

Multiple
Sclerosis
Trust

MS

Primary progressive multiple sclerosis



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We hope you find the information in this book helpful. 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.

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MS Trust information service

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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

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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
	<ul style="list-style-type: none">• Clinical features• Pathological features	
4.	Diagnosing primary progressive MS	17
	<ul style="list-style-type: none">• Diagnostic criteria• Tests that may be carried out to diagnose primary progressive MS	
5.	Symptoms of primary progressive MS	26
	<ul style="list-style-type: none">• Symptoms commonly seen in primary progressive MS• Other symptoms that may be experienced• Complications and other concerns	
6.	Progression	34
	<ul style="list-style-type: none">• Predicting how primary progressive MS might progress	
7.	Management of primary progressive MS	37
	<ul style="list-style-type: none">• Drug treatment• Rehabilitation medicine• Prevention of complications• General health and wellbeing• Complementary and alternative therapies• Self-management	

- 8. Trials in primary progressive MS 49**
- Challenges to trials in primary progressive MS
 - Past and current trials
 - Questions still to be answered
 - Future research priorities
- 9. Living with primary progressive MS 57**
- The MS journey
 - Impact of diagnosis
 - Coping with increasing disability and maintaining independence
 - Maintaining social relationships
 - Working with primary progressive MS
 - Needing more help
 - Final thoughts
- 10. Sources of support 78**
- 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 (periods where symptoms flare up) and remissions (periods of good or complete recovery); instead their symptoms gradually worsen from the outset (progression) – this is known as primary progressive MS. It is estimated that 10-15,000 people have primary progressive MS in the UK.
- Around 10-15% of people with primary progressive MS experience a single relapse at some point, where their symptoms flare up and then resolve, but the predominant feature of their MS is progression.
- 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 – where the severity and frequency of relapses decrease or even stop altogether, and symptoms gradually worsen.
- 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, a protective reaction mounted by the immune system in response to damage to the tissues, is seen to a lesser extent in primary progressive MS lesions than in relapsing remitting or secondary progressive MS, but there is more damage to the nerves themselves. It is this nerve damage that is thought to be a major contributor to the non-remitting, 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 other conditions 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 generally 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 a person make the best of, and maintain, functions that will help them manage their symptoms and live life as fully as possible.
- It is important a person makes the most of both their physical and emotional wellbeing to enable them to do the things they enjoy as well as the things they have to do.

There is no preventative action that could or should have been taken to prevent a person 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)
- **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 following the results of an international survey, the National Multiple Sclerosis Society Advisory Committee on Clinical Trials of New Agents in MS 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 periods of remission, instead progression occurs from the outset. The rate at which progression occurs differs 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. A small proportion of people with primary progressive MS do experience a single relapse at some point in their condition, where symptoms rapidly worsen in the absence of an infection, this is then followed by recovery back to where the individual was before the relapse.

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 have worsened slightly following a relapse, there isn't a marked increase in the level of a person's disability between relapses.

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'; with early attacks that are so mild that they are dismissed or overlooked, or the 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, the rate of progression is difficult to predict for 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, as they do not experience relapses which 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

Primary progressive MS is typically diagnosed when people are in their 40s or 50s, though it can be diagnosed earlier, or later, than this. Relapsing remitting MS is generally diagnosed at a younger age, usually between the ages of 20-40, with the average age at diagnosis being 30.

Comparing primary progressive with secondary progressive MS, the age of onset is similar, i.e. 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

Similar numbers of men and women are affected by primary progressive MS, whereas with relapsing remitting MS more women than men are diagnosed.

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, become more noticeable and result 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

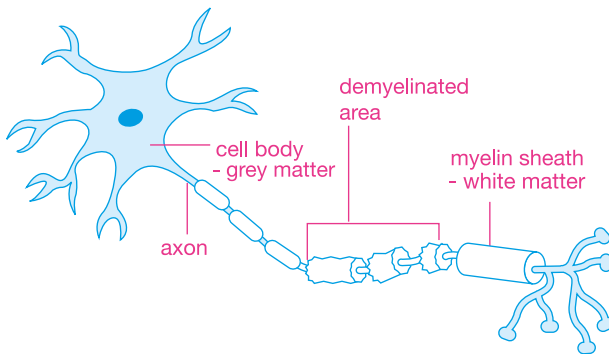
Distribution of lesions in the central nervous system

The central nervous system (the brain and spinal cord) is made up of white matter and grey matter:

- white matter consists mainly of nerve axons, the long slender fibres that conduct electrical impulses away from the nerve cell body. Axons are surrounded by a protective coating, the myelin sheath
- grey matter consists predominantly of the nerve cell bodies.

