

Primary progressive MS exposed



Alison Whittam



Primary progressive MS exposed

Alison Whittam, Information Officer, MS Trust

The MS Trust is a UK charity for people with MS, their family and friends. The MS Trust Information Service offers a personalised enquiry service; produces a wide range of publications including Open Door, a quarterly newsletter; and provides web based information.

Thank you

The MS Trust would like to thank the following health professionals for their expert advice during the development of this book:

- Professor Alan Thompson, Consultant Neurologist, National Hospital for Neurology and Neurosurgery, London
- Professor Chris Ward, Consultant in Rehabilitation Medicine, Derby Hospitals NHS Trust, Derby
- Dr Belinda Weller, Consultant Neurologist, Western General Hospital, Edinburgh
- Mary Fraser, MS Specialist Nurse, Addenbrooke's Hospital, Cambridge
- Alison Smith, Rehabilitation Specialist Nurse MS, Derby Hospitals NHS Trust, Derby

We would also like to thank everybody with primary progressive MS who completed our questionnaire. Particular thanks go to Alan, Edith, Linda and Tony for reviewing the book and providing invaluable comments. Without your help this book would not have been possible.

Alison Whittam

Primary progressive MS exposed

ISBN 1-904156-21-5

© 2011 Multiple Sclerosis Trust

Registered charity no 1088353

This publication will be reviewed in three years.

All rights reserved. No part of this book may be reproduced, stored in a retrieval system or transmitted in any form by any means, electronic, electrostatic, magnetic tape, mechanical, photocopying, recording or otherwise without the written permission of the publisher.

Foreword

I have been associated with the MS Trust since 2001, so I was delighted when I heard they were working on a publication for people with primary progressive MS. My predominantly one sided PPMS was provisionally diagnosed in 1997 after several years of being last on long walks. I failed to get any fitter or faster and developed a tendency to trip over my left foot and to overbalance. Two years later I was referred to Queen Square where a further MRI scan, lumbar puncture and an array of tests gave me the confirmation I needed. I have a biomedical background - after studying Zoology, I taught at Edinburgh's Napier College and served on its Court, I also worked in medical research prior to raising our two children and carrying out my husband's constituency correspondence – so the diagnosis came as no real surprise to me and everything seemed to slot into place. However, that didn't make it any easier to come to terms with.

A dozen years on, aspects of body malfunction that horrified me in the abstract are now an everyday feature. One hand performs most of the functions of two and I have only recently stopped driving my adapted car as I cannot get in and out unaided. I still go up stairs easily, down backwards and keep the trigeminal neuralgia at bay with an amazing cocktail of drugs. Because of my background, researching my MS came naturally and I have been only too aware of how difficult it is to readily access information about the primary progressive condition. This book will be an essential eye-opener to the newly diagnosed and valuable to those of us who need to keep up to date with our present or changing circumstances.

Edith Rifkind

Patron MS Trust

Contents

1.	Key information at a glance	6
2.	What is primary progressive MS?	9
3.	How primary progressive MS differs from the other forms of MS	12
	Clinical features	
	Pathological features	
4.	Diagnosing primary progressive MS	17
	Diagnostic criteria	
	Tests that may be carried out to diagnose primary progressive MS	
5.	Symptoms of primary progressive MS	26
	Symptoms commonly seen in primary progressive MS	
	Other symptoms that may be experienced	
	Complications and other concerns	
6.	Progression	34
	Predicting how primary progressive MS might progress	

7. Management of primary progressive MS	37
Drug treatment	
Rehabilitation medicine	
Prevention of complications	
General health and wellbeing	
Complementary and alternative therapies	
8. Trials in primary progressive MS	48
Challenges to trials	
Past and current trials	
Questions still to be answered	
Future research priorities	
9. Living with primary progressive MS	53
The MS journey	
Impact of diagnosis	
Coping with increasing disability and maintaining independence	
Working with primary progressive MS	
Maintaining social relationships	
Final thoughts	
10. Sources of support	77
Other MS Trust publications you may find helpful	
Online support groups	
Benefits and legal advice	
Getting around	
Equipment and adaptations	
Relationship support	
Health and social care information and carer support	
Holidays	

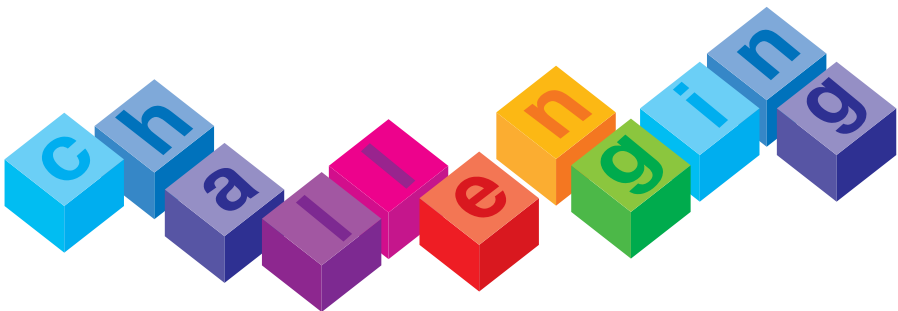
1. Key information at a glance

- For about 10-15% of people diagnosed with MS, there is an absence of relapses and remissions and progression occurs from the outset – this is known as primary progressive MS. It is estimated that 10-15,000 people have primary progressive MS in the UK.
- The rate at which progression occurs varies widely between individuals. At times it may be at a standstill, or it may even improve very slightly for a short period. For others the progression is more rapid. However, onset is typically slow with symptoms steadily increasing over a period of months or years.
- The majority of people with primary progressive MS are diagnosed in their 40s and 50s; this is often about the same age that someone with relapsing remitting MS typically moves into the secondary progressive stage of their MS.
- About the same number of men and women are affected by primary progressive MS.
- In the majority of people with primary progressive MS the spine is predominantly affected by MS lesions. Although there can also be lesions in the brain, they tend to be smaller and fewer in number than in the other types of MS.

- Inflammation is less dominant in primary progressive MS than in relapsing remitting or secondary progressive MS, but results in more damage to the nerves themselves. It is this nerve damage that is thought to be a major contributor to the non-relapsing, progressive nature of primary progressive MS.
- Diagnosing primary progressive MS can be a long and frustrating process because there are many other conditions with similar symptoms. Establishing a diagnosis is essentially a process of elimination, ruling things in or out, with the neurologist trying to ensure that there is no alternative explanation for the symptoms and findings. They will also want to see firm evidence of at least one year of progression to secure a diagnosis.
- In more than 80% of people with primary progressive MS, the first symptom experienced is a stiffness and/or weakness in both legs that gradually worsens.
- Although people with primary progressive MS don't have relapses, it is possible to experience an acute deterioration in function due to other factors such as pain or an infection.
- There is currently no disease modifying therapy available that can alter the course of primary progressive MS, but trials are ongoing. Management focuses on three key areas: symptom management; prevention of complications; and promoting general health and wellbeing.
- It is unusual for steroids to be prescribed for primary progressive MS, but sometimes a one-off course may be given if a person begins to experience a period of deterioration. As with the other types of MS, there is no evidence that steroids alter the long-term course of primary progressive MS.

- Rehabilitation therapies, such as physiotherapy, may be beneficial. They aren't curative, but they can help you make the best of, and maintain, functions that will help you manage your symptoms and live your life as fully as possible.
- It is important to make the most of both your physical and emotional wellbeing to enable you to do the things you enjoy as well as the things you have to do.
- Historically, primary progressive MS has been the least studied of the different clinical types of MS. Trials have been more challenging for many reasons: inadequate diagnostic criteria; the relative rarity of the condition; use of measures of effectiveness that are not relevant or sensitive to the slow changes seen in primary progressive MS; and the processes that need targeting, neuroprotection and repair, are more testing.

There is no preventative action that you could or should have taken to prevent you from developing primary progressive MS.



2. What is primary progressive MS?

MS is a complex and unpredictable condition that varies widely from person to person and does not follow a set pattern. However, there are a number of 'types' of MS that have been used to try to broadly group individuals in accordance with how the condition has developed.

Originally MS was classified into two main clinical subtypes:

■ relapsing remitting MS

characterised by periods when symptoms flare up (relapses) followed by periods of good or complete recovery (remissions); and

■ progressive MS

where progressive deterioration occurs over months or years.

However, over the years clinical evidence pointed to there being three main subtypes of MS. As well as clinical differences being noted within the group with progressive MS, there were also found to be differences on magnetic resonance imaging (MRI) scans of the brain. Therefore, in 1996, it was proposed that the group of people with progressive MS should be further sub-divided to distinguish between those with progression that occurred primarily from the outset of the condition - **primary progressive MS**, and those whose progression followed on secondarily after an initial relapsing remitting course - **secondary progressive MS**.

This classification is now widely recognised and the agreed definitions are as follows:

Primary progressive MS (PPMS):

“Disease progression from outset with occasional plateaus and temporary minor improvements allowed.”

For about 10-15% of people diagnosed with MS, there is an absence of relapses and remissions and progression occurs from the outset. The rate at which progression occurs varies widely between individuals. At times it may be at a standstill, or it may improve very slightly for a short period. For others the progression is more rapid.

Relapsing remitting MS (RRMS):

“Clearly defined disease relapses with full recovery or with sequelae* and residual deficit upon recovery; periods between disease relapses characterised by a lack of disease progression.”

[*sequelae - after effects of the condition.]

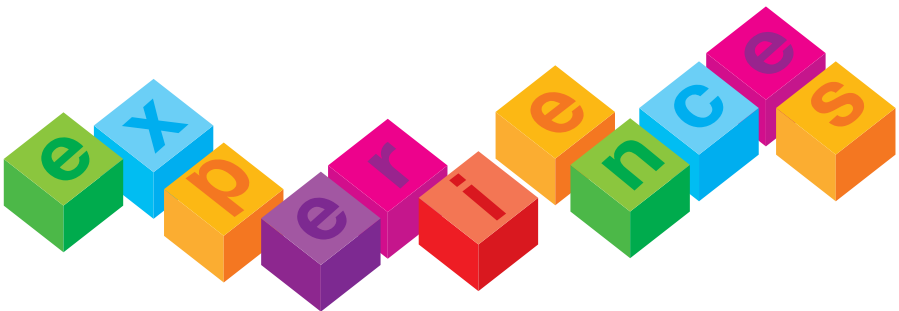
The majority of people initially follow a relapsing remitting course of MS. On average relapses (a significant worsening or re-occurrence of a symptom, or group of symptoms, that lasts for more than 24 hours in the absence of an infection) occur once or twice a year. This is followed by a period of recovery when symptoms become less severe or disappear altogether. Although symptoms may worsen gradually over time, there isn't a marked increase in the level of a person's disability.

Secondary progressive MS (SPMS):

“Initial relapsing remitting disease course followed by progression with or without occasional relapses, minor remissions, and plateaus.”

People with relapsing remitting MS mostly go on to develop a progressive form of the condition. Whilst the severity and frequency of relapses decrease or even stop altogether, the level of permanent disability increases over time.

Each of these ‘types’ of MS encompasses a wide range of experiences of MS. Even with these definitions in place it can still be difficult to determine exactly what type of MS an individual has, particularly at the point of diagnosis and it may only become apparent over time as the condition develops.



3. How primary progressive MS differs from the other forms of MS

There has been a lot of debate over the years as to whether the different types of MS represent different points on a broad spectrum within one condition, or whether they are in fact separate conditions in their own right. One school of thought is that primary progressive MS may actually be exactly the same as secondary progressive MS, but that the initial relapsing remitting phase is ‘missing’ or ‘clinically silent’. It could be that early attacks may have been mild and dismissed or overlooked, or that lesions have occurred in areas that have not given rise to symptoms. However, to date there is no consensus on whether primary progressive MS is a different condition or not.

