

First edition
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What is
primary progressive
MS?

This booklet is available in large print and audio CD. For either of these formats, contact the MS Society information team: 020 8438 0799 (Weekdays 9am-4pm, except Tuesday, 9am-3pm) or infoteam@mssociety.org.uk

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Where this symbol  appears in the booklet, the MS Society or another organisation can provide more information on a particular subject – more details are in the ‘Further information’ section at the back of this booklet.

As with the diagnosis of any form of multiple sclerosis (MS), it can be a shock to be told you have primary progressive MS. Whether your symptoms are obvious or barely visible to others, your condition has been given a label which opens up questions as well as providing answers.

You might want to find out everything you can straight away, or you might feel like taking it slowly. There's no 'right way' to react, but knowing a little about your type of MS and what it might bring can be helpful for planning ahead, managing the symptoms and adapting to changes over the years.

As well as this booklet, you might want to read *Just diagnosed: an introduction to MS* or *What is MS?*. For these free booklets or for further information, visit our website or get in touch with the MS Society information team. 

You can call the MS national Helpline to talk to someone about any aspect of living with MS. Freephone 0808 800 8000 (Monday to Friday, 9am-9pm) or email helpline@mssociety.org.uk. The helpline is for anyone affected by MS – people with MS, their family, friends, carers and colleagues.

What is primary progressive MS?

Primary progressive MS affects about 10 to 15 per cent of people diagnosed with MS.¹ It is called this because from the first (primary) symptoms it is progressive. Symptoms gradually get worse over time, rather than appearing as sudden attacks (relapses).

In primary progressive MS, early symptoms are often subtle problems with walking, which develop – often slowly – over time. People with primary progressive MS rarely have relapses and this is the most obvious difference with the more common relapsing remitting MS. Whatever symptoms someone experiences, the way they progress can vary – from person to person and over time. So, although in the long-term symptoms might get gradually worse, there can be long periods of time when they seem to be staying level, with no noticeable changes.

This type of MS is usually diagnosed in people in their forties or fifties – older than the average age for relapsing remitting MS – but it can be diagnosed earlier or later than this.

Equal numbers of men and women have primary progressive MS. This is different to relapsing remitting MS, where more women than men have the condition.

Progressive relapsing MS

A few people have progressive MS from the start, but also have occasional relapses. The progression is still the main cause of symptoms gradually getting worse, but because there are sometimes relapses this can be described as ‘progressive relapsing’ MS.

What symptoms can it cause?

People with primary progressive MS can experience many of the same symptoms as those with relapsing remitting MS, such as fatigue, muscle stiffness and sensory changes (which might be painful, cause numbness or ‘pins and needles’, for example). As with all types of MS, symptoms vary from person to person. Some symptoms, however, are more common than others.

Most people with primary progressive MS – around 80 per cent – develop problems with their walking.¹ At first, the changes may be so slight that they are hard to notice: perhaps a slight difficulty running or climbing stairs, or tripping up, particularly if not concentrating on walking. Some people find their walking gets more difficult the further they walk, and they need to rest when their legs start to feel weaker. Over time (usually many years), this can develop into progressive stiffness and weakness of both legs.

Why are problems with the legs so common in this type of MS?

A likely reason is that primary progressive MS tends to involve the spinal cord – a vital part of the nervous system for controlling this area of the body. This could also be why symptoms of the bladder, bowel and sexual function can sometimes accompany problems with the legs – these parts of the body also rely heavily on messages from the spinal cord.

People with primary progressive MS can also experience problems with memory or thinking (known as ‘cognitive’ symptoms), though usually any symptoms are mild. Studies into these symptoms in different types of MS have not shown clear differences.¹

Managing any of these symptoms is a case of finding what works best for you (see page 7).

Diagnosing primary progressive MS

The diagnosis of any form of MS can take a long time. It is not uncommon for a diagnosis to take several months, and frustratingly it can take even longer. A range of other possible causes need to be explored and many different tests need to be carried out. To make things more difficult, MS doesn't always show itself in the same way in each person.

In addition, as people get older they are more likely to have other conditions which could cause mobility and walking difficulties. As primary progressive MS is most often diagnosed in people in their forties and fifties, this can also make it harder to reach a diagnosis.

You may be given a diagnosis of 'MS' before it's clear what type it is. Although you may still feel 'in limbo', waiting for answers, it shouldn't stop you getting help to manage your symptoms, so discuss this with your doctors.