The white matter passes messages between different areas of grey matter, the role of myelin is to protect the nerve axon and help speed the messages along.

Nerve cell



In MS, the myelin sheath surrounding the axons is attacked by the immune system and is damaged or destroyed - this is called demyelination. Once the attack stops, scar tissue can form over the area of damaged myelin, called a plaque or lesion, which can be seen on an MRI scan. 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. The number and size of lesions in the spinal cord in primary progressive MS mirrors that seen in secondary progressive MS.

Of all the groups of people with MS, those with primary progressive MS have the largest discrepancy between the number of lesions seen and their level of disability, so what else could be responsible for the degree of disability associated with primary progressive MS?

Inflammation versus axonal damage

When tissues in the body are damaged or become infected, the immune system responds by causing a protective reaction known as inflammation. Although inflammation is seen in the lesions in primary progressive MS, it is more typically seen in the early stages of the condition, and it is present to a much lesser extent than in both relapsing remitting and secondary progressive MS.

It has been suggested that the inflammation seen in progressive MS may be of a different nature to that seen in relapsing remitting MS. One theory is that inflammation in progressive MS may be driven by B cells (white blood cells which are programmed to recognise a specific virus or bacterium), rather than by T cells (white blood cells that have a role either in attacking and destroying infected cells, or that can produce molecules that tell other elements of the immune system how to act).

Although there is less inflammation, there is more damage to the nerves themselves in primary progressive MS lesions than is seen in either relapsing remitting or secondary progressive MS. Nerve damage, or neurodegeneration, can also be seen in areas of white and grey matter that actually appear to be normal on an MRI scan. These subtle abnormalities in the normal appearing white and grey matter have been shown to increase over time. It is widely believed that this diffuse damage to the nerves 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 repair (or 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 also 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 can be even more challenging as symptoms usually develop 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. These conditions 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 more than 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 an individual's clinical history and a neurological examination. With the advent of MRI in 1983, the neurologist Charles Poser formulated the 'Poser criteria'. These were the first criteria that could be used to determine whether a person had possible, probable or definite MS and were used for nearly 20 years.

In 2001, the 'McDonald criteria' were proposed by another neurologist, Ian McDonald, which were more specific, more sensitive and allowed for earlier diagnosis in many cases. They also included criteria specifically for diagnosing primary progressive MS. These criteria have subsequently been further simplified. 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.

Diagnostic criteria for primary progressive MS (2010 Revisions to the McDonald Criteria)

Primary progressive MS may be diagnosed in subjects with:

1. One year of disease progression (determined retrospectively or prospectively).
2. Plus **two** of the following criteria:
 - evidence for dissemination in space in the brain based on one or more T2 lesions in at least one area characteristic for MS
 - evidence for dissemination in space in the spinal cord based on two or more T2 lesions in the cord
 - a positive cerebrospinal fluid (CSF) test.

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
- 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, because 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, so the dissemination in space aspect of the criteria can be supported more readily.

Excluding early relapse activity

For some people with MS, early attacks or relapses might have been so mild that they were overlooked or dismissed at the time, or they may have happened so long ago they have been forgotten about. A thorough clinical history needs to be taken to determine whether early relapses actually 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 the normal brain changes that are seen as we age. 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 worsening stiffness and/or weakness in the legs (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
- 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 other conditions 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 one year of progression to secure a diagnosis.

Tests that may be carried out to diagnose primary progressive MS

Several tests can be used to help secure a diagnosis of primary progressive MS, though the majority of people will only have to undergo a few of these tests. 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

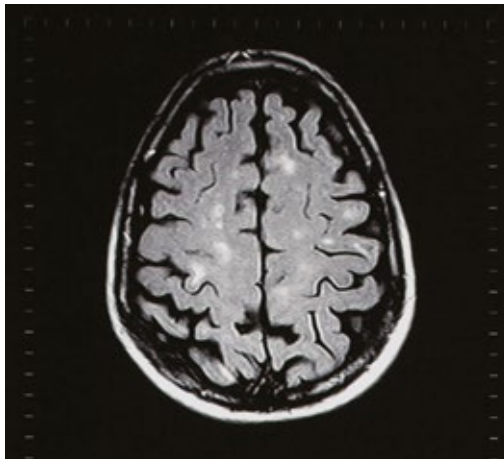
The neurologist may carry out simple tests on movement, balance, coordination, reflexes and vision that can suggest whether MS is a cause of symptoms. These tests may also indicate, or give clues to, where in the central nervous system the damage has occurred.

