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Factsheet

Series for healthcare professionals

A Guide to Multiple System Atrophy for: General Practitioners

This document serves as a guide to general practitioners working with people with multiple system atrophy (MSA). It draws on available literature in MSA, Parkinson's disease and other atypical Parkinsonism disorders. It does not cover aetiology, epidemiology, neuropathology and medical management in any depth.

The Multiple System Atrophy Trust (MSA Trust) produces a series of specialist MSA factsheets for health professionals to enable them to manage symptoms and improve the treatment people with MSA receive. Other factsheets can be found on our website: www.msatrust.org.uk.

The Multiple System Atrophy Trust (MSA Trust) is the only charity working in the UK and Ireland specifically to support people with MSA. As well as helping people who have MSA, the Trust supports anyone affected by the disease, including carers, families, friends and health professionals.

The Trust employs MSA Health Care Specialists, a Social Welfare Specialist, manages a telephone and email advice service and runs a network of Support Groups. We provide up-to-date literature for people affected by MSA and for health professionals. We also fund vital research to find the cause, and one day, cure for MSA.

To ensure services are accessible to everyone, the Trust is committed to providing services for people affected by MSA free of charge. The MSA Trust is a charity funded entirely on voluntary donations.

The MSA Trust is always keen to receive feedback about the information we produce, please email support@msatrust.org.uk with any comments.

Definition

MSA is a progressive neurological disorder which leads to a shortened lifespan. By definition MSA is a palliative condition. There is no cure or treatment to ameliorate the progression of MSA. Management is totally focussed on managing each individual's specific symptoms and maximising their quality of life. It is associated with the degeneration of nerve cells in the cerebellum, brain stem and basal ganglia.

Alpha-synuclein, a protein structure that gathers in glial inclusion bodies, is seen in three areas of the brain on examination after death. This degeneration causes the symptoms of MSA, which include movement and balance, urogenital, speech, swallowing, bowel, bladder, blood pressure, breathing and temperature control difficulties.

MSA is characterised by a combination of the following features:

- parkinsonism (muscle rigidity with or without tremor and bradykinesia)
- ataxia (poor coordination / unsteady walking)
- autonomic dysfunction

MSA may be further identified by two subtypes depending upon the predominant motor presentation at the time of diagnosis:

- MSA-C - indicating primarily cerebellar symptoms and composed of gait ataxia, limb kinetic ataxia, scanning dysarthria as well as cerebellar ocular disturbances (sometimes called sporadic olivopontocerebellar atrophy)
- MSA-P - indicating primarily parkinsonian symptoms and is dominated by progressive akinesia and rigidity (sometimes called striatonigral degeneration)

Autonomic dysfunction is present in both MSA subtypes.

Epidemiology

The overall prevalence of MSA is estimated at around 5 cases per 100,000 people. This means roughly 3,000 – 4,000 people in the UK are living with MSA at any one time. The most common age of onset is mid to late fifties, though it can occur at any age between 30-75 years old.

It affects both genders but has been found to be slightly more common in males with around 55% of MSA cases occurring in men.

The underlying aetiopathogenesis is still unknown, no specific risk factors have been identified and it is thought a complex interaction of genetic and environmental factors seems likely. Several studies have looked at occupational and daily habits, including exposure to pesticides, metals, cleaning products and a history of farming but no study has confirmed an increased risk. To date there is no evidence to indicate that MSA is an hereditary condition.

Disease duration

Mean survival from symptom onset is between six and nine years with some people living beyond fifteen years after onset. Many people have a much shorter life span than the published average, by the time diagnosis is made they may be 2-3 years into the disease journey. No two people will present with the same symptoms or the same rate of progression. There is little difference in duration of living with MSA between MSA-P and MSA-C, although motor deterioration is often more rapid in MSA-P.

Numerous autonomic dysfunction symptoms presenting early are thought to be indicative of poorer prognosis.

Initial presentation

The most common motor sign of MSA is the appearance of an "akinetic-rigid syndrome" (a slowness of initiation of movement) which commonly leads to a misdiagnosis of Parkinson's disease. People with MSA-C present with balance, co-ordination and speech problems.

Both men and women often experience problems with their bladders including urgency, frequency, nocturia, incomplete bladder emptying, or retention. Erectile dysfunction is an early symptom in male patients and is almost always present.

Also, at the early stages postural stability is compromised and around one in five people living with MSA will experience falls in their first year of symptoms.

