Multiple System Atrophy (MSA)

is the current name for disorders once known individually as striatonigral degeneration, sporadic olivopontocerebellar atrophy, and the Shy-Drager syndrome.

Multiple System Atrophy (MSA) is a progressive neurological disorder that affects adult men and women. MSA is caused by degeneration of nerve cells in specific areas of the brain. This cell degeneration causes problems with movement, balance and automatic functions of the body such as bladder control.☐☐

How common is MSA?☐☐

Until recently MSA was thought to be a very rare disorder. As we learn more about the condition, it becomes easier to recognise and diagnose. Recent research suggests that MSA affects 5 people per 100,000. This means that there may be up to several thousand people with MSA at any time in the country.☐

☐Who has MSA?☐☐

MSA usually starts between the ages of 50-60 years, although it can affect people younger and older than this. MSA does not appear to be hereditary and is not infectious or contagious. It is a sporadic disorder that occurs at random.

☐What does MSA mean?☐☐

The term Multiple System Atrophy was first used in 1969, although it has been called many things including; olivopontocerebellar atrophy, striatonagral degeneration, Parkinson's plus and Shy-Drager Syndrome. The confusion caused by this variety of names led to an international consensus by medical experts in 1996 to use the term Multiple System Atrophy (MSA).☐☐Broken down MSA stands for:

Multiple - many☐
System - brain structures that control different functions☐
Atrophy - cell shrinkage or damage☐☐

This means that cells are damaged in different areas of the brain that control different body functions. The three areas affected are the basal ganglia, cerebellum and brain stem. These areas are responsible for movement, balance and automatic body functions like bladder control.

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How are the nerve cells in the brain damaged in MSA?☐☐

Nerve cells in the affected areas of the brain shrink (atrophy). This can sometimes be seen on MRI scans. When brain tissue is examined under a microscope, structures called glial inclusion bodies can be seen; they contain a protein called alpha-synuclein. It is the presence of these inclusion bodies in the movement, balance and automatic control centres of the brain that confirms a diagnosis of MSA.☐

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Why do nerve cells become damaged?☐☐

It is still unclear as to exactly why the cells become damaged in MSA. MSA is not contagious. It does not appear to be inherited, although there may be a predisposition within the genetic make-up of an individual for cells to become damaged. What triggers the damage process to begin is unknown. This is a focus of ongoing research.☐

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What are the first signs of MSA?☐☐

For men, the first sign is often erectile dysfunction (unable to achieve or sustain an erection). Both men and women often have early bladder problems: urgency, frequency, incomplete bladder emptying or an inability to pass urine (retention). These problems may sometimes be incorrectly attributed to the ageing process or to prostrate disease in men.☐☐Other early problems can be feeling stiff and slow and finding movement difficult, feeling dizzy when standing up, fainting, difficulty turning in bed, and changes in writing. Some people become clumsy or unsteady when walking.☐☐However these early symptoms could also be due to a range of other diseases which need to be excluded before a diagnosis of MSA can be made.☐

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What happens next?☐☐

MSA is a progressive disease - this means that your symptoms will change over a period of time and you will need more help to care for yourself. The speed of these changes is difficult to predict as people with MSA experience the disease differently. Some people feel they cope better when they know what lies ahead. You can discuss the future with your specialist or the nurses.

☐What else happens in MSA?☐☐

There are three groups of symptoms. They are sometimes called :

- parkinsonism (slow, stiff movement),
- cerebellar (co-ordinating movement and balance)
- autonomic (automatic body functions)

Parkinsonism

- feeling slow and stiff when moving
- difficulty turning in bed
- difficulty in starting to move
  writing becoming small and spidery

Cerebellar

- feeling clumsy, dropping things
- finding it difficult to fasten buttons
- feeling unsteady in crowds
- unable to balance without support
- difficulty writing
  slurred speech

Autonomic

- bladder problems
- difficulty with erections
- feeling dizzy or fainting (blood pressure problems)
- pain around neck and shoulders
- constipation
- cold hands and feet
  inability to sweat

Other problems
- weakness in arms and legs
- heightened emotional response, laughing or crying
- restless sleep
- nightmares
- noisy breathing during the day, snoring at night
- unintentional sighing
- swallowing problems, difficulty chewing, choking episodes
- weak, quiet voice

Having a diagnosis of MSA does not mean you will experience all of these symptoms.

