MSA Trust

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 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 three specialist nurses, 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 premature death. 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 at post mortem examination in three areas of the brain. This cell degeneration in the cerebellum causes problems with movement, balance and autonomic functions of the body.

MSA is characterized by a combination of the following features:

- parkinsonism (muscle rigidity +/ 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 universal in both MSA subtypes.

The term multiple system atrophy was proposed in 1969 by Graham and Oppenheimer who recognised substantial clinicopathological overlap between three existing conditions: sporadic olivopontocerebellar atrophy, striatonigral degeneration and Shy-Drager syndrome (autonomic failure).

A variant with combined features of MSA and Lewy body dementia may also exist.

Epidemiology

The overall prevalence of MSA is estimated at around 5 cases per 100,000 people. This means roughly 3,000 people in the UK are living with MSA at any one time. Adult onset is on average in the sixth decade in the mid to late fifties. However, onset can be between 35-75 years of age but is not known to occur under 30 years of age.

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 etiopathogenesis 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. MSA is also not thought to be hereditary.

Disease duration

Mean survival from symptom onset is between six and nine years with some people living beyond fifteen years after onset. No two people will present with the same symptoms or the same rate of progression. There is little difference in duration between MSA-P and MSA-C, although progression in MSA-P is more rapid.

Initial presentation

The most common first 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.

Other signs at onset include problems with balance (cerebellar ataxia) followed by genitourinary 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 single diagnostic test. 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:

- Erectile dysfunction
- Bladder disturbance
- Postural hypotension
- Poor response to Levodopa
- Cold extremities
- Severe dysphonia, dysarthria, dysphagia
- Inspiratory sighing/stridor
- Rapid progression leading to wheelchair dependency

Symptoms

The three features of MSA produce a variety of symptoms and a patient with MSA can encounter many, if not all, of them at some stage of disease progression. The list of symptoms includes:

- Slurred speech, dysphonia, dysarthria
- Difficulty negotiating spaces, unsteadiness
- Falls
- Limb ataxia, intention tremor, difficulty dressing and eating
- Erectile dysfunction
- Bladder dysfunction-urgency, frequency, nocturia, retention
- Constipation
- Unintentional sighs

- Stridor
- Snoring
- Sleep apnoea
- Oxygen de-saturation
- Antecollis (abnormal neck flexion)
- Vivid dreams, REM sleep disorder and lashing out in sleep
- 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
- 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
- Cognitive slowing
- Depression and anxiety

Key management problems

These include:

- 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 food ingestion, after medication (such as L-Dopa), after exertion or whilst straining to empty the bladder or bowels.
- 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
- Urinary disturbances, including recurrent infections.
- Constipation, often severe and refractory to standard approaches.
- Difficulties with swallowing that may necessitate a feeding gastrostomy

- Breathing impairment because of stridor or apnoea, and recurrent respiratory tract infections which complicate aspiration

How can the GP support a patient with MSA?

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:

- 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
- Conduct a regular patient review at practice meetings.
- Provide a named GP within the practice, where possible, or mark the patientôs computer record with appropriate disease Read Code.
- Investigate, treat and manage any new problems as would be done for someone without a diagnosis of MSA. Although MSA is not treatable in itself, each symptom can be managed to an extent.
- Provide prompt treatment of probable infections, particularly urinary and chest infections.
  - Infections and stress will cause all MSA symptoms to worsen rapidly
  - Prescribe urinary multistix for home testing if regular urine infections occur
- 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 Parkinsonôs medications (see separate medication information sheet).

Multi disciplinary team (MDT)

Management of MSA will require input from a multi-disciplinary team. The GP will need to refer the patient to the following at some stage:

- **Neurologist –** ideally a movement disorder specialist or Parkinsonôs specialist
- **Parkinson's Nurse Specialist** ï most will support person living with MSA and have knowledge of symptom control and local support
- **Speech & Language Therapist** ï at early onset of dysarthria and especially those with choking and stridor. Also communication and swallow assessment, advice and support. Advise on possible need for Percutaneous Endoscopic Gastrostomy (PEG) Tube.

- **Physiotherapist** ï needed from early stages to maintain muscle strength and can target specific training such as: turning in bed, exercises on bed or chair to prevent postural hypotension
- **Occupational Therapist** ï advise and plan adaptations to living accommodation, plan for wheelchair dependency and potential hoist transfers and to promote independence
- **Continence Nurse Advisor** ï 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** ï 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** ï support for family and carers and offer respite and/or day-care
- **Dieticians** ï especially for those with food-induced hypotension
- **MSA Trust –** for specialist nurse advice and information for patients, carers, family and professionals; and access to local support groups

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