

What is the Cause?

The cause of dystonia is not known at this time. 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 primary or secondary. In primary dystonia there is no evidence of any identifiable cause for the dystonic symptoms. Primary cervical dystonia may be genetic and typically affects adults. Widespread epidemiological studies are needed, but estimates suggest that primary cervical dystonia affects no less than 4,000 people in Canada. Cervical dystonia may occur in the context of early-onset generalized dystonia, which is often associated with a mutation in the DYT1 gene.

Secondary dystonias are caused by specific structural or metabolic causes and are usually associated with additional neurological symptoms. The most common causes of secondary 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. Secondary 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. The total number of Canadians living with secondary dystonia is not known.

What Is DMRF Canada?

The Dystonia Medical Research Foundation Canada (DMRFC) is a registered non-profit Canadian charity. DMRFC funds medical research toward a cure, promotes awareness and education, and supports the well being of affected individuals and families. DMRF Canada works in partnership with the Dystonia Medical Research Foundation in the United States to ensure funding of the best and most relevant dystonia medical research worldwide and partners with the Canadian Institute of Health Research (CIHR) in funding excellent dystonia research in Canada.

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Cervical Dystonia/Spasmodic Torticollis

**DYSTONIA OF THE
NECK MUSCLES**



What is Cervical Dystonia?

Dystonia is a neurological movement disorder, and *cervical dystonia* is a specific form that affects the head and neck. Cervical dystonia produces patterned, repetitive muscle contractions in the neck. These muscle contractions cause abnormal movements and awkward postures of the head, neck, and sometimes shoulders.

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



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 which is a separate movement disorder.

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?

One of the most effective treatments for cervical dystonia is regular botulinum neurotoxin injections to the affected muscles. It is unfortunate that a therapy that has improved the lives of countless individuals with cervical dystonia cannot shed the term “toxin” from its name. Understandably, the idea of injecting a “toxin” into the body may cause concern.

Botulinum neurotoxin injections have decades of research demonstrating that they are a safe and effective medical therapy. Botulinum neuro-toxin has been approved for use in Canada since 1990. 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)

In recent years, selective denervation surgery is used less frequently for cervical dystonia patients. Deep brain stimulation has been explored for the treatment of cervical dystonia with promising results.

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 primary cervical dystonia is more likely to respond to a sensory trick than someone with secondary cervical dystonia.

How is This Going to Affect My Daily Life?

Living successfully 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 kind of doctor who is typically qualified to treat cervical dystonia is a neurologist who specializes in movement disorders.

What Support is Available?

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