MSA Trust

Factsheet

Series for healthcare professionals

A guide to Multiple System Atrophy for: Speech and Language Therapists

This document serves as a guide to Speech and Language Therapists 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. Further reading on these topics and others can be accessed via the list of resources (including the MSA website), given at the end of the document.

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 four 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.

Introduction

MSA is a rare progressive neurological disorder that affects adult men and women and leads to premature death. Currently, there is no known cause or cure. MSA causes degeneration or atrophy of nerve cells in several (or multiple) areas of the brain which results in problems with movement, balance and autonomic functions of the body such as swallowing, bowel, bladder and blood pressure control.

Globally, around five people per 100,000 have MSA which equates to almost 3,000 people living with MSA in the UK [1]. Parkinson's disease is about 45 times more common, affecting about 200 per 100,000 in the UK [2].

MSA usually starts between the ages of 50-60 years, but it can affect people younger and older. MSA does not appear to be hereditary although current research is examining whether or not there is a genetic predisposition to develop the disease. It affects both sexes equally [7].

Neuropathology

MSA falls within the entity of the spectrum of oligodendrogliopathies . The mechanisms underlying the condition and the factors that trigger MSA onset are yet to be established. Environmental and dietary influences have been cited [4] however definitive cause and risk factors are yet to be established. Symptoms of MSA are a manifestation of the accumulation of alpha-synuclein in glial inclusion bodies, originating in the striatonigral, olivopontocerebellar and central autonomic (brain stem) regions. People diagnosed with MSA will eventually present with an overlap of symptoms, as illustrated in Figure 1.

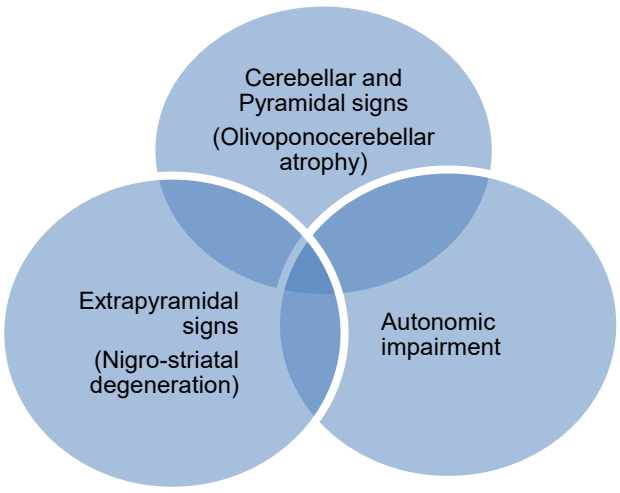

Figure 1 - MSA signs and symptoms (*Adapted from Swan* [5])

Clinical Diagnosis of MSA

The term MSA was first used in 1969 - prior to this it had previously been known as Shy - Drager Syndrome. The first diagnostic criteria for MSA were proposed in 1989 [7] and Second Consensus Criteria were defined in 2008 [8] which define three levels of certainty of the diagnosis—possible, probable and definite MSA.

Distinguishing MSA from idiopathic Parkinson's disease is still problematic, with both presenting with abnormal DAT scans. Occupational Therapists need to be aware of clinical features that characteristically distinguish symptoms of MSA from other Parkinsonism syndromes including PD, although these can be hard to discern in the early stages (see Table 1).

The key distinguishing clinical signs at diagnosis are [7]:

1. Autonomic failure which includes orthostatic hypotension and bladder dysfunction (with erectile dysfunction in men)
2. Poor response to levodopa (may receive partial or transient benefit)
3. Akinetic rigid parkinsonism (present in 58% of cases) or cerebellar ataxia (29%)

Multiple System Atrophy	**Parkinson's Disease**
a-synuclein pathology Cerebellum, brainstem and basal ganglia affected Bradykinesia (MSA-P) or Gait ataxia (MSA-C) Movement/intention tremor Impaired posture; forward, side and antecollis Autonomic failure, usually early symptoms – constipation, urinary and sexual dysfunction, orthostatic hypotension Usually bilateral/symmetrical presentation Partial, limited or unsustained response to levodopa Early dysphonia, dysarthria and dysphagia Respiratory symptoms include stridor, snoring, apnoea, respiratory insufficiency, vocal cord paralysis, obstructive and central apnoea Dystonia including antecollis Early falls Cerebellar ataxia Fatigue Median Survival 3-9 years Emotional lability	a-synuclein pathology Basal ganglia affected Bradykinesia and rigidity Resting and 'pill-rolling' tremor Unilateral early symptoms Impaired posture; tend to lean forward Festinating gait Micrographia Hypophonia Speech and swallow impairment later Good response to L-Dopa Constipation Falls (later, uncommon in early disease) Survival can be decades
Progressive Supranuclear Palsy	**Cortico-Basal Degeneration**
Tau-opathy Mid brain, basal ganglia and brain stem affected Early postural instability and falls, usually backwards, retrocollis Tremor less common, intentional/postural if present Vertical gaze palsy Bilateral involvement Dysarthria and dysphagia Fatigue Emotional lability Progressive reduced cognitive processing, lack of insight, behavioural and personality changes L-Dopa response limited Median survival 5-7 years	Tau-opathy Frontal parietal cortex and basal ganglia affected Bradykinesia Focal asymmetrical limb apraxia Early speech apraxia, aphasia Unilateral myoclonus Lateral tilt Cognitive impairment, loss of judgement and motor planning, frontal lobe dementia No autonomic involvement Median Survival 7-9 years

Table 1: Common and distinguishing features for PD, MSA, PSP and CBD (*Courtesy of Katie Rigg, MSA Nurse Specialist, Northeast UK)*

