all of the following symptoms, or in any particular order. You may have additional symptoms that are not listed here, as the disease affects everyone differently. You may have another unrelated problem which also requires medical attention, so ask your GP if you have any concerns.

Typical symptoms you may experience with PMA are:

- weakness and wasting of muscles in the legs, arms, hands and body
- flail arm syndrome (where the arm muscles waste and become limp and cannot be moved voluntarily)
- fatigue
- muscle cramps and pain
- muscles twitching and rippling beneath the skin (known as fasciculations)
- clumsiness
- breathing difficulties
- weight loss.

See section 3: *What help can I get?* for more information about managing these symptoms.

Can I still drive?

It is a legal requirement for you to inform the DVLA and your insurance company of your diagnosis of PMA, as it may affect your ability to drive. Seek advice from your GP, consultant or nurse.



For further information about driving and transport see Information sheets:

12A – *Driving*

12B – *Choosing the right vehicle*

12C – *Travel and transport*

3: What help can I get?

PMA affects everyone differently, but the impact of the disease can be frustrating and limit your independence. However, equipment and therapies can help you to stay independent for as long as possible and improve your quality of life.

Although with PMA you may have more time to plan for your future needs, it is worth arranging things as early as possible as equipment and waiting lists can cause lengthy delays.

Before making any decisions about equipment and aids, it is important to be assessed by an occupational therapist. Ask a member of your health and social care team for a referral if you do not already have one. Private purchases can be costly and may not suit your needs or your home.

An assessment with an occupational therapist also enables you to discuss ways of adapting your routines and methods to help maximise your independence. Over time you may develop your own solutions and techniques, which can be very effective, but check with your occupational therapist for advice on safety.



For more information about using equipment with MND, see:
Information sheet 11C – *Equipment and wheelchairs*

What can be done about muscle weakness?

Muscle weakness caused by MND cannot be reversed by exercise. Gentle regular exercise may assist with flexibility and range of movement of your joints, helping to maintain unaffected muscle function and support posture and balance. Gentle muscle stretching can also relieve cramps.

Ask your GP or health and social care team for a referral to a physiotherapist experienced in MND. They can recommend a suitable exercise programme to meet your needs. It is not advised to over-exercise with any form of MND, as this may lead to fatigue.



For information about physiotherapy with MND see:
Information sheet 6A – *Physiotherapy*

Using a range of aids and equipment can help you to manage daily living more independently. Ask your GP or a member of your health and social care team for a referral to an occupational therapist for an assessment.



For more information about using equipment with MND, see:
Information sheet 11C – *Equipment and wheelchairs*

What can be done about fatigue?

It might be helpful to take on the idea that you have a 'bank account' of energy to spend each day. If you overspend your energy one day, you may feel extremely tired the next.

Plan to do valued and essential tasks first. Delay or get help for other tasks as needed.
Try to:

- listen to your body and pace yourself
- prioritise tasks, manage your time and be flexible with routines
- use equipment to help you do things more easily
- talk to your physiotherapist, occupational therapist and dietitian about ways to manage your fatigue
- plan a rest day before and after a particularly busy time, such as a family event
- avoid excessive exercise routines.



For more detailed information, see:
Information sheet 11D – *Managing fatigue*

What can be done about muscle cramps?

Getting plenty of rest may help to ease painful cramps and help you recover better if you are injured after a trip or fall.

Talk to your health and social care team for advice. If painful cramps are persistent, your GP, consultant or specialist nurse may be able to offer medication for cramps and muscle spasms. A referral to a palliative care or pain clinic can be helpful.

Physiotherapy can also provide some relief, although not all physiotherapists will have worked with MND before. The specialist skills and experience of a neurological physiotherapist, either from the NHS or an independent practice, can provide a more in-depth approach to specific neuromuscular problems such as those seen in PMA/MND.



For more detail, see:
Information sheet 6A – *Physiotherapy*
Information sheet 6C – *Managing pain*

What can be done about twitching and rippling sensations?

Ask your GP, neurologist, MND team or specialist nurse, as medication may help to relieve this symptom.

What can be done about accidents and falls?

PMA affects muscle function in arms, legs or both. This can cause problems with dexterity and grip, and falls in the case of leg weakness. Daily routines may become increasingly difficult, which could lead to risk of injury. Referral to an occupational therapist is important, as you may be able to access advice and equipment to help you.

As PMA progresses, you may need to consider using walking aids or a wheelchair to get around. Your GP, MND team, physiotherapist or occupational therapist can refer you to an appropriate service for support and advice.



For more information about wheelchairs, see:
Information sheet 11C – *Equipment and wheelchairs*

What can be done about breathing difficulties?

PMA can weaken the muscles used in breathing. You may notice the following signs or symptoms:

- feeling breathless, even when resting or lying flat
- shallow or rapid breathing
- difficulty clearing mucus or phlegm due to a weak cough
- repeated or lengthy chest infections
- a weak sniff
- disturbed sleep or feeling tired when you wake up
- nightmares or hallucinations
- morning headaches.

If you notice any of these symptoms, contact your health and social care team for advice. This is a good time to ask for referral to your local respiratory team, so you can be assessed and discuss options for treatment and care.




For more information, see:
Information sheets 8A-8D – our information range about breathing


What can I do about weight loss?

As your muscles reduce in mass, you may lose weight. Your dietitian can advise you how to maintain your body weight and ensure you are getting all of the nutrition your body needs.

If you experience difficulties swallowing, ask your GP for a referral to a speech and language therapist and a dietitian who will be able to assess your ability to swallow safely and give you information about the types of diet, supplements or aids that might support you to maintain a healthy diet.

