CHOICES

PRIMARY PROGRESSIVE MS (PPMS)

What is Primary Progressive MS?

Approximately 10% of people with MS are told they have Primary Progressive MS – a form of MS where from the first or primary symptoms, the progression of the disease is continuous from the outset. Symptoms get continually worse over time rather than having attacks and relapses.

Primary Progressive MS is usually diagnosed in people in their 40s and 50s (older than the average age for diagnosis of relapsing remitting MS) although in some cases it can be diagnosed in those younger or older than this.

How does Primary Progressive MS differ from Relapsing Remitting MS?

The most obvious difference between Primary Progressive MS (PPMS) and the more common Relapsing Remitting MS (RRMS) is the absence of relapses.

Nerve cell body
Axon (nerve fibre)
Nucleus
Myelin sheath
Nerve-cell ending
NERVE CELL
Taken from the BMA Illustrated Medical Dictionary

RRMS is caused by inflammatory attacks on the myelin – a protective layer which covers the cells of the central nervous system. With PPMS there is much less of this inflammation and people with PPMS tend to have fewer brain lesions than those with RRMS. Any lesions they do have tend to have fewer inflammatory cells. PPMS does however tend to cause more axonal damage.

Generally, those with PPMS tend to have more lesions on the spinal cord than in the brain.

Just as many men as women have Primary Progressive MS whereas there are approximately three times as many women with Relapsing Remitting MS as men.

What are the symptoms of Primary Progressive MS?

The symptoms of Primary Progressive MS are much the same as any other form of MS and these include:

- Fatigue
- Optic Neuritis (Loss of vision in one eye, blurred or double vision)
- Weakness of limbs
- Reduced co-ordination
- Balance problems
- Numbness, pins & needles
- Spasticity (stiffness or spasms in muscles)

Multiple Sclerosis Resource Centre

Multiple Sclerosis Resource Centre Tel: 01206 226500 or email: info@msrc.co.uk
www.msrc.co.uk   MS Counselling Service: 0800 783 0518 & press1

As with any form of MS, some symptoms are more common than others and it is rare for any two people with MS to have the exact same symptoms. However, because those with Primary Progressive MS tend to have more lesions on the spinal cord, problems with walking are very common. In a recent survey we conducted, 90% of people with PPMS said they had some form of mobility problems. This usually starts with slight problems, which could include difficulty getting up the stairs or running, tripping up if not paying attention when walking or needing to rest their legs as they start to get weaker. Another common symptom affecting mobility is drop foot - a difficulty or inability to properly lift the toe of the foot. A functional electrical stimulation (FES) device can be used to help alleviate this symptom. An FES stimulates the muscles which lift the foot, effectively doing what the nerve should do. It can be used on one leg or both.

Generally however, there appears to be no clear difference between the symptoms in different types of MS.

How is Primary Progressive MS diagnosed?

It can take some time to get a positive diagnosis of any form of MS. This is because the common symptoms of MS can also be due to many other conditions. If MS is considered a possibility, you will be referred to a neurologist who will look at your medical history and examine you. You are then likely to undergo a number of tests including:

Magnetic Resonance Imaging (MRI) Scan – this will identify any areas of the brain or spinal cord that may have scarring.

Lumbar Puncture – A lumbar puncture (also know as a spinal tap) is used to take a sample of spinal fluid to test for abnormalities.

Evoked Potentials – simple tests that measure the time it takes for the nerves to respond to electrical stimulation.

The evidence required for a positive diagnosis of Primary Progressive MS according to the McDonald criteria:

Positive result from a sample of spinal fluid

and

Dissemination in space demonstrated by:

- MRI evidence of 9 or more T2 brain lesions
- or 2 or more spinal cord lesions
- or 4-8 brain and 1 spinal cord lesion
- or positive result from Evoked Potentials test together with 4-8 MRI lesions

or positive result from Evoked Potentials together with less than 4 brain lesions plus 1 spinal cord lesion

and

Dissemination in time demonstrated by:

- MRI

or continued progression for 1 year

This means that diagnosis of Primary Progress MS can be a lengthy process,

especially if symptoms have only just begun.