“I did not understand how different PPMS is to the other types and nobody really explained this. I sometimes think that PPMS is a different disease entirely. I would have liked to know more, but felt that professionals shied away from giving me this information because they were wary of the emotional impact.”

Fiona, Surrey

There is a common misconception that primary progressive MS is the ‘worst’ type of MS. However, just as there is a large variation seen in the number and severity of relapses between people with relapsing remitting MS, so too the rate of progression varies widely between

those with primary progressive MS. Although undoubtedly for some people progression can be extremely rapid, for others it can be slow or at a virtual 'standstill', whilst others may reach a plateau after a period of steady progression.

“Slowly over a period of six years my walking has deteriorated. In the past few months it has taken a sudden down slide and has become more noticeable to others around me.”

Vanessa, Lancashire

Many people with primary progressive MS actually consider the condition to be less unpredictable than relapsing remitting MS, where relapses of varying length and severity can occur very suddenly with no prior warning.

The following sections highlight some of the principal differences between primary progressive, relapsing remitting and secondary progressive MS.

Clinical features

Age at onset

Diagnosis of primary progressive MS tends to be at an older age, typically ten years later than relapsing remitting MS, so most people are diagnosed in their 40s and 50s, though it can be diagnosed earlier (and later) than this. Primary progressive MS is virtually unheard of in children.

Typically relapsing remitting MS is diagnosed between the ages of 20-40 years, with the average age at diagnosis being 30. However, children as young as five have been diagnosed with relapsing remitting MS.

Comparing primary progressive with secondary progressive MS, the age of onset is similar, ie someone with primary progressive MS tends

to develop symptoms at around the same age that someone with relapsing remitting MS typically moves into the secondary progressive stage of their MS.

Gender

More women than men are diagnosed with relapsing remitting MS. This gender difference is not seen in primary progressive MS; similar numbers of men and women are affected.

Symptoms at onset

In more than 80% of people with primary progressive MS, the first symptom experienced is a stiffness and/or weakness in both legs that gradually worsens – the medical term for this is ‘spastic paraparesis’. Initially this may manifest as minor problems such as tripping on kerbs or when climbing stairs, but can gradually worsen over time and become more noticeable resulting in walking difficulties. Bladder and bowel function can also be affected and there may be sexual problems such as erectile dysfunction or loss of sensation.

“I have muscle spasms in my legs. I have experienced a loss of libido.”

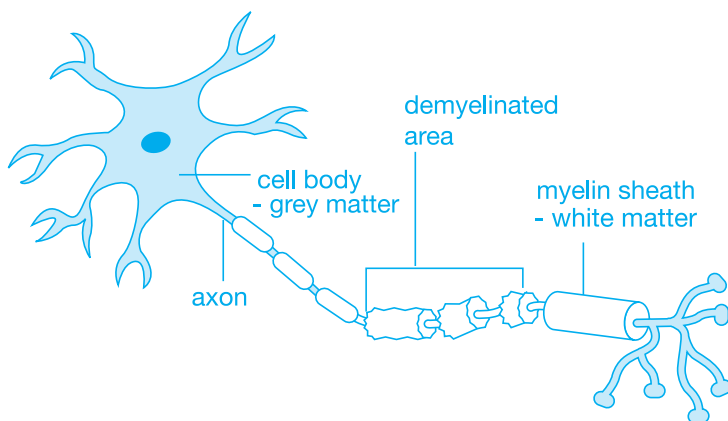
Christine, Wiltshire

In relapsing remitting MS, it is more likely that sensory symptoms such as pins and needles or numbness, or acute visual loss due to optic neuritis will be experienced at the onset.

Pathological features

Distribution of lesions in the central nervous system

The central nervous system is made up of white matter and grey matter. White matter makes up around 60% of the brain volume and consists mainly of myelinated axons. This is the tissue through which messages pass between different areas of grey matter, which makes up the remaining 40% of the brain. The grey matter consists predominantly of the nerve cell bodies.



A classic sign of MS are lesions in the central nervous system (brain and spinal cord), which indicate areas where the myelin sheath surrounding the nerve axons has been damaged or destroyed. In 80-90% of people with primary progressive MS it is the spinal cord that is predominantly affected by lesions, whereas in relapsing remitting and secondary progressive MS the majority of lesions are found in the brain. Although there can be lesions in the brain in primary progressive MS, they tend to be smaller and fewer in number than in the other types of MS.

Despite a clear increase in disability in people with primary progressive MS, very few new lesions are found over time, on average there are 3.3 new lesions per year. The average number of new lesions for someone with secondary progressive MS is 18.2 per year. The pattern of lesions in the spinal cord in primary progressive MS mirrors that seen in secondary progressive MS.

Inflammation versus axonal damage

Although inflammation is present in primary progressive MS, it is seen to a much lesser extent than in both relapsing remitting and secondary progressive MS, and it is more typically seen in the early stages of the condition. It has recently been suggested that progressive MS may be caused by inflammation, but that the inflammation is of a different nature to that seen in relapsing remitting MS. One theory is that progressive MS may be driven by plasma B cells, rather than by white blood cells in the immune system known as T cells.

MS lesions are seen in the white matter of the central nervous system, where demyelination of the nerve axons has occurred. As has already been mentioned, both the size and location of these lesions is different in primary progressive MS to the other types of MS, but of all the groups of people with MS those with primary progressive MS have the largest discrepancy between the number of lesions and their level of disability, so what else could be responsible for the degree of disability associated with primary progressive MS?

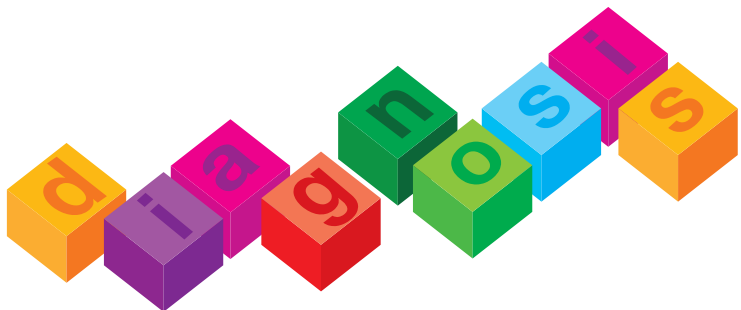
Although there is less inflammation associated with the lesions, there is more degeneration of the nerve axons themselves within primary progressive MS lesions than either relapsing remitting or secondary progressive MS. This loss of nerve cells, or neurodegeneration, is also seen within the white and grey matter that appears to be normal on an MRI scan. These subtle abnormalities in the normal appearing matter have been shown to increase over time. It is widely believed that this diffuse neurodegeneration throughout the central nervous system is a major factor in the development of the irreversible progressive disability seen in primary progressive MS.

In the early stages of relapsing remitting MS some remyelination of the lesions can occur leading to periods of remission. However, in primary progressive MS it has been shown that there is extensive damage to the cells responsible for making myelin (oligodendrocytes) which could contribute to the non-remitting course of the condition.

4. Diagnosing primary progressive MS

Diagnosing MS can be a long, difficult and frustrating process. Not only does MS vary widely from person to person, but there are also many other conditions which have similar symptoms to MS that need to be ruled out as a possible cause. Therefore features that are typical of MS, as well as those that are not characteristic, need to be recognised in the diagnostic process.

Securing a diagnosis of primary progressive MS has problems all of its own. Symptoms usually occur slowly over a long period of time and may initially be overlooked. As we age there is more general 'wear and tear' on the body and we are more prone to conditions such as cervical spondylosis (degeneration of the vertebrae and discs in the neck), trapped nerves, and slipped discs, which can give rise to symptoms similar to those seen in primary progressive MS, as can the menopause and weight problems. This can make diagnosis a lengthy process, with people often seen by other specialist services such as orthopaedics, vascular surgeons and physiotherapists, before they are referred to a neurologist.



“I was first referred to an orthopaedic registrar (three month wait) by my doctor with a possible prolapsed disc, MRI revealed a lesion on my spinal cord and I was referred on to neurology. A further three month wait was curtailed by seeing my neurologist as a private patient. Getting the diagnosis was a bit of an anti-climax...I think the real tough part was the waiting and being told early on that the lesion indicated something unpleasant.”

Clive, Berkshire

If you were diagnosed with MS a decade ago, it may have taken some time before it was confirmed as primary progressive MS. However, due to increased awareness and better diagnostic criteria, you are now more likely to be told if it is primary progressive MS at the time of diagnosis.

“I more or less knew I had MS, but to hear the doctor say it was heartbreaking. I asked the doctor which type of MS, but it was several months before they told me I had PPMS.”

Debbie, Norfolk

“I was diagnosed with MS in 2003; I was informed by my new specialist in 2008 I had PPMS, but only after asking.”

John, Cumbria

Diagnostic criteria

The diagnostic criteria for MS have been gradually evolving over the last 50 years. Originally they focused on a patient's clinical history and a neurological examination. With the advent of MRI, in 1983 the 'Poser criteria' were formulated.

In 2001, the 'McDonald criteria' were proposed which were more specific, more sensitive and allowed for earlier diagnosis in many cases. They also included criteria specifically for diagnosing primary progressive MS, which were further simplified in a revision of the criteria in 2005.

Diagnostic criteria for PPMS (2005 Revisions to the McDonald Criteria)

1. Evidence of at least one year of disease progression (determined retrospectively or prospectively).
2. Plus **two** of the following:
 - a positive brain MRI;
 - a positive spinal cord MRI;
 - a positive cerebrospinal fluid (CSF) test.

A further revision of the criteria is expected in 2011 and it is anticipated that the criteria will continue to evolve over time as medical knowledge advances.

To make a diagnosis of MS, a neurologist is looking for evidence that several areas of the central nervous system have been affected; this is known as 'dissemination in space'. They are also looking for evidence that symptoms have arisen at two or more different times; this is known as 'dissemination in time'. As explained later, this can be difficult to establish in primary progressive MS. The following must also be considered when trying to diagnose primary progressive MS:

- excluding early relapse activity;
- MRI abnormalities are less specific and less common for primary progressive MS than other types of MS;

- distinguishing the condition from other progressive syndromes; and
- ruling out co-existing illnesses that may have overlapping symptoms.

Meeting the diagnostic criteria for ‘dissemination in space and time’

In relapsing remitting MS, relapses occur at distinct points in time and provide the evidence for dissemination in time. Because these discrete attacks are absent in primary progressive MS, it can be more difficult to establish that symptoms have occurred at different times.

It is also more difficult to demonstrate clinically that different parts of the central nervous system have been involved in primary progressive MS. In primary progressive MS often only one area or ‘system’ of the body is affected. Although walking problems can be associated with bladder and sexual problems, these symptoms are all caused by lesions in just one area - the spinal cord. This can make it difficult to show that different areas of the central nervous system have been affected. In contrast two or more areas or ‘systems’ in the body are usually affected in relapsing remitting MS, for example vision, bladder function, cognition and/or altered sensations. These different symptoms are caused by lesions occurring in different areas of the central nervous system and support the dissemination in space aspect of the criteria.

Excluding early relapse activity

For some people with MS, early attacks or relapses might have been so mild that they were overlooked or dismissed in the mist of time, or they may have happened so long ago they have been forgotten. A thorough clinical history needs to be taken to determine whether early relapses have occurred, which could mean that a diagnosis of secondary progressive MS is more appropriate.

Even in clear cut primary progressive MS, around 10-15% of people experience a single relapse at some point during the course of their MS; but overall the predominant feature is progression.

Magnetic resonance imaging (MRI) abnormalities are less specific and less common

Complications can arise when interpreting the MRI scans of people with suspected primary progressive MS. Given that primary progressive MS is generally diagnosed at a later age, it can be difficult to distinguish lesions caused by MS from normal brain changes caused by ageing. The lesions that are seen in the brain in primary progressive MS are also usually smaller and fewer in number than those seen in relapsing remitting and secondary progressive MS.

Distinguishing the condition from other progressive syndromes

The most common presenting symptom in primary progressive MS is a progressive spastic paraparesis, but this is also seen in many other progressive conditions with several different origins, some examples of which are listed below:

- cord compression – cervical spondylosis;
- hereditary conditions – Friedrich’s ataxia;
- metabolic conditions – B12 deficiency;
- inflammatory conditions – neurosarcoidosis;
- infections – syphilis, HIV; and
- degenerative conditions – motor neurone disease.

If the picture is not clear, additional tests may be carried out to exclude conditions whose symptoms can mimic those of MS.

“I had a switched on GP who referred me quickly to a neuroconsultant who is an expert in MS. At the same time she arranged investigations of all presenting symptoms to eliminate other possibilities.”

Maggie, Yorkshire

Co-existing illnesses

As a person ages they become more susceptible to conditions such as type 2 diabetes, osteoporosis, cancer, depression, memory loss or dementia, and cardiovascular disease. Some of the symptoms of these conditions overlap with those of MS and may hamper the diagnosis.

Establishing a diagnosis is essentially a process of elimination, ruling things in or out, with the neurologist trying to ensure that there is no better explanation for the symptoms and findings than a diagnosis of primary progressive MS. They will also want to see firm evidence of at least one year of progression to secure a diagnosis.

Tests that may be carried out to diagnose primary progressive MS

The following is a list of the tests that may be used to help secure a diagnosis of primary progressive MS. The majority of people will only have to undergo a few of these tests, but if symptoms are atypical, or results from initial tests such as MRI are inconclusive, further tests such as visual evoked potentials, a lumbar puncture or blood tests may be needed.

“Results were confusing from my initial tests, so I went for a second opinion and [further] tests.”

Mahesh, London

Clinical history

This is a discussion about a person’s general health, medical history and any previous symptoms, or problems, they have experienced.

Neurological examination

The neurologist may carry out simple tests on movement, balance, coordination, reflexes and vision that can suggest whether MS is a

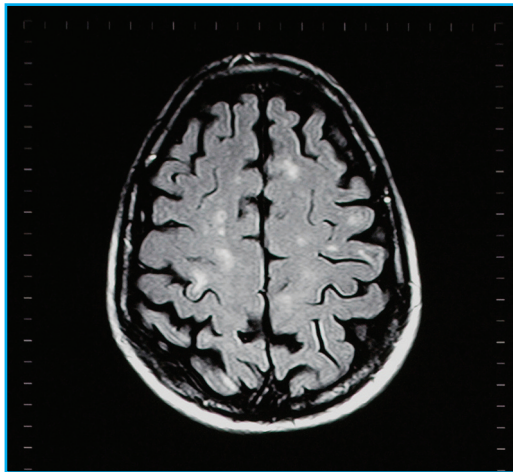
cause of symptoms. These tests may also indicate, or give clues to, where in the central nervous system the damage has occurred.

Magnetic resonance imaging (MRI)

Since it was first introduced in the 1980s, MRI has become a key tool in the diagnosis of MS. Magnetic fields and radio waves are used to create images of the brain and/or spinal cord. Two types of scan may be carried out.

The main type of scan used in the diagnostic process is known as a T2 image. This is used to determine the total number of lesions or overall disease burden; lesions appear as bright spots on the scan which indicate areas where the myelin sheath has been damaged or destroyed. Changes in the brain due to the ageing process also appear as bright spots on a T2 scan, which is why it can be difficult to distinguish the small lesions seen in primary progressive MS from the normal changes seen as a person ages.

MRI scan of the brain



Since the spinal cord is primarily affected in primary progressive MS, it would be expected that an MRI of both the brain and spinal cord

would be carried out, whereas in the other forms of MS a spinal MRI may not be necessary. Spinal imaging can also be useful in excluding other conditions.

Although less useful for diagnosis than a T2 image, the other type of image that can be taken is a T1 image. Areas where nerve cells have been permanently lost or damaged appear as dark areas on this type of scan. A T1 scan can also show areas of swelling (oedema) that aren't permanent. A contrast dye called gadolinium can be used during a T1 MRI to enhance the sensitivity of the scan. Gadolinium enhancement (GdE) shows areas of active inflammation indicating new lesions or lesions that are becoming larger. However, as inflammation is less common in primary progressive MS than relapsing remitting or secondary progressive MS, gadolinium enhanced T1 images are less useful.

“In a funny way, I was almost pleased to get an MRI scan that proved I’d got it, lumbar puncture and other tests had proved negative.”

David, Leicestershire

Visual evoked potentials

Evoked potential tests are used to measure the speed of electrical messages along sensory nerves to the brain. Visual evoked potentials (VEPs) are the most commonly used in the diagnosis of MS, they measure the speed of messages sent from the eyes to the brain in response to a flashing chessboard pattern on a computer screen. A delay in the speed of the message can indicate damage to the optic nerve, even if there aren't any obvious visual problems.

Lumbar puncture

A hollow needle is inserted into the base of the spine and a small amount of the cerebrospinal fluid (CSF) that surrounds the brain and spinal cord is drawn off. The number of white blood cells in the CSF

may then be counted. The number of white blood cells a person with MS has can be as much as seven times higher than would be normally expected. However, a count higher than this would more likely be due to an infection, rather than MS.

In MS, immune cells cross the blood brain barrier and attack myelin, this results in higher levels of a protein called immunoglobulin (IgG) being present in the CSF than would be expected. Tests can be carried out on the CSF to see if this has happened. The amount of IgG in the CSF can be compared to that in plasma from a blood sample, giving a value known as the IgG index.

In another test, samples of CSF and plasma are placed on a gel and a voltage is applied (electrophoresis). Proteins of the same size bunch together and are visualised as 'bands' on the gel. The presence of two or more bands in the CSF, but not in the plasma sample, is indicative of disease activity. The bands are known as oligoclonal bands and are seen in around 80% of people with primary progressive MS. Although highly suggestive of MS, it is not a definitive test and the presence of oligoclonal bands doesn't mean that someone definitely has MS. Therefore a lumbar puncture is more typically used to support a diagnosis if other tests have proved inconclusive.

An increased IgG index or evidence of oligoclonal bands is classed as a positive CSF result.

Blood tests

There is currently no blood test that can determine whether a person has MS or not. However, blood tests may be performed to rule out other conditions or an infection.

5. Symptoms of primary progressive MS

As with other forms of MS, a wide variety of symptoms can be experienced in primary progressive MS, some are more common than others, but not everyone will experience all of the symptoms described.

Symptoms commonly seen in primary progressive MS

Mobility problems

As the lesions are predominantly found in the spinal cord in primary progressive MS, the most common symptoms experienced are associated with mobility. The majority (83%) of people experience a gradual stiffness or weakness in both of their legs (spastic paraparesis) which makes walking increasingly difficult. However, a smaller proportion of people (6%) find that their primary progressive MS is 'one-sided', affecting the arm and leg on one side of the body. This is known as hemiparesis and it can be confused with stroke. Hemiparesis can lead to problems with writing and feeding if the dominant arm is affected.

“My left side has been impaired, making life very difficult. I’ve had to change from being left handed to right handed.”

Linda, Essex

Weakness in the limbs is due to poor conduction of messages by the damaged nerves, rather than a problem with the muscles themselves. Weakness can result in the dragging of a leg or dropped foot (the inability to lift the foot and toes when swinging the leg forward whilst walking). Weakness can lead to other problems such as pain in the hips and poor balance as people often alter the way they walk to compensate for any difficulties. It can become harder to walk longer distances.

“I have difficulties walking because of a ‘dropped foot’, this is now much improved by use of a functional electrical stimulator which I have had for 15 months.”

Alan, London

Spasticity and spasms

Spasticity is a condition in which muscle tone becomes greatly increased. When spasticity occurs in a limb, signals from the brain are interrupted and the muscle remains in a shortened contracted state, causing the limb to feel painful, stiff, tight and difficult to move. Spasticity is especially common in the legs and can make it difficult to walk. The severity of spasticity can vary widely from mild muscle stiffness to severe and painful muscle spasms.

Bladder and bowel problems

Lesions in the spinal cord can also affect the function of the bladder and the bowel. Typical bladder problems experienced in MS are:

- frequency – an increase in the number of times a person needs to urinate;
- urgency – the feeling of having to empty the bladder immediately, an inability to ‘hold on’;
- hesitancy – difficulty in starting to urinate;
- incontinence – the inability to hold urine in the bladder until an appropriate time.

Damage to nerves controlling the process of passing food through the digestive system can result in bowel problems. Constipation can arise as a result of a disruption of messages to the colon or rectum so the body does not realise a stool needs to be passed. If a person is inactive, this can slow the digestive process leading to constipation, as can a lack of fluid in the diet. The other bowel problem that can occur is faecal incontinence, the inability to control bowel movements. A continence specialist can give advice if you are experiencing bladder or bowel problems.

“I have to know where all the toilets are in any location I am visiting and I know that this can be frustrating for those with me.”

Tony, Kent

Sexual difficulties

The nerves of the reproductive organs originate in the spinal cord, so sexual difficulties can occur in primary progressive MS. This can be a distressing symptom and one that can be particularly difficult to talk about. Erectile dysfunction is common in men, whilst women may experience discomfort during intercourse due to a lack of lubrication. Other symptoms can include a loss of libido, altered sensations in the genital area or a change in the intensity and frequency of orgasms. Sexual difficulties can be compounded by other symptoms of primary progressive MS such as pain, bladder and bowel problems, fatigue and depression. The menopause and some medications can also affect sexual function.

Fatigue

Fatigue is one of the most common symptoms of MS. The cause of it is not well understood and it cannot be linked to a particular area of damage within the central nervous system. For some people fatigue can be the most disabling of symptoms, affecting their ability to work

and interrupting their normal activities. The fatigue experienced in primary progressive MS is not proportional to the amount or level of physical demands; simply getting out of bed, showering and dressing can be exhausting for some people. The level of fatigue experienced does not necessarily reflect the severity of a person's MS and it can be their only symptom. Other factors such as poor or interrupted sleep, low mood, stress, infections, some medications and heat can also add to fatigue.

“Fatigue is an increasing problem – not necessarily related to levels of activity, and therefore completely unpredictable. I have occasional mild bladder and bowel problems and pain which is definitely aggravated by the wrong kind of exercise.”

Maggie, Yorkshire

Other symptoms that may be experienced

Although the spinal cord is predominantly affected in people with primary progressive MS, lesions can be found in the brain and so symptoms caused by these lesions may still occur, although they are more typically seen in people with relapsing remitting MS. They include problems with thinking (cognition), balance issues, visual problems, sensory problems, tremor, speech and swallowing difficulties, depression and anxiety.

Cognitive problems

Cognitive problems seen in primary progressive MS can include difficulties with attention, memory, spatial reasoning and verbal fluency. In the past cognitive problems were considered to be rare in primary progressive MS. Although seen to a lesser extent than in secondary

progressive MS, it is now acknowledged that problems with thinking are more common than previously thought, with as many as a quarter of people with primary progressive MS affected.

Cognitive problems can be particularly challenging as often the person with MS does not recognise, or accept, that they have an issue. This can be difficult for other family members as the person with MS can be perceived as being uncaring or insensitive.

“[You] should be made aware of certain cognitive problems; I was a teacher and I suddenly found it almost impossible to learn the names of the 250 pupils I taught each week. I didn’t know why at the time and am certain that many colleagues thought I wasn’t up to the job.”

