

Not if you have BLEPHAROSPASM!

What is it?

Blepharospasm, a form of focal dystonia, is a chronic, unremitting, bilateral (both eyes), forcible closure of the eyelids. It is a variably progressive neurologic dysfunction in the motor control center of the brain. It is due to involuntary muscle contractions caused by misfiring of neurons within the central nervous system and involves the fifth and seventh cranial nerves.

Who gets it?

The most common starting age is the forties to sixties, though a few patients have been diagnosed in their twenties, and some in their seventies. About two or three women are affected for every one man. Estimates of incidence vary from 1 in 10,000 to 1 in 25,000.

How does it start?

Most commonly the patient notices some or all of the following symptoms: increased blinking, dry eyes, "tired eyes," aversion to bright lights and difficulty reading and watching television.

How does it progress?

Sometimes the spasms that start in the eyelid muscles extend further down into the lower face, causing uncontrolled contractions of the jaw or tongue. This condition is called oromandibular dystonia (Meige Syndrome). Additional extensions of the spasms can affect the throat or neck. When the voice box is involved, the condition is called spasmodic dysphonia. Over weeks or months the symptoms tend to get worse. At its worst, the patient cannot

willfully open his or her eyes and is essentially blind. Relaxing and waiting a few minutes will sometimes allow the eyelids to pop open spontaneously, but those periods of sight cannot be counted on. The threat of another spasm coming on without warning makes driving impossible, and even walking becomes dangerous.

Is it temporary?

Unfortunately, no. Though a very small percentage of cases dissipate spontaneously, the great majority of patