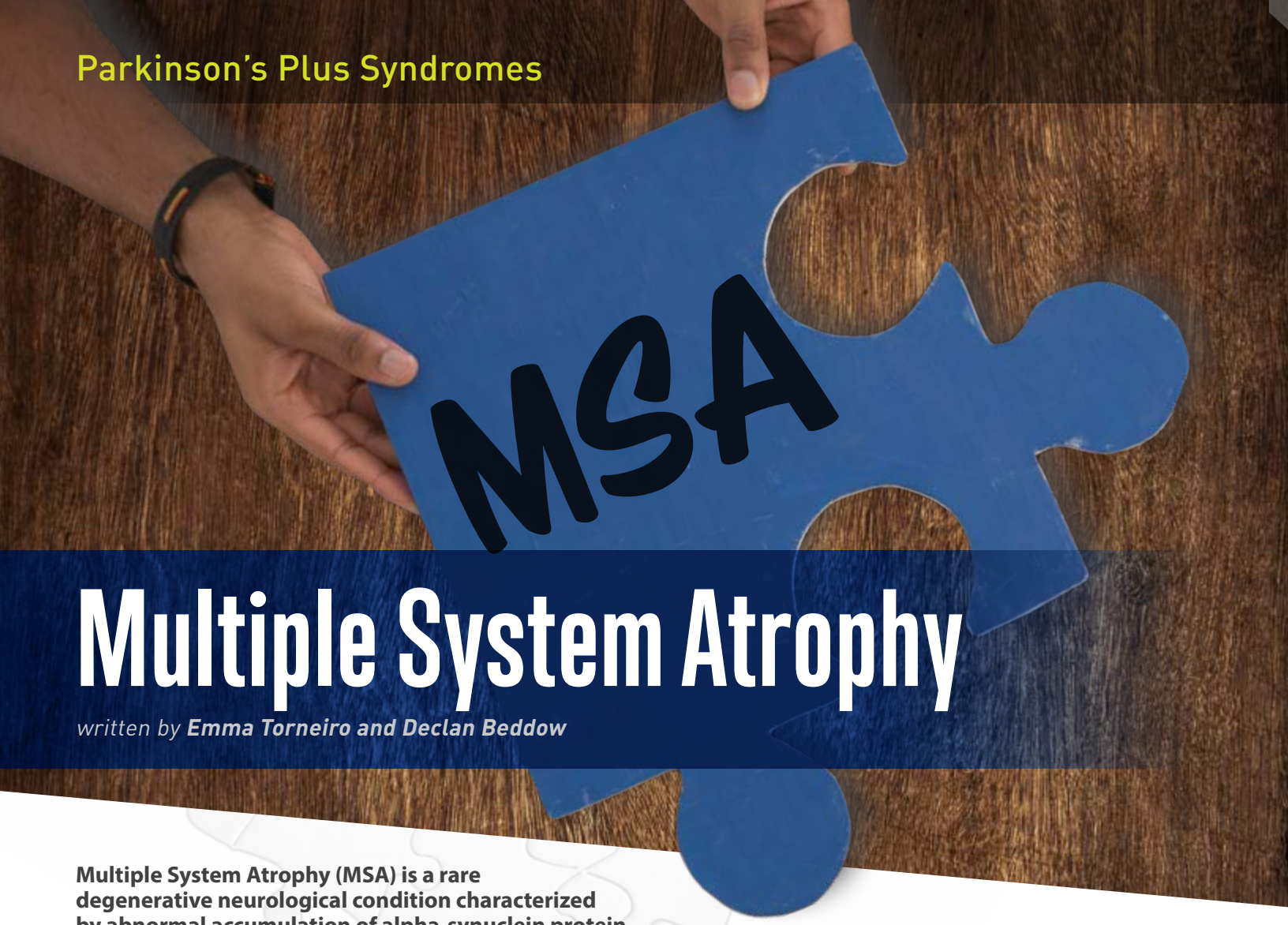

Multiple System Atrophy

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Multiple System Atrophy (MSA) is a rare degenerative neurological condition characterized by abnormal accumulation of alpha-synuclein protein (synucleinopathy1,2) in brain cells.

The alpha-synuclein protein is responsible for the transmission of messages between our brain cells and bodies[3]. What does this really mean? The alpha-synuclein protein creates clusters in the brain cells and disrupts the transportation of vital messages for bodily functions and movements. In MSA, shrinkage (atrophy) also occurs in parts of the brain responsible for movement and control. The onset of symptoms stems from the progressive loss of nerve cells in the brain and spinal cord[4,5]. The atrophy and protein accumulation lead to the motor, autonomic, speech, vision, and cognitive symptoms of MSA. The disease was initially described in 1969 as the Shy-Drager Syndrome stemming from the early contributions of the physicians Dr. Milton Shy and Dr. Glenn Drager.

The three types of MSA are Parkinsonian, Autonomic, and Cerebellar, which are differentiated by their associated symptoms. The Parkinsonian type (MSA-P) describes the presence of typical Parkinson symptoms (e.g., stiffness, rigidity, bradykinesia) with a degree of cerebellar dysfunction. The Autonomic type (MSA-A) describes dysfunction of autonomic nervous system (e.g., blood pressure, respiration, urination, digestion) with symptoms impacting involuntary functioning. Lastly, the Cerebellar type (MSA-C) describes symptoms connected to coordination, speech, and symptoms impacting voluntary movements.

There are many overlapping symptoms between MSA-P, MSA-A, and MSA-C, leading to the general classification of MSA. Similarly, to Parkinson disease (PD), the cause of MSA is unknown and there is currently no cure for the disease. The disease occurs sporadically, as it currently lacks genetic or environmental risk factors.