Magnetic resonance imaging (MRI)

MRI uses magnetic fields and radio waves 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. T2 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 in the brain 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. 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. These 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

In MS, immune cells cross the blood brain barrier – a layer of cells that usually keeps cells in the blood from coming into

contact with the central nervous system - and attack myelin, this results in higher levels of a protein called immunoglobulin (IgG) being present in the cerebrospinal fluid (CSF) which surrounds the brain and spinal cord than would be expected.

A hollow needle is inserted into the base of the spine and a small amount of the CSF is drawn off. 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. The number of white blood cells in the CSF may also 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 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

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. None of the following symptoms are exclusive to primary progressive MS.

Symptoms commonly seen in primary progressive MS

Mobility problems

The majority of people experience a gradual stiffness and/or weakness in both of their legs 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

Weakness in the limbs is due to poor conduction of messages by the damaged nerves in the spinal cord, 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, the residual tension in your muscles when they are completely relaxed, 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 affect the function of the bladder and/or 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 the 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. Psychological issues such as body image or self-esteem can also affect sexual function in MS. Other issues not related to MS, such as the menopause and some medications, can also contribute to sexual difficulties.

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. They include problems with thinking (cognition), balance issues, visual problems, sensory problems, tremor, speech and swallowing difficulties, depression and anxiety.

Cognitive problems

Cognitive problems can include difficulties with attention, memory, spatial reasoning and the ability to find words (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 individual 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

Balance and posture (cerebellar syndrome)

The cerebellum is the area of the brain that controls movement, balance and posture. This area is affected in around 9% of people with 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.

Vision and hearing (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. Damage in this area can lead to problems such as double vision (diplopia) or a flickering or jerking of the eyeballs on movement known as nystagmus. Damage to nerves in the brainstem is seen in around 1% of those with primary progressive MS.

Optic neuritis

Optic neuritis is the inflammation of the optic nerve – which carries visual information from the eye to the brain – 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

Neuropathic (or nerve) pain can be experienced in MS as a result of damage to the nerves. Types of nerve pain include

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

Musculoskeletal pain can be experienced if there is damage to the muscles, tendons, ligaments or soft tissue. This type of pain can be caused by alterations in the way an individual walks, or if the limbs are stiff and kept in a fixed position for long periods of time. Musculoskeletal pain often arises as a result of spasticity or weakness.

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 usually don't have relapses, it is possible to experience a temporary acute deterioration in function due to other factors such as pain or an infection.

Complications and other concerns

Some symptoms can arise as a consequence of the problems that MS brings, 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 (see section 7).

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 also create concerns around emotional, social and financial issues. This may be as a result of the psychological adjustments you have to make to come to terms with living with a progressive condition, possible changes of role within the family or 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 affected 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

In primary progressive MS, often only one area or ‘functional system’ of the body is affected. For example mobility and bladder difficulties often go hand in hand, as they are both caused by lesions in the spinal cord. Progression usually occurs within a single system for an individual, so if problems are predominantly with mobility, systems associated with areas such as vision and cognition will tend to be less affected. However, there are some individuals who will have more than one system affected.

Although the rate of progression in any individual is unpredictable, 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 between the two types. It was found they are in fact very similar, with the average rate that symptoms increase 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 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 in people with primary progressive MS:

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 Canadian study showed that the longer a person with primary progressive MS took to reach a score of 3.0 on the scale, the more likely it was they had a slower rate of progression in the future.

2. **The number of symptoms experienced at onset** – individuals whose symptoms are caused by damage to three or more different areas (or systems) 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 system, 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 have a slower rate of progression.
5. **Amount of inflammation in the early stages** – people who had primary progressive MS for less than five years and had active lesions (as shown by gadolinium enhancement - 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

The management of primary progressive MS 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
- promoting general health and wellbeing to attain the best quality of life possible.

There is currently no disease modifying therapy that is available that can alter the course of primary progressive MS.

