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:

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?

Bulbar:	Used to describe symptoms relationg to swallowing and speech. 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. When muscles start to lose their peripheral nerve supply, this can be detected by abnormal electrical activity.
Emotional lability:	Also known as pseudo-bulbar affect. This refers to laughing or crying in situations that feel inappropriate to you. 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 that produces detailed images of the inside of the body. It is used to rule out anything abnormal 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 peripheral 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.
Peripheral nerves:	Nerves in your body that exist outside of the brain and spinal cord. With MND, the focus is on the motor nerves that control movement.
Urinary urgency:	Inability to wait to use the toilet (urinate) when your bladder feels full.

1: What is PLS?

PLS is a neurological disease. This very rare form of MND has symptoms that progress slowly over many years. It represents 3 in 100 cases of MND.

The symptoms of PLS typically start when people are aged over 40, but younger adults can be affected.

"Like myself at age 38, many people in my PLS community were diagnosed in their late 30s."

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 more independent than with other forms of MND. You may still require some care support depending on the way your symptoms progress. Each individual with PLS may have different needs.

"Including more than 13 years of volunteering service, I'm still relatively able with PLS. I can manage for several nights at home by myself."

PLS usually begins in the legs, but some people develop problems with the muscles used for speech and swallowing. Over time, it can also affect the arms and hands.

Not all symptoms always happen to everyone with PLS, and they do not usually develop at the same time.

For detailed information about MND, see our main guide: *Living with motor neurone disease* – to help manage possible symptoms with all forms of MND.

Living with PLS can be challenging, due to the ongoing impact on your communication, mobility and independence. Ensuring that your needs are well met can help to reduce 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?

It is not known what causes PLS, and research is ongoing. It is thought that a combination of environmental and genetic factors play a part.

Environmental factors are things that we experience in our lives, that may have an effect. These could be different for each person and in combination. This makes it very difficult to know what could have triggered the disease.

Genetic factors are held within our own cells, which use genes to carry information. This information controls how our bodies grow and then develop over time. Genes may have some impact with PLS, but this form of MND 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, rather than another form of MND or a different condition. You may have had symptoms for up to four years before a definite diagnosis can be made by a neurologist.

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

- 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 (cerebrospinal fluid or CSF), known as a lumbar puncture.

How will PLS affect my life expectancy?

PLS is disabling, but the slow progress of symptoms means that it is not necessarily life-shortening. Most people live a typical lifespan following diagnosis, but this depends on the speed at which symptoms get worse and any other medical conditions.

Symptoms and speed of progress become clearer over time. This means a PLS diagnosis is occasionally reviewed and changed to amyotrophic lateral sclerosis (ALS). ALS is the most common form of MND. It progresses more quickly than PLS and is usually life-shortening.

2: What are the symptoms?

PLS usually begins in the lower limbs. For example, you might experience stiffness in the way you walk. If you run, your movement may not feel as fluid as before.

Other symptoms may impact speech, which may feel as though it takes more effort and sound slurred

> *"At first, most people in my PLS group noticed it in their legs, due to problems with walking and balance. However, over time most of us had problems with arms and hands to varying degrees."*

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 needs medical attention, so ask your GP if you have any concerns.

Some of the symptoms you may experience with PLS are:

- problems with balance
- stiffness and weakness of muscles, especially in the legs
- muscle spasms and cramps
- being easily startled (known as hyperekplexia)
- slurring of your speech with less frequent swallowing (that may cause drooling)
- slurring of your speech
- emotional lability
- bladder urgency
- fatigue.

The symptoms may cause you to experience:

- a higher number of trips and falls
- difficulties walking
- pain in your neck, back, legs and other muscles
- problems with speaking clearly
- emotional responses, such as laughter or crying that feel out of place
- difficulties with daily tasks, such as washing, dressing and cooking.

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 co-ordinator.

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

3: What help can I get?

Although PLS affects everyone differently, its impact can be frustrating. Equipment and therapies can help you stay independent for as long as possible and improve quality of life.

Ask your GP for a referral to an occupational therapist (OT). They can assess your needs, advise on what equipment may be helpful, and help you continue with daily routines independently for as long as possible.

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 buying any equipment and aids, get an independent assessment of your needs from an OT. Private purchases can be costly and may not suit your needs or your home. An 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.

For more information on what equipment or aids may be helpful, see:
Information sheet 11C – *Equipment and wheelchairs*

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 co-ordinator may be able to offer medication to relax your muscles and relieve cramps and spasms. If your pain is persistent, you can ask to for a referral to a pain clinic or palliative care team. Physiotherapy can also provide some relief.

For more detail, see:
Information sheet 3D – *Hospice and palliative care*
Information sheet 6A – *Physiotherapy*
Information sheet 6C – *Managing pain*

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:
Information sheet 6B – *Complementary therapies*

What can be done about muscle weakness?

Muscles already weakened by PLS cannot recover by exercise, but gentle regular exercise can:

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

"I've found, as have many others, that regular physio is of great benefit with PLS. After doing no exercise for 20 years, I gradually improved my distance and speed through treadmill sessions."

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 and help you avoid unnecessary fatigue.

For information about physiotherapy with MND see:
Information sheet 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.

"I noticed difficulties with speech a year from diagnosis. Within five years, only my family could interpret my speech. I still try to speak short sentences with my wife and she can sometimes figure out what I'm saying if it's in context."

Ask your GP or wider health and social care team for a referral to a speech and language therapist (SLT) for an assessment. They can advise you about techniques and equipment to help with speech problems.