MSA phenotypes

The criteria used most often to classify MSA recognise two main phenotypes [7]. In general, people present with predominance of Parkinsonian features (MSA-P) or predominance of cerebellar features (MSA-C), however symptoms overlap. Almost all people with MSA develop autonomic symptoms. . In Western hemisphere cohorts approximately 80% of cases are predominantly MSA-P. The contrary is true for Eastern hemisphere populations. This may be due to be to racial genetic differences and cultural characteristics. MSA is a rapidly progressing, multi-organ disorder leading to severe disability. It has been established that the prognosis is poorer in people who present with early autonomic dysfunction. People who have been classified as MSA-P are more likely to have greater functional decline [49]

MSA-P

The motor symptoms characteristic of MSA-P are similar to those observed in typical Parkinson's disease and include rigidity, bradykinesia, tremor and poor balance. However, autonomic symptoms can also predate motor symptoms in MSA-P.

MSA-C

Cerebellar symptoms in MSA originate from the trunk spreading to the lower limbs which eventually affect gait. Gait ataxia, limb kinetic ataxia and scanning dysarthria as well as cerebellar occulomotor disturbances are typical motor symptoms of this phenotype.

Treatment strategies

There is currently no consensus on the stages of disease progression in MSA, nor is it clearly defined. Average survival is close to a decade from symptom onset [8], although this is a guide only. Treatment varies for each stage of the disease and Speech and Language Therapists are urged to use their clinical reasoning skills based on the knowledge of the neuropathology of the disease. Patient centred goals should be realistic and appropriate, and multi-disciplinary intervention is key to the provision of a quality service. Medical management is based on symptom alleviation, most notably bradykinesia and orthostatic hypotension [40]. The table below summarises key medical interventions.

Symptom	Intervention
Parkinsonism	L-Dopa (40-60% of MSA patients will initially respond)
Ataxia	Physiotherapy and Occupational therapy
Orthostatic hypotension	Non-pharmacological, TEDS, abdominal binder, fluids, small meals, poitioning and exercise advice. Pharmacological – fludrocortisone, midodrine
Neurogenic urinary tract dysfunction	Non pharmacological strategies, medications and catheterisation. Urology or continence team involvement.
Constipation	Exercise, diet, fluids and medications. Continence team involvement.
Erectile dysfunction	Specialist referral to andrology or sexual dysfunction clinic
Breathing problems	Physiotherapy, respiratory team, sleep studies, CPAP, tracheostomy
Dystonia/pain	Physiotherapy, positioning, support, massage, medications, Botox
Camptocormia	Physiotherapy and Occupational therapy support
REM Sleep disorder	Sleep studies, clonazepam, melatonin
Depression	Non-pharmacological, psychology and psychiatry support, pharmacological interventions

Table 2: Key symptoms and medical management Adapted from: Flabeau et al [9]

Speech and Language Therapy

Speech and Language Therapists (SLTs) play a vital role in assessing and providing therapeutic input for MSA patients with communication, swallowing and voice difficulties. Research surrounding the efficacy of speech and language therapy intervention with MSA is still limited and a SLT may only see a few clinical cases over their entire career, due to the rarity of the condition [6].

A SLT must apply a patient centred and holistic approach when working with this client group [7]. The differential diagnosis of MSA is still problematic and early symptoms can often be likened to those seen in patients with Parkinson's disease. Both MSA and Parkinson's disease present with progressive dysarthria, progressive dysphagia and reduced volume of voice. As a result of their similarities, a SLT could consider studies carried out with Parkinson's disease patients to help inform their early interventions.

Family members will often play a key role in supporting interventions, especially towards the advanced stages of the disease. They will often be responsible for supporting meal times and encouraging the person to use SLT techniques to aid communication during everyday life.

Liaison with specialist nurses, physiotherapy, occupational therapy, dietetics, neurology and their medical team will be crucial to ensuring the person receives the right level of care. Working as part of a multi-disciplinary team is also a factor that is very important when working with people who have a progressive neurological condition [8].

However, people with MSA will often exhibit symptoms of impaired communication or swallowing difficulties much earlier than those with Parkinson's disease [50]. Earlier dysphagia symptoms are directly correlated with a generally shorter life expectancy [50]. Once communication, swallowing or voice difficulties emerge, they are also likely to decline more rapidly than those who are diagnosed with Parkinson's disease [51]. This can cause distress and requires the Speech and Language Therapist to be able to take an informed approach to carrying out any interventions. Reviews may need to be more frequent and as such people with MSA may not be discharged from care depending on their local health authority's policies and procedures. It is also important to ensure they know exactly how to contact their Speech and Language Therapist if things suddenly change.

Communication Difficulties in MSA

As MSA is a type of movement disorder, people diagnosed with it often develop a progressive motor speech disorder. The communication difficulty will differ from person to person depending on the MSA phenotype. People with MSA will present with a decreased ability to articulate compared to those with Parkinson's disease and also fewer syllables per second [10]. Penner, Miller & Walter's [10] results (see Table 1) demonstrate the significant differences between the impact of hypokinetic dysarthria (as often seen in Parkinson's disease) and ataxic or spastic dysarthria (usually seen in people with MSA or PSP). This can make their speech particularly slow and they will require increased amount of time for personal expression. Studies that have investigated motor speech disorders in Parkinson's disease reveal that up to 90% of patients diagnosed with the condition will have some degree of a motor speech disorder [11,12].