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 individuals to, and liaising with, other members of the specialist team
- **neurologist** – makes the diagnosis of MS and determines what medical treatment and rehabilitation may be appropriate
- **consultant in rehabilitation medicine** – works with therapists and nurses to provide medical advice and coordinate rehabilitation services for individuals with a combination of symptoms and disabilities

- **MS specialist nurse** – often the first point of contact after diagnosis to discuss any 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 – a group of health professionals that are usually based in a GP surgery or health centre, includes practice nurses and district nurses
- **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
- **dietitian** – 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

None of the disease modifying treatments that are used in other forms of MS have been found to be effective in primary progressive MS. Instead drug treatment is aimed at alleviating the symptoms of MS as best as possible to enable individuals to carry out normal day to day living.

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. 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, Sativex
- **Bladder problems:** oxybutinin, tolterodine, desmopressin, botulinum toxin
- **Bowel problems:** docusate, lactulose, macrogol, bisocodyl, loperamide
- **Sexual difficulties:** sildenafil, vardenafil, tadalafil
- **Fatigue:** amantadine
- **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, for example fatigue, so they may not be tried until other avenues have been explored such as a fatigue management course. 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 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

Steroids are sometimes used in relapsing remitting MS to speed up recovery during a relapse, it is unclear how they work but they are thought to act by suppressing the immune system and/or reducing fluid accumulation around the sites of nerve damage. Sometimes a one-off course may be given in primary progressive MS if a person begins to experience a period of deterioration, particularly if their movement is predominantly affected, but it is generally accepted that steroids do not usually

have a role in primary progressive MS. Steroids are rarely used in primary progressive MS because they can in fact make things worse, there have been some reports of a sudden deterioration after the use of intravenous steroids in some cases of advanced progressive MS. There is also no evidence that steroids can alter the long-term course of primary progressive MS.

“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.

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 exercises suggested by a physiotherapist at home, 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

There is a huge range of possible therapy interventions. They are generally grounded in a person'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

Like other treatments for MS, rehabilitation is not curative, but it can help an individual discover abilities they didn't realise they still had. It can help make the best of, and maintain, functions in order to manage symptoms, promote independence and above all maintain individual choice whatever the 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 a person 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 aims 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 sitting 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 an individual’s work place and develop strategies to help maintain, or facilitate a 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 medication, 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 dietitian may be involved if there are nutritional problems, for example if a person has 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.

Prevention of complications

Some of the symptoms of MS can make you more prone to complications 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 MS. For those who use catheters it is important that a good, clean technique is taught and followed to reduce the risk 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 individuals keep moving regularly by repositioning themselves if they are in a wheelchair/in bed on a regular basis, will minimise the risk of pressure sores forming.

General health and wellbeing

It is important for individuals to make the most of both their physical and emotional wellbeing to enable them to live life to the fullest and do the things they enjoy as well as the things they have to do.

Eating healthily and exercising regularly can help maintain a healthy weight, preserve muscle strength and flexibility, minimise fatigue and improve 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 to lifestyle are small. As well as potentially helping some MS symptoms, lifestyle factors such as taking some form of exercise, eating and drinking sensibly and not smoking can reduce 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. Less aggressive treatments might be able to be employed than would be needed if these conditions are detected at a later stage.

MS symptoms can worsen when you have an infection, so if a urinary tract, chest or other infection is suspected 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 are also beneficial in terms of overall wellbeing.

Further information on maintaining 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 mindfulness, relaxation or meditation techniques. Much of the evidence for the use of CAMs 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.

Self-management

Being diagnosed with primary progressive MS sets you on a 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!

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.

8. Trials in primary progressive MS

Historically, primary progressive MS has been the least studied of the different clinical types of 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. In 2012, the **Progressive MS Alliance** was formed (previously known as the International Progressive MS Collaborative). This is a global initiative, involving the collaboration of organisations from around the world, who are working together to find answers that will lead to new treatments and ultimately, end progressive MS.

In 2013, over 170 MS researchers, clinicians and industrial partners gathered to discuss challenges and strategies to accelerate innovative research focused on progressive MS. Five key areas of research were identified:

- developing better experimental models for progressive MS
- identifying outcome measures to be used in trials that are better able to show the effects treatments are having on progression
- identifying new, or existing drugs currently used for other conditions, that could be effective therapies for progressive MS
- clinical trial strategies that give an early indication whether an intervention will be effective (known as proof-of-concept)
- symptom management and rehabilitation.