What are they looking for?

To diagnose any type of MS, a neurologist must decide that there has been MS activity in different parts of the brain or spinal cord, at different times. They also need to be confident that no other condition could be causing these symptoms.

What tests are used?

There is not a single test that can prove MS. Instead, a neurologist has to build a picture from the evidence they find when you are examined, and from what you describe about your symptoms. There are a number of tests, including:

- an MRI (magnetic resonance imaging) scan of the brain and spinal cord
- ‘evoked potentials’ (measuring the speed of messages passing to and from the brain)
- a lumbar puncture (an injection to test the fluid around the spinal cord)

Just as important are the symptoms you tell them about and the smaller changes they might pick up if they test your balance, your reactions or your eye movements.

Is a lumbar puncture still necessary?

For many people with primary progressive MS, a lumbar puncture is necessary to get the right diagnosis. With modern MRI techniques, evidence of relapsing remitting MS can often be seen without the need for a lumbar puncture. But the added information from a lumbar puncture is often still needed to diagnose primary progressive MS.

It is possible in some cases to make a diagnosis without a lumbar puncture if two MRI scans, at least a month apart, show new signs of MS activity.² However, this might not clearly show MS in every case and two MRI scans are rarely carried out in this way.

The difficulty of interpreting MRI scans

The brain MRI has to be interpreted with care for a number of reasons:

- People with primary progressive MS often have very few lesions in the brain – usually far fewer than people with relapsing remitting and secondary progressive MS.
- 40 to 50 years of age is the most common age for diagnosing primary progressive MS. People this age and upwards can have areas on a scan which show up brightly and look like lesions but in fact are part of the normal ageing process.
- Similar lesions can be seen in other neurological conditions.

Is it really the same condition as other types of MS?

Because of the different way it affects people and the different MRI features, it has been suggested that primary progressive MS may be a completely different condition to relapsing remitting MS. Most experts, however, think it is more likely that these two forms of MS are at different ends of the same spectrum.

Differences

MRI scans of the brain show fewer lesions in people with primary progressive MS compared with relapsing remitting MS. There may be other forms of inflammation happening and other processes involved in the brain which cause progression.

Similarities

No clear differences have been found in genes or the way the immune system works in people with the different types of MS.^{1,3} In fact, in those rare families in which there are two or more people with MS, it is not unusual to find that they have different types of MS.

Many people with relapsing remitting MS eventually develop secondary progressive MS. With both secondary and primary progressive MS, people's symptoms – on average – progress at a similar rate, suggesting that similar processes are involved.¹

Managing primary progressive MS

By its very nature, primary progressive MS is a changing condition. The progression of symptoms is often very slow, but such changes will happen over time. Because the way that it progresses varies from person to person and over time, there are still uncertainties about how MS might affect you, but knowing a little of what to expect can help with planning for the future – making sure change, when it happens, can be managed as smoothly as possible.

There is help available for the practical and lifestyle changes you and those close to you might experience, but this can be easier to access when you need it if you've checked out what's available earlier. Find out about your local social services and about any financial benefits you might be entitled to. There is also support for carers – partners, family members or friends who help look after someone with MS. The MS Society has more information on these and many other topics related to living with MS. You might also want to contact the local Society branch, who often have a good knowledge of local services. 

There are many treatments, including drug treatments, which can help manage symptoms. You should always discuss any treatment options, whether 'traditional', 'complementary' or 'alternative', with your GP, neurologist or other qualified health care professional. When a prescription medicine is discussed with you, there should be a clear explanation of the potential risks, side effects and benefits. The same discussion should take place when other therapies are being considered. Anything which might have the potential to improve symptoms might also be potent enough to cause side effects.

Wellbeing

Many people find that healthy eating and appropriate exercise help them to manage their MS symptoms. Keeping as fit and healthy as possible might make it easier to deal with the changes MS brings about. This kind of 'self management' is many people's natural response to changes in health – making the best of what they can do and being as prepared as possible to adapt when necessary.

There are health and social care professionals, including MS specialist nurses, physiotherapists, psychologists, dietitians and occupational therapists, who can help with this and make sure you find the best strategies to manage your MS. Your GP or neurologist can make appropriate referrals if you are not in touch with these specialist professionals.