The diagnostic process for MSA involves an examination of personal medical history, symptoms, autonomic testing (primarily blood pressure), assessment of bladder functioning, and brain imaging scans (e.g., MRI for shrinkage or PET for metabolic function)[4]. Due to the scans being unable to differentiate between PD and MSA, many individuals initially receive a PD diagnosis. The diagnosis typically changes upon the recognition of faster progression by the healthcare team.

The Facts

- The average age of onset is 54
- Life expectancy is 7-10 years following symptom onset
- Prevalence is 6 people per million per year

Motor Symptoms

- Parkinsonian symptoms
- Difficulty initiating movement
- Action or coarse tremor, resulting in irregular movements
- Myoclonus (*rapid, jerky, and irregular movements in limbs or trunk*)[3]
- Earlier detection of postural instability comparative to PD
- Cerebellar Dysfunction
  - Incoordination in limbs, posture, and gait
  - Gait ataxia (*wide, irregular steps*)
  - Unsteady balance
  - Increased falls
- Pyramidal Tract Syndrome (*decrease in fine motor coordination*)[2]
- REM Sleep Behavior Disorder (*vividly acting out dreams*)
- Contractures (*chronic shortening of muscles around joints limiting movement*) in hands or limbs
- Pisa Syndrome (*curvature of spine or leaning towards one side*) similar to the Leaning Tower of Pisa[2,4]
- Antecollis (*a position where one is not able to keep their head up and looking forward, rather head is bent forward looking at the floor, forward flexion of neck*)[4] and inability to keep head lifted
- Camptocormia (*a "hunched" forward position, forward flexion of spine*)[2]
- Dystonia

Autonomic Dysfunction Symptoms

- Fluctuations in blood pressure
  - Orthostatic Hypotension (*low blood pressure when changing positions*)
  - Supine Hypertension (*high blood pressure when laying down*)[1]
  - Nocturnal Hypertension (*high blood pressure when sleeping*)
  - Fainting and Light-headedness
- Urinary incontinence, retention, or urgency
- Erectile dysfunction, impotence, bladder dysfunction, and constipation
- Nocturnal Stridor (*narrowing or obstruction of upper airways outside of chest cavity identified by strained high-pitched sound*)[2] or obstructive sleep apnea
- Anhidrosis (*reduced production of sweat, tears, and saliva*)[2] leading to heat intolerance and dry skin
- Inability to regulate body temperature, leading to color and temperature changes in hands and feet
- Inability to control emotions and expression of emotions (*laughing or crying at inappropriate times*)
- Irregular breathing patterns and heart rate patterns[3]
- Insomnia
- Involuntary sighing or gasping[4]

Vision Symptoms

- Blepharospasm (*twitching of eyelids*)
- Vertical Gaze Palsy (*limitations of eye gaze upwards or downwards*)
- Nystagmus (*involuntary, rapid, and jerky movements of eyes*)[6]
- Impaired vestibular-ocular reflex[6]
- Reduced accuracy of vision
- Double or blurred vision
- Visual hallucinations

Speech Symptoms

- Dysarthria (*difficulty with speech*)
- Slurred speech
- Dysphagia (*difficulty swallowing*)
- Hypophonia (*soft speech*)

Cognitive Symptoms

- Mild Cognitive Impairment
- Difficulties with Executive Functioning (*organizing information, understanding instructions, and decision-making processes*)

Treatment Options

There is no specific medication to treat MSA or slow the progression of symptoms, therefore treatments target management of symptoms and related conditions. The Parkinson's medication, Levodopa Carbidopa, may initially improve motor functions (slowness and stiffness) with benefits being limited as disease progresses. The healthcare team can also prescribe additional medications to manage the treatable conditions of MSA (e.g., erectile dysfunction, bladder control, blood pressure, myoclonus, dystonia).

The main treatment options for MSA focus on using therapies for management of symptoms and improve quality of life. The recommended therapies include physiotherapy for issues with balance and gait, occupational therapy for assessment of mobility aids and improvements to functional abilities, and speech therapy for challenges with speech, swallowing, and communication. The use of a dietitian is beneficial for help with proper nutrition, finding easy to swallow diets, and managing blood pressure. Finally, psychologist or social workers can provide support for the non-motor symptoms associated with the diagnosis.

Resources

1 *Multiple system atrophy (MSA). (2021, May 21). Mayo Clinic. Retrieved March 2022 from https://www.mayoclinic.org/diseases-conditions/multiple-system-atrophy/symptoms-causes/syc-20356153*

2 *Multiple System Atrophy. (n.d.). National Organization for Rare Diseases (NORD). Retrieved March 2022 from https://rarediseases.org/rare-diseases/multiple-system-atrophy/*

3 *Golbe, L.I. (2019). MSA: Some Answers. CurePSP. https://www.psp.org/wp-content/uploads/2021/04/2019-MSA-SOME-ANSWERS-BROCH_web.pdf*

4 *Multiple System Atrophy Fact Sheet. (2022, April 15). National Institute of Neurological Disorders and Stroke. Retrieved March 2022 from https://www.ninds.nih.gov/health-information/patient-caregiver-education/fact-sheets/multiple-system-atrophy-fact-sheet*

5 *Multiple System Atrophy. (2014, November). National Institute of Neurological Disorders and Stroke. Retrieved March 2022 from https://web.archive.org/web/20160406203452/http:/www.ninds.nih.gov/disorders/msa/multiple_system_atrophy_pamphlet.pdf*

6 *Armstong, R.A. (2014). Visual signs and symptoms of multiple system atrophy. Clinical and Experimental Optometry, 97(6), 483-491. https://doi.org/10.1111/cxo.12206*