CMS Council for Medical Schemes

CMScript

Member of a medical scheme?
Know your guaranteed benefits!

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Dystonia is a neurological condition characterised by involuntary and sustained muscle spasms as a result of incorrect signals from the brain. These muscle spasms tend to force affected parts of the body into abnormal movements or postures. The condition may affect speech, sight and mobility but not intellect. Living with dystonia can be painful and debilitating, as well as embarrassing and stigmatising.

Causes

Causes vary and include gene mutations, brain injuries and tumors, inborn errors of metabolism, exposure to drugs or chemicals.

Classification

Dystonia syndromes are classified along three axes, namely, cause, age of onset and body distribution

Causes

- Inherited dystonias are genetic in origin through gene mutations.
- Acquired dystonia have a specific cause such as brain injury and tumors, encephalitis, exposure to drugs or chemicals.
- Some of the causes are idiopathic (meaning that the cause is unknown)

Age at onset

Symptoms may first appear in infancy (birth to 2 years), childhood (3–12 years), adolescence (13–20 years), early adulthood (21–40 years), or late adulthood (>40 years)

Body distribution

The classification of involvement is:

- Focal – involves a single body part such as blepharospasm (forceful closure of the eye), oromandibular dystonia (forceful contractions of the face, jaw, and/or tongue), and dystonia of neck muscles, laryngeal dystonia, and writer's cramp.
- Segmental – affects two or more adjacent parts of the body. For example, cranial dystonia (blepharospasm with lower facial and jaw or tongue involvement).
- Multifocal - affects two or more unrelated body parts
- Generalized - affects most or the whole body
- Hemidystonia – half of the body is affected

The body distribution may change over time with progression to the involvement of previously uninvolved sites.

http://www.nature.com/nrn/journal/v9/n3/images/nrn2337-f1.jpg

Diagnosis

Dystonia can be difficult to diagnose because of its variable presentation, variation of causes and coexistence with other movement disorders. The diagnosis includes making sure that common treatable causes of abnormal body movement are ruled out. That is, one may need to do electrolytes, calcium and magnesium, thyroid functions and stop medication to ensure that Dystonia is not from a treatable cause. The core manifestation of this condition is abnormal postures and involuntary muscle spasms with or without tremors. The diagnosis is therefore primarily based on clinical presentation or signs and symptoms related to dystonia.

Structural brain imaging (MRI) is required in generalised or hemidystonia and if there are any features to suggest that there may be an identified neurological condition, such as a focal brain lesion (abnormal tissue in the brain).

Neurophysiological tests are not routinely recommended for the diagnosis or classification of dystonia. However, multiple simultaneous electromyography (EMG) which is a test done to record

the electrical activity of the muscles may contribute to the clinical assessment by showing characteristic features of dystonia.

2 Treatment

Oral medication
These medicines are often used in the treatment of dystonia. Drugs such as anticholinergics, dopamine depleting agents, benzodiazepines, anti-epileptics and baclofen are used in the treatment of the condition. However, patients may be addicted to benzodiazepines.

Chemical denervation
Botulinum toxin (Botox) injections are injected into the affected muscles and are considered to have revolutionised the treatment of dystonia. The effect of the toxin wears off, therefore the injections are repeated every 12 weeks. Treatment is symptomatic.

Surgical options
These include peripheral denervation, intrathecal baclofen and deep brain stimulation (DBS). Where all other surgical treatments have failed to provide adequate improvement, DBS is considered a good option. In this procedure, two fine electrodes are inserted into the brain powered by a battery implanted in the chest. The electrodes send a pulse that changes the signals from the brain that cause the involuntary muscle spasms.

Cognitive Behavioural Therapy (CBT)
Currently, CBT is considered an experimental treatment as there is little research evidence about the use of this therapy in dystonia. The principles on which CBT is based suggest that it may be helpful in managing the condition.