For those who present with MSA-P, their speech will often appear to be relatively similar to those with Parkinson's disease. Their speech will often present with a hypokinetic dysarthria but may also contain spastic or ataxic elements [12, 13, 14, 15, 16]. The presentation of the hypokinetic dysarthria is what makes differential diagnosis of the conditions so difficult. However, therapists should note that dysarthria latencies for MSA and Progressive Supranuclear Palsy (PSP) are on average 24 months post-diagnosis compared to 84 months for those with Parkinson's disease [50]. For those with MSA-C, their speech is more likely to exhibit increased co-ordination difficulties commonly associated with ataxic dysarthria [12]

PEA – word produced	Idiopathic Parkinson's disease (IPD)	Multiple System Atrophy (MSA)	Progressive Supranuclear Palsy (PSP)	IPD vs MSA vs PSP
First sec	5.0	3.0	4.0	F=2.0891n.s.
Mid 3 s	15.0	10.0	9.0	F=18.102 p<0.00001
Last sec	4.5	3.0	3.0	F=0.06202 p<0.1
Total	25.0	22.0	19.0	F=4.21 p<0.05

Table 1: "Number of completed syllables in syllable repetition" [10).].

Common Features Impairing Communication in MSA

- Dysarthria; specifically hypokinetic dysarthria, ataxic dysarthria, spastic dysarthria or a mixture (mixed dysarthria).
- Dysprosody
- Dysphonia; may present with a hoarse voice or have reduced loudness
- Progressive limb weakness; impacting on the use of gesture
- Masked facial expression (orofacial dystonia); reducing non-verbal communication
- Increased or decreased rate of speech
- Dysfluency
- Expressive and receptive language difficulties
- Increasing cognitive difficulties, negatively impacting on conversational discourse
- Plan for people with MSA to lose the ability to speak.

Assessing Communication Difficulties in MSA

The following aspects should be taken in to consideration when assessing a person with MSA:

1. **Dysarthria** – A dysarthria assessment should include auditory-perceptual clinical assessment and instrumental assessment if appropriate or available [17, 12]. A perceptual assessment may include the use of informal rating scales or formal tools such as the The Frenchay Dysarthria Assessment (FDA) [18]. Standardised reading passages such as 'The Grandfather Passage' may also be a useful resource to help collect auditory-perceptual information. Any findings from the perceptual assessment should help to inform whether instrumental assessment would be beneficial. In the clinical setting, it is unlikely that an instrumental assessment of the person's dysarthria would drastically change the clinical management.
2. **Receptive Language** – A person with MSA may find processing complex auditory or written information increasingly difficult, due to increasing cognitive difficulties [19, 20, 21]. Simple orientation tasks and assessing their ability to follow instructions of varying complexity should help to establish their current receptive capabilities [22]. Formal assessments of receptive language may be attempted, but may not be appropriate depending on the stage of the MSA.
3. **Expressive Language** –Is also affected due to the increasing cognitive impairments. A person with MSA experiences increasing difficulties with word finding and speech initiation, due to progressive neurological deterioration [23, 22]. The rate of speech may also be slower compared to someone with Parkinson's disease [24].
4. **Dysphonia –** A perceptual voice analysis tool such as the GRBAS [25] may be useful to record the impact of MSA on the patient's vocal output. Pitch is often increased in people with MSA [24] there may be increased hoarseness and breathiness on phonation [26]. Due to the interconnectivity between dysarthria and dysphonia, the term dysarthrophonia can be used to describe the presentation.

5. **Inspiratory stridor –** Can be defined as the presence of "turbulent airflow through a partially obstructed airway" [27] this symptom increasingly impacts on the MSA patient's ability to breathe effectively [12]. Therefore, there will be an impact on their communication and connected speech output due to reduced respiratory support. The patient should seek medical support from ENT if inspiratory stridor is identified (ibid), as they may require surgery such as a tracheostomy procedure.

Potential Interventions to Support Communication

Direct evidence based research into appropriate communication interventions for people with MSA is an area that is significantly lacking in research. A recent Cochrane review into Speech and Language Therapy interventions for people with Parkinson's disease was unable to conclude which interventions were most efficacious for speech disorders [28].

Despite this fact, many of the features with which someone with MSA will present can be seen in Parkinson's disease; this indicates that interventions used for Parkinson's disease may be effective. The Speech and Language Therapist should bear in mind the stage of MSA, in terms of appropriate intervention. For advanced stage MSA, direct therapeutic input is unlikely to be beneficial due to the increasing cognitive impairment [8]. In relation to Alternative Augmentative Communication (AAC) use, due to the oral-facial dystonia and visuo-spatial difficulties MSA patients often encounter, AAC options may be limited; especially in the advanced stages.

- **AAC** - following appropriate assessment. If AAC is deemed appropriate, the Speech and Language Therapist should consider using both low technology and mid/high technology aids. A referral can be made to the local Assistive Communication Service if access to mid/high technology aids is unavailable in the patient's current service. They may also require an alternative access assessment to enable use of adaptive keyboards for example.
- **Oral motor exercises** - can be used to help maintain strength in the laryngeal, lingual and facial muscles [29]; however, improvement may be inconsistent over time due to the neurodegenerative nature of the disease [30].
- **Compensatory dysarthria strategies** - appropriate pacing, spelling out words and applying appropriate environmental controls. For example, minimising external noise or ensuring face to face communication occurs.
- **Encouraging the use of deeper breathing techniques** – such as diaphragmatic breathing [31] which can help increase subglottic pressure and increase volume [32].
- **Communication support groups** – Group approaches have found to be a beneficial way to implement therapy for people with Parkinson's disease and have been shown to improve intonation and body language measures [3, 33].
- **Vocal Exercises** –A formal training programme such as Lee Silverman Voice Therapy LOUD programme (LSVT®) technique 34,35] may be beneficial in the earlier stages of the condition. However, there have been no direct studies in MSA to test this hypothesis as in Parkinson's disease [31].

