



2C


Primary lateral sclerosis (PLS)

Information for people with or affected by primary lateral sclerosis

Primary lateral sclerosis (PLS) is a very rare form of motor neurone disease (MND) that progresses slowly. If you have been diagnosed with PLS, your needs may be different to those with other types of MND. This information sheet looks at ways to manage the symptoms and how to access further support.

The content is split into the following sections:

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

 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.



The MND Association has been certified as a producer of reliable health and social care information.

www.england.nhs.uk/tis

What do the words mean?

Bulbar:	used to describe swallowing or speech symptoms. The word refers to the bulb-shaped part of the brainstem that contains motor neurones needed for swallowing, speaking and chewing.
Dysarthria:	slurring of speech.
Dysphagia:	difficulties with swallowing. This may be liquids, solids or both.
Electromyogram (EMG):	a test that uses thin needles to measure the activity in the muscles to find abnormalities or damage.
Emotional lability:	also known as pseudo-bulbar affect. It causes laughing or crying, sometimes at inappropriate times. It can be difficult to stop and often does not reflect how you feel.
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 abnormalities in the brain and spine that might produce the same symptoms as PLS.
Nerve conduction test:	this test uses small patches placed on the skin to check the speed of electrical signals in the nerves.
Palliative care:	these specialists work to improve the quality of life for people with life-shortening conditions. This can include symptom control and support for any psychological, social, spiritual, religious or other needs.
Urinary urgency:	inability to wait to use the toilet when your bladder feels full.

1: What is PLS?

PLS is a rare form of MND where symptoms progress slowly over a number of years. It typically affects people aged over 50, but it can affect younger people too. As with all types of MND, there is no cure or specific treatment for PLS, but symptoms can be managed to improve your quality of life. With PLS, you are likely to remain independent longer than with other forms of MND.

PLS affects the nerve cells that send messages from the brain down the spinal cord and on to the muscles. This causes weakness, especially in the legs, and stiffness in the muscles and joints (known as spasticity). Some people develop weakness and stiffness in the muscles used for speech and swallowing too.

Not all symptoms necessarily happen to everyone and they do not usually develop at the same time.



For detailed information about MND, see:

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

Living with PLS can lead to many frustrations due to the ongoing impact of the disease on your communication, mobility and independence. Ensuring that your needs are well met can help to reduce this frustration. Equipment, advice and support are available to assist with many aspects of daily living. See section 3: *What help can I get?* for more information.

What causes PLS?

The causes of PLS are not known, and research is ongoing. 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. PLS is not thought to be an inherited condition.

How is PLS diagnosed?

Like all types of MND, there is no single test to diagnose PLS, but an experienced neurologist can identify the possibility based on symptoms and physical examination. They will first rule out other conditions that might cause similar symptoms, such as multiple sclerosis, or something causing damage to the brain or spinal cord. It can take time to be sure that you have PLS and most people will have had symptoms for 3 to 4 years before a definite diagnosis can be made by a neurologist.

If your GP thinks you may have a form of MND or another neurological condition, you will be referred to a neurologist who may arrange the following:

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

How will PLS affect my life expectancy?

PLS progresses only very slowly, and many people with the disease live for 10 to 20 years and occasionally longer. However, it is important to note that occasionally people thought to have PLS have their diagnosis changed to a different form of MND, such as amyotrophic lateral sclerosis (ALS), as the progression of their symptom become clearer. ALS is the most common form of MND, and progresses more quickly than PLS.

2: What are the symptoms?

The early symptoms of PLS are very similar to 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 can affect people differently. You may also have an unrelated problem, which requires medical attention, so ask your GP if you have any concerns.

Some of the symptoms you may experience with PLS are:


- problems with balance
- weakness of muscles, especially in the legs
- muscle spasms and cramps
- slurring of your speech
- swallowing difficulties, occasionally resulting in drooling
- emotional lability
- fatigue
- bladder urgency.