How will Primary Progressive MS develop?

Many people think that the progression of PPMS will be rapid, their symptoms will increase and their condition will worsen quickly. This is not always the case. For some people their progression will be rapid; however, generally PPMS onset is slow and symptoms steadily increase over a number of years. The way PPMS progresses varies from person to person. Although in the long-term symptoms do generally get worse, it can take quite some time for this to happen and there may be times when there are no noticeable changes to the progression.

How can Primary Progressive MS be treated?

Drug Therapies

The disease-modifying drugs (DMDs) currently available to Relapsing Remitting MS patients are unfortunately not effective in treating Primary Progressive MS. This is because they target inflammation, reduce relapses and reduce the number of new lesions forming. As PPMS causes less inflammation, a more gradual loss of nerve fibres and fewer lesions, the DMD's available are not effective in slowing the neural damage that takes place in those with PPMS.

There are however ways in which you can manage your symptoms. There are a number of Drug Therapies available to treat your symptoms. The most commonly used ones are detailed below:

Spasticity

Baclofen - Baclofen is a muscle relaxant which treats spasms, cramping and tightness of muscles. Common side effects are drowsiness and a feeling of muscle weakness. It is usually administered orally; however, for severe spasticity that cannot be managed with oral medication, it can be administered by an implanted pump - Intrathecal Baclofen.

Dantrolene - Dantrolene is also a muscle relaxant which is taken orally. Common side effects include dizziness and diarrhoea or constipation.

Bladder Dysfunction

Oxybutynin - Oxybutynin is used to treat urinary frequency, urgency and incontinence and works by relaxing the muscles of the bladder and stopping sudden muscle contractions (spasms). This helps control the release of urine. Common side effects include dry mouth, constipation & dizziness. It is usually administered orally.

Detruisitol (Tolterodine) - Detruisitol also works by controlling the muscle spasms of the bladder. Common side effects include dry mouth, constipation & dizziness. It is usually administered orally.

Fatigue

Amantadine - A G.P. can prescribe Amantadine for fatigue. It is an antiviral medication which can improve muscle control and reduce muscle stiffness. It is general-

ly well tolerated by people with MS but it can cause insomnia, nausea, dizziness, loss of appetite and dryness of the mouth.

Modafinil - Modafinil is generally used to treat narcolepsy. It is not specifically licensed for use in MS; however it can be prescribed by a Neurologist. Research has shown that it can be an effective treatment for fatigue in some people. It is generally well tolerated, with only mild side effects such as headaches.

Tremor
Clonazepam - Clonazepam is an anticonvulsant which is usually taken orally to treat tremor. Common side effects include drowsiness and dizziness.

Pain
Gabapentin - Gabapentin is an anticonvulsant drug which is used to treat neuropathic pain (pain resulting from damage to the nervous system). It is taken orally and common side effects are drowsiness and muscle tremor

Pregabalin (Lyrica) - Pregabalin works in the brain and the nerves to help relieve nerve pain. It is taken orally and common side effects include dry mouth and constipation.

For more detailed information about drug treatments available, please refer to our Choices leaflet 'Drug Therapies' .

Low Dose Naltrexone (LDN)

LDN has been used in the treatment of MS in the USA since 1985 but it is relatively new in the United Kingdom. Naltrexone is a drug referred to as an opiate antagonist. Its normal use is to treat opiate drug addicts addicted to drugs such as heroin; however, when taken to treat MS, the dose is much smaller. Those taking it have experienced a range of benefits, including reduced spasm and fatigue, improvements in bladder control, heat tolerance, mobility, sleep, pain, tremor and other symptoms. The two main symptoms that appear to improve most significantly are muscle spasm and fatigue.