Diagnosis and Red Flags

There is no diagnostic test. Alongside the cerebellar and the parkinsonism symptoms there must be autonomic dysfunction to make the diagnosis of MSA. Referral to a neurologist or physician for further evaluation (that should include assessment of autonomic function), is needed. The investigations, which range from autonomic function tests, imaging of the brain and evaluation of urological function, should ideally be performed to confirm the diagnosis, exclude other overlapping disorders, and provide an evaluation of function that will aid management.

This difficulty of diagnosing MSA and the difficulty in discriminating it from Parkinson's disease has led to the creation of red flags to act as warning signs that may raise the clinical suspicion of MSA.

These include:

- Early bladder issues- urgency/frequency/nocturia/retention or combination often before movement problems
- Early erectile dysfunction often before movement problems
- Balance issues-falls are common much earlier than would be seen in Parkinson's
- Bradykinesia- may look like Parkinson's disease
- Speech may become quiet or slurred particularly when fatigued
- Swallow - may complain of coughing or choking, especially on fluids
- No resting tremor, but may be an intention tremor and myoclonus of fingers
- Low mood or increased anxiety, emotional lability later
- Postural hypotension, especially after eating, intercourse or standing from sitting/lying
- Disturbed sleep/sleep apnoea/headache on waking/low oxygen saturations
- Stridor

Symptoms

The combination of symptoms a person with MSA may present with arise from alphasynuclein damage in the 3 brain areas previously mentioned: basal ganglia cerebellum and brain stem. A person with MSA can encounter many, if not all, of the symptoms at some stage of disease progression.

It is important to note that significant deterioration of a person's symptoms over a period of a few days is highly indicative of an acute clinical cause and will resolve on treatment of that cause e.g. infection.

The list of symptoms includes:

- Slurred speech, dysphonia, dysarthria
- Difficulty negotiating spaces, unsteadiness
- Falls
- Limb ataxia, intention tremor, difficulty dressing and eating
- Sexual dysfunction
- Bladder dysfunction-urgency, frequency nocturia, retention
- Constipation
- Unintentional sighs
- Stridor
- Snoring
- Sleep apnoea
- Oxygen de-saturation
- Vivid dreams, REM sleep disorder and lashing out in sleep
- Antecollis (abnormal neck flexion)
- Postural hypotension, which can be difficult to treat and includes;
    - a drop of at least 20mmHg systolic on standing
    - dizziness, falls and fatigue
    - blurred vision
    - coat hanger pain (in trapezius muscles due to poor perfusion of blood)
    - altered consciousness

Helpful strategies for postural hypotension

- Raise the head of the bed during sleep to prevent postural hypotension on rising
- Increase fluid and salt intake (particularly water will help recover from a sudden drop in BP)
- Do calf pump exercises before attempting to stand
- Eat small meals more frequently
- Avoid hot temperatures, dehydration

Cognitive problems may arise, including:

- Increased emotional response with inappropriate laughing/crying (emotional lability)
- Cognitive slowing, but no frank dementia
- Depression and anxiety
- Changes in mental state, e.g. sudden confusion or having hallucinations, are strong indicators the person has an infection or acute clinical episode that needs treating.

Key management problems

These include:

- Urinary disturbances, including recurrent urine infections.
- Severe postural hypotension resulting in fainting and other symptoms such as dizziness on standing, and neck pain. This is often worse in the morning, after a hot shower, after food ingestion, after medication (such as levodopa and other anti-parkinsonian medications), after exertion or whilst straining to empty the bladder or bowels.
- Breathing impairment because of stridor or apnoea. Recurrent respiratory tract infections may be exacerbated by aspiration. Vocal cord paralysis can occur so a person with MSA presenting with stridor needs to be seen urgently and urgent referral to ENT.
- Worsening parkinsonian features especially rigidity, that is increasingly refractory to anti-parkinsonian drugs
- Worsening cerebellar features that in combination with postural hypotension make these patients prone to falls and also injury
- Constipation, often severe and refractory to standard approaches.
- Difficulties with swallowing that may necessitate a discussion about the potential for insertion of a PEG/RIG gastrostomy feeding tube.

How can the GP support a patient with MSA?

The GP is crucial in maintaining an overview to balance management options of new symptoms against the other co-morbidities the person has and medication they are on. Management of MSA is about working with the patient, and their carer, to alleviate the problems associated with each symptom; and so improve the quality of life.

