

What is Blepharospasm?

Dystonia Educational Series



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What is Blepharospasm?

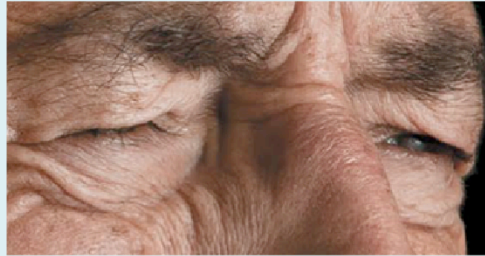
Blepharospasm is a focal dystonia characterized by repetitive, sustained contractions of the muscles around the eyes and eyebrows. This results in involuntary eye closure.

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Blepharospasm is a form of focal dystonia characterized by involuntary eye closing. There is repetitive sustained contractions of the muscles which close the eyes and often associated muscles which cause moving of the nose and eyebrows.

Symptoms

- spectrum of severity ranges from increased blinking to becoming functionally blind
- involuntary closure of the eyelid can occur
- symptoms always in both eyes
 - one eye may be affected more than the other



Overview of BOTOX® for cervical dystonia and blepharospasm. (2012) Botox Medical. Allergan, Inc.

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Blepharospasm usually begins in middle age, typically around the ages of 40 to 60 though it may begin in patients who are younger or older. These symptoms begin gradually, over several months and begin with excessive blinking usually associated with light sensitivity or eye irritation. The symptoms may remain isolated to just the eye or may spread to involve the lower face resulting in facial grimacing or abnormal jaw clenching or opening. When the upper and lower face are both involved, the disorder is sometimes referred to as Meige's syndrome. If the symptoms also spread to involve the neck, with associated cervical dystonia, this disorder is referred to as craniocervical segmental dystonia.

Characteristics

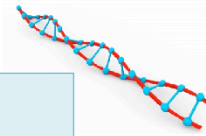
- symptoms can spread to other parts of the body, such as the jaw, mouth, neck over time
 - small percentage of patients experience this
- may occur as part of generalized dystonia
- patients may have light sensitivity

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The severity of the symptoms range from simply increased blinking resulting in minor embarrassment to pronounced involuntary eye closing with sustained spasms, which may render the patient functionally blind and interfere with day-to-day activities including reading, watching TV, and driving. Blepharospasm always affects both eyes, though one eye may be affected more than the other.

Causes

- no genes have been directly linked to isolated blepharospasm
- acquired
 - parkinsonism syndrome (PD, PSP)
 - drug induced
 - rarely due to focal brain lesion (from a head injury)

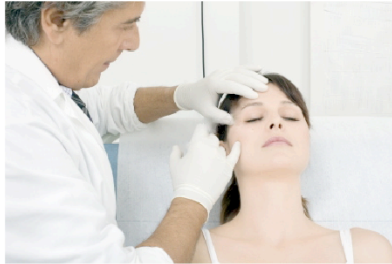


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Most isolated blepharospasm is said to be idiopathic, meaning we do not know the underlying cause and no single gene has been directly linked to isolated blepharospasm. Blepharospasm may occur though as part of a more widespread genetic dystonia for which various genes have been identified. Isolated blepharospasm may be acquired and occur as a result of exposure to different medications, especially drugs which block the chemical dopamine such as certain drugs to prevent nausea and vomiting or drugs for psychosis. Blepharospasm may also occur in Parkinson's disease or atypical parkinsonian syndromes such as progressive supranuclear palsy. Rarely isolated blepharospasm may occur due to focal brain lesion such as from a head injury or a tumor.

Diagnosis

- there is no way to test for blepharospasm
- diagnosis based on clinical examination and documented history by a specialist



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Diagnosis is based on clinical history and physical examination, typically by a specialist such as an ophthalmologist or neurologist. There are no blood tests that can be used to confirm the diagnosis.

Treatment for Blepharospasm

Purpose of treatment is to reduce contractions of the eyelid and other affected muscles around the eye.