Examples of AAC Aids

Low Technology Aids:

- **Simple alphabet charts** – These can support a person with dysarthria, however various font sizes and colour combinations should be assessed as vision can become impaired.
- **A personalised communication passport** – to provide unfamiliar adults with an overview of the diagnosis and associated difficulties.
- **Basic self-care communication books** – for members of the multi-disciplinary team and family to use with the patient.

High Technology Aids:

- **Light-writers** – to improve overall intelligibility, however the patient's dexterity should be taken in to consideration.
- **Amplifiers** – to help support voice projection.
- **iPad** – may be beneficial for some patients in the early stages of MSA, but might not always be the most efficient form of communication depending on the patient's current impairments. Such forms of AAC require a good level of cognition and fine motor skills to operate successfully.

- Electronic communication aids, eye gaze technology and switches, environmental control and computer access assessments may be helpful at different stages, depending in individual need and ability.

Summary of the Key Communication Points:

- People with MSA can experience **an earlier onset of dysarthria** compared to people diagnosed with Parkinson's disease [13].
- The **dysarthria** is also likely to be **more severe**. (ibid)
- There is a **higher incidence of mixed dysarthrias** in MSA, involving a combination of: ataxic, spastic and hypokinetic [10]
- People with MSA will generally **use fewer syllables per second** in their speech due to the **increased bradykinesia** (ibid, see Table 1)
- **Intelligibility** is likely to be difficult due to **co-ordination difficulties** of the lingual musculature and general muscle atrophy [36]
- They may experience **increased respiratory difficulties** due to inspiratory stridor [7]
- **Cognitive decline** in advanced MSA will significantly impact the use of AAC and overall communication ability.

Swallowing Difficulties in MSA

Research studies reflect that there is a high incidence of dysphagia occurring in people with MSA [37]. The oral stage of swallowing involves the person being able to masticate the bolus adequately and then propel the bolus posteriorly to the pharynx [38]. In a person with MSA, the propulsion of the bolus is likely to be delayed and extremely effortful; resulting in a significantly prolonged oral stage of the swallow [39]. This puts them at higher risk of aspiration as the reduced co-ordination may lead to a premature spillage of bolus.

Due to similarities seen in PSP, MSA and Parkinson's disease, they are often compared with each other in research studies; however, the onset of dysphagia may differ. Dysphagia latencies are typically 67 months post diagnosis for people with MSA, compared to 42 months in PSP and 130 months in Parkinson's disease [50]. Although, people with PSP are more likely to experience dysphagia symptoms earlier than MSA patients, MSA patients experience dysphagia significantly sooner than people with Parkinson's disease.

Common Features Impairing Swallowing in MSA

As with other Parkinson plus diseases, people with MSA are likely to experience progressive oropharyngeal dysphagia. This can be caused by any of the following factors and a person may present with all of these symptoms:

- **Coughing or choking** on food or fluids
- **Increasing bradykinesia**; making independent feeding increasingly difficult
- **Poor hand to mouth co-ordination**
- **Decreased lip seal** and **anterior loss of saliva**
- **Fasciculation of the tongue**; this can make manipulation of a bolus within the oral cavity very difficult. It can also impact the anterior to posterior propulsion of a bolus.
- **Rigidity**; impacting on the muscles required for adequate mastication
- **Reduced appetite**
- **Impact of medication**; if MSA medication is not taken on time, the swallow function may deteriorate

A person with MSA-C is more likely to find the co-ordination of a bolus more difficult due to their ataxia.

Assessing Swallowing Difficulties in MSA

Any person who presents with a dysphagia should be thoroughly assessed in accordance with current RCSLT dysphagia clinical guidelines.

A Speech and Language Therapist should consider:

- Carrying out a thorough cranial nerve examination –weakness in all of the cranial nerves involved in swallowing is likely
- Alertness and positioning of the patient prior to assessment; you may need to access your physiotherapy team to aid with positioning
- Previous levels of oral intake and any reported difficulties

- Any previous dysphagia recommendations that are in place
- Current respiratory status
- Carrying out a formal clinical bedside evaluation of swallow
- Referring for instrumental analysis of swallow as appropriate; however, a procedure such as video-fluoroscopy may not be appropriate in the advanced stages
- Any advanced care planning which may be in place for the patient; when considering alternative feeding measure.

Potential Interventions to Support Swallowing

- **Oro motor exercises** – lingual exercises may help improve co-ordination and strength of swallow, chewing sugar free gum may also be beneficial in the early stages. However, chewing gum may also increase saliva production which a patient may struggle to control.
- **Appropriate saliva management** – Some people with MSA may struggle to control excess saliva and this can have a negative impact on swallowing. A Speech and Language Therapist should consider this and seek medical input if felt appropriate.
- **Modified diet consistencies** – patients will often require softer diet consistencies and/or thickened drinks as the condition advances.
- **Swallow therapy exercises** – such as a chin tuck, the Masako manoeuvre or Supraglottic swallow technique if cognitively able. Swallow therapy exercises for people with MSA are unlikely to be appropriate in the latter stages, as it is likely that their swallow will continue to deteriorate alongside general cognitive decline. As with dementia patients, the RCSLT [8] does not recommend direct therapy exercises when working with people who have cognitive impairments. An individual capacity assessment should be made for each person with MSA, as cognition is usually largely preserved, but executive function disorder may be present.
- **Carer training** – (as with other progressive neurological conditions,) carers often play a vital role in ensuring safe delivery of nutrition and hydration by following SLT recommendations.
- **Environmental adaptations** – adaptive cutlery or plate guards to aid meal times and maintain independence and dignity for as long as possible. Smaller meals little and often may also help with fatigue. Liaise with Occupational Therapists or Dietitians.
- **Joint working** – An MDT approach could be used to advise appropriate positioning during meal times.
- **Alternative methods of feeding** – A nasogastric tube or a percutaneous endoscopic gastrostomy (PEG) to ensure adequate hydration and nutrition if appropriate (this should be discussed as part of a MDT). PEG for fluid intake may be a particularly relevant option if the person has symptoms of postural hypotension. Due to the progressive nature of the condition, the concept of alternative methods of feeding may be discussed within the early stages of MSA.
- **Medication** - If a patient is struggling with swallowing solids, they may need their medication altered to a more suitable form. The SLT could advise this to the patient's GP or speak to pharmacy directly.

