

## What is the Cause?

Scientists believe dystonia symptoms result from improper signals in the nervous system that cause muscles to contract involuntarily. Researchers and doctors do not yet fully understand the neurological mechanisms that cause this abnormal muscle contraction.

Cervical dystonia may be isolated (primary) or acquired (secondary). In isolated dystonia there is no evidence of any identifiable cause for the dystonic symptoms. Isolated cervical dystonia may be genetic and typically affects adults. Widespread epidemiological studies are needed, but estimates suggest that isolated cervical dystonia affects no fewer than 40,000 people in the United States. Cervical dystonia may occur in the context of early-onset generalized dystonia, which is often associated with a mutation in the DYT1 gene.

Acquired dystonias are caused by specific structural or metabolic causes and are usually associated with additional neurological symptoms. The most common causes of acquired cervical dystonia are physical trauma (about 10% of reported cases) and drug reaction (less than 10% of reported cases). Drug-induced cervical dystonia is most commonly attributed to neuroleptic medications. Acquired dystonia may occur at various ages, depending on the causative event or condition. Children with cerebral palsy may have secondary dystonia symptoms including cervical dystonia.

## What is Cervical Dystonia?

*Dystonia* is a movement disorder characterized by uncontrollable muscle contractions that cause involuntary, often repetitive, body movements and/or postures.

*Cervical dystonia* is a specific form that affects the head and neck. Cervical dystonia produces excessive muscle contractions in the neck. These muscle contractions cause involuntary movements and awkward positions of the head, neck, and sometimes shoulders.

Cervical dystonia may cause the:

- Neck to twist or tilt to the side
- Head to tip forward or back
- Shoulder to elevate toward the ear
- Neck to shift away from the midline of the body

Cervical dystonia is among the most common forms of dystonia seen in movement disorder clinics. Cervical dystonia is sometimes referred to as *spasmodic torticollis*.

## What Support is Available?

The Dystonia Medical Research Foundation ([www.dystonia-foundation.org](http://www.dystonia-foundation.org)) can provide educational resources, self-help opportunities, contact with others living with cervical dystonia, volunteer opportunities, and connection to the greater dystonia community.

## What is the DMRF?

The Dystonia Medical Research Foundation (DMRF) is a 501(c)3 non-profit organization that has served the dystonia community since 1976. The DMRF funds medical research toward a cure, promotes awareness and education, and supports the well being of affected individuals and families.

The DMRF can put you in touch with others with dystonia for networking and support.

### To learn more about dystonia and the DMRF, contact:

#### Dystonia Medical Research Foundation

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Chicago, Illinois 60601-1980

**Phone:** 312-755-0198

**Toll free:** 800-377-DYST (3978)

**Email:** [dystonia@dystonia-foundation.org](mailto:dystonia@dystonia-foundation.org)

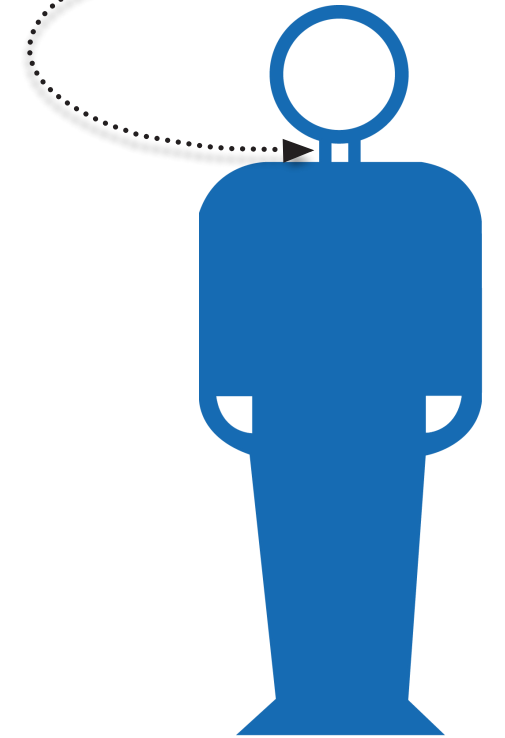
**Web:** [www.dystonia-foundation.org](http://www.dystonia-foundation.org)

*Connect with the DMRF on Facebook, Twitter, and YouTube.*

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# Cervical Dystonia

## DYSTONIA OF THE NECK MUSCLES



## Are There Different Forms or Varieties?

Yes. Cervical dystonia symptoms may be sustained (“tonic”), causing abnormal postures of the head and neck, or jerky (“clonic”), causing abrupt head movements. Many individuals have a combination of sustained and jerky movements. Jerky movements may resemble a tremor, and sometimes dystonia may be confused with essential tremor.