For more information on speech difficulties with MND, see:
Information sheet 7C – *Speech and communication support*
Information sheet 7D – *Voice banking*

What can be done about swallowing difficulties?

With PLS, swallowing can become more difficult. You may experience occasional coughing or even a choking sensation, but this usually passes without the need for support. However, this can lead to a slightly increased risk of inhaling small pieces of food into your lungs (known as 'aspiration'), which can sometimes lead to chest infections.

Ask your GP or wider health and social care team for a referral to a speech and language therapist (SLT). Your SLT can assess your ability to swallow safely and advise on consistencies of food and drink.

Also ask for a referral to a dietitian, who can provide guidance about diet and supplements. This may be helpful if swallowing difficulties lead to weight loss, dehydration or lack of energy.

"I have not been able to drink normally for over 20 years, but I still eat normally, albeit slowly."

With PLS, you are likely to be able to maintain your eating and drinking needs without having to top up with tube feeding. This is where liquid feeds are passed through a tube inserted through the outer wall of your stomach. However, if you feel tube feeding could help, ask for referral to an SLT and a dietitian.

For more information about swallowing, eating and drinking, see:
Information sheet 7A – *Swallowing difficulties*
Information sheet 7B – *Tube feeding*
Our guide – *Eating and drinking with motor neurone disease*

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 for 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:
Information sheet 7A – *Swallowing difficulties*

Will I be affected by emotional lability with PLS?

Emotional lability (also known as pseudobulbar affect) is a symptom sometimes seen in MND, where people experience crying or laughing that feels inappropriate to them. 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 and assume that crying means you are depressed.

"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 emotions see our guide:
Emotional and psychological support

"The more people that know this problem is due to my disease, the less of a problem it feels to me."

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 you experience changes to the way you think and learn, 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 choices and consequences carefully
- concentrate and take in new information
- recognise other people's feelings
- finish tasks.

What can be done about fatigue?

It might be helpful to take on board the idea that your energy is like a battery. If you use a lot of energy one day, you may need to rest and 'recharge' the next day. Do essential tasks first. Delay or get support 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 or 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.

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

What can be done about bladder urgency?

Some people with PLS experience a strong, sudden urge to urinate (known as urinary urgency) caused by over-activity of the muscles that control the bladder.

"Frustratingly, urinary urgency issues are very common for those of us with PLS, although bowel issues seem to be rare."

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 likely to have 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 advice.

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

How can I contact other people with PLS?

See *Further information* towards the end of this sheet for details about our MND Connect helpline. They can guide you to our local branches and groups, where you can meet others with MND, but not specific to PLS.

As the PLS form of MND is generally 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 PLS, and another slower progressing form of MND, usually known as progressive muscular atrophy (PMA). This is not an MND Association email group, but if you would like to join, contact our helpline, MND Connect for guidance.

You can also join our online forum at: **https://forum.mndassociation.org** which provides a safe space for people with or affected by MND to share experiences and support. This will include posts from people affected by all forms of MND.

You can also find a variety of MND or PLS groups on social media. This includes facebook, twitter and other platforms, which may have a local, national or global presence, allowing for wider peer support. Search for PLS or primary lateral sclerosis to see what's available. However, these groups may not be moderated.

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 information about benefits, entitlements and other support.

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

Health and Social Care Northern Ireland (NHS Northern Ireland)

Online information about NHS services in Northern Ireland.

Email: through the website contact page
Website: **http://online.hscni.net/**

Health in Wales

Online information about NHS services in 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 UK

The main online reference for the NHS.

Website: **www.nhs.uk**

NHS 111

The NHS 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 and some areas of Wales)

NHS Direct Wales

Health advice and information service for Wales.

Telephone: 0845 4647 (or 111 if available in your area)
Website: **www.nhsdirect.wales.nhs.uk**

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 information are available on request from

Email: infofeedback@mndassociation.org

or write to:

Information feedback, MND Association, Francis Crick House, 6 Summerhouse Rd, Moulton Park, Northampton NN3 6BJ

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Further information

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

3A – *MND care centres and networks*

3D – *Hospice and palliative care*

6A – *Physiotherapy*

6B – *Complementary therapies*

6C – *Managing pain*

7A – *Swallowing difficulties*

7C – *Speech and communication support*

7D – *Voice banking*

11C – *Equipment and wheelchairs*

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

Eating and drinking with motor neurone disease (MND) – our guide to help people with MND adapt how they eat and drink, if needed. It includes information, tips and easy-swallow recipes

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

Making the most of life with MND – our booklet on quality of life with MND focuses on how adapting your approach may help you continue doing the things you want to do

Emotional and psychological support - our guide on identifying difficult emotions and how to get support.

Changes to thinking and behaviour - our guide about the changes to thinking some people with MND may experience.

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 802 6262
Email: mndconnect@mndassociation.org
MND Association, Francis Crick House, 6 Summerhouse Rd,
Moulton Park, Northampton NN3 6BJ

MND Association website and online forum

Website: **www.mndassociation.org**
Online forum: **https://forum.mndassociation.org** or through the website

We welcome your views

We'd love to know what you think we're doing well and where we can improve our information for people with or affected by MND, or Kennedy's disease. Your anonymous comments may also be used to help raise awareness and influence within our resources, campaigns and applications for funding.

To feedback on any of our information sheets, access our 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, Francis Crick House, 6 Summerhouse Rd,
Moulton Park, Northampton NN3 6BJ

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