Inspiratory Stridor

Inspiratory stridor is a symptom that is likely to emerge towards the latter stages of the condition [40]. This is likely to have a **negative impact on the safety of the swallow**, but also **affect vocal output** [41]. There is currently mixed research as to why this occurs in people with MSA. One theory is that it is caused by vocal fold adductor paralysis [42, 43]. Increased neurogenic atrophy within the intrinsic laryngeal muscles specifically, the posterior crico-aryteniod muscle, will also directly affect voicing ability [44]. Another theory is that poor vocal fold adduction is in fact caused by vocal fold dystonia [45]; which some studies suggest can be reduced by the use of Botox [46]. This worsening respiratory condition may significantly increase the patient's chances of developing aspiration pneumonia due to reduced airway protection.

Speech and Language Therapists may notice that their patients with MSA will have more breathing difficulties and increased dysphonia than those with Parkinson's disease. This will therefore mean **decreased vocal power** and a significantly **increased risk of aspiration during swallowing** [43]. The small apnoeic period that is experienced during swallowing may also be increased, causing greater fatigue during eating and drinking activities.

Speech and Language Respiratory Care Specialists are emerging within the clinical field [8]. However, due to the significant lack of these clinical roles within clinical teams, the Speech and Language Therapist should liaise with their respiratory team if the stridor becomes particularly intrusive on activities of daily living. Appropriate respiratory measures could then be applied if necessary.

Speech and Language Therapy in action: Case Example 1

Background

Patient X was a 66 year old lady diagnosed with probable MSA-P who was referred to her local Speech and Language Therapy service 12 months post-diagnosis. She was referred by her GP for increasing difficulties with her speech. X had reported no concerns regarding her eating and drinking ability, yet she was now avoiding high risk consistencies such as nuts and toast. It was also taking her longer to finish her meals and her husband supported her with this and other activities of daily living.

X was seen in an outpatient clinic for assessment, alongside her husband who was her main carer. X was predominantly wheelchair bound as the condition had significantly impacted on her mobility over the past 12 months. X was a smoker and had a pre-existing chronic obstructive pulmonary disease, which impacted on her breath support during conversation.

Assessment

An oral motor cranial nerve assessment was carried out with X to view any significant neurological impairment. The assessment revealed that she had reduced range of movements in her jaw (cranial nerve V), lips (cranial nerve VII), soft palate (cranial nerves IX & X) and tongue (cranial nerve XII). Although mild, these symptoms were negatively impacting on her ability to produce clear speech. Laryngeal movement was also significantly reduced on tactile assessment and her voice was markedly weak in presentation. The therapist therefore carried out a perceptual analysis of her voice using the GRBAS scale (Hirano, 1981).

Her results are depicted as follows:

Grade	Roughness	Breathiness	Asthenia	Strain
2	2	1	1	1

It is not uncommon for voice and speech to also be particularly affected in MSA due to a decline in oro lingual strength [13, 10]. The literature suggests that in people with a Parkinsonian related disease, it is not uncommon for their palatal movement to be significantly reduced on phonation [47]. She was asked to read 'The Grandfather Passage', [48], which revealed how her intelligibility decreased with utterance length, as expected from the literature.

In a quiet one to one environment in a clinic, she was largely intelligible however; this was likely to decrease significantly in a noisier environment.

Her dysarthria could be characterised as a mixed dysarthria with spastic and hypokinetic elements.

Management

X was encouraged to implement the following advice to help minimise the effects of her dysarthria. Her husband was also present to help implement the strategies:

- Avoid particularly loud or noisy environments
- Ensure people face you directly when talking with them
- Use shorter sentences or key words instead of long complex sentences
- Repeat key words if necessary or rephrase
- Spelling out words or using topic cues (or combination of both) can be useful to help people understand you
- Check that someone has thoroughly understood you
- Consideration was given towards using simple alphabet charts and speech apps, but was felt not to be appropriate at the time of assessment.

Outcome

X was still a functional and competent communicator who only required minimal environmental considerations and prompts for clearer articulation. At the time of assessment, X was still able to converse on the telephone. However, if she had been struggling with this she may have been advised to consider using a pre-prepared phrase in order for unfamiliar listeners to give her more time to speak. For example 'I have a difficulty with my speech, please be patient'. This could be facilitated by her husband and other family and friends. X was provided with recommendations to take away and they were advised that if they felt they had deteriorated further, to contact the speech and language therapy department.

Reflection

One point that X and her husband both commented on was the rapid deterioration of mobility

and general motor function (which in this case was impacting her speech). Over just 12 months, X had gone from being completely independent to requiring support for most activities of daily living.

The emotional impact of this was important for the Speech and Language Therapist to be able to empathise with and having prior knowledge about the rate of deterioration helped support the session and patient.

This may also reinforce the benefit of early discussions about future communication needs depending upon how much the person with MSA knows and understands about the course of their condition.