Following a further meeting in 2013, the Alliance announced two funding initiatives and put out a request for research proposals. Projects were due to be selected and funded in early 2014. More information on the Progressive MS Alliance can be found at www.endprogressivems.org

Challenges to trials in primary progressive MS

Accurate diagnosis

Recruitment to trials was hindered in the past because the older diagnostic criteria were inadequate at differentiating between the types of MS. The introduction of new criteria 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

The relative rarity of primary progressive MS has caused problems for research trials:

- 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 are determined by looking at data that charts the natural progression of MS, this is known as natural history data. 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 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

- 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 outcome measures typically used in trials for relapsing remitting MS have limited suitability in primary progressive MS trials. They are either not relevant, for example measures to document number of relapses, or they 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 progression

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 strategies to protect the nerve axons from further damage, and to repair nerve damage that has already occurred, also need to be considered.

“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 of the disease modifying treatments that are used in relapsing remitting MS in people with primary progressive MS, including interferon beta 1a, interferon beta 1b, glatiramer acetate, mitoxantrone and riluzole. The majority of these drugs are designed to inhibit the inflammation that is thought to cause damage to the myelin of the nerves. Because inflammation is less of a feature in primary progressive MS, these drugs are of less relevance and have generally been shown not to be effective. The trials have been justified because although there is less inflammation seen than in the other forms of MS, there is some inflammation.

- One trial of glatiramer acetate for primary progressive MS was stopped early due to a lack of effect. However, following a more detailed analysis of the data, it was suggested that the drug 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.

People with primary progressive MS have also been included in trials of cladribine, cyclophosphamide, haematopoietic stem cell transplantation, intravenous immunoglobulins, methotrexate and pifenidine; 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.

The majority of disease modifying therapies in MS target the T cells of the immune system, which have a role in recognising and killing 'foreign' invaders. However, it is now being

increasingly recognised that the antibody-producing B cells of the immune system may also have a role in causing inflammation and eventually nerve damage in MS. Therapies that target B cells have started to be tested and have led to improvements in people with MS. The influence of antibodies on tissue damage is also being investigated. B cells have been found in areas of the brain associated with progressive MS, so it may be that treatments that target B cells 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.
- A small study tested riluzole (a glutamate receptor blocker), which is used to treat a form of motor neurone disease. Glutamate is a chemical involved in transmitting messages between nerve cells, excessive glutamate can lead to the loss of nerve axons. The study looked to see whether riluzole could protect the nerves in MS, but no definite effect was found.
- Another trial involving people with primary progressive MS was the CUPID (Cannabinoid Use in Progressive Inflammatory brain Disease) trial. This trial looked to see whether tetrahydrocannabinol (THC), one of the chemical compounds found in cannabis, could slow the development of disability in progressive MS. As well as finding out whether THC was an effective treatment, the researchers hoped that this study would also give further insight into how best to carry out research into primary progressive MS. Overall, the study found no evidence to support an effect of THC on MS progression. However, there was some evidence to suggest a beneficial effect in participants at the lower levels of disability at the start

of the study, but further studies looking at larger numbers of people would be needed to confirm this. One of the main findings of the trial was that the study participants progressed at a slower rate than had been expected, which may have affected the researchers ability to detect clinical change. This finding has implications for the design of future studies.

- The treatment natalizumab (Tysabri) is currently used in some people with relapsing remitting MS. A small study has looked at the effect of the drug in progressive MS and included some patients with primary progressive MS. The study looked to see whether markers of inflammation and tissue damage were reduced rather than at clinical changes. The drug was shown to reduce inflammation in the space around the brain and spinal cord (intrathecal space) and tissue damage. Further studies are required to see if these findings translate into a clinical effect.
- Fingolimod (Gilenya) is a treatment licensed for relapsing remitting MS, studies indicated that as well as its effects on cells of the immune system, it may also lessen nerve cell damage and enhance the repair of myelin, therefore it might also be of value in primary progressive MS. A study began in 2009 to evaluate whether fingolimod is effective at delaying disability progression in people with primary progressive MS. The study is due to finish in September 2014.
- Masitinib is a drug that targets mast cells, which are involved in allergy and inflammation, reducing the immune response and inflammation. In a small study of the drug in primary progressive MS, participants showed an improvement on the Multiple Sclerosis Functional Composite score (MSFC). The MSFC assesses walking ability, hand and arm coordination, and mental function. Although improvements were seen,

they were not statistically significant, but a larger study is now under way to see if the results can be replicated. The study is expected to fully complete in December 2015.

- A monoclonal antibody that targets a marker on the surface of B cells called ocrelizumab is also being tested in primary progressive MS to see if it can slow progression. B cells are now thought to influence the abnormal immune response that causes the immune system to attack myelin. This trial is not due to finish until 2017.