☐What is the treatment for MSA?☐☐

Treatment for MSA involves a combination of medication, specialised equipment and the use of therapists to manage individual symptoms. Your treatment will usually be managed by a neurologist or physician who has specialised in movement disorders or autonomic disorders (a specialist). The complex nature of MSA means that the best treatment is gained from a multidisciplinary team approach. A multidisciplinary team is a group of health and social care professionals.☐☐Some members of the multidisciplinary team may not previously have treated anyone with Multiple System Atrophy. Don't be discouraged by this. MSA be can provide them with information and advice. All they need is to be enthusiastic about your care.☐☐Any of these members of a multidisciplinary team may be involved in your care:☐☐

What medication can I take?☐☐

Medication regimes will vary depending on your symptoms. Your specialist will prescribe the best combination to meet your needs. Here are some of the medications commonly used in MSA.☐

☐**Movement problems** - drugs to help stiffness and slowness are the same drugs used in Parkinson's disease (PD). They are not as effective in MSA and can make blood pressure problems worse. It may take time to find what suits you best.
- L-Dopa (Madopar or Sinemet)
- Amantadine (Symmetrel)

Blood pressure problems - drugs to control the fall in blood pressure can be effective in reducing dizziness, fainting and falls. These drugs may cause high blood pressure when lying down. It is advisable to raise the head of your bed and have regular blood pressure checks.

- Fludrocortisone (Florinef) - A steroid, taken in very small doses; may cause ankle swelling.
- Ephedrine - Works quickly to raise the blood pressure, usually taken three times a day.
- Midodrine (Gutron) - Works quickly to raise the blood pressure (within 30-60 minutes), usually taken three times a day. Only prescribed by specialists.

Bladder problems - drugs can reduce urgency and frequency problems
- Oxybutinin (Ditropan) - improves bladder control
- DDAVP (Desmopressin) - used to reduce the production of urine overnight

Antibiotics - should be prescribed at the first signs of infection. Any infection in someone with MSA can worsen symptoms like postural hypotension dramatically.☐

☐**What about complementary medicine?**☐☐
There is no evidence to recommend a specific therapy in MSA but some people find complementary medicine useful. Discuss with your specialist or GP before trying any of these. Bear in mind that very few of these therapies are funded on the National Healthcare Service. Use a therapist who is a member of the appropriate governing body e.g. The Homeopathic Association. For further advice contact the Institue of Complementary Medicine.☐

☐**What about the future?**☐☐
Many of the ideas and plans you had for your future will change once you have MSA. People adjust to this change in different ways. There is no simple recipe to help you cope but there are lots of people out there who can help you, and not least of these is the MSA support group.☐☐Thinking ahead and talking things through with your family can help ease the path in front of you.☐☐Progress in treating MSA is steady and hopeful as increased awareness and understanding of this unique disorder gains momentum.

Frequently asked questions

How is MSA different from Parkinson's disease?☐☐Many people with MSA may have had a diagnosis of Parkinson's disease (PD) initially. Both MSA and PD cause stiffness and slowness in the early stages. The additional problems like swallowing and dizziness that develop in MSA are unusual in early PD. These additional problems, together with loss of certain nerve cells and the presence of inclusion bodies, make MSA a separate and distinct disorder from PD.☐
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Will my children get MSA?☐☐There is no evidence that MSA is an hereditary condition.☐
☐What about dementia? Is this part of MSA?☐☐Dementia is not a symptom of MSA. However, some people may find that their thinking and memory can become slower.

Why has no-one heard of MSA?Specialists in neurology have only recently begun to understand MSA and realise that it is not as rare as they once thought. The information learnt about MSA takes time to filter down to other professionals. Try not to be worried if professionals that you meet have not heard of MSA. They can become experts by caring for you.Be prepared to cope with questions about MSA by

- having a standard answer ready, e.g. "it's like Parkinson's disease but I have extra problems such as low blood pressure"
- having the Italian MSA information leaflets handy to show your family and friends
- giving your therapists our contact details, so they can get further information themselves

I get dizzy when I move. The dizziness may be due to your blood pressure falling when you stand up or change position. This drop in blood pressure is called Postural Hypotension. In addition to medication there are several things you can do to relieve this symptom. These include increasing your fluid and salt intake as well as calf exercises to help your blood flow. It is important that you avoid sudden changes in position like rising to your feet and plan your periods of activity for later in the day. Medication to help blood pressure can be very effective.