Case Example 2

Referral

A patient has been referred to the Speech and Language Therapy department following concerns relating to the safety of their swallow. The patient has a known probable diagnosis of MSA and is in the latter stages of the condition. The patient is 80 years old and has a supportive family. The medical team have requested a review is carried out whilst he is an inpatient in order to carry out an assessment and provide recommendations.

Dysphagia Assessment

Prior to assessment consent should be obtained from the patient, if the patient is felt to lack capacity to do this an assessment may be carried out in the patient's best interests.

In line with clinical dysphagia guidelines set out by the RCSLT, an oro motor assessment may be carried out in order to observe the impact of muscle weakness on the swallowing mechanism. It is likely that cranial nerves V, VII, X, XI and XII will all be directly affected and therefore reduce the safety and co-ordination of the swallow.

A clinical evaluation of swallow may then be carried out with the patient if appropriate. This can involve trialling modified consistency diets and establishing the safest form of oral intake for the patient. If the patient is thought to be acutely unwell, it may be appropriate for the medical team to consider alternative forms of nutrition and hydration, such as a nasogastric tube (NGT). However, it would also be important to consider their current respiratory status, due to the known co-morbidity of inspiratory stridor in some patients with MSA.

Dysphagia Management

Swallow therapy exercises such as a chin tuck or supraglottic swallow could reduce the risk of aspiration [38, 7] and improve co-ordination. However, it is unlikely that a patient would be able to retain and use these strategies effectively at every mealtime if they were in the latter stages of MSA. Instead, a modified consistency diet may be considered (if available) with support and assistance from carers. Consideration towards appropriate saliva management should also be discussed if the patient was not already receiving treatment.

Other Considerations

Ataxic symptoms and problems with overall co-ordination can be a particular problem for patients with probable MSA [49]. It can result in an uncoordinated swallow, especially during the oral stage [39]. MSA-P patients can experience on/off periods with medication; the

impact of which is likely to affect a patient's ability to communicate, swallow and mobilise [2].

As people with MSA often require regular medication [51], if they are seen to be struggling with solid tablet dose medications, an alternative form should be considered e.g. liquid or dispersible.

The Speech and Language Therapist can therefore advise the patient's GP or medical team to liaise with a Pharmacist to identify the appropriate alternatives available.

Summary

At present the exact nature of how quickly a swallow may deteriorate in a person with MSA is unclear. Recommendations may therefore be set with the knowledge that Speech and Language Therapy can be contacted if a review is required. For most patients with MSA, it is unlikely that a referral onto an instrumental dysphagia assessment (such as video fluoroscopy) would change the dysphagia management.

If X's swallow was seen to deteriorate further and they were aspirating on a range of consistencies, alternative feeding may be considered dependent on the individual [8]. This is the use of either a nasogastric tube (NGT; acute periods) or a percutaneous endoscopic gastrostomy (PEG; chronic periods) in order to maintain nutrition and hydration needs. Some researchers support the use of PEG insertion in MSA patients with severe dysphagia. They suggest that if a person with MSA suffers from severe dysphagia, a PEG may help reduce the risk of aspiration, provide adequate calorie intake and ensure medications are being received [52, 7, 51].

However, Gazulla et al [53] stated that although a PEG may reduce the risk of aspiration on food and fluids, a patient with MSA may still potentially aspirate on their own saliva. The cognitive decline associated with MSA may also mean that a patient would be at a high risk of pulling out a NGT or PEG, as often seen in patients with dementia [54,55]. It may only prolong a poor quality of life, so these factors would need to be thoroughly discussed with the patient, the family and medical team. Due to the poor prognosis for patients with MSA, alternative feeding may not be considered appropriate and a decision to "feed at risk" may be made by the medical team.

If it was decided that a patient was not appropriate for a PEG placement, an appropriate management plan for the family should be put in place. A report from Speech and Language Therapy may also be required in order to support appropriate care funding if required.

What next?

Currently, there are still no direct studies assessing appropriate therapeutic interventions for dysphagia in MSA [7]. However, parallels may be drawn between MSA and Parkinson's disease dysphagia management strategies due to the similar nature of their difficulties. Attention to MSA-specific characteristics such as laryngeal stridor and ataxia will be important for the Speech and Language Therapist to consider in relation to future dysphagia management.

Written by Lauren Gray, Speech and Language Therapist with special thanks to support from Gloucestershire Care Services Speech and Language Therapy department and members of the RCSLT.

This Version Date 12/19 | Review Date: 12/22 | Version: 2.0

The Trust's contact details:

MSA Trust, 51 St Olav's Court, Lower Road, London SE16 2XB
T: 0333 323 4591 | **E**: support@msatrust.org.uk | **W**: www.msatrust.org.uk

MSA Trust Nurse Specialists:

Samantha Pavey **T**: 0203 371 0003 | **E**: samantha.pavey@msatrust.org.uk

Katie Rigg **T**: 01434 381 932 | **E**: katie.rigg@msatrust.org.uk

Jill Lyons **T**: 01934 316 119 | **E**: jill.lyons@msatrust.org.uk

Emma Saunders **T**: 0330 221 1030 | **E**: emma.saunders@msatrust.org.uk

Social Welfare Specialist:

Jane Stein
01404 44241
jane.stein@msatrust.org.uk

Feedback: Your feedback helps us ensure we are delivering information to the highest standard. If you have any comments or suggestions please complete a short survey by following the links from our website: www.msatrust.org.uk or by contacting us at support@msatrust.org.uk.

References

1. A. Schrag, Y Ben-Shlomo, and NP Quinn. Prevalence of Progressive Supranuclear Palsy and Multiple System Atrophy: a cross sectional study. *The Lancet.* 2006; 354: 1771-1775.

