

UNDERSTANDING

Benign Essential Blepharospasm
& Hemifacial Spasm



University of Michigan
Kellogg Eye Center

Benign Essential Blepharospasm (BEB) and Hemifacial Spasm (HFS) are uncommon, chronic, and disabling medical conditions. Both disorders result in constant and uncontrollable blinking which interferes with the performance and enjoyment of many day-to-day activities and may even render a patient functionally blind and occupationally handicapped. Sometimes the blinking is so emotionally unsettling that patients may become desperate, frustrated, and angry.

Because BEB and HFS are relatively uncommon medical problems, patients may see several doctors before they are correctly diagnosed. Once diagnosed, however, most patients are able to find at least temporary relief from their symptoms and return to normal activities.

Physicians on the Eye Plastic, Orbital and Facial Cosmetic Surgery Service and the Neuro-Ophthalmology Service at the University of Michigan Kellogg Eye Center specialize in the treatment of BEB and HFS.

NORMAL BLINKING

The eyes normally close either by a *simple* blink or by a *tonic* blink. A simple blink is defined as an involuntary, gentle touching of the upper to the lower lid and is used regularly to lubricate the cornea or front of the eye.

Tonic blinking is a more forceful and animated muscular contraction. It keeps irritative substances, such as soap, out of the eyes. Unlike a simple blink, the tonic blink tightly squeezes the eyes shut as a result of the contraction of several facial muscles. It often causes a downward movement of the brow.

When the frequency of tonic blinking increases and the relaxation phase is prolonged, it is termed *clonic activity* or *spasm*, as is seen with both BEB and HFS. Prolonged contraction of the muscles may contribute to headaches and eyestrain. Some patients also may experience drooping of the eyebrows.



Simple blink

Most commonly BEB begins in the mid 50s, but may occur in adults of any age and affects women more frequently than men. In the beginning, there is increased blinking with sunlight, excessive air pollution, wind, noise, movement of the head, or stress. This progresses to involuntary spasms, often occurring initially on only one side, but ultimately affecting both sides.

The excessive and frequent simple blinking, combined with tonic contractions of the eyelids, can then lead to spasms of eye closure for no obvious reason. The eyes may stay closed for prolonged periods, opening only for a short time, then clamping shut again. This activity produces tight eyelid closure, frowning of the forehead, as in frowning, and downward movement of the brow.



Tonic blink

Hemifacial spasm affects only one half of the face and does not progress to the other side. Unlike BEB, it is present during sleep and patients and their families are usually aware of this. The disorder is commonly associated with some evidence of damage to the facial

(VIIth) nerve. This may express itself either by mild weakness of some of the muscles of facial expression on that side or by mass action of the facial muscles on that side of the face. *Mass action* means that when contracting a facial muscle, adjacent muscles also receive some stimulation. Several examples could be movements around the mouth when forcefully closing the eye or narrowing of the eye opening when pursing the lips.

Many patients with BEB and HFS are evaluated by a large number of specialists before the correct diagnosis is established. Frustration and lack of an explanation for their symptoms may lead to anxiety and/or depression. Diagnosis of BEB is based on the patient's history and observed characteristics of the spasm. The unwary doctor may wrongly attribute the patient's symptoms to anxiety or depression, thus delaying the correct diagnosis and management.

The physical and biochemical causes of BEB are unknown. Although various theories have been proposed, none unify all the clinical observations and biochemical hypotheses. For instance, one theory regards BEB as a recurrence of a primitive, infantile reflex. When a fetus is less than six months of age, there is a reflex blinking in response to stimulation around the eye. Yet, this occurs in the absence of a well-formed eye so the reflex is not a protective one.

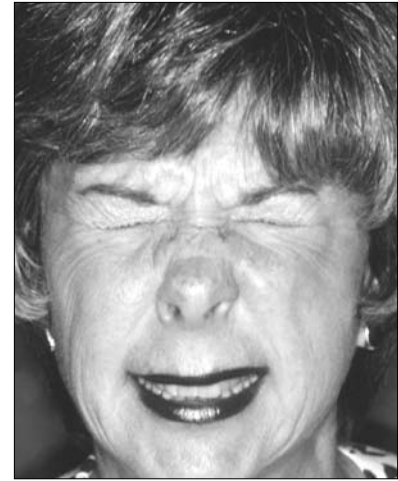
This purposeless reflex activity is referred to as *primitive* because it disappears with age. When primitive reflexes return in the elderly, they are almost always accompanied by a loss of intellectual function and the presence of other primitive reflexes. When a patient develops BEB, however, it is unaccompanied by other signs of primitive reflex activity and intellectual function remains intact. Thus, many physicians reject this theory.