Linda, Northamptonshire

Cerebellar syndrome

The cerebellum is the area of the brain that controls movement, balance and posture. This area is affected in around 9% of cases of primary progressive MS. If lesions occur in this region it can result in problems with dizziness and vertigo, balance, difficulties with coordination particularly when walking, or tremor. Speech can also be affected.

Brainstem syndrome

The brainstem is the lower part of the brain located above the start of the spinal cord. It is involved with functions including the coordination of vision and hearing, so damage in this area can lead to visual problems such as double vision (diplopia) or a flickering or jerking of the eyeballs on movement known as nystagmus. A brainstem syndrome is seen in around 1% of those with primary progressive MS.

Optic neuritis

Optic neuritis is the inflammation of the optic nerve and it can cause progressive visual loss. This is estimated to occur in 1% of people with primary progressive MS. Optic neuritis can also lead to impaired colour vision and blurred vision that cannot be corrected with glasses.

Pain and sensory symptoms

For many years MS was considered to be a pain free condition. However, it is now widely acknowledged that neuropathic (or nerve) pain can be experienced in MS as a result of damage to the nerves. Examples of some of the types of nerve pain that can occur in primary progressive MS are sensory symptoms such as pins and needles, feelings of burning or crawling, numbness and tightness. Trigeminal neuralgia, a sharp excruciating pain in the face that can be triggered by simple actions such as talking, smiling or eating, can occur if the facial nerves are affected.

“The left side of my face is rather immobile and I have left-sided trigeminal neuralgia. This is the worst aspect of the condition for me.”

Catherine, Edinburgh

Speech and swallowing difficulties

Speech and/or swallowing difficulties can arise if there is damage to the nerves controlling the diaphragm, vocal cords, lips, tongue or nasal cavity. This can lead to problems such as slurring or controlling the volume of speech, known as dysarthria. If damage occurs to areas of the brain controlling thought, memory and attention, this can lead to problems in finding words or forming sentences, known as dysphasia. A lack of coordination of the muscles involved in speech can also lead to problems with swallowing, this is known as dysphagia.

Depression and anxiety

Depression and anxiety is more common in people with MS than in the general population. It may be caused directly by MS damage to the nerves in the brain. It can also be a side effect of some medications, or arise as a consequence of having to live with the demands of a chronic condition – this is known as a reactive depression.

“I struggled with depression for a year before seeking help. I had to stop working almost as soon as I was diagnosed and have had great difficulty in adjusting to that – I still do struggle with it.”

Mel, Warwickshire

Although people with primary progressive MS don't have relapses, it is possible to experience an acute deterioration in function due to other factors such as pain or an infection.

Complications and other concerns

The symptoms already described can occur directly as a result of the condition, but symptoms may also arise as a consequence of the problems that MS symptoms bring, rather than being caused directly by the MS itself. These may include the development of pressure sores due to a lack of mobility, falls due to walking or balance problems, muscle pain as a result of added strain on the back or legs caused by altering the way you walk, or weight problems if there are mobility or swallowing issues. Good management of the condition should help prevent many of these complications from arising.

It is important to remember that not all the symptoms you experience are necessarily connected to your MS. If you experience anything unusual you should still see an appropriate health professional.

“It can be easy to blame various symptoms on MS, but it should be emphasised that these problems are just as likely to be a different medical problem. Don’t take anything for granted and don’t be fobbed off by doctors.”

Hilda, Surrey

Living with primary progressive MS can create concerns around emotional, social and financial issues due to factors such as the psychological adjustments you have to make to come to terms with living with a progressive condition, possible changes of role within the family and maintaining employment. More information about living with MS can be found in section 9 of this book.

One thing to keep in mind is that there is no preventative action that you could or should have taken to prevent you from developing MS.



6. Progression

It is a common misconception that the course of primary progressive MS can be predicted and that a diagnosis implies a rapid downhill course for everyone. This has fuelled the belief that primary progressive is the ‘worst’ type of MS. In reality, just as with other types of MS, considerable individual variation is seen in the course of primary progressive MS.

Whilst for some people progression can be very rapid, primary progressive MS onset is typically slow with symptoms steadily increasing over a period of months or years. The changes can be very subtle over a long period of time and some people experience long periods of relative stability. Indeed some consider the condition to be more ‘predictable’ than relapsing remitting MS, where a person can be struck down by a disabling relapse at any time without any prior warning.

“I have seen a slow but sure deterioration over the two years since I was diagnosed. I’m still walking, working full time, but I get more tired.”

Rosemary, W Sussex

Progression usually occurs in the same functional ‘system’ of the body, for example mobility and bladder function, both of which are caused by lesions in the spinal cord, whilst other systems such as vision and cognition tend to be unaffected in the majority of people.

Researchers have compared the course of primary progressive and secondary progressive MS to see if there are any major differences between the rates of progression. It was found they are in fact very similar, with the average rate of deterioration being the same. The time at which benchmarks such as needing assistance to walk are reached, were found to be related to the age of the person and were independent of any previous relapse history of the person with secondary progressive MS. This means that on average, regardless of the number of relapses they had previously experienced, a person who entered the secondary progressive phase at the age of 45 would progress at a similar rate to someone with primary progressive MS who experienced their first symptoms at the age of 45.

Predicting how primary progressive MS might progress

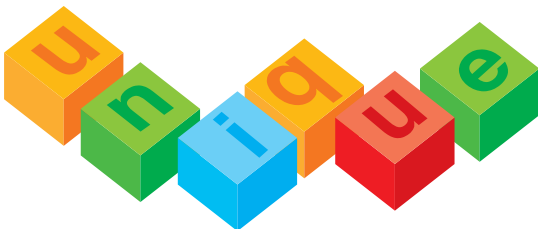
To see if it is possible to predict how someone's MS might progress studies have been carried out to look at the natural progression of primary progressive MS in several different populations. These are known as natural history studies. One such study published by Canadian researchers found that although 25% of their study group needed assistance to walk seven and a half years after their symptoms began, 25% were still able to walk unaided after living for 25 years with the condition.

The following factors have been found to influence the rate of progression:

1. **Initial rate of progression** – the Expanded Disability Status Scale (EDSS) is used in MS as a method of quantifying and monitoring changes in the level of disability over time. The first major 'landmark' on this scale is a score of 3.0 – 'mild' disability, but able to walk without any assistance. The longer a person takes to reach a score of 3.0 on the scale, the more likely it is they will have a slower rate of progression in the future.

2. **The number of symptoms experienced at onset** – someone whose symptoms are caused by damage to three or more different areas within the CNS, for example problems with walking, vision and cognition, are at risk for a higher rate of progression than someone with problems associated with damage in only one area, such as walking.
3. **Gender** – men generally progress at a faster rate than women.
4. **Number of brain lesions** – those with a lower number of brain lesions and less brain atrophy (a decrease in the amount of tissue in the brain) tend to do better.
5. **Amount of inflammation in the early stages** – people who had primary progressive MS for less than five years and had gadolinium enhancement of their lesions (a marker of inflammation) were found to have a greater level of disability and more brain atrophy than those without enhancing lesions.

Although the above factors have been shown to influence the rate of progression, it must be emphasised that every person is unique and these predictors will not hold true for everyone with primary progressive MS. There will be some men who progress at a slow rate and similarly women who progress more rapidly than the average. There will be people who reach a plateau in their condition after initially having a swift deterioration and so on.



7. Management of primary progressive MS

There is currently no disease modifying therapy that is available that can alter the course of primary progressive MS. Instead management focuses on the following three key areas:

- symptom management – this may either be drug treatment, rehabilitation, or a combination of the two;
- prevention of complications; and
- promoting general health and wellbeing to attain the best quality of life possible.

The symptomatic treatments and rehabilitation strategies used in primary progressive MS are the same as for other forms of MS. A multidisciplinary team of specialist health professionals may be involved in your treatment, these might include:

- **GP** – can help management of some symptoms, GPs also have a role in referring you to, and liaising with, other members of the specialist team;
- **neurologist** – involved in the diagnostic procedure and determining what medical treatment and rehabilitation may be appropriate;
- **consultant in rehabilitation medicine** – works with therapists and nurses to provide medical advice and coordinate services for people with a combination of symptoms and disabilities;
- **MS specialist nurse** – often the first point of contact to discuss your clinical issues, they also have a role in referring to other members of the specialist team to ensure continuity of care and liaising with the GP and primary care team;

- **rehabilitation nurse** – has a similar role to an MS specialist nurse, but is affiliated with a consultant in rehabilitation medicine rather than a neurologist;
- **physiotherapist** – involved in the assessment of movement to help improve mobility and may have a role in the management of symptoms such as pain and spasticity. They can also provide advice on exercise;
- **occupational therapist (OT)** – involved in assessing a person’s ability to perform daily activities. They have a role in teaching fatigue management strategies and can provide aids or adaptations if they are needed;
- **continence advisor** – specialises in managing bladder and bowel problems;
- **speech and language therapist (SLT)** – involved in assessing problems with speech and swallowing;
- **dietician** – assesses a person’s nutritional needs and provides advice on managing diet, particularly if symptoms have resulted in substantial weight loss or gain, or feeding has become difficult.

Drug treatment

The fact that there is currently no disease modifying drug therapy available that can alter the course of primary progressive MS is often a source of frustration for people with this type of MS. Instead drug treatment is aimed at alleviating the symptoms of MS as best as possible to enable you to carry out normal day to day living.

“One of my frustrations is that there appears to be no treatment for PPMS. I have tried LDN without success and currently take amantadine which has a mild effect on the fatigue.”

Robin, Surrey

It is impossible to mention all of the different drugs for all of the different symptoms in this book, but listed below are some of the drugs that are most widely prescribed for different symptoms. These are the same for primary progressive MS as for all other types of MS. Some treatments are only available in specialist centres and not all are licensed for use in MS but they may be prescribed off-licence at the discretion of the doctor/neurologist in some circumstances. GPs can be reluctant to prescribe some treatments unless they have been specifically recommended by a neurologist.

- **Spasticity and spasms:** baclofen, gabapentin, tizanidine, diazepam, clonazepam, dantrolene, carbamazepine, intramuscular botulinum toxin, intrathecal baclofen, intrathecal phenol, cannabis-based medicines;
- **Bladder problems:** oxybutinin, tolterodine, desmopressin, botulinum toxin;
- **Bowel problems:** docusate, lactulose, macrogol, bisocodyl, loperamide;
- **Sexual difficulties:** sildenafil, vardenafil, tadalafil;
- **Fatigue:** amantadine, modafinil;
- **Pain and sensory symptoms:** amitriptyline, pregabalin, imipramine, nortriptyline, carbamazepine, gabapentin;
- **Depression:** fluoxetine, citalopram, sertraline, amitriptyline, imipramine, duloxetine, phenelzine sulphate.

For some people tolerating the side effects of some of these medications, which can include nausea, sleepiness, weakness, or diarrhoea, can be a problem in itself.

There are some symptoms of primary progressive MS for which there are no specific drug treatments, for example speech problems and weakness. For some symptoms drugs may be of only limited benefit. Sometimes a drug aimed at another symptom or condition can be prescribed to give some symptom relief, for example anticonvulsant drugs (used in epilepsy) and antidepressants are often used to treat

nerve pain in MS as one of their effects is to alter the chemical transmission of pain thereby reducing pain symptoms.

“[I have tried] an assortment of drugs which all seemed to have unfortunate side effects (sleepiness, diarrhoea). I now have botox injections into my right leg which I feel is quite beneficial. I should have treatment every three months, but it is always four to five months before I get ongoing treatment.”

Sue, Derbyshire

Steroids and PPMS

Whilst steroids are sometimes used in relapsing remitting MS to speed up recovery during a relapse, it is generally accepted that steroids do not have a role in primary progressive MS. Although it is unusual for steroids to be prescribed for primary progressive MS, sometimes a one-off course may be given if a person begins to experience a period of deterioration, particularly if their movement is predominantly affected. However, there is no evidence that steroids can alter the long-term course of primary progressive MS and in some cases of advanced progressive MS a sudden deterioration has been reported after the use of intravenous steroids.