2. Parkinson's UK website https://www.parkinsons.org.uk/information-and-support/what-parkinsons accessed 14th August 2019

3. Haw C, Trewhitt T, Boddy E, Evans J. Involving carers in communication groups for people with Parkinson's disease. (1999). *Parkinson's Disease: Studies in Psychological and Social Care.* BPS Books, Leicester, UK: Percival R, Hobson P (Eds).

4. PA Hanna, JJ Jankovic and JB Kirkpatrick. Multiple System Atrophy: The putative causative role of environmental toxins. *Arch Neuro* 199; 56:90-94.

5. Swan L, Dupont J. Multiple system atrophy. *Phys Ther*. 1999;79:488–494.

6. Schrag, A., Y. Ben-Shlomo, and N. P. Quinn. "Prevalence of progressive supranuclear palsy and multiple system atrophy: a cross-sectional study." *The Lancet* 354.9192 (1999): 1771-1775.

7 . Wenning, G., Fanciulli, A. (2014). *Multiple System Atrophy*. Austria: Springer-Verlag Wien.

8. Royal College of Speech and Language Therapists. (2005). *Clinical Guidelines: Disorders of Feeding, Eating, Drinking & Swallowing (Dysphagia).* RCSLT Clinical Guidelines. London: Speechmark.

9. Flabeau, O, Meissner, G and Tison, F (2010). Multiple System Atrophy: current and future approaches to management. *Therapeutic Advances in Neurological Disorders*. 2010;3(4): 249-263.

10. Penner, H., Miller, N., & Walters, M. (2007). Motor speech disorders in three parkinsonian syndromes: a comparative study. In *16th International Congress of Phonetic Sciences (ICPhS)*.

11. Dronkers, N.F. (1996). A new brain region for co-ordinating speech articulation. *Nature, 384, 159.*

12. Duffy, J. R. (2013). *Motor speech disorders: Substrates, differential diagnosis, and management*. Elsevier Health Sciences.

13. Kluin, K.J., Gilman, S., Lohman, M., Junck, L. (1996). Characteristics of the dysarthria of multiple system atrophy. Archives of Neurology 53, 545-548.

14. Knopp, D. B., Barsottini, O. G. P., & Ferraz, H. B. (2002). Multiple system atrophy speech assessment: study of five cases. *Arquivos de neuro-psiquiatria*, *60*(3A), 619-623.

15. Hartelius, L., Gustavsson, H., Astrand, M., Holmberg, B. (2006). Perceptual analysis of speech in multiple system atrophy and progressive supranuclear palsy. *JMSLP 14, 241-247.*

16. Rusz, J., Bonnet, C., Klempíř, J., Tykalová, T., Baborová, E., Novotný, M., Rulseh, A. and Růžička, E. (2015). Speech disorders reflect differing pathophysiology in Parkinson's

disease, progressive supranuclear palsy and multiple system atrophy. *Journal of neurology*, *262*(4), pp.992-1001.

17. Murdoch, B. E. (1998). *Dysarthria: A physiological approach to assessment and treatment*. Nelson Thornes.

18. Enderby, P., & Palmer, R. (1983). Frenchay Dysarthria Assessment (Austin, TX: PRO-ED). *CIT0007*.

19. Grossman, M. (1999). Sentence processing in Parkinson's disease. *Brain and cognition*, *40*(2), 387-413.

20. Murray, L.L. (2008). Language and Parkinson's disease. *Annual Review of Applied Linguistics, 28, 113–127.*

21. Spaccavento, S., Del Prete, M., Loverre, A., Craca, A., & Nardulli, R. (2013). Multiple system atrophy with early cognitive deficits: A case report. *Neurocase*, *19*(6), 613-622.

22. Walsh, B., & Smith, A. (2011). Linguistic complexity, speech production, and comprehension in Parkinson's disease: Behavioral and physiological indices. *Journal of Speech, Language, and Hearing Research*, *54*(3), 787-802.

23. Lieberman P, Kako E, Friedman J, Tajchman G, Feldman LS, Jiminez EB. (1992). *Brain Lang. 43(2), 169-89*.

24. Huh, Y. E., Park, J., Suh, M. K., Lee, S. E., Kim, J., Jeong, Y., ... & Cho, J. W. (2015). Differences in early speech patterns between Parkinson variant of multiple system atrophy and Parkinson's disease. *Brain and language*, *147*, 14-20.

25. Hirano, M. (1981). *Clinical examination of voice* (Vol. 5). Springer.

26. Skodda, S., Grönheit, W., Mancinelli, N., & Schlegel, U. (2013). Progression of voice and speech impairment in the course of Parkinson's disease: a longitudinal study. *Parkinson's Disease*, *2013*.

27. Haydock, S., Whitehead, D., & Fritz, Z. (Eds.). (2014). *Acute Medicine*. Cambridge University Press.

28. Herd, C. P., Tomlinson, C. L., Deane, K. H., Brady, M. C., Smith, C. H., Sackley, C. M., & Clarke, C. E. (2012). Speech and language therapy versus placebo or no intervention for speech problems in Parkinson's disease. *The Cochrane Library*.

29. Huckabee, M. L. (1992). Oral pharyngeal dysphagia: application of EMG biofeedback in the treatment of oral pharyngeal dysphagia. *Electromyography: applications in physical therapy. West Chazy, NY: Thought Technology*.

30. Schulz, G.M. and Grant, M.K., (2000). Effects of speech therapy and pharmacologic and surgical treatments on voice and speech in Parkinson's disease: a review of the literature. *Journal of communication disorders*, *33*(1), pp.59-88.