 For more information, see:
Information sheet 7A – *Swallowing difficulties*


Some people with MND will choose to have a feeding tube. This is a way of passing fluids, specially prepared liquid feed and medication straight into your stomach through a tube in your abdomen. Your dietitian and speech and language therapist can advise you about your options.

 For more information, see:
Information sheet 7B – *Tube feeding*

Will PMA affect the way I think?

Up to half of people MND experience changes in how they think, although this is less likely with PMA.

These cognitive changes affect people in different ways. For many people these changes are subtle and have little or no effect on daily life. For some however, these changes may be more apparent and intensive support may be needed to manage daily routines.

 For more information about changes to thinking and behaviour with MND, see:
Information sheet 9A – *Will the way I think be affected?*

If you, or those close to you, are concerned about changes to your thinking and behaviour, contact your health and social care team for advice. You may wish to share information sheet 9A with them, as they may not be familiar with the links between MND and changes to thinking.

How can I contact other people with PMA?

The PMA form of MND can be a longer term condition. You may find it beneficial to share experience and knowledge with others in a similar situation.

An email based support group has been developed for people living with PMA, and other slower progressing forms of MND.

If you would like to join the group, contact our helpline, MND Connect. See *Further information* in section 4: *How do I find out more?* for contact details.

4: How do I find out more?

Useful organisations

We do not necessarily endorse any of the following organisations, but have included them to help you begin your search for further information. The contact details are correct at the time of print, but may change between revisions. If you need help to find an organisation or have any questions, contact our MND Connect helpline (see *Further information* at the end of this sheet for details).

GOV.UK

Online government advice on a variety of financial benefit and welfare topics, including support for disabled people.

Email: email addresses are provided on the website, related to each enquiry
Website: **www.gov.uk**
www.nidirect.gov.uk (Northern Ireland)

Health in Wales

Information on NHS services in Wales, including a directory of the Welsh health boards.

Email: through the website contact page
Website: **www.wales.nhs.uk**

MND Scotland

MND Scotland provides support and information for people affected by MND in Scotland.

Address: 2nd Floor, City View, 6 Eagle Street, Glasgow G4 9XA
Telephone: 0141 332 3903
Email: info@mndscotland.org.uk
Website: **www.mndscotland.org.uk**

NHS Choices

The main online reference for the NHS.

Address: Customer Service, Richmond House, 79 Whitehall, London SW1A 2NL
Email: through the website contact page
Website: **www.nhs.uk**

NHS 111

The NHS online/telephone service in England for urgent, but not life-threatening medical help or advice. Available 24-hours a day, 365 days a year.

Telephone: 111

NHS Direct Wales

Health advice and information service for Wales.

Telephone: 0845 4647

Website: **www.nhsdirect.wales.nhs.uk**

NHS Northern Ireland

Information on NHS services in Northern Ireland. This is an online service only.

Email: through the website contact page

Website: **www.hscni.net**

NI Direct

Providing government information for Northern Ireland on a variety of financial benefit and welfare subjects, including health services and support for disability.

Email: through the website contact page

Website: **www.nidirect.gov.uk**

National Institute for Health and Care Excellence (NICE)

Provides national guidance and quality standards to improve health and social care, including a guideline on MND.

Address: 10 Spring Gardens, London SW1A 2BU

Telephone: 0845 003 7780

Email: nice@nice.org.uk

Website: **www.nice.org.uk/guidance/ng42**

References

References used to support this information are available on request from

Email: infofeedback@mndassociation.org

or write to:

Care Information feedback, MND Association, PO Box 246, Northampton NN1 2PR

Acknowledgements

We would like to thank the following for their kind assistance with the review of this content:

Jean Waters, a person living with MND

Tim Williams, Consultant Neurologist & Associate Clinical Lecturer in Neurology, Royal Victoria Infirmary

Gitana Zvikaite, MND Care Centre Coordinator, Barts and The National MND Care Centre

Further information

You may find these information sheets from the MND Association helpful:

3A – *MND care centres and networks*

6A – *Physiotherapy*

6B – *Complementary therapies*

7A – *Swallowing difficulties*

8A – *Support for breathing problems*

9A – *Will the way I think be affected?*

9C – *Managing emotions*

11C – *Equipment and wheelchairs*

11D – *Managing fatigue*

12A – *Driving*

12B – *Choosing the right vehicle*

We also provide the following guides:

Living with motor neurone disease – our main guide to help you manage the impact of the disease

Caring and MND: support for you – comprehensive information for unpaid or family carers, who support someone living with MND

Caring and MND: quick guide – the summary version of our information for carers

You can download most of our publications from our website at **www.mndassociation.org/publications** or order in print from the MND Connect helpline, who can provide further information and support.

MND Connect can also help locate external services and providers, and introduce you to our services as available, including your local branch, group, Association visitor or regional MND Association staff.



MND Connect

Telephone: 0808 8026262 (Free to all mobiles and landlines)

Email: mndconnect@mndassociation.org

MND Association, PO Box 246, Northampton NN1 2PR

MND Association website and online forum

Website: **www.mndassociation.org**

Online forum: **<http://forum.mndassociation.org>** or through the website

We welcome your views

Your feedback is really important to us, as it helps improve our information for the benefit of people living with MND and those who care for them.

If you would like to provide feedback on any of our information sheets, you can access an online form at: www.smartsurvey.co.uk/s/infosheets_1-25

You can request a paper version of the form or provide direct feedback by email: infofeedback@mndassociation.org

Or write to:

Information feedback, MND Association, PO Box 246 Northampton NN1 2PR



Last revised: 01/17
Next review: 01/20
Version: 4

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