What is Cervical Dystonia?

Dystonia Educational Series

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Cervical dystonia is a form of abnormal movement characterized by abnormal head and neck position caused by involuntary contraction of neck muscles. This may result in sustained abnormal position or jerky repetitive movements mimicking tremor.
Cervical dystonia more commonly effects women than men and usually begins in middle age, typically between the ages of 30 and 60 though it may begin in younger individuals or in the elderly. The head tends to presume a dominant position with either turning, tilting, or shifting of the neck to one side or the other. The movements are a result in sustained abnormal posturing, jerky tremulous-like movements or a combination of both sustained posture plus jerky movements.

**Characteristics of Cervical Dystonia**

- Tends to pull head to one side or the other.
- movements either sustained, jerky, or a combination of the two

**Age of Onset:**
- Any age, most commonly affects those of middle age (30-60 years old)
A variety of different abnormal head postures may be seen. Torticollis refers to turning of the head to the right or left. Laterocollis means tilting of the head to the right or left.
Retrocollis refers to pulling of the head backward, and anterocollis tilting of the head forward. In addition, the head may shift laterally or to the sides toward one shoulder or the other or may shift forward - like a duck may move their head forward or backward. Forward and backward shift of the neck is referred to as sagittal shift.
The abnormal contractions of the neck may also involve the shoulder resulting in shoulder elevation or forward deviation. Jerky tremulous-like movements often occur and the majority of patients have associated neck and shoulder pain. Pain in some patients may be even more disabling than the abnormal movements. Symptoms of cervical dystonia gradually come on over one to two years and then tend to stabilize. Remission is seen in only a small percentage of patients, and even in those in whom remission occurs, the symptoms typically come back. Cervical dystonia can occur by itself and is then considered a focal cervical dystonia or can be part of more widespread dystonia. When cervical dystonia is combined with dystonia involving the face, it is referred to as craniocervical segmental dystonia. Cervical dystonia may also occur as part of widespread dystonia involving several limbs and the trunk, in which case it is part of generalized dystonia.
When cervical dystonia remains focal and does not involve other areas of the body, it is generally idiopathic; meaning we do not know the underlying cause. It is thought that in most of these cases, there is an underlying genetic cause, but we have not yet identified the gene. Recently, three different genes have been identified that may cause familial isolated cervical dystonia, but it is thought that these genes account for only a very small percentage of patients who have focal cervical dystonia. Cervical dystonia may also occur due to acquired causes such as exposure to dopamine-blocking drugs or rarely due to head or neck trauma. There is some controversy as to whether or not posttraumatic cervical dystonia is the same disorder as cervical dystonia, which comes on unassociated with trauma.
The majority of patients with cervical dystonia are initially misdiagnosed as having a stiff neck or arthritis involving the neck. When jerky or tremulous movements are present resulting in dystonic head tremor, patients are often misdiagnosed as having essential head tremor. Given the fact that no specific tests are available to make the diagnosis and the diagnosis based on careful history and examination, it is important to see a neurologist or other physician who specializes in movement disorders to be certain of having an accurate diagnosis.
Unfortunately, no cure is available for focal cervical dystonia and all of our treatments are aimed at reducing symptoms and improving quality of life. Specifically, the available treatments may improve the abnormal head positioning and jerky movements, reduce pain, and help to reduce development of secondary complications such as accelerated neck arthritis. There are three main forms of treatment: Botulinum toxin injections are the standard first-line treatment. Oral medications may be used typically in patients who do not have an adequate response to botulinum toxin injections alone. Focal surgery to cut different nerves and muscles or deep brain stimulation may be used in patients with severe dystonia who do not have an adequate response to more conservative treatments.
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