Cervical dystonia may cause the:

- Neck to twist to the side (“torticollis”)
- Head to tip forward (“anterocollis”)
- Head to tip back (“retrocollis”)
- Head to tilt toward the ear (“laterocollis”)
- Shoulder to elevate toward the ear
- Neck to shift away from the center of the body

Most people have a combination of the movements listed above. Estimates suggest that up to 75% of people with cervical dystonia experience pain in the neck and shoulder area. Headaches are also frequently reported.

It is not unusual for individuals with cervical dystonia to have hand tremors.

## What Treatments are Available?

Botulinum neurotoxin injections are typically the first line treatment for cervical dystonia. The injections are given intramuscularly, often with electromyography (EMG) or ultrasound guidance, and must be repeated every three to six months.

Although a medication with the word “toxin” in the name may seem confusing, botulinum neurotoxin injections have decades of research and clinical experience demonstrating that they are a safe and effective medical therapy. Botulinum neurotoxin has been approved for use in the United States since 1989. Several brands of botulinum neurotoxin are commercially available.

A multitude of oral medications have demonstrated some benefit, but no single drug has proven effective for a majority of patients. The categories of drugs reported to help relieve symptoms include:

- Anticholinergic medications such as Artane® (trihexyphenidyl) and Cogentin® (benztropine)
- Dopaminergic drugs such as Sinemet® or Madopar® (levodopa) and dopamine agonists
- Muscle relaxants such as baclofen
- Benzodiazepines such as clonazepam and Valium® (diazepam)

Surgical procedures such as deep brain stimulation or selective denervation may be recommended in select cases.

Individuals with cervical dystonia may be at increased risk for depression and anxiety disorders, so addressing emotional and mental health may be an important part of the treatment strategy. Gentle physical therapy with a physical therapist who specializes in neurological disorders may preserve/improve range of motion and help reduce pain. For some individuals, specially constructed cervical braces may be useful to improve position of the head and/or serve as a substitute for a sensory trick (see below).

Complementary therapies should be explored, and regular relaxation practices may improve discomfort, pain, and general well being.

## How is This Likely to Change or Progress Over Time?

Symptoms usually plateau and remain stable within a few years of onset. The symptoms, however, may continue to change or fluctuate in severity over time. In approximately one-third of people with cervical dystonia, the symptoms spread to nearby body areas such as the face, jaw, shoulder, and/or arm.

Cervical dystonia symptoms are often partially relieved by a “sensory trick” (also known as geste antagoniste) such as gently placing a hand on the chin, other areas of the face, or back of the head. A person with isolated (primary) cervical dystonia is more likely to respond to a sensory trick than someone with acquired (secondary) cervical dystonia.

## How is This Going to Affect My Daily Life?

Living well with cervical dystonia is possible. The early stages of onset, diagnosis, and seeking effective treatment are often the most challenging. The symptoms may vary from mild to severe, and symptoms often fluctuate from day to day. Cervical dystonia often causes significant physical pain and fatigue. Individuals may have to learn new ways to accomplish daily tasks.

Individuals living with dystonia are strongly encouraged to:

- Seek out the best medical care.
- Educate yourself about dystonia and treatment options.
- Develop a multi-layered support system of support groups, online resources, friends, family, and mental health professionals, if needed.
- Investigate complementary therapies.
- Get active within the dystonia community.

## How Do I Find the Best Doctor?

The type of subspecialist with the expertise to diagnose and treat dystonia is typically a neurologist with special training in movement disorders.