Some people find the Expert Patients Programme helpful – a self management course for people with long-term conditions which includes tried and tested techniques for managing symptoms and daily life. Their details are at the back of this booklet. The MS Society also runs a number of information days (called 'Living with MS') around the country. Some of these are specifically for people with primary progressive MS. Visit www.mssociety.org.uk or call 020 8438 0700 for details of your nearest course.

Rehabilitation

A rehabilitation programme can help people remain independent. It covers a wide range of techniques, but the aim of each is to help someone make the best use of their abilities.⁴ When symptoms change, different people might be best placed to help. For example, a physiotherapist and occupational therapist can often help to minimise the effects of muscle weakness or unsteadiness on your feet. A dietitian might help you find ways to eat healthily if you are less mobile.

Neurological multi-disciplinary teams (combining many different specialist roles) are beginning to be established in the UK. The MS Society is working to place MS nurses and other MS specialist roles in health and social care throughout the UK, and to develop multi-disciplinary teams for MS. For details of services in your area, speak to your MS nurse, GP, neurologist or the MS Society information team.

For rehabilitation to work best, people with MS and their families also need to play an active part – learning about MS and ways to manage it. It is important that everyone is involved in making the right choices of how to manage symptoms and that any suggested treatments are well explained: How long will a treatment last? What inconvenience or side effects might it cause? What benefits can be expected?

Steroids

Steroids are not recommended for treating primary progressive MS. Steroids can speed up recovery from relapses, but where there are no distinct relapses, they have not been shown to be beneficial.⁵

Drugs to slow the progression

Historically there has been very little research carried out into progressive types of MS. This is for a number of reasons, including the fact that the processes behind MS progression

have not been understood as well as the processes behind relapses. In addition, the technology available has not been advanced enough to do the kind of research needed to find treatments for progressive types of MS.

However, things are beginning to change. For example, the MS Society has recently begun working with leading research experts to launch a Clinical Trial Network which focuses on developing therapies to protect nerve fibres. It's thought that if nerve fibres can be protected, progression might be slowed down or even stopped. This project is in its early stages, but should in a few years give a clearer focus to primary progressive MS research.

A drug called lamotrigine is already being investigated in a trial to see if it can protect nerve fibres and slow progression. Results from this study are expected in 2009.

The Society has also invested more than £3 million in new research centres in Cambridge and Edinburgh which are investigating the use of adult stem cells, along with other techniques to protect nerve fibres and promote the growth of 'myelin', the protective sheath around nerve fibres. These are all areas of research which hope to lead on to treatments for progressive forms of MS.

Another study, jointly funded by the Medical Research Council, MS Society and MS Trust is the CUPID trial. This is looking at how effective cannabis extract might be for protecting nerve fibres.

The MS Society is by no means the only funder of research in this area. The pharmaceutical company Novartis, for example, are planning a trial to look at the possible benefits of a drug – taken as a tablet – called fingolimod, for people with primary progressive MS. Recruitment for trial participants is expected to start in early 2009.

For more information about any of these areas of research, see www.mssociety.org.uk/research (the 'Research we fund', 'In the spotlight' and 'Potential therapies' sections) or contact the MS Society information team. 

i Further information

Further reading

Living with progressive multiple sclerosis: overcoming the challenges by Patricia K. Coyle and June Halper. Published by Demos Medical Publishing (Second edition 2008). This American book includes chapters on managing the social, economic and medical aspects of progressive MS.

Coping with Multiple Sclerosis: a practical guide to understanding and living with MS by Cynthia Benz and Richard Reynolds. Published by Vermillion (Second edition 2005). An informative and encouraging book to help people find ways to manage their MS.

Multiple Sclerosis: the questions you have – the answers you need by Rosalind C. Kalb. Published by Demos Medical Publishing (Fourth edition 2008). This American book looks into many of the topics people affected by MS raise, and offers information about what MS is, how to treat it, self management and practical solutions.

MS Society publications

The MS Society has publications on a wide variety of topics, including information for people just diagnosed, types of MS, managing relapses, and social services. For a publications list and order form visit the website www.mssociety.org.uk or call 020 8438 0799 (Weekdays 9am-4pm, except Tuesday, 9am-3pm). A range of Factsheets are also available to download from our website, or call the information team.

MS Society website and magazine

Keep up to date with news relating to MS with the MS Society website www.mssociety.org.uk and members' magazine, *MS Matters*. Details about *MS Matters* are on the web and in the Society's publications list.