I am unsteady on my feet. This may be due to problems with balance. People often feel they need to use furniture for support. Physiotherapists and occupational therapists can provide advice and equipment to help you move safely.

I feel stiff and find it difficult to get moving. Medication used in controlling Parkinson's disease can help reduce stiffness. Blood pressure medication may also help some people. Physiotherapy can also prove useful with tips to get you moving. This is important as inactivity will worsen the problem.

Stairs are difficult; I feel unsafe. Going up and down stairs requires mobility and balance - both can be affected in MSA. A physiotherapist can help improve your safety on the stairs. At some point a stair lift may become advisable and social services can advise on funding for this. An occupational therapist can help you to plan your living space to minimise the need to the use the stairs.

My partner says I stop breathing at night. Stopping breathing at night (sleep apnoea) is sometimes associated with snoring. If you feel tired and lethargic during the day, it may mean that you have low oxygen levels because of sleep apnoea. Sleep studies may be done to see how this symptom is affecting you and it may be necessary for you to have some breathing assistance at night from a piece of equipment called CPAP. This can help improve your quality of sleep and can travel with you wherever there is

electrical power. Other breathing problems in MSA are deep sighs, and noisy breathing.☐☐

I laugh and cry at the silliest things.☐☐This common problem in MSA is not a sign of dementia. Even when people cry frequently it may not be a sign of depression but if excessive crying is bothering you, anti-depressants may help.
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I**'m getting really constipated.**☐☐Constipation is common in MSA. It is to be avoided where possible as straining will affect your blood pressure. It is important to drink plenty of fluids and include lots of fibre in your diet. Daily medication to prevent constipation is often needed to maintain a regular bowel habit.
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I am having problems being intimate with my partner☐☐Erectile dysfunction is common in men with MSA. There are various treatments that help maintain intimacy between couples but may reduce spontaneity. Seeking help on this issue is important and you should not be embarrassed to ask your doctor.
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What is a sphincter EMG?☐☐This is a specialised test that can help with diagnosing MSA by assessing the nerve supply to the sphincter (outlet) of the bladder or bowel. This involves inserting a needle into the muscle; some people find this uncomfortable. It is not widely available.
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My bladder never feels completely empty.☐☐Your bladder may not empty completely and, if you are leaving any behind (a residual volume), this can cause urine infections. Bladder surgery is very unlikely to improve this problem but a continence advisor can help you learn techniques such as intermittent self catheterisation to make you more comfortable.
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I keep having to go to the toilet at night.☐☐People with MSA produce less urine during the day and more at night. If this problem is resulting in disturbed sleep, medication (DDAVP) can help relieve the symptoms by reducing urine production overnight. You may also consider using a commode or urinal at the bedside to reduce the effort of getting to the bathroom. This is especially important if you have postural hypotension or are prone to falls.
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What sort of diet should I be on?☐☐There is no special diet recommended for MSA, although eating a balanced diet, with fibre, protein, and fresh fruit and vegetables is important. If you have postural hypotension, there is some dietary advice to prevent the drop in blood pressure in the postural hypotension. Drinking plenty of liquids is important too.
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I cough and choke after meals.
☐This could be a sign that you have a problem swallowing. Coughing is a normal reaction to help prevent food going into your lungs and causing infection. A speech therapist can assess the strength of your swallow and together with a dietician will advise on the best type of food for you.

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What is a gastrostomy tube?

☐If you are continually losing weight, doctors may suggest a gastrostomy tube. This is a small tube, called a PEG, that is inserted into your stomach through your abdomen during a minor procedure. This tube will provide your nutritional and fluid requirements and is hidden under clothes when not in use. A special liquid diet is fed at times that fit into your daily activities. Medication can also be given through the gastrostomy tube. Although the prospect of this tube may be daunting, it can be a good way to help control symptoms, and is reversible. Many people with a gastrostomy tube are still able to eat small amounts of food for pleasure.

http://www.youtube.com/watch?v=atQGkK0zW2s&feature=fvw

This 3D animation reveals the placement of a percutaneous endoscopic gastrostomy - PEG , or "feeding tube" in patients who are unable to take food by mouth for an extended period of time.

It demonstrates the two main surgical techniques for PEG tube placement.

1. The "pull" method,
2. The "push" method. ANH00005

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I am worried about my partner having to look after me.

☐At some point in all our lives we will need someone to help look after some of our everyday needs. With MSA this point may come sooner than anticipated. Professionals within the health and social care systems can help your partner to look after you. A Carers Assessment is available through the local social services. Contact your local Social Services Adult Disability Team.