The GP can:

- Provide prompt treatment of probable infections, particularly urinary and chest infections.
    - Infections and stress will cause all MSA symptoms to worsen rapidly
    - People with MSA do not always have a raised temperature if they have an infection.
- Prescribe urinary multistix for home testing if regular urine infections occur or consider long-term prophylactic antibiotics or rescue packs for the person's home
- Provide a named GP within the practice, where possible, or mark the patient's computer record with the appropriate disease Read Code.
- Investigate, treat and manage any new problems as would be done for someone without a diagnosis of MSA. The cause may not be due to MSA but a common treatable cause. Although MSA is not treatable in itself, there are management options for many of the symptoms caused by MSA.
- Help the MSA patient understand that MSA is a palliative condition. That:
    - It is incurable
    - It is a deteriorating and progressive condition
    - It has an unpredictable end of life
    - Encourage and facilitate person with MSA to have discussions about future care planning.

    - Ensure MSA is put on the death certificate
    - Referral to Community Matron / Long Term Conditions service / District Nurse service for regular monitoring and access to proactive interventions in a timely way.
- Liaise with the specialist team and palliative care services for advice and management of symptoms
- Support for the carer
    - A carer-friendly policy within the practice will help.
    - Caring for someone with MSA will become a 24-hour nursing care role
    - Carers can become exhausted and suffer burnout.
    - Regular respite is essential to allow the carer to continue in this role.
- Medication - some symptoms may be suitable for management by medication
    - Most antiemetic drugs should be avoided as anti-sickness medications can interfere with Parkinson medications, an exception being domperidone.
    - Similarly, some antidepressants are contra-indicated with Parkinson's medications (see separate medication information sheet).
    - Specialist teams can be contacted for advice and support, please contact the MSA Trust Health Care Specialists at any time if you have any questions or concerns.

Multi-disciplinary team (MDT)

Management of MSA will require input from a multi-disciplinary team. Timely referrals to the following services are essential to maintain the safety and quality of life of a person with MSA:

- **Neurologist –** ideally a movement disorder specialist or Parkinson's specialist. Other specialists may include **Urology**, **Andrology**, **Respiratory, Gastrology** and **Ear, Nose and Throat** services.
- **Parkinson's Nurse Specialist** – most will support person living with MSA and have knowledge of symptom control and local support.
- **Speech & Language Therapist** – at early onset of dysarthria and dysphagia especially those with caqhoking and stridor. Also, communication and swallow assessment, advice and support. Advise on possible need for supplemental tube feeding.
- **Physiotherapist** – needed from early stages to maintain muscle strength posture management, and can target specific training such as: turning in bed, exercises on bed or chair to prevent postural hypotension. Maintain and improve lung volume and cough clearance ability.
- **Occupational Therapist** – advise and plan adaptations to living accommodation, plan for wheelchair dependency and potential hoist transfers and to promote independence, as well as provide aids to assist with activities of daily living at every stage.
- **Bladder and Bowel/Continence Service** – teach intermittent self-catheterisation, eventually indwelling (urethral or supra pubic) catheter, manage constipation problems.
- **Community Matron –** to manage complex needs and case manage/signpost where possible.
- **Social Services** – Local Authority OT for advice on housing adaptations. To arrange package of care at home or nursing home and support for the carer.

- **Palliative Care Outreach Team –** symptom control and quality of life improvement.
- **Hospice** – support for family and carers and offer respite and/or day-care.
- **Dietitians** – especially for those with food-induced hypotension or those losing weight.
- **MSA Trust –** for MSA Health Care Specialist advice, Social Welfare Specialist advice and information for patients, carers, family and professionals and access to local support groups.

The Trust's contact details

We have MSA Health Care Specialists that support people affected by MSA in the UK and Ireland. If you would like to find the MSA Health Care Specialist for your area, contact us on the details below or use the interactive map here – https://www.msatrust.org.uk/support-for-you/hcps/.

T: 0333 323 4591 | **E**: support@msatrust.org.uk | **W**: www.msatrust.org.uk

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Disclaimer

This factsheet is intended for qualified professionals. The information provided is for guidance purposes only and should be used alongside other relevant research, professional standards and individual clinical or professional judgement. Circumstances for people affected by MSA vary greatly, and professionals should not rely on this material alone when providing support to people.

References for this information sheet are available by contacting support@msatrust.org.uk.

Your feedback helps us ensure we are delivering information to the highest standard. If you have any comments or suggestions, please contact us at support@msatrust.org.uk.