

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 the abnormal muscle contraction.

Oromandibular dystonia may be isolated (primary), meaning that it is the only apparent neurological disorder, with or without a family history. Cases of inherited oromandibular/cranial dystonia have been reported, often in conjunction with generalized dystonia. Oromandibular dystonia may also be acquired from secondary causes such as drug exposure or disorders such as Wilson's disease. Drug-induced dystonia often first manifests as symptoms in the facial muscles

Oromandibular dystonia should not be mistaken for temporomandibular joint disease (TMJ), which is an arthritic condition.

What is Oromandibular Dystonia?

Dystonia is a neurological movement disorder characterized by persistent or periodic muscle contractions that cause abnormal, often repetitive, body movements, postures, or both. The movements are usually patterned and twisting, and may resemble a tremor.

Oromandibular dystonia, sometimes referred to as *cranial dystonia*, is a specific form characterized by forceful muscle contractions of the face, jaw, and/or tongue. The muscle contractions cause involuntary movements that interfere with opening and closing the mouth and may affect chewing and speech.

The dystonic contractions of the muscles may cause:

- Mouth to open involuntarily
- Jaw to clench shut
- Bottom jaw to shift toward one side or to the back
- Involuntary movements in the face and lips
- Involuntary movements in the tongue

Oromandibular dystonia symptoms usually begin later in life, between the ages of 40 and 70 years, and appear to be more common in women than in men.

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:

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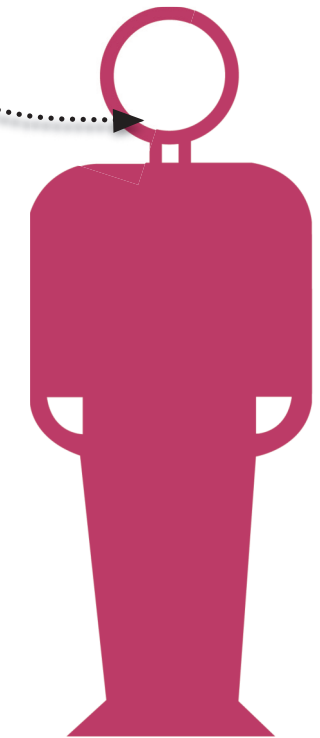
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Oromandibular Dystonia (Cranial Dystonia)

· DYSTONIA OF THE
JAW, FACE & TONGUE



**DYSTONIA
MEDICAL
RESEARCH
FOUNDATION**

Are There Different Forms of Oromandibular Dystonia?

Oromandibular dystonia symptoms may cause the jaw to open, the jaw to close, the jaw to deviate from center, the jaw to retract back into the head, or a combination of these movements. Another term used to describe oromandibular dystonia is *cranial dystonia*. (Cranial dystonia is a broad description for dystonia that affects any area of the face and mouth.) Dystonia that affects the face and lip muscles of musicians who play wind instruments is called *embouchure dystonia*. Dystonia that specifically affects the tongue is called *lingual dystonia*.

Oromandibular dystonia may occur with additional dystonia symptoms. Oromandibular dystonia is often associated with dystonia of the neck muscles (cervical dystonia/spasmodic torticollis), eyelids and brow (blepharospasm), or larynx (spasmodic dysphonia/laryngeal dystonia). When oromandibular dystonia occurs with blepharospasm, it may be called *Meige's syndrome*. Sometimes symptoms of oromandibular dystonia are task-specific and occur only during certain activities such as speaking or chewing. Paradoxically, in some people, activities like speaking and chewing reduce symptoms. Difficulty in swallowing is a common aspect of oromandibular dystonia if the jaw is affected, and spasms in the tongue can make it difficult to eat and swallow.

What Treatments are Available?

Treatment for oromandibular dystonia must be highly customized to the individual. Several oral medications have been studied to determine benefit for people with oromandibular dystonia. About one-third of people's symptoms improve when treated with oral medications such as Klonopin® (clonazepam), Artane® (trihexyphenidyl), diazepam (Valium®), tetrabenzine, and/or Lioresal® (baclofen).

Although the symptoms may vary from person to person, many people with oromandibular dystonia experience some reduction of spasms and improvement of chewing and speech after injection of botulinum neurotoxin into the masseter, temporalis, and lateral pterygoid muscles. The injections may be given 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.

Botulinum neurotoxin injections are most effective in jaw-closure dystonia, while treating jaw-opening dystonia may be more challenging. Botulinum neurotoxin injections may also be an option for lingual dystonia. Side effects such as swallowing difficulties, slurred speech, and excess weakness in injected muscles may occur, but these side effects are usually temporary and relatively well tolerated.

Oromandibular dystonia may respond to the use of sensory tricks to temporarily reduce symptoms. For

example, gently touching the lips or chin, chewing gum, talking, biting on a toothpick, wearing an orthodontic device or mouth guard, or placing a finger near an eye or underneath the chin may cause symptoms to subside temporarily. Different sensory tricks work for different people, and if a person finds a sensory trick that works, it usually continues to work.

Speech and swallowing therapy may lessen spasms, improve function, strengthen unaffected muscles, and facilitate easier speech and swallowing. Regular relaxation practices may benefit overall well being.

Will My Symptoms Change or Progress Over Time?

Individuals with oromandibular may first notice symptoms as a twitching or tightness in the jaw or face. Symptoms evolve over time and then typically stabilize. Symptoms are typically worsened by stress and may improve after sleep (though symptoms of acquired oromandibular dystonia may persist in sleep).

How is This Going to Affect My Daily Life?

Living well with oromandibular dystonia is possible. The early stages of symptom 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. Symptoms may interfere with eating and drinking, and fatigue is common due to the exertion required to speak or maintain positioning of the jaw. Individuals may have to learn new ways to accomplish daily tasks.

Individuals living with oromandibular dystonia are strongly encouraged to:

- Seek out the best medical care.
- Educate yourself about oromandibular 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.

What Kind of Doctor Treats Oromandibular Dystonia?

The kind of doctor who is typically trained to diagnose and treat oromandibular dystonia is a neurologist who specializes in movement disorders. Ongoing treatment may require consultations with multiple specialties. For example, a neurologist or otolaryngologist (ear, nose, and throat specialist) may administer botulinum neurotoxin injections. A speech-language pathologist may conduct swallowing therapy.

What Support is Available?

The Dystonia Medical Research Foundation can provide educational resources, self-help opportunities, contact with others living with oromandibular dystonia, volunteer opportunities, and connection to the greater dystonia community.