“I did have some steroid treatment initially, but was told I shouldn’t have any more.”

John, Oxfordshire

Rehabilitation medicine

The aim of rehabilitation medicine is to use all means available to help an individual to live their life as fully as possible, minimising the impact of physical, psychological or social complications. Rehabilitation is something that should accompany medical treatment, rather than being thought of as a last resort. Ideally rehabilitation management should be an ongoing rather than an episodic process, although there may be periods of intense therapy.

There is a huge range of possible therapy interventions. They are generally grounded in a patient's desire to continue to live their life in a certain way, or participate in a given activity, aiming for a clear goal. Optimum use of medication can be enhanced by positive lifestyle changes, psychological support if it is required and the coordination of specific therapies to meet individual needs.

“[It is] eight to nine years since I felt my first symptoms, I am still on my feet thanks to a regime of exercise and medication.

Charles, W Sussex

Although rehabilitation is not curative, it can help you discover abilities you didn't realise you still had. It can help you make the best of, and maintain, functions in order to manage symptoms, promote independence and above all maintain individual choice whatever your level of disability – therefore it is vital to ask for help sooner rather than later. A wide variety of strategies and therapies may be employed to help you find new ways to do things that have started to become difficult.

Some people with primary progressive MS will have access to a consultant in rehabilitation medicine and their team, whilst others will access services through their GP, neurologist or MS specialist nurse. Below are some of the rehabilitation therapies that may be used to help manage MS symptoms.

Physiotherapy

This is probably the most widely recognised of the rehabilitation therapies. A physiotherapist can carry out an assessment and design individual stretching and exercise programmes to help with a variety of symptoms such as mobility problems, spasticity and spasms, weakness, tremor, balance problems and dizziness. They can also teach relatives or carers to support a physiotherapy programme.

“A course of regular physiotherapy has helped with my core stamina and strength and I am beginning to manage my diet and fitness a lot better now.”

Tim, Dorset

Occupational therapy

Occupational therapy is designed to make day to day living more manageable. It can involve finding ways to save energy and minimise fatigue. Equipment may be provided to help with daily activities, for example grab rails in the bathroom, perching stools to enable you to sit whilst preparing food, or specialist cutlery to assist with eating. An occupational therapist will also be involved in assessing for mobility equipment such as sticks, crutches, walkers and wheelchairs if required. An occupational therapist may also be able to assess your work place and develop strategies to help you maintain or return to employment.

“Some while ago I did see an occupational therapist who gave me various aids.”

Pat, Suffolk

Continence advisor

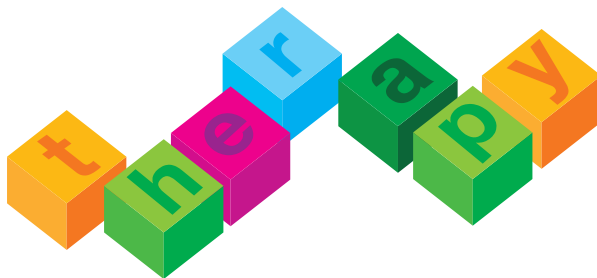
Continence specialists assess and advise people who have bladder or bowel problems. This can include advice about oral medications, teaching techniques such as clean-intermittent catheterisation or devising bladder retraining programmes.

Speech and language therapy

For those with difficulties with speaking, a speech and language therapist can assess the problem and devise exercises to strengthen and improve the muscles involved in speech and breathing. They can also advise on aids for communication. If swallowing difficulties are encountered, a speech and language therapist can suggest ways to minimise or avoid problems.

Nutritional therapy

A dietician may be involved if there are nutritional problems, for example if you have lost or gained a significant amount of weight. This may be due to mobility issues, loss of appetite, or feeding or food preparation difficulties. They can also give advice on different food textures and alternative methods of feeding if swallowing becomes difficult.



How the multidisciplinary team might work together towards a clear goal is demonstrated by the following example:

An individual wants to go to their daughter's wedding, but has problems with spasms as well as bladder and bowel issues. The neurologist or consultant in rehabilitation medicine might advise on controlling the spasms, a community physiotherapist might help the individual find better ways of getting in and out of a car, a specialist nurse or continence advisor could work with them on timing and management of their bladder and bowels, and one or more of the rehabilitation team could be involved in helping the person and their family develop more confidence.

Unfortunately, it can be difficult to access rehabilitation therapies on the NHS, particularly on an ongoing basis. In an ideal world everyone would have regular access to physiotherapy, but in reality a course of physiotherapy is typically for a fixed number of weeks and you may be limited to the number of courses you can access in a year. There can be an element of self-management in between treatment episodes, in terms of maintaining the motivation to carry on doing the suggested exercises or finding other resources that may be available to you, for example a local exercise class.

“There is a GP referral scheme to a local fitness centre in my area and I found it very helpful receiving a tailored workout regime. Unfortunately, I didn't stick with it.”

Alan, Highlands

Prevention of complications

Some of the symptoms of MS can make you more prone to complications such as bladder infections or pressure sores, which can worsen pre-existing symptoms or trigger new ones. The specialist knowledge of MS nurses, consultants in rehabilitation medicine and district nurses is particularly important in anticipating and preventing complications in progressive conditions.

Bladder infections can be particularly problematic in people who use catheters and it is important that a good, clean technique is taught and followed to reduce the likelihood of infection.

In advanced primary progressive MS maintaining the integrity of the skin is an important aspect of self care, otherwise what starts as a minor skin irritation could result in a pressure sore. Keeping the skin as clean as possible and well moisturised, and ensuring you keep moving regularly by repositioning yourself if you are in a wheelchair/bed on a regular basis, will minimise the risk of pressure sores forming.

General health and wellbeing

It is important to make the most of both your physical and emotional wellbeing to enable you to live your life to the fullest and enable you to do the things you enjoy as well as the things that you have to do.

Eating healthily and exercising regularly can help maintain a healthy weight, preserve muscle strength and flexibility, minimise fatigue and improve your mood amongst other things. However, symptoms such as spasticity and fatigue can make trying to do this on a regular basis seem even more of a challenge, but it is important to try and do as much as you are capable of, even if the changes are small. As well as potentially helping some of your MS symptoms, lifestyle factors such as taking some form of exercise, eating and drinking sensibly and not

smoking can reduce your susceptibility to other conditions such as type 2 diabetes, cancer and cardiovascular disease.

“The best therapy for me is exercise and doing things that I can get a feel good factor from.”

Tim, Dorset

Regularly attending preventative health screening programmes, such as those for breast, cervical, prostate and bowel cancer, as well as blood pressure and cholesterol checks, means that problems can potentially be caught early and less aggressive treatments might be able to be employed than if detected at a later stage. MS symptoms can worsen when you have an infection, so if you suspect you have a urinary tract, chest or other infection it is important to get it treated sooner rather than later. Regular dental check ups to help maintain good oral health and avoid complications such as gum disease, tooth decay and infections, which can arise as a result of taking some medications, are also beneficial in terms of your overall wellbeing.

Further information on maintaining your wellbeing can be found in section 9.

Complementary and alternative therapies

Many people with MS find that using complementary and alternative medicines (CAMs) not only helps to relieve some symptoms, but can also help to improve their sense of wellbeing. A diagnosis of MS may leave you feeling that you have lost control of your body, knowing that you are doing something for yourself can in itself be a positive step and give you back some sense of control.

People try many different therapies or approaches including: yoga, Pilates, Tai Chi, reflexology, massage, aromatherapy, acupuncture, homoeopathy, dietary changes, supplements, herbal remedies,

hyperbaric oxygen therapy, and relaxation or meditation techniques. Much of the evidence for the use of CAMs in MS is based on individual experiences rather than hard and fast scientific evidence. This is because it can be hard to conduct clinical trials that measure the effect of CAMs, especially in such a variable condition as MS.

“My nurse referred me to a homoeopathic hospital for my emotional problems; so far I have had good results from this.”

Tim, N Lanarkshire

“Acupuncture has improved my bladder. By doing these extra [things], it makes me feel good that I’m making the effort to combat my condition.”

Pauline, W Midlands

If you are considering trying a complementary or alternative therapy there are a few things to bear in mind. No therapy can currently offer a cure for MS, so be wary of anyone who makes this claim. CAMs are perceived as being ‘natural’ and ‘safe’, but this is not always necessarily true – some CAMs can interact with conventional therapies, so it is worth letting the health professionals involved in your care know if you are using any CAMs. It is also sensible to check that your CAM practitioner is properly trained and qualified to offer the therapy – are they registered with their professional association or regulatory body if they have one? Finally make sure you are aware of the potential cost of any treatments and how long they are likely to last.

8. Trials in primary progressive MS

Historically, primary progressive MS has been the least studied of the different clinical types of MS. The lack of trials, along with the fact that there is no treatment available that can modify the course of primary progressive MS, has understandably led to people feeling like they are on the periphery of research into new treatments, especially when compared to those with relapsing remitting MS. However, the situation has slowly begun to improve in the last decade since both the definition of primary progressive MS and the diagnostic criteria have been tightened. The increase in the amount of research being carried out, along with more information being produced that is tailored specifically for people with primary progressive MS, should in time help overcome the perception of being the ‘forgotten few’.

Challenges to trials

Accurate diagnosis

Recruitment to trials has been hindered in the past because of inadequate diagnostic criteria. The introduction of new criteria now makes it easier to give an accurate diagnosis of primary progressive MS earlier in the course of the condition, so it should be easier to identify participants for future clinical trials.

Relative rarity

Data that looks at the natural progression of MS is used to determine the number of people that are needed for a trial and how long the trial

should run to show that a treatment has a significant effect on the condition. Because primary progressive MS is a relatively rare condition, in the past the natural history data has tended to be combined with that of people with secondary progressive MS, so data solely on primary progressive MS has been limited. Earlier trials also tended to look at 'progressive MS' in its entirety, so they included people with secondary progressive MS rather than exclusively looking at the effects of a treatment in primary progressive MS.

The relative rarity of primary progressive MS means that even with today's improvements in diagnosis, recruiting the numbers needed for large multi-centre trials, particularly in the later stages of the drug development process, can be problematic.

Appropriate measures of effectiveness

The measures typically used in trials for relapsing remitting MS have been found to have limited suitability in primary progressive MS trials. They are either not relevant, or are not sensitive enough to the slow changes that occur in primary progressive MS. The outcome measures chosen to be used in future trials designed for primary progressive MS are going to be of utmost importance. It is thought that markers of tissue destruction, particularly those for axonal loss, may be more clinically relevant in primary progressive MS than the measures that have been used in the past. The outcome measures used in trials need to be able to show what effect treatments have on slowing progression or preventing further disability.

Targeting

In relapsing remitting MS, treatments have been aimed at modulating the immune system to reduce the number of relapses. In primary progressive MS, treatments need to be focused on targeting a different and more challenging process, that of progression itself. This means that neuroprotection and repair need to be considered, rather than, or as well as, immune modulation.

“Having PPMS, I have often felt like a ‘poor relation’ in the MS world...it is good that at last more attention is being given to PPMS which...presents particular challenges and an uncertain future.”

Maggie, Yorkshire

Past and current trials

There have been trials of several disease modifying treatments (DMTs) specifically for people with primary progressive MS including interferon beta 1a, interferon beta 1b, glatiramer acetate, mitoxantrone and riluzole. The majority of DMTs target inflammation; therefore they are of less relevance in primary progressive MS and have generally been shown to have a lack of efficacy. The trials have been justified because although there is less inflammation seen than in the other forms of MS, there is some inflammation.

More detailed analysis of data from a trial of glatiramer acetate for primary progressive MS that was stopped early due to a lack of efficacy, suggested that it may in fact have been effective in the small number of participants who had gadolinium enhancing lesions (a marker for inflammation) and that some of the men involved in the study may also have had some benefit. Some of the studies of the beta interferons and mitoxantrone have also shown a trend towards a slower progression rate, but they were not conclusive.

The majority of DMTs target the T-cells of the immune system, which have a role in recognising and killing ‘foreign’ invaders. It may be that treatments that target the B-cells of the immune system, which produce antibodies, may be of more benefit in primary progressive MS. Rituximab, a drug that reduces the number of B-cells, has been tested in primary progressive

MS. Although the overall trial results were disappointing, further analysis of sub-groups within the trial suggested that younger people with evidence of inflammation did have some response to the drug.

People with primary progressive MS have also been included in trials of cladribine, cyclophosphamide, haematopoietic stem cell transplantation, intravenous immunoglobulins, methotrexate and pirfenidine; these trials also included people with other forms of MS. None of these treatments were definitively proven to modify the course of primary progressive MS.

It has been suggested that a drug called fingolimod (Gilenya), which has been trialled in relapsing remitting MS, may have a direct effect on nerve repair (neuroprotective) and therefore could also be of value in primary progressive MS. A study began in 2009 in primary progressive MS to evaluate whether fingolimod tablets taken daily are effective at delaying disability progression. This is a three year trial and results are not expected until 2013 at the earliest.

Another trial involving people with primary progressive MS is the CUPID (Cannabinoid Use in Progressive Inflammatory brain Disease) trial. This trial is looking to see whether tetrahydrocannabinol (THC), one of the chemical compounds found in cannabis, can slow the development of disability in progressive MS. As well as finding out whether THC is an effective treatment, the researchers hope that this study will also give further insight into how best to carry out research into primary progressive MS. The trial is expected to complete in 2012.

Questions still to be answered

There are a multitude of questions about primary progressive MS that still remain unanswered, including:

- What is the significance of the lower levels of inflammation seen?
- Is the relationship between inflammation, demyelination and axonal loss different in primary progressive to the other types of MS?

- Are the axons more vulnerable in people with primary progressive MS?
- Why are the oligodendrocytes more susceptible to damage?
- To what extent do the abnormalities seen in both the normal appearing white and grey matter contribute to disability?
- Are some sub-groups of people with primary progressive MS more responsive to a certain treatment than others, for example those with more inflammation, or men more than women?

Future research priorities

Future research for primary progressive MS needs to primarily focus on neuroprotection and repair. Neuroprotection is aimed at protecting the axons from damage to prevent/slow further progression, whilst strategies that promote remyelination and regeneration are needed to encourage the recovery/repair of axons that have already been damaged.

Some of the areas that are currently being researched for primary progressive MS are:

- sodium channel blockers, eg lamotrigine - neuroprotection;
- glutamate agonists, eg riluzole - neuroprotection ;
- cannabinoids - neuroprotection;
- plasticity of the brain – the ability to re-route messages around damaged areas of the brain; and
- rehabilitation strategies to maintain function.

For the most up-to-date information, including drugs in development and trials that are recruiting; see the research pages on the MS Trust website: www.mstrust.org.uk/research

Primary progressive MS has encouraged researchers to focus on neuroprotection and repair, which should prove beneficial for all forms of MS, not just primary progressive MS.

9. Living with primary progressive MS

A diagnosis of primary progressive MS can introduce an element of uncertainty into virtually all aspects of a person's life, in terms of changing physical capabilities, the sense of loss for your life before MS and the unknowns that the future may hold. Although it is important to be a full participant in decisions relating to managing and treating your primary progressive MS, it can be all too easy to focus purely on managing your physical symptoms. It is just as important that you don't neglect your emotional health, which also has a major influence on your overall quality of life.

As previously mentioned, looking after yourself in terms of eating healthily, exercising when possible and attending 'wellness' check-ups where appropriate can improve your general wellbeing and have a positive impact on some symptoms, such as anxiety and depression. The following section looks at the emotional, social and financial impact of living with primary progressive MS in more detail.

The MS journey

The needs of people with primary progressive MS can be very different to those of people with the other types of MS. Because primary progressive MS is typically diagnosed at a later age, it is more likely that you are well-established in your working and home life. You may be anticipating a second career path, possible retirement, or taking up a new skill or activity. You may have been looking forward to getting fit, relaxing or travelling

more. If you have children they may be older and nearing adulthood. Or it may be that you regret not having had children and know that it is too late. If you have to give up work you may worry about whether you are ever going to be able to return to the job market in the future and how you are going to fill your days. Also if your financial situation changes significantly your concerns may centre on how you are going to cope and whether you will be able to maintain a reasonable standard of living. This can be especially worrying if you have a family and have historically been the main breadwinner.

The changing nature of primary progressive MS means that you may periodically have to adapt or readjust to your condition in different ways at different times. This can be emotionally draining in itself and can result in you feeling like life is a succession of emotional highs and lows, not only for yourself, but also the rest of the family as your expectations and plans change over time.

There are several points in your journey with primary progressive MS where you can find yourself having to rethink who you are and what your role is within your family, social or working life, these include:

- at diagnosis;
- when adjusting to new or worsening symptoms which may threaten or restrict your independence;
- if you decide to stop working;
- if any meaningful activity you enjoy is being affected.

These 'milestones' can all be triggers for changes to your emotional wellbeing, social confidence, or financial security.

Impact of diagnosis

Everyone has a picture of themselves; this might be a 'professional' view of yourself in terms of your job, it could be your role within your family or social circle, or a combination of all these things. A diagnosis of primary progressive MS means that this image could change and you have to redefine yourself in the future.

Everybody's reaction to diagnosis and their way of dealing with it is different and there is no right or wrong way to react. For some there is a sense of relief that at last they have a name to put to the cause of their symptoms, or that it is not something 'worse' like the brain tumour they had feared it might be. All the trips to the doctors have finally been justified and for some people who suspected they had MS there can in fact be a sense of closure. However, for many people a diagnosis of primary progressive MS will come as a great shock, especially if they had not considered MS, or if the diagnosis was given in an insensitive way. Whether primary progressive MS was suspected or not, the diagnosis can still be potentially devastating and some of the most common reactions include anger, fear, anxiety or even guilt. You may find you are uncertain of what you are feeling, or that your feelings fluctuate.

“The shock of being diagnosed is not easy to explain, we knew a little bit about MS, but nothing about PPMS. Lots of things run through your mind. How are we going to cope, the children, our future, and our lives will never be the same.”

Alison, Surrey

If diagnosis has taken a long time, or you have felt that health professionals haven't been honest with you or have withheld information, it is not unusual to feel angry. You may feel angry for no clear reason at all. Many people will wonder “Why me?” The fear of an unknown future, especially one where we are not able to control events, leads to anxiety: “Will I progress quickly and become very disabled?”; “Will I have to stop work”; “Am I going to lose my independence?”; “How are we going to cope?”

Unfortunately there aren't any hard or fast answers to any of these questions as everybody is affected differently, so it is not unusual to get completely overwhelmed at times. Often people feel a sense of guilt, they wonder if there is something they could have done that would have stopped them getting primary progressive MS, or if their children will get MS too. They worry that they are letting people down, that they are not contributing as they should be, or that they may become a burden in the future. These feelings are known as 'problem effects' and can make a situation more difficult than it need be.

“Firstly I felt relief that there was a diagnosis, but was very fearful as there was very little information about the possible course of the illness. I felt there was a great reluctance to ‘give bad news’, which hindered frank and free discussion.”

Catherine, Edinburgh

“When the diagnosis came around my life kind of imploded and I found it hard to re-engage with myself let alone other people. I felt like I had let everyone in my life down and the guilt almost got the better of me.”

Tim, Dorset

Grieving

It is only natural to need a period of time to come to terms with the diagnosis and many people liken it to bereavement. Just like bereavement there are different stages to work through and there is no set time for working through this process, it will be different for everybody. The first stage is the initial shock; feelings of numbness, relief, anger. The second stage is the process of mourning, the reality

of the diagnosis and what it might mean begins to sink in. Often people feel low, irritable or angry at this time. They can become withdrawn from family and friends and can be prone to sudden outbursts of tears or to mood swings. This is the time to grieve for your former self. The final stage is when you begin to come to terms with the diagnosis and begin to see life in a more positive way again. Although you may always feel a sense of loss for your former self, you slowly and gradually learn to accept and live with your MS. There can be some overlap between these different stages and you may find you move from one stage onto the next and then back again. Undoubtedly some days will seem worse than others, but only once you have gone through this process are you able to move on and begin the process of redefining yourself.

It is very important to remember that acceptance is not giving in or giving up. Resignation to a situation takes no effort, but adapting and accepting requires effort and work.

“My husband cried on the way home from the hospital and I was unable to. But looking back it was good as he came to terms with it and took over the day to day issues which allowed me to grieve in my own time. I still have not come to terms with it and probably never will.”

Alison, Surrey

“At the moment I’m slowly getting back on track, although the illness presents its challenges on a day by day basis, through hard work and determination I am now living with and managing my condition.”

Tim, Dorset

Impact on the family

It is important to recognise that a diagnosis of primary progressive MS doesn't just have an impact on the person who is diagnosed. The whole family is affected and will have different reactions and ways of dealing with the news – just like you, so remember there is no right or wrong way for them to react. Some may bury their heads in the sand and pretend it hasn't happened, whilst others want to find out everything they can about the condition. Some family members can go into 'take over' mode and try to do everything for you. Although this can be useful in the short term, giving you time to come to terms with the diagnosis, it may not be beneficial if they keep doing it in the long term. It can become a source of tension if you feel they are discouraging you from doing things that you feel you are still capable of. Sharing concerns and agreeing solutions is a healthy process. Those who love you will naturally want to help you and it can be hurtful if you constantly 'test' your family or reject their support.

“My husband found it very difficult to accept, and although he is now supportive has never read anything about MS and I do wonder at his reaction when I get really bad.”

Christine, Wiltshire

“This has been a rollercoaster and my family has suffered as a result. I have had periods of anger and others of just weeping.”

Tim, N Lanarkshire

Other members of the family may have different and potentially conflicting coping mechanisms to your own, which can be a challenge in itself. It can

be difficult to cope with the reactions and emotions of others when you are still coming to terms with your own feelings. The most important thing is to keep communicating and make sure everyone shares their feelings as much as they are able; hopefully this can prevent minor issues and niggles from escalating into major problems.

“My wife is brilliant and I seem to be good at looking at the positives of what I can do, not the negatives of what I can’t do. I think many people struggle to do this.”

Tim, Dorset

“My family have been very supportive, my son found it hard at first, but is coming to terms with it now.”

Debbie, Norfolk

Adapting to the presence of primary progressive MS in your life can be emotionally exhausting. It is important to acknowledge your feelings about having MS and how it influences your thoughts and how you behave. However, it is important not to let MS become the whole picture and to dominate all your thoughts, feelings and behaviours. Finding the right balance can be tricky and if you, or your family, find it is difficult then talking to an outsider can help. This may be through a support group of other people with MS, which can be in the form of face-to-face groups or online forums. However, support from other people with MS isn't for everybody, the prospect of meeting other people with more advanced MS than you can be frightening for some, also not everybody wants to join a 'club'. Some people find impartial professional help, such as formal counselling, to be beneficial.

“I’ve needed to draw on all my resources and have leaned heavily on my partner. I’ve realised that the ‘poor me’ response takes me nowhere and that realisation has been helpful and empowering. I’ve sought out emotional support from health professionals with mixed success.”

Alan, Highlands

“Belonging to MS clubs and organisations is so beneficial. Exchanging information and learning how to cope with this debilitating condition, keeps me going.”