Treatment Options:

- botulinum toxin injections
 - surgery
- oral medications

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Treatment is aimed at reducing involuntary eye closing so as to improve vision and improve quality of life and activities of daily living. There are three main sorts of treatments: Botulinum toxin injections, local surgery on the muscles surrounding the eyes, and use of oral medications. Botulinum toxin injections are the treatment of choice and helpful in the vast majority of patients. Surgery involving removal of the muscles which close the eyes is typically reserved for patients who do not respond well to botulinum toxin injections or who have adverse effects associated with this treatment. Oral medications including trihexyphenidyl, benzodiazepines such as clonazepam, or other medications such as baclofen are usually not prescribed since these medications are poorly effective and botulinum toxin injections are highly effective.

Botulinum Toxin

- safe and effective
- onset of benefits: a few days
- peak effects last about 12 weeks on average
- about 90% of patients obtain very good relief from the injection



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Botulinum toxin injections are helpful in about 90% of patients in substantially reducing involuntary blinking or sustained eye closure. Botulinum toxin injections are considered the standard treatment and two forms of botulinum toxin have been approved by the FDA for treatment of patients with blepharospasm as being safe and effective. The injections are usually performed in several sites, typically four or five, as the patient in the picture on this slide. Injections usually involve the orbicularis oculi close to the mid margin to weaken this muscle while attempting to avoid spread of the toxin to the eye opening muscle, which is located in the center of the upper lid. Injections may also be performed between the eyebrows in order to reduce involuntary wrinkling of the brow which is often associated with blepharospasm. The injections usually take effect within a few days with the peak effect in approximately one to two weeks. The effect is not permanent and lasts on average about 12 weeks, though the duration of benefit varies substantially from patient to patient from approximately 8 to 20 weeks. The dose and distribution of the injections needs to be customized for each individual patient in order to maximize benefit and minimize adverse effects. Injections need to be done periodically because the effect wears off.

Side Effects of Botulinum Toxin

- dry eyes
- excessive weakening with incomplete eye closure
- drooping of eyelids (ptosis)
- double vision (diplopia)
- blurred vision may occur due to dry eye or keratitis



Side effects tend to improve within two weeks

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The most common adverse effect is dryness of the eyes caused by reducing both involuntary and voluntary blinking which is important in lubricating the surface of the eyes. The injections may also reduce secretion of tears from the lacrimal gland which is located close to the site of the injections. The effect of the injections may result in excessive weakening of eyelid closing and as a result patient may have incomplete eye closure which can result in exposure of the cornea, causing irritation and keratitis. Keratitis may result in blurred vision. Patients are typically instructed on careful eye care techniques including use of lubricant eye drops and an ointment as well as potentially keeping the eyes shut during the night if the eyes are closed incompletely. Spread of the injected toxins to the eyelid opening muscles can result in drooping or ptosis and spread of the toxin to the muscles adjacent to the eyeball which moves the eye can result in double vision or diplopia. All of the adverse effects are temporary and typically resolve in a few weeks.

Surgery - Myectomy

- can be an option for patients who don't obtain relief from botulinum toxin injections
- improves 75-80% of patients with blepharospasm
- removes muscles around the eye that are involved in eye closure

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Myectomy refers to selected removal of the eyelid closing muscles in order to weaken eyelid closing and reduce involuntary eye blinking and sustained closure. This surgery is typically reserved for patients who experience intolerable adverse effects from botulinum toxin injections such as repeated droopiness of the eyelids, despite adjustment of the pattern and dose of the injections. Rarely, patients receiving injections may develop resistance or antibodies to the toxin which result in loss of benefit from the injections. Patients receiving myectomy typically experience about 75% reduction in symptoms. Patients who receive this surgery who continue to respond to botulinum toxin injection but experienced adverse effects may often continue to receive a lower dose and altered pattern of injections in order to further improve the involuntary eyelid closing which may remain to a small extent after surgery.

Myectomy Side Effects

- swelling of the eyelid
- numbness of the forehead
- decreased eyelid closure, though this improves with less swelling

Eyelid function and appearance will usually improve within 6 months to a year

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Transiently patients may experience swelling of the eyelid, numbness of the forehead, and decreased eyelid closing due to weakening of eyelid closure and the location of the surgical incisions. Usually the eyelid function and appearance improves over several months. Reduced eyelid closure is usually managed with lubricant eye drops and ointment similar to how incomplete eye closure is managed following botulinum toxin injections.

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