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Who can give me help in getting things sorted out at home?

☐There are many people who will be involved in your care either via the GP and hospital or through social services and other voluntary organisations. Contact the Adult Disability Team at your local Social Services department for an assessment.

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I'm worried about the financial implications of having MSA.

☐Having MSA may mean that you will have to stop working sooner than you anticipated - maybe before you can claim your pension. There is a range of benefits and tax allowances that are available whether you are still working or are unable to work. The amount of benefit available to you will be assessed on your individual circumstances and the level of your disability. For a comprehensive assessment of your entitlement contact your local Social Services department.☐☐

Why has palliative care been mentioned?

☐Palliative care specialists manage symptoms in a wide range of illnesses. Palliative care teams include specialist nurses, doctors and hospices. They may also be able to offer individualised respite care. Hospice care is designed to involve the whole family.

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Can I travel abroad on holidays?

☐Travel abroad is possible with the correct arrangements in place. Travel companies will need to be made aware of your special requirements and you will need to ensure the accommodation is suitable.☐☐Take care in the sun, especially if you have postural hypotension.

Research :
European MSA Study Group
The European Multiple System Atrophy Study Group (EMSA-SG) was founded in January 1999.☐☐It represents a consortium of scientific investigators from academic and research centres in Europe and Israel who are committed to clinical trial activity and other research studies aimed at improving the treatment of MSA.☐☐To find out more about EMSA and their current research projects across Europe visit their website at **Fout! De hyperlinkverwijzing is ongeldig.**

Research : Getting involved in research
Trial Participation☐☐Your specialist may be involved in research that you can take part in. Trials may be into medication and therapy for MSA, or may be investigating the diagnostic process and progression of the disease.

☐**Brain Donation**☐☐Brain donation is an important area of neurological research. This is a sensitive subject to discuss for many health professionals as brain donation can only occur after death and this discussion seems at odds with their work to manage symptoms and improve the daily lives of their patients. Mrs. Rita Schouppe, Spokesperson of the World MSA Day organization, who talks to families about brain donation feels that "donation, which may help others in the future, can offer patients and relatives the opportunity to salvage some meaning from an otherwise confusing situation".☐☐Brain donation has enabled the discovery of alpha synuclein (a protein) in the glial inclusion bodies which is peculiar to MSA, and improved the accuracy of diagnosis. Brain donation has also been incorporated (voluntarily) into some clinical research trials to help identify and evaluate any benefit from the treatment to the brain itself.☐☐For a list of Brain Banks please contact your specialist or your local support group
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DNA Donation☐☐Another type of research that members often ask about is genetic research. MSA is not thought to be hereditary; it does not appear to be passed on through family generations. However through the study of genes and DNA, which carry information about our characteristics and body processes, we may be able to unlock some important information about MSA. There are some blood sample DNA banks set up as part of clinical research trials, also some of the Brain Banks may take samples when you register. Most Brain Banks will look at DNA as part of their examination of donated tissue.☐☐For further information contact your specialist or your local MSA Support Groupe

Multiple System Atrophy (MSA)

is the current name for disorders once known individually as striatonigral degeneration, sporadic olivopontocerebellar atrophy, and the Shy-Drager syndrome.

MSA is a progressive neurodegenerative disorder characterized by symptoms of autonomic nervous system failure (such as lightheadedness or fainting spells, constipation, erectile failure in men, and urinary retention) combined with tremor and rigidity, slurred speech, or loss of muscle coordination.

MSA affects both men and women, primarily in their 50s. It can progress swiftly or slowly, but people with MSA generally survive for 9 years after the appearance of symptoms. There is no remission from the disease.

There is no specific treatment for nerve degeneration in MSA.

Levodopa, used to treat rigidity and tremor in Parkinson's disease, may offer some help. However, striatonigral degeneration does not respond well to levodopa.

Dopamine and anticholinergic drugs may be prescribed to treat spasms.

Orthostatic hypotension may be treated with flucortisone and other drugs that raise blood pressure.

Increased dietary fiber intake or use of laxatives may relieve constipation, and drugs or a penile implant may help with male impotence.

A routine of stretching and exercise can help retain muscle strength and range of movement.

An artificial feeding tube or breathing tube may be surgically inserted for management of swallowing and breathing difficulties.

MSA is a progressive disorder and most patients have a life expectancy after diagnosis of about 10 years or less.