Hilda, Surrey

“I have, rightly or wrongly, avoided joining local MS groups. I feel that I want to be amongst ‘normal’ people as long as possible and also think that I could quickly sink into a state of self-pity if I perceived myself as an invalid.”

Jane, Leicestershire

Coping with increasing disability and maintaining independence

Most people view ‘normality’ as being able to do what they want, when they want. Being able to go to places, enjoying social and intimate relationships and maintaining independence and dignity are

important to everybody. In the face of increasing disability it can be difficult to maintain a sense of normality.

“Everything goes too fast for me – is it MS, old age, or both? I get so frustrated, always remembering how much more quickly I could do things in the ‘good old days’ when I walked normally, held down a job and could venture anywhere, anytime.”

David, Leicestershire

“I sometimes feel like I am sitting on a ‘time bomb’. Will I stay the same or will the disease suddenly accelerate? I try not to think too much about the future and live day to day.”

Jean, Cornwall

When faced with a new or worsening symptom, you may find yourself going through the whole grieving process again. Fears for the future resurface: “What impact are these new changes going to have on my relationships?”; “How am I going to maintain my dignity in the light of difficult symptoms?”; “Am I going to need to use a wheelchair?” These fears can result in heightened stress, anxiety and when there is no motivation to re-invest in life there is a real risk of developing depression.

“There are times when I could sit and cry, but there’s nothing to be ashamed of for feeling that way.”

Vanessa, Lancashire

“I find life progressively difficult but try to be positive, which is the only real way to face MS I feel. I can’t tap dance any more – but I couldn’t in the first place!”

Robin, Norfolk

Depression

Depression is different to the lows or blues that everybody experiences at times; it is a persistent and pervasive low mood that lasts for a period of two weeks or more. The causes of depression are not fully understood and anyone can develop it any time in their lives. People with a long-term condition such as MS are more at risk for developing depression, as are older people who tend not to have the sense of immortality we have in our 20s and 30s. It is important to recognise that you may be at increased risk of depression, particularly at diagnosis and times of significant change in your condition. Depression can affect your sleep, you can lose interest in food, find it difficult to be motivated and this all has a negative impact on your energy levels and general wellbeing. Depression is treatable, so if you are able to recognise that it may be a problem, something can be done to help. The first step is to go to your GP and talk about it.

“I am profoundly depressed ...my self-esteem is on the floor and I simply can’t bear the sight of myself. The person I was is lost.”

Fiona, Surrey

Depression varies in its severity. Some cases are mild and may respond to ‘talking therapies’, these help you identify and overcome the negative thought patterns associated with depression. In more severe cases of depression, drug treatment may be recommended. Antidepressants can also be used in combination with counselling.

“Counselling has helped me climb out of periods of depression, and to come to terms with enforced changes in lifestyle ... it’s painful at times, but very beneficial having someone with professional skills and emotionally uninvolved, to help you identify problems and steer you towards dealing with them.”

Maggie, Yorkshire

Medication and mood

Emotional and cognitive changes, such as depression, mood swings, and problems with memory or word finding can all be associated with coming to terms with living with a long-term condition. However, they can also be side effects of the medications that are used to treat the symptoms of primary progressive MS, for example antidepressants and steroids can affect mood. It is important to seek help if you think that any of your treatments are having a negative effect on your mood. Tell your health professionals about all the medications you are taking as there may be alternatives that suit you better. Sudden withdrawal of some medications, for example baclofen for spasticity, can also lead to anxiety so seek advice if you are considering stopping any treatments.

Stress and anxiety

Stress is a normal part of everybody’s life, it can’t be eliminated completely, but it can be managed to some extent to lessen its impact. Learning to manage stress is particularly important for people with MS who often report that their symptoms feel worse when they are under increased stress.

“I suffer from anxiety attacks. I think this may be due to worrying about what I can no longer do.”

Mandy, Nottinghamshire

The first step in managing stress is identifying what causes you to become stressed. Maybe it is new social situations, looming deadlines at work or the fact you are not working, or even large family gatherings. Often the event is not the primary cause of stress, but rather our reaction to it. In the majority of cases it will be impossible to avoid the stressful situation altogether, but it may be possible to develop strategies to manage the circumstances better. Prioritisation is important. Although we all have to do the things that are necessary, are there other things that can wait? Can you make the time to do the things you enjoy and give your life more balance? Maybe you need to be more assertive and learn to say “No!” in certain circumstances. Perhaps you could relax your standards a little bit if you have a tendency to aim for perfection all the time. Stress and anxiety can be helped with techniques such as yoga, Tai Chi, meditation or relaxation. Taking five minutes out to perform some breathing exercises may help put things back into perspective.

“I have learned to meditate which helps me mentally and helps me to cope.”

Christine, Wiltshire

Maintaining independence

Needing to use adaptive equipment, such as a stick or wheelchair, or aids to help with day to day living such as getting in and out of the bath, may become a reality if your ability to carry out everyday tasks is reduced. This can be difficult to come to terms with as they are very visible indicators of your condition, which previously you may have managed to keep fairly well hidden. Choosing not to use assistive equipment, perhaps due to embarrassment or an inability to accept help, could result in you becoming withdrawn and socially isolated, which impacts negatively on your wellbeing. Try to look on using aids as a way of maintaining your independence, rather than as a negative thing. Aids can have an important role in allowing you to retain social

contact and do the things that you enjoy doing for a longer time. Maybe you thought you were going to have to stop walking the dog, but using a stick or mobility scooter could be a way of continuing to take pleasure in this activity.

“Recently my physio gave me a walking stick, I was reluctant to take it due to embarrassment as I’m 33, but I use it when I really have to, to give me a little more independence. When shopping I know my partner would steer me away from clothes/shoes, now I can totter over with my stick!”

Laura, Essex

Although it is not probably something you particularly want to think about, especially if you are currently managing well, it can be beneficial to plan ahead and learn about options for resources and services that you may need in the future; then if you do have a crisis it may be less stressful as you already have the relevant information to hand.

“In my experience, NHS and social services tend to be reactive rather than proactive, so one needs to know what to ask for. Other people’s hard-won knowledge is vital.”

Michael, Surrey

Changing family roles

Things you have always done may become more difficult, or you may have to give up some things you have always done. Maybe there are things you had anticipated that you would do in the future, such as helping care for your parents or a partner as they age, or looking after grandchildren, that may no longer be possible. A partner or other family

member may have to take on these tasks and it can be stressful whilst everyone learns to adapt. It is important when family roles change to keep the communication channels open to keep potential conflict to a minimum, especially as individual feelings may be out of sync. A partner may feel overburdened; whilst you may feel frustrated that they are taking over and doing things that you still want to do. A breakdown in communication means problems are unlikely to get resolved.

“My husband tends to do too much for me and is unwilling to leave me to do simple things – he won’t let me assist with meal preparation, even though I could for instance chop vegetables.”

Linda, Northamptonshire

Recognising and addressing problems can make a major difference to the dynamics of your relationships. Some people find it useful to get the whole family together to set out their expectations and develop coping, communication and problem solving strategies, but this approach doesn’t suit everybody. Others opt for external help, counselling can work on improving coping and problem solving skills, as well as communication skills and increasing self-awareness. Remember, relationships are the responsibility of everyone in the relationship, not just the person with MS.

A major role change that may have to be considered at some point is whether a partner or other family member is going to take on a caring role. This is not something that everyone is willing, or able, to take on. They may not be able to face the intimate tasks that caring can involve, or it may be that they are the main breadwinner in the family and need to keep working. In these circumstances it can be better for everyone concerned if professional help is sought. Nobody should have to be a carer if they don’t want to be.

“I took the decision from an early time that I was not going to keep my complaint hidden and most people have been supportive. However, my wife was not able to fully accept the situation and we were divorced.”

Richard, Gloucestershire

If a partner chooses to take on a caring role, whether this is on a full or part-time basis, there are lots of things to consider. It can be easy to lose your identity as a couple as some partners begin to feel more like a parent than a partner, especially if they are providing regular hands-on care. It can also be a struggle to maintain sexual attraction in this situation which can result in a loss of intimacy. This may be compounded if sexual dysfunction is experienced as part of the MS. Fatigue can also be a big issue, both for the person with MS and the caregiver, and romance may be the last thing on your mind. There are also potential financial implications if a partner has to reduce their working hours, or stop working to spend more time caring, so make sure you are aware of any benefits that you are entitled to claim.

“I am frightened by what is likely to happen to my mobility and how dependent I will become on my husband.”

Julie, Surrey

Taking care of a loved one can be both physically and emotionally draining. The caregiver often neglects their own health and wellbeing as they are so preoccupied with looking after the other individual. There is also the potential to become socially isolated if they have a full-time caring role. It is important they take time out where possible to do something they enjoy and have some balance in their own life if they are to avoid physical, mental or emotional burnout.

For some people, very advanced primary progressive MS can mean they have a level of disability that requires more specialised care than a relative is able to provide. Although there may be care services that can help a person remain in their own home as long as possible, in some circumstances the level of care required may mean they have to move out of the family home. This is never an easy decision for anybody to make, but planning ahead and finding out about the options, resources and services that are available can make it a less stressful experience if it ever has to be faced.

Working with primary progressive MS

For the majority of people, work is a big part of their lives. It provides a means to be financially independent, gives structure to your day and is a source of social contact outside your immediate circle of family and friends. It also forms part of your identity, and often your self-esteem and confidence is tied up with your 'professional image' of yourself.

When you are initially diagnosed with primary progressive MS it can be a big shock and everything can seem rather overwhelming. However, at this early stage it is impossible to determine how your MS is going to affect you and if, or how, your symptoms might impact on your ability to continue working. The effect symptoms have will also vary depending on the type of work that you do, for example, for those with a desk bound job that requires them to be creative, problems with walking may not be too much of an issue, whilst difficulties with concentration and memory could prove more challenging.

“I still work full-time although [my] mobility is poor and slow.”

Vanessa, Lancashire

Although it can be difficult to see how you may cope in the future, making a decision regarding work at a time of crisis may not prove to be the best choice in the long term. Many people find that if they take the decision to stop working at diagnosis, or when they experience a sudden deterioration in their condition, that they come to regret it later. It may be more prudent to wait until the crisis is over and you are in a better position to explore all the options available to you.

With the exception of a few professions, such as the armed forces, you are not obliged to inform your employer about your condition unless there are specific health and safety risks. However, if you choose not to disclose your MS, it makes it difficult to access any adjustments that might enable you to stay in the workplace for longer or make your job more manageable. An employer who is sensitive to your needs with some knowledge of MS, including the impact of 'invisible' symptoms such as fatigue; and who is willing to make accommodations such as allowing you to reduce or change your hours, work from home, or change your role if appropriate is key to your working successfully with primary progressive MS. It may be useful to seek advice and support from an occupational therapist, MS nurse, colleague or union representative if you decide to disclose your diagnosis, wish to request changes to your working conditions or are experiencing difficulties at work. It is also useful to be knowledgeable about your employment rights, particularly the Equality Act (which has replaced the Disability Discrimination Act).

“My ability to carry out my work deteriorated to the point that I took early retirement as it was too unsafe for me to carry on as a Building Inspector.”

Richard, Gloucestershire

“I am wheelchair dependent and get tired easily, but I still manage to work four days a week.”

Elizabeth, W Midlands

For some people, choosing to stop working could be the right decision. This has implications both for the person with primary progressive MS and the rest of the family. You may wonder “What am I going to do with my life now?” The loss of routine and identity that work provides can be difficult for some people, especially if your social circle begins to contract. You may find that there are changes of role in your relationships; a partner may have to go back to work or increase their hours. Conversely, if a partner decides to take on some caring responsibilities they themselves may have to stop work or reduce their hours.