"I take 3mg of LDN daily. I think it helps, though the symptoms are still there of course. On the odd occasion I forget to take it the pain is considerably worse the next day" Isabella

For more detailed information about LDN, please refer to our Choices leaflet 'LDN'.

Complementary Therapies
Many people with MS have benefited hugely from using complementary and other therapies. Complementary therapies can help MS; sometimes improvement is dramatic, sometimes less so. In the majority of cases people generally *feel* better.

Some of the more common therapies include:

Acupuncture - the use of very fine needles, which puncture the skin to reach an acupuncture point. Acupuncture can be used to treat pain and tension, spasticity eye problems, bladder urgency and fatigue.

Chiropractic - gentle manipulation to treat disorders of the joints, ligaments and muscles. In MS, it can help improve the function of the joints, relieve pain and muscle spasm.

Reflexology - a complementary therapy where gentle pressure is applied to specific areas of the feet or hands, pressing what are know as reflex points which encourage the body to heal itself naturally. By working on these points, blockages can be released to help restore the free flow of energy around the body. Tension can also be eased and circulation improved.

HDOT (High Dose Oxygen Therapy) - breathing pure oxygen while under increased air pressure. Many people find relief from their symptoms such as poor balance, fatigue, mobility problems, incontinence and speech problems. Many people benefit from a reduction in the severity of symptoms and improved quality of life.

"I use HDOT once a week and this has enabled me to virtually stop using painkillers on a regular basis and now I only use them when really necessary. Although this treatment was not prescribed as it is deemed as therapeutic but not a proven one, it works for me." Kathy

For more detailed information about alternative or complementary therapies, please see our Choices leaflet 'Complementary & Other Therapies'

Exercise

Most people with MS, regardless of their degree of disability, can benefit from some exercise. Many people with MS think they cannot exercise because they will become too fatigued but providing you exercise sensibly your energy levels are likely to increase. Good forms of exercise include Physiotherapy, Swimming, Yoga, Tai Chi and Pilates.

"I used to be very active and climb and mountaineer, which of course I can't do anymore but I can go to the gym as I can sit between exercises – no treadmill of course but I can do enough to feel like at least I'm getting some exercise." Philip

For more information about these forms of exercises and how they could help you please read our Choices leaflet 'Exercise'.

Diet & Nutrition

A number of people with MS have found that by having a restricted diet, they can manage some of their MS symptoms. A popular choice is 'Ashton Embry's Best Bet Diet (BBD)' which aims to minimise the chance of inflammation and autoimmunity in

MS by avoiding certain foods and by taking certain supplements. It is also recommended that you undertake an ELISA test to test for food intolerances. There is an extensive section about this diet on our website; however, a short summary is listed below.

Avoid:

- Dairy
- Gluten
- Legumes
- Refined sugar
- Eggs and Yeast

Take various supplements including Calcium, Magnesium, Omega 3 and Vitamin E.

For more information, see our Choices leaflet 'Diet and Nutrition'

Vitamin D

Some research suggests that there is a link between MS and Vitamin D deficiency. It is important Vitamin D3 is taken and not D2. This is because D3 is the natural form and not a synthetic supplement. It is recommended that a daily Vitamin D3 supplement of 5000IU is taken. However, exposure to ultraviolet B (UVB) radiation in sunlight is the most effective way to boost Vitamin D supply and as little as 15 minutes in the sun a day will increase Vitamin D levels without increasing the risk of cancer. Below is an extract from a recent consensus statement released by a number of societies including Cancer Research UK:

" *This consensus statement represents the unified views of the British Association of Dermatologists, Cancer Research UK, Diabetes UK, the Multiple Sclerosis Society, the National Heart Forum, the National Osteoporosis Society and the Primary Care Dermatology Society.*

Vitamin D is essential for good bone health and for most people sunlight is the most important source of Vitamin D. The time required to make sufficient Vitamin D varies according to a number of environmental, physical and personal factors, but is typically short and less than the amount of time needed for skin to redden and burn. Enjoying the sun safely, while taking care not to burn, can help to provide the benefits of Vitamin D without unduly raising the risk of skin cancer. Vitamin D supplements and specific foods can help to maintain sufficient levels of Vitamin D, particularly in people at risk of deficiency. However, there is still a lot of uncertainty around what levels qualify as "optimal" or "sufficient", how much sunlight different people need to achieve a given level of Vitamin D, whether Vitamin D protects against chronic diseases such as cancer, heart disease and diabetes, and the benefits and risks of widespread supplementation."

A positive attitude

Many people that we have spoken to who have PPMS have said that a positive attitude has helped them to deal with the challenges that PPMS presents.

"My PPMS stops me from working, and is disabling, but has afforded me many

"My PPMS stops me from working and is disabling, but has afforded me many reasons to be thankful for it! It has enabled me to understand people and life much better. MS has enabled me to be more tolerant, both with myself and other people." Susan

"The main thing is to remain positive and keep at least trying to do things that are important to me – a challenge in itself sometimes." Philip

New Developments

There are some ongoing trials for new treatments of Primary Progressive MS:

Fingolimod (Gilenya) – Fingolimod is a new oral treatment under trial in the UK to treat both Relapsing Remitting MS and Primary Progressive MS. It has already been approved in the US & Canada for the treatment of Relapsing Remitting MS and is currently being tested for effectiveness for Primary Progressive MS. Fingolimod blocks white blood cells called lymphocytes from circulating in the body, preventing them from reaching and causing damage to the brain, spinal cord and optic nerves. The current study (INFORMS) to evaluate whether Fingolimod is effective in delaying disability progression in those with PPMS suggests that Fingolimod may have a direct effect on nerve repair. The study is not due to finish until December 2013.

CUPID (Cannabinoid Use in Progressive Inflammatory brain Disease Trial) – The CUPID trial is a long-term research trial looking at whether one of the active ingredients in cannabis can slow the progression of Primary Progressive MS. It will also assess the long-term safety of cannabis-based medicines. The trial involves 493 people around the UK and is due to end in 2012.

What assistance am I entitled to?

Financial Support

If you find that you condition is worsening and you are unable to work or can not continue the job you were doing, then you can claim for certain benefits including Disability Living Allowance if you are unable to work or Employment Support Allowance if you have limited work capability.

For more information about these and other benefits that you may be entitled to please see our Choices leaflet 'Benefits & MS'

You may find that if your condition worsens, your home is no longer suitable for you and you may need to make adaptations, or you may need additional care and support. Social Services have a duty under the Community Care Act 1990 to make a health and social care assessment of your needs and also those of your carer.

You have a legal right to be able to obtain care services support which:

(1) Enable you to live in your own home
(2) Enable you to retain as much independence as possible, whether at home or in residential care
(3) Are tailored to your individual needs

For more information, see our Choices leaflet 'Social Services'.

Common Misconceptions

PPMS is the worst type of MS – PPMS is not the 'worst' type of MS. The progression rate of PPMS varies greatly from person to person and although it is true that for some people, the condition can progress very rapidly, for others progression is gradual. Many of those with primary progressive MS feel their condition is less unpredictable than Relapsing Remitting MS, where a relapse can happen at any time and can vary in severity and length.

"The decline has been very gradual. I manage my symptoms by keeping positive and being lucky enough to have a nice home and lovely wife helps! I find reflexology weekly helps my speech." Ralph

I will end up in a wheelchair – Not everyone with PPMS will have to use a wheelchair. Although there are those that have had to use a wheelchair full time within a number of years, there are also people who have had PPMS for over 20 years who still do not need to use a wheelchair.

"I have had PPMS for 13 years. I can walk short distances using sticks, for longer distances I use a manual wheelchair. I have physio every Thursday which I believe helps. I also do daily exercises at home." Alan