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 damage to the nerve axons different in primary progressive to the other types of MS?
- Are the axons more vulnerable in people with primary progressive MS?
- Why are the myelin-producing cells (oligodendrocytes) more susceptible to damage?
- To what extent do the abnormalities seen in 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 or slow further progression, whilst strategies that promote remyelination and regeneration are needed to encourage the recovery and repair of axons that have already been damaged.

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

- plasticity of the brain – the ability to re-route messages around damaged areas of the brain
- rehabilitation strategies to maintain function.

For the most up-to-date information on drugs in development 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 – changing physical capabilities, the sense of loss for 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 physical symptoms. It is just as important that you don't neglect your emotional health.

As previously mentioned, looking after your health in terms of eating a balanced diet, exercising when possible and attending wellness check-ups where appropriate can improve 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.

If you have to stop working, you may worry about whether you will be able to return to the job market in the future, how you will cope financially and whether you will be able to maintain your current standard of living. This can be particularly worrying if you

have a family and have historically been the main breadwinner – the needs of children will differ depending on whether you have a young family, or your children are older and nearing adulthood.

You may have been anticipating a second career path, taking up a new skill or activity, or if you are older possibly considering retirement. Plans for getting fit, relaxing or travelling more may have to be altered.

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, for example if you are no longer able to drive.

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

“I felt that the diagnosis gave me membership of a club – not a club I would elect to join, but one that did offer some benefits. I now knew what was wrong with me and could plan accordingly.” Clive, Berkshire

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 if 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. This 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. 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 at 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 changes, such as depression or mood swings, can be associated with coming to terms with living with a long-term

condition like MS. However, they can also be side effects of the medications that are used to treat symptoms, for example 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 perhaps large family gatherings. Sometimes the thought of the situation causes us to become stressed, rather than the situation itself.

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, mindfulness, 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 certain tasks – maybe altered sensations in the hands make preparing and cooking food more challenging, or balance issues mean you are no longer able to play football with the children. 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 trying whilst everyone learns to adapt.

It is important when family roles change to keep the communication channels open, this will help 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.

Maintaining social relationships

Everyone recognises the importance of human contact. Having good family and social networks and things to look forward to are 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 on the whole 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 telling friends about hidden symptoms such as fatigue. 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.

“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

Working with primary progressive MS

For many people work is a big part of their life. 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 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 support to help you to stay in the workplace for longer or make your job more manageable.

The following can be key to your working successfully with MS. An employer who is:

- sensitive to your needs, with some knowledge of MS, including the impact of 'invisible' symptoms such as fatigue
- willing to make accommodations such as allowing you to reduce or change your hours, work from home, or change your role if appropriate.

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 find out about your employment rights, particularly the Equality Act, which legally protects people from discrimination in the workplace and in wider society.

**“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. However, you need to carefully consider the

implications, both for you and your family, before reaching that decision. 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 worries. 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 health professionals such as 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 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

Needing more help

If your MS becomes more advanced, you may need to consider 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.

If a partner chooses to take on a caring role, whether this is on a full-time 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 be a struggle to maintain an interest in sex 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 both of 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 may neglect 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 and make caring a successful experience.

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.

Final thoughts

“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
- Managing your bladder: a guide for people with MS
- Managing your bowels: a guide for people with MS
- MS and me: a self-management guide to living with MS
- MS explained
- Tips for living with MS

Factsheets

- Cognition
- Depression
- Diet
- Functional electrical stimulation (FES)
- Pain
- Spasticity and spasms

Exercise resources

- Move it for MS: a DVD of exercises for people living with MS
- 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

Disability Benefits helpline

Advice on Disability Living Allowance (DLA)
Tel: 0845 712 3456

Attendance Allowance helpline

Advice on Attendance Allowance (AA)
Tel: 0345 605 6055

Personal Independence Payment helpline

Advice on Personal Independence Payment (PIP)
Tel: 0845 850 3322

Department of Work and Pensions

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

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.gov.uk/blue-badge-scheme-information-council

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

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.nhs.uk/CarersDirect/guide/practicalsupport/pages/overview.aspx

Carers Direct helpline

Provides information to support carers

Tel: 0300 123 1053

Carers Trust

Provides information, advice and services for people caring at home for a family member or friend

www.carers.org

Carers UK

Provides information and advice to carers

www.carersuk.org

Holidays

Open Britain

A guide to accessible accommodation in the UK

www.openbritain.net/

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 holiday search facility for people with a disability

www.disabilityholidaysguide.com/

About the author

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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.

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Primary progressive MS

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