MS Helpline

The award-winning MS Helpline offers confidential emotional support and information to anyone affected by MS, including family, friends, carers, newly diagnosed or those who have lived with the condition for many years. Information about MS is available in over 150 different languages by speaking to a Helpline worker via an interpreter. Call freephone **0808 800 8000** (Monday to Friday, 9am-9pm, except bank holidays), or email **helpline@mssociety.org.uk**

MS Society National Centre, Information Centre

Based at the MS National Centre in London, the Information Centre is equipped for visitors to read or locate books and journals or view videos and DVDs. The Information Centre also runs an information line 020 8438 0799 (Weekdays 9am-4pm, except Tuesday, 9am-3pm), which you can call to request publications, research articles or other information about MS. Search our library database at www.mssociety.org.uk/library

Local information centres

There are MS Society local information and support centres in many locations around the country. These centres are staffed by volunteers who can help you with information about MS and services in your area. Call 020 8438 0799 for the details of your nearest centre.

Local branches

The MS Society has a network of over 300 local branches across the UK. The branches – run by trained volunteers – provide information about MS and local services, a chance to meet others affected by MS and take part in a range of activities. For more information check the MS Society website or call 020 8438 0759.

Other organisations

Expert Patients Programme

Six-week courses to help people with a long-term condition maintain their health and improve their quality of life. Led by people who themselves live with a long-term condition. To find out about courses in your area, go to www.expertpatients.co.uk or ask your GP or MS nurse for details.

National MS Therapy Centres

A federation of therapy centres around the UK. They offer a variety of therapies, often including physiotherapy.

PO Box 126
Whitchurch SY14 7WL
Telephone 0845 367 0977
www.ms-selfhelp.org

Useful contacts



A large rectangular area filled with a light pink background and a grid of small, evenly spaced pink dots, intended for writing contact information.

References

- 1 Miller, D. H. and Leary, S. M. (2007) Primary progressive multiple sclerosis. *The Lancet Neurology*, 6, 903-11.
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- 5 National Institute for Clinical Excellence (2003) NICE Clinical Guideline 8. *Multiple Sclerosis: Management of multiple sclerosis in primary and secondary care*. London, NICE.

Authors and contributors

With thanks to Professor Alan Thompson and all the people affected by MS who contributed to this publication.

Disclaimer: We have made every effort to ensure that the information in this publication is correct. We do not accept liability for any errors or omissions. The law and government regulations may change. Be sure to seek local advice from the sources listed.

Suggestions for improvement in future editions are welcomed.

Please send them to infoteam@mssociety.org.uk

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Multiple Sclerosis Society

Multiple sclerosis (MS) is the most common disabling neurological disorder affecting young adults and we estimate that around 85,000 people in the UK have MS. MS is the result of damage to myelin – the protective sheath surrounding nerve fibres of the central nervous system. This damage interferes with messages between the brain and other parts of the body.

For some people, MS is characterised by periods of relapse and remission while, for others, it has a progressive pattern. For everyone, it makes life unpredictable.

The MS Society is the UK's largest charity dedicated to supporting everyone whose life is touched by MS. It provides respite care, a freephone MS Helpline, grants for home adaptations and mobility aids, education and training, specialist MS nurses and a wide range of information.

Local branches cater for people of all ages and interests and are run by people with direct experience of MS.

The MS Society also funds around 70 vital MS research projects in the UK.

Membership is open to people with MS, their families, carers, friends and supporters. You can help the work of the MS Society by:

- becoming a member
- making a donation
- offering your time as a volunteer

Contact information

MS National Centre
372 Edgware Road
London NW2 6ND
Telephone 020 8438 0700

MS Society Scotland
National Office
Ratho Park
88 Glasgow Road
Ratho Station
Newbridge EH28 8PP
Telephone 0131 335 4050

MS Society Northern Ireland
The Resource Centre
34 Annadale Avenue
Belfast BT7 3JJ
Telephone 028 9080 2802

MS Society Cymru
Temple Court
Cathedral Road
Cardiff CF11 9HA
Telephone 029 2078 6676

National MS Helpline
Freephone 0808 800 8000
(Monday to Friday, 9am-9pm)
Website www.mssociety.org.uk

The Multiple Sclerosis Society of Great Britain and Northern Ireland is a charity registered in England and Wales (207495) and Scotland (SC016433)