The symptoms may cause you to experience:

- a higher number of trips and falls
- pain in your neck, back, legs and other muscles
- difficulties managing daily tasks such as washing, dressing, cooking etc
- difficulties walking
- problems with eating and drinking.

Can I still drive?

It is a legal requirement for you to inform the DVLA and your insurance company if you have been diagnosed with PLS, as it may eventually affect your ability to drive. However, it may still be possible to drive, so seek advice from your GP, consultant, specialist nurse or MND care coordinator.

 For further information about driving and MND, see:
Information sheet 12A – *Driving*

3: What help can I get?


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

With PLS you have time to plan for your future needs, but it is worth planning ahead as waiting lists for equipment can cause lengthy delays. Before making any decisions about equipment and aids, get an independent assessment from an occupational therapist (OT). Private purchases can be costly and may not suit your needs or your home. An independent assessment also enables you to discuss ways of adapting your routines and methods to help you maximise your independence.


Over time you may develop your own solutions and techniques, which can be very effective, but check with your OT for advice on safety.

What can be done about stiff, tight muscles or 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. Your GP, consultant, specialist nurse or MND care coordinator may be able to offer medication to relax your muscles and relieve cramps and spasms. If your pain is persistent, you can be referred to a pain clinic or palliative care team. Physiotherapy can also provide some relief.

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

Some people find massage helpful to ease stiff or painful muscles. Get advice from your GP or consultant to ensure this is suitable for you. Massage should be provided by an experienced, registered therapist.

 For more information about massage and other complementary therapies, see:
6B – *Complementary therapies*

What can be done about muscle weakness?

Muscle weakness cannot be reversed by exercise. Gentle regular exercise may:

- assist with flexibility and range of movement in your joints
- help maintain unaffected muscles
- support posture and balance.

Ask your GP or health and social care team for a referral to a relevant 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:
6A – Physiotherapy

What can be done about my foot dragging?

If you experience 'foot drop' where your foot drags, ask your physiotherapist or OT for guidance. This may include a referral to an orthotics department who can provide you with appropriate splints and footwear to help support your foot and ankle.

As PLS progresses, you may need to use walking aids or a wheelchair to get around. Your GP, physiotherapist or OT can refer you to an appropriate service for support and advice.

What can be done about speech difficulties?


PLS may affect the muscles in your tongue, face and throat, making it difficult to speak. Your speech may slur and become unclear.

Ask your GP or MND clinic staff for a referral to a speech and language therapist for an assessment. They can advise you about techniques and equipment to help with your speech problems.

What can be done about swallowing difficulties?

With PLS, swallowing can become more difficult and you may experience occasional coughing. This can lead to a small increased risk of inhaling small pieces of food into your lungs, which can lead to chest infections. Swallowing difficulties can also lead to weight loss, dehydration and lack of energy.


Ask your GP or MND clinic staff for a referral to a speech and language therapist and a dietitian who will be able to assess your ability to swallow safely. They can provide information about diet, consistencies of food, drink and supplements to help you maintain a healthy diet.

 For more information about swallowing:
7A – Swallowing difficulties

What can be done about drooling?


If your swallowing is affected by PLS, saliva can gather in the mouth and throat and lead to drooling, which may be embarrassing. However, drooling is less common in people with PLS than other forms of MND.

Talk to your health and social care team as various options can be offered. This can include advice on medication to help dry up saliva and suction machines for clearing out the mouth.


 For more information about problems with saliva, see:
7A – Swallowing difficulties

Will I be affected by emotional lability with PLS?

Emotional lability (also known as pseudo-bulbar affect) is a symptom sometimes seen in MND, where people experience crying or laughing inappropriately. Although it is not experienced by everyone, this symptom is more common in people diagnosed with PLS. The crying or laughing can be difficult to stop and you may feel out of control. This can be distressing for you and those close to you, who may not understand.