Referrals

Physiotherapy
In focal dystonia's, the use of rehabilitative physiotherapy in treating the condition is well developed and structured. It aims to give patients as much independence as possible and also helping in correcting the affected function through specific interventions. Despite therapeutic handling methods being useful for generalised dystonia's, the dystonic posture or movement tends to return when the therapist stops the treatment.

Pain management
Pain resulting from dystonia can be in the muscles affected by spasms, or in joints where bone surfaces rub together due to twisting of posture and limbs. The resulting intractable pain sometimes dominates the patient's life and might be unresponsive to medication. Sometimes such patients are referred to a pain specialist for pain management.

Speech and language therapy
For patients with oromandibular and generalised dystonia's with articulation difficulties, mouth and swallowing exercises help to reduce the risk of choking. For those with spasmodic dysphonia, techniques to help them speak include breathing exercises and ways to make best use of the voice and sound, albeit with limited effect.

Occupational therapy
Occupational therapy can help people with dystonia with practical ways of dealing with everyday tasks allowing them to live as independently as possible at home, at work or in their studies. Support includes identifying ways of dealing with difficult tasks and recommending alterations or adaptations in the home, school or workplace environment.

Podiatry
Patients may experience gait problems and struggle to look after their own feet because of poor mobility, poor dexterity or problems caused by uncontrollable muscle spasms. Podiatrists help them to address these problems using foot orthotics to assist with gait problems.

Psychiatrist / psychologist / counsellor
Dystonia is not a mental health condition but it can precipitate severe depression and anxiety due to pain, stigma, employment difficulties and social isolation. Psychological therapies and counselling therefore play an important role in managing the condition.

Genetic Counselling
Adults with genetic forms of dystonia who are considering having children may have concerns about their children also developing dystonia. They may decide to seek genetic counselling to help inform their decision making.

What is covered under PMBs?

Dystonia is a PMB condition under Diagnostic Treatment Pair (DTP) code 341A. The DTP refers to Basal ganglia, extra-pyramidal disorders; other dystonia's NOS. The treatment component specified for this DTP is initial diagnosis; initiation of medical management.

All medical schemes are required by law to pay for the diagnosis, treatment and care costs of PMB conditions as prescribed. In case of dystonia, the medical schemes are required to pay for the initial diagnosis; initiation of medical management as PMB level of care.

Initial diagnosis includes all the tests done to confirm or exclude the condition whilst initiation of medical management applies to the first prescription of medication.

Whilst the disease is debilitating and requires continuous care to improve functionality, it is covered as a prescribed minimum benefit for initial diagnosis and initiation of medical management. Some schemes may fund for continuation of care from discretionary risk pool benefits, day to day benefits or from the medical savings account. In cases where members do not have cover and cannot afford out of pocket payments for continuation of

care, state hospitals must be considered for continuation of care. It is very important to confirm with the medical scheme about the benefits available for the condition. If the doctor deems it necessary for the medication, tests or procedures to be done that the medical scheme does not normally fund, the doctor should write a clinical motivation to the scheme for payment to be considered as PMB only if the requests relate to the initial diagnosis and initiation of medical management.

References

The Dystonia Society. 2011. Dystonia: A guide to good practice for health and social care professionals. London
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PMBs

Prescribed minimum benefits (PMBs) are defined by law. They are the minimum level of diagnosis,treatment, and care that your medical scheme must cover – and it must pay for your PMB condition/s from its risk pool and in full. There are medical interventions available over and above those prescribed for PMB conditions but your scheme may choose not to pay for them. A designated service provider (DSP) is a healthcare provider (e.g. doctor, pharmacist, hospital) that is your medical scheme's first choice when you need treatment or care for a PMB condition. You can use a non-DSP voluntarily or involuntarily but be aware that when you choose to use a non-DSP, you may have to pay a portion of the bill as a co-payment. PMBs include 270 serious health conditions, any emergency condition, and 25 chronic diseases; they can be found on our website by accessing the link provided (www.medicalschemes.com/medical_schemes_pmb/index.htm).

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