31. Sackley, C. M., Smith, C. H., Rick, C., Brady, M. C., Ives, N., Patel, R., & Clarke, C. (2014). Lee Silverman voice treatment versus standard NHS speech and language therapy versus control in Parkinson's disease (PD COMM pilot): study protocol for a randomized controlled trial. *Trials*, *15*(1), 213.

32. Fawcus, M., 2013. *Voice disorders and their management*. London: Springer.

33. Martinez-Martin, P., Rodriguez-Blazquez, C. and Forjaz, M.J., 2012. Quality of life and burden in caregivers for patients with Parkinson's disease: concepts, assessment and related factors.

34. Fox, C., Ebersbach, G., Ramig, L., & Sapir, S. (2012). LSVT LOUD and LSVT BIG: behavioral treatment programs for speech and body movement in Parkinson disease.

35. Tsanas, A., Little, M., Fox, C., & Ramig, L. O. (2014). Objective automatic assessment of rehabilitative speech treatment in Parkinson's disease. *Neural Systems and Rehabilitation Engineering, IEEE Transactions on*, *22*(1), 181-190.

36. Wenning, G. K., Ben-Shlomo, Y., Hughes, A., Daniel, S. E., Lees, A., & Quinn, N. P. (2000). What clinical features are most useful to distinguish definite multiple system atrophy from Parkinson's disease?. *Journal of Neurology, Neurosurgery & Psychiatry*, *68*(4), 434-440.

37. Vogel, A.P., Fendel, L., Paige Brubacher, K., Chan, V. and Maule, R., 2015. Dysphagia in spinocerebellar ataxia and multiple system atrophy-cerebellar. *Speech, Language and Hearing*, *18*(1), pp.39-43.

38. Logemann, J.A. (1998). *Evaluation and Treatment of Swallowing Disorders.* Second Edition. Austin: PRO-ED Inc.

39. Fernagut, P. O., Vital, A., Canron, M. H., Tison, F., & Meissner, W. G. (2011). Ambiguous mechanisms of dysphagia in multiple system atrophy. *Brain*, awr185.

40. Flabeau, O., Meissner, W. G., Ozier, A., Berger, P., Tison, F., & Fernagut, P. O. (2014). Breathing variability and brainstem serotonergic loss in a genetic model of multiple system atrophy. *Movement Disorders*, *29*(3), 388-395.

41. Lalich, I. J., Ekbom, D. C., Starkman, S. J., Orbelo, D. M., & Morgenthaler, T. I. (2014). Vocal fold motion impairment in multiple system atrophy. *The Laryngoscope*, *124*(3), 730-735.

42. Isozaki, E., Shimizu, T., Takamoto, K., Horiguchi, S., Hayashida, T., Oda, M., & Tanabe, H. (1995). Vocal cord abductor paralysis (VCAP) in Parkinson's disease: difference from VCAP in multiple system atrophy. *Journal of the neurological sciences*, *130*(2), 197-202.

43. Kurisaki, H. (1999). Prognosis of multiple system atrophy--survival time with or without tracheostomy. *Rinsho shinkeigaku, Clinical neurology, 39(5), 503-507.*

44. Yokoji, I., Nakamura, S., & Ikeda, T. (1997). [A case of progressive supranuclear palsy associated with bilateral vocal cord abductor paralysis]. *Rinsho shinkeigaku, Clinical neurology*, *37(6), 523-525.*

45. Vetrugno, R., Liguori, R., Cortelli, P., Plazzi, G., Vicini, C., Campanini, A., & Montagna, P. (2007). Sleep-related stridor due to dystonic vocal cord motion and neurogenic tachypnea/tachycardia in multiple system atrophy. *Movement disorders*, *22*(5), 673-678.

46. Merlo, I. M., Occhini, A., Pacchetti, C., & Alfonsi, E. (2002). Not paralysis, but dystonia causes stridor in multiple system atrophy. *Neurology*, *58*(4), 649-652.

47. Love, R.J., Webb, W.G. (2001). *Neurology for the Speech-Language Pathologist (4th Ed)*. Butterworth: Heinemann.

48. Darley, F. L., Aronson, A. E., & Brown, J. R. (1975). Motor speech disorders (3rd ed.). Philadelphia, PA: W.B. Saunders Company.

49. Wenning, G. K., Geser, F., Krismer, F., Seppi, K., Duerr, S., Boesch, S., ... & European Multiple System Atrophy Study Group. (2013). The natural history of multiple system atrophy: a prospective European cohort study. *The Lancet Neurology*, *12*(3), 264-274.

50. Müller, J., Wenning, G. K., Verny, M., McKee, A., Chaudhuri, K. R., Jellinger, K.,& Litvan, I. (2001). Progression of dysarthria and dysphagia in postmortem-confirmed parkinsonian disorders. *Archives of neurology*, *58*(2), 259-264.

51. Fanciulli, A., Wenning, G.K. (2015). Multiple system atrophy. *New England Journal of Medicine, 372, 249-63.*

52. Mehanna, R., & Jankovic, J. (2010). Respiratory problems in neurologic movement disorders. *Parkinsonism & related disorders*, *16*(10), 628-638

53. Gazulla, J., Berciano, J., Gilman, S., Wenning, G. K., & Low, P. A. (2015). Multiple-system atrophy. *The New England journal of medicine*, *372*(14), 1374-1374.

54. Finucane, T.E., Christmas, C., Travis, K. (1999). Tube feeding in patients with advanced dementia: a review of the evidence. *JAMA, 282, 1365-70.*

55. Kim, Y (2001) To Feed or Not to Feed: Tube Feeding in Patients with Advanced Dementia, *Nutrition Reviews, 59, 86-88*