 *“It is so difficult to explain that my crying does not mean I am depressed, in fact I more often cry about happy things”.*

In many cases the symptom eases over time. If the responses are persistent and causing distress, talk to your GP as medication is available that may help. If your GP is not familiar with emotional lability, try to talk to an appropriate member of your health and social care team with experience of neurological conditions.


 For more information about emotional lability, see:
9C – Managing emotions


Will PLS affect the way I think?

You are less likely to be affected by significant changes to thinking and behaviour with PLS than with other forms of MND. For most, the changes are subtle and have little or no effect on daily life or the ability to make decisions.

If the way you think and learn changes, you may find it becomes more difficult to:

- make and carry out plans
- think about what you need or want to do
- do activities in the right order
- do more than one thing at a time
- process information and solve problems
- consider options and consequences carefully when making decisions
- concentrate and take in new information
- recognise other people's feelings
- finish tasks.

 For more details about how MND can affect thinking and behaviour, see: 9A – *Will the way I think be affected?*

 For information about supporting someone affected by these changes, see: 9B – *How do I support someone if the way they think is affected?*

What can be done about fatigue?

It might be helpful to take on board the idea that you have a 'bank account' of energy to spend each day. If you overspend on energy one day, you may feel extremely tired the next. Plan to do valued and essential tasks first. Delay or get assistance 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 OT 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.

What can be done about bladder urgency?

Some people with PLS experience a strong, sudden urge to urinate (urinary urgency) caused by muscles in the bladder tightening. Medication may help with this symptom, so ask your GP or neurologist for advice. They can also rule out other causes such as bladder infections.

What can be done about falls?

PLS often affects balance early on and, with additional weakness of the leg muscles, you may be more prone to falls. This can make day-to-day tasks difficult, and you could injure yourself.

Ask your GP to refer you to an OT who may be able to provide you with equipment and advice to reduce the risk of falls. Your physiotherapist can also advise you and your carer on how to manage falls if they happen.

As PLS progresses, you may need to use walking aids or a wheelchair to get around. Your GP, physiotherapist or OT can refer you to an appropriate service for support and advice.

How can I contact other people with PLS?

The MND Association has developed an email based support group for people living with PLS, and another slow form of MND known as PMA. As PLS is generally a longer term condition, you may find it beneficial to share experience and knowledge with others in a similar situation.

If you would like to take part in the email based support 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, 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 welfare topics, including support for people with disabilities.

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. This is an online information service only (see also NHS Direct Wales).

Email: through the website contact page

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

MND Scotland

MND Scotland provides care, information and research funding for people affected by motor neurone disease 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, including information on continuing healthcare.

Address: Customer Service, Richmond House, 79 Whitehall, London SW1A 2NL

Telephone: 0207 210 4850

Email: through the website contact page

Website: **www.nhs.uk**

NHS 111

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

Telephone: 111 (England)

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 welfare subjects, including health services and support for disability.

Email: through the website contact page

Website: www.nidirect.gov.uk

References

References used to support this document are available on request from:

Email: infofeedback@mndassociation.org

Or write to:

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

Acknowledgements

Many thanks to:

Helen Madden, MND care coordinator, Bristol MND care centre

Tracy Thomas, MND Care Network Co-ordinator, South West Peninsula Network

Professor Martin Turner, Consultant Neurologist, Oxford University Nuffield Department of Clinical Neurosciences

Debra Warren, MND Care Centre Co-ordinator, Nottingham Care Centre

Professor Tim Williams, Consultant Neurologist, Royal Victoria Infirmary

Professor Adrian Wills, Consultant Neurologist, Nottingham University Hospitals NHS Trust

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

9B – How do I support someone if they way they think has been affected?

9C – *Managing emotions*
11D – *Managing fatigue*
12A – *Driving*

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 care development adviser.



MND Connect

Telephone: 0808 802 6262

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.surveymonkey.com/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

Created: 10/15

Next review: 10/18

Version: 4

Registered Charity No. 294354

Company Limited by Guarantee No. 2007023

© MND Association 2018