For the majority of people there will be some financial implications to stopping work, which can lead to additional stress. Planning ahead may help lessen financial concerns. Don't just resign - get advice, this could include finding out if taking early retirement on medical grounds is an option and familiarising yourself with any benefits that you are entitled to receive. Organisations such as the Citizens Advice Bureau, or your MS nurse, may be able to help you with any application forms.

Of course stopping work doesn't have to be a negative thing. It may give you the time to do something at last that you've always wanted to do, such as take up a leisure activity, new hobby or volunteering opportunity. Maybe you have wanted to spend more time with family or friends, but work got in the way. You may find that through pursuing new interests your social network actually increases.

“I had to take early retirement because of the MS. Since then my social life has blossomed ... I am up front with my limitations and find everyone is helpful. I have made many new friends and acquaintances.”

Christine, Wiltshire

Maintaining social relationships

Everyone recognises the importance of human contact, having good family and social networks and something to look forward to is central to our sense of wellbeing. Although some casual acquaintances and friends may drop out of your lives if it becomes increasingly difficult to do things together, the people who are most important to you are likely to be supportive and remain good friends.

“One of the joys of the condition has been the kindness and consideration I have received from so many people. Amazing!”

Jane, Leicestershire

“You just have to work round the disability; our friends have been very supportive.”

Glynis, Lancashire

It is important to remember that there is a poor understanding of MS amongst the general population, so people tend to make assumptions about what a person with MS can and can't do. Learning to ask for help when it is required, or how to decline offers that aren't needed tactfully, can be a skill in itself. You may also want to develop strategies

to deal with unwanted or intrusive questions, or to acknowledge your limitations in a way that you feel comfortable with.

“Lurching in to a gathering on a rollator, I am the spectre at the feast. They all want to help or feel sorry – I just want to blend in unobtrusively.”

David, Leicestershire

I hate friends asking if I'm alright, can I manage? Treat me as normal – I'll tell you if I can't or don't want to do something.”

Rosemary, W Sussex

Consider disclosing hidden symptoms such as fatigue to friends. Constantly turning down last minute invitations to social occasions might mean the invitations eventually dry up, but if your friends are aware of the impact fatigue has on you they may not only be more accepting if you decline or pull out at the last minute, they also might consider giving you more notice to enable you to conserve energy so you are more likely to be able to attend a night out. Don't forget you can also suggest doing something that is easier for you to manage, for example if going to your favourite restaurant is proving difficult, maybe you could invite everyone round for a takeaway instead, or if you have enjoyed long walks in the past suggest a route that is more accessible for you so you don't miss out.

“I really miss walks with friends. My MS limits the sort of things we can do together. Yesterday four of us went to the beach and the other three had a long walk without me – we all felt it.”

Alan, Highlands

“Sometimes I’m sad at what I no longer have, but this is balanced by being able to enjoy the slower, simpler things in life.”

Janet, Worcestershire

For many people being diagnosed with a long-term condition such as primary progressive MS provides them with an opportunity to reassess or reflect on their life. Although it invariably means life may not turn out as you had planned, you may find that it makes you more self aware. It can give you a chance to focus on the people and things in life that are most important to you. Your appreciation of your relationships with both family and friends and the smaller things in life may be enhanced as a result of your diagnosis. It may give you an opportunity to change career path, live life at a slower pace, try a new activity, or make new friends and acquaintances.

Final thoughts

Being diagnosed with primary progressive MS sets you on a lifelong journey, along the way there will be periods where the water is calm and periods when it seems choppy. Learning to find and use all the resources available to you will help you manage your MS more effectively and live your life as fully as possible. Some of these resources will be internal, like your inner strength, problem solving or setting yourself goals. Others will be external like finding the information you need and being aware of what services are available to you at any particular point in your journey. This is known as self-management and you are probably doing it all the time without even realising it. Self-management can help you face the challenges that living with primary progressive MS can bring by enabling you to better control the things you can control and deal with those you can't.

Self-management doesn't mean 'going it alone'. Support from family, friends and the health professionals you are in contact with will help you make the decisions about lifestyle and treatments that are right for you. There are also free courses, such as the Expert Patient Programme which is available in England and Wales, that can help provide support and develop your self-confidence so you feel more in control.

The internet can also be a valuable self-management tool as it can be useful for 'fishing' for information. However, it is important to bear in mind that the quality of sites, particularly where health-related information is concerned, does vary widely in terms of the accuracy, consistency and independence of the content. Your MS nurse may be able to point you in the direction of some reliable sites. Another benefit of the internet is that it can be a good way to maintain social contact if you are finding it increasingly difficult to get out and about. Also by being able to carry out tasks like online shopping, it can help you save time and energy so you can do some of the more fun things in life!

“I haven't an illness, it's an inconvenient condition and I feel the main way to help yourself is a positive mental attitude.”

Gillian, Flintshire

“When I have a problem I put the kettle on and make a cup of tea. Over the past years I must have drunk buckets of tea! Sometimes you have to take two steps back and look at the bigger problem before moving two steps forward.”

John, Cumbria

“Obviously there are many different challenges to overcome; the worst is dealing with the news, but also the effect on the rest of the family and friends. The main thing you have to accept is your life does not stop, it just runs a different path. Laughter and the future are still there, they do not stop.”

Alison, Surrey

“I live life in a fast manner; it’s just that at the moment I have moved into the slow lane.”

Vanessa, Lancashire

“I would advise anyone to read all the information available and share this with your family and friends. They will be your strength and support along with the professionals who will be there to guide and advise you.”

Rosalind, Cardiff

“It all comes good in the end, after a period of acceptance.”

Martin, Dorset

“My best experiences of health care professionals since being diagnosed with primary progressive MS have been those where I’ve felt listened to and understood. I’ve come away feeling stronger, clearer. I’ve been helped to process my bundle of losses leading me towards the goal of acceptance, re-framing, discovery of new abilities, new senses of autonomy, perhaps restored or new faith, hope even. So don’t tell me there is nothing you can do for someone with primary progressive MS!”

Alan, Highlands

10. Sources of support

Other MS Trust publications you may find helpful

Books

- Are you sitting comfortably? A self-help guide to good posture in sitting
- At work with MS
- Falls: managing the ups and downs of MS
- Living with fatigue
- MS and me: a self-management guide to living with MS
- MS explained
- Tips for living with MS

Factsheets

- Bladder problems
- Bowel problems
- Cognition
- Depression
- Diet
- Fingolimod
- Functional electrical stimulation (FES)
- Low dose naltrexone (LDN)
- Pain
- Spasticity and spasms

Exercise resources

- Move it for MS: a DVD of exercises for people living with MS. £1
- Exercises for people with MS: an online resource. The exercises are arranged in categories based both on how they are done and the problems they address www.mstrust.org.uk/information/exercises/

Websites

- The MS Trust website: a reliable source of information about symptoms, treatments and more www.mstrust.org.uk
- MS services: a map of local services and MS specialist centres www.mstrust.org.uk/information/services
- StayingSmart: a website designed for people who want to understand how MS can affect thinking, memory and concentration www.stayingsmart.org.uk

Online support groups

MS People UK

An online community for those with MS, their families and friends
www.ms-people.com

Jooly's Joint

An online community of people who live with MS
www.mswebpals.org

Benefits and legal advice

Benefits Enquiry Line

Confidential advice and information on the range of benefits available and how to claim them **Tel: 0800 882 200**

Disability Benefits Helpline

Advice on Disability Living Allowance and Attendance Allowance
Tel: 08457 123 456

Department of Work and Pensions

The government department that oversees the benefits system, the website has information on the range of benefits available www.dwp.gov.uk

Citizens Advice Bureau

Offers free, confidential, impartial and independent advice, including benefits, legal matters and employment rights www.citizensadvice.org.uk

Equality and Human Rights Commission

The Equality and Human Rights Commission is a good source of advice if you feel you may have been discriminated against at work or elsewhere www.equalityhumanrights.com

Getting around

Blue Badge Scheme

Information on the Blue Badge Scheme, including eligibility, where it can be used and how to apply for a Blue Badge

www.direct.gov.uk/en/DisabledPeople/MotoringAndTransport/Bluebadgescheme/DG_4001061

Shopmobility

A federation of more than 250 schemes providing scooters and wheelchairs for shoppers with mobility problems www.shopmobilityuk.org

Motability

A national UK charity that helps disabled people and their families become more mobile www.motability.co.uk

Forum of Mobility Centres

A network of independent organisations who offer information, advice and assessment to individuals with a medical condition that may affect their ability to drive www.mobility-centres.org.uk

Equipment and adaptations

Disabled Living Foundation

Provides information on disability equipment, day to day household gadgets, new technologies and training techniques www.dlf.org.uk

Assist UK

Provides impartial information about products and equipment for easier living through more than 40 local centres www.assist-uk.org

Remap

Federation of volunteer organisations that provide custom made aids for people with disabilities www.remap.org.uk

AbilityNet

Offers advice, support, assessment of needs and the supply of adapted computer equipment for people with any sort of disability www.abilitynet.org.uk

Foundations

Has information on local Home Improvement Agencies, which can offer help with finding funding and managing home improvements www.foundations.uk.com

Relationship support

Relate

Offers advice, relationship counselling, sex therapy, workshops, mediation, consultations and support face-to-face, by phone and through the website www.relate.org.uk

British Association for Sexual and Relationship Therapy

Professional body for sexual and relationship therapists. Can provide a list of accredited therapists in your area www.basrt.org.uk

www.mstrust.org.uk

Health and social care information and carer support

Arranging health and social care

Includes information on accessing health and social care assessments, direct payments, professional carers and help with health costs

www.direct.gov.uk/en/DisabledPeople/HealthAndSupport/ArrangingHealthandSocialCare/index.htm

Caring for someone

Information to support carers, including caring and support services, carers' rights, looking after yourself, respite and care homes

www.direct.gov.uk/en/CaringforSomeone/index.htm

Princess Royal Trust for Carers

Provides information, support and practical help to carers through more than 100 independently managed Carers Centres

www.carers.org

Crossroads Association

Provides practical support to carers in the home and with respite care

www.crossroads.org

Carers UK

Provides information and advice to carers www.carersuk.org

Holidays

RADAR

Produces a book called The Open Britain Guide, a guide to accessible accommodation in the UK and Ireland

<http://radar-shop.org.uk/Range.aspx?id=1>

Tourism for All

Provides information on accessible accommodation of different types in the UK and abroad, includes a directory to search accessible accommodation, attractions, respite care and services

www.tourismforall.org.uk/

Disability Holidays Guide

Provides information and a holidays search facility for people with a disability www.disabilityholidaysguide.com/

MS Trust information service

Helping you find the information you need

We hope that you have found this information helpful. The MS Trust offers a wide range of publications, including a newsletter **Open Door**, which provides an ongoing update on research and developments in MS management. In addition it contains articles from people with MS and health professionals.

For a full list of MS Trust publications, to sign up for Open Door and much more visit our website at www.mstrust.org.uk

If you would like to speak with someone about any aspect of MS, contact the MS Trust information team and they will help find answers to your questions.

Freephone

0800 032 3839

(Lines are open Monday – Friday 9am-5pm)

email

infoteam@mstrust.org.uk

write

MS Trust

Spirella Building, Letchworth Garden City
Herts, SG6 4ET

All our services are provided free within the UK. If you have found this information valuable, please help us to help others; any donation, however small, is much appreciated.

Please contact the MS Trust information team if you would like any further information about the reference sources used in the production of this publication.

telephone: 0800 032 3839



Multiple Sclerosis Trust
Spirella Building, Bridge Road
Letchworth Garden City
Hertfordshire SG6 4ET

T 01462 476700

E info@mstrust.org.uk

www.mstrust.org.uk

Registered charity no. 1088353