Some researchers blame BEB on a biochemical imbalance in the central nervous system. They liken it to Parkinsonism, because in a *few* instances, there is evidence of damage to the same part of the brain, the basal ganglia, that is damaged in Parkinsonism. Furthermore, some patients with Parkinsonism also show the symptoms and signs of BEB. A family history of Parkinsonism or other dystonia is more common in patients with BEB than in the general population. It is extremely *uncommon*, however, for BEB to be the initial sign of Parkinsonism, and there is little likelihood that the patient with BEB will develop Parkinsonism. Certain biochemical abnormalities have been identified consistently in patients with Parkinsonism that are not present in patients with BEB.

Hemifacial spasm is caused in most patients by an artery that is too close to the facial (VIIth) nerve, causing trauma to the nerve with each pulse of blood flow in the artery.

RATIONALE OF TREATMENT FOR BENIGN ESSENTIAL BLEPHAROSPASM

Since BEB is characterized by excessive activity of the facial nerve, the nerve that supplies the facial muscles, treatment is aimed at interrupting the innervation of the muscles or weakening the muscles directly. This can be done by each of the following procedures: chemical denervation of the orbicularis muscle with Botox (botulinum-A toxin); surgical removal of the eyelid-closing muscles (orbicularis oculi, procerus, and corrugator muscles); or surgical interruption of the facial nerve supply to the eyelid-closing muscles.



*Benign essential
blepharospasm*

RATIONALE OF TREATMENT FOR HEMIFACIAL SPASM

Although BEB has no curative treatment, HFS does. The procedure is a neurosurgical decompression of the nerve. It is accomplished by exposing the nerve and placing padding between the nerve and the adjacent artery. The procedure is successful in eliminating the hemifacial spasm in about 85% of patients. There is a small risk of hearing loss on the operated side. We encourage people with HFS to consult a neurosurgeon who performs this procedure. Getting complete information about the procedure allows the patient to make an informed

decision about his or her therapy. Treatments used for BEB may also be used for HFS in people who elect not to have decompression surgery, but the BEB treatments do not treat the cause of the spasm, only the peripheral manifestations. The result may be satisfactory to the patient, but is less complete than with definitive neurosurgical treatment of HFS.

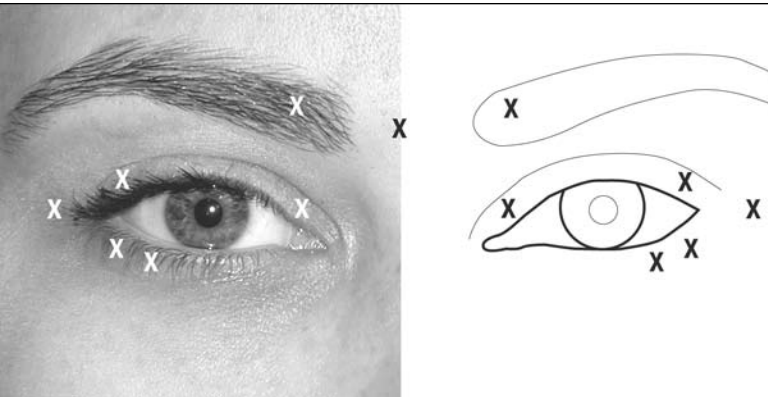
BOTULINUM TOXIN THERAPY (BOTOX, DYSPORT, MYOBLOC)

Botulinum toxin injection has become the initial treatment of choice for BEB, and, for those opting not to have neurosurgical management, for HFS. The bacteria *Clostridium Botulinum* produces several toxins that interfere with the release of acetylcholine from the nerve ending. Acetylcholine is the chemical that passes from nerve to muscle to activate muscular contractions. If acetylcholine does not reach its destination on the muscle membrane, then the muscle does not contract. Injecting minute amounts of toxin into the orbicularis oculi, corrugator, and procerus muscles thus weakens them. It slows the rate and force of blinking and effectively and safely reduces blepharospasm for the duration of the drug's action, usually 3-4 months.



Left hemifacial spasm

Botulinum toxin in spoiled ingested food causes food poisoning and can lead to muscle weakness. However, ***the injections given for facial spasm contain extremely minute, safe quantities of toxin***: the medication binds strongly to the nerve endings near the site of injection with little or no absorption into the blood stream. Effects of the toxin are seen only near the sites of injection. A very small needle is used to administer a predetermined amount of Botox to various areas of the eyelids and brows. ***Because the injection needle does not come near the eye itself***, there is no danger of puncturing the eye.



Botox injections are easily administered in the physician's office and produce significant relief of facial spasm in greater than 90% of patients, usually within 3 days. There is no difference in the duration of effect between the first and later injections. The time of reinjection is determined primarily by patient preference, with most patients

requesting reinjection when they again experience difficulty functioning, but before a complete return of the pretreatment symptoms. For those achieving less than desired degrees of benefit, the dose of the Botox can be increased on subsequent injections.

The side effects of botulinum toxin are a direct response to local injection: some drying of the eyes from reduced blinking is common; drooping of an upper eyelid occurs about 6% of the time; double vision in some direction of gaze occurs about 3% of the time; sagging of a lower eyelid is a rare occurrence. All side effects are transient, usually lasting 2-6 weeks, a shorter time than the beneficial effects of the injection. Local lubricating agents (eye drops and ointment) can be used when dry eye symptoms (irritation, sensitivity to light) are present.

Although botulinum toxin provides relief of eyelid spasm for a high percentage of patients, surgical treatment still has a place in the management of BEB. Surgery is useful for those who do not adequately respond to Botox injections, those who cannot afford it, those who have difficulty getting transportation for the shots, and those who do not choose to have repeated injections. These procedures on the eyelid-closing muscles can also be used for patients with HFS who do not elect definitive neurosurgery as described under "Rationale of treatment for hemifacial spasm."

The initial surgical procedure of choice, *myectomy*, is usually done in two stages. The upper eyelid-closing muscles are excised and the brow fixed or suspended to the bone above the brow as the first step. This procedure provides permanent relief in about

SURGICAL
PROCEDURES
USED TO TREAT
BEB AND HFS

40% of patients. The other 60% require the second stage, excision of the lower eyelid-closing muscles. About 80% of patients have relief of spasm after the second stage. The side effects of myectomy may include numbness of the forehead, shrinking of eyelid skin, dry eyes, decrease in eyebrow hair, and, if the lower eyelids have surgery, a lowering or pulling away of the lower eyelids.

An older procedure, *differential section of the VIIth nerve*, is held in reserve for those patients who do not get an adequate response from other treatments. This procedure involves excising portions of the nerve that innervates the muscles that close the eyelid. The potential side effects of this procedure are more significant than with myectomy. They include decreased facial expression, which usually improves within six months, drooping of the corner of the mouth, and drooping of the lower eyelid. The latter two often spontaneously improve, but may require surgical correction. Differential section of the seventh nerve has a long-term success rate of 50%. Nevertheless, differential section of the seventh nerve may succeed when everything else has failed.

Patients who would like to make an appointment for diagnosis and treatment of Benign Essential Blepharospasm or Hemifacial Spasm may call the Eye Plastic, Orbital and Facial Cosmetic Surgery Service or the Neuro-ophthalmology Service at the University of Michigan Kellogg Eye Center. The number to call between 8 a.m. and 5 p.m. Monday through Friday, is: 734.763.9142.

APPOINTMENTS
AND
INFORMATION

EYE PLASTIC, ORBITAL AND FACIAL COSMETIC SURGERY SERVICE

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Additional Resources

The following organizations provide reputable and up-to-date information for patients

Benign Essential Blepharospasm (Information Page from NIH)

http://www.ninds.nih.gov/health_and_medical/disorders/blepharospasm.htm

Benign Essential Blepharospasm Research Foundation

www.blepharospasm.org

Essential Blepharospasm and Hemifacial Spasm from the American Society of Ophthalmic Plastic and Reconstructive Surgery

<http://www.asoprs.org/Pages/blepharospasm.html>

Hemifacial Spasm Association

<http://hfs-assn.org/>

Hemifacial Spasm (Information Page from NIH)

http://www.ninds.nih.gov/health_and_medical/disorders/hemiface_doc.htm

Kellogg Eye Center at the University of Michigan

www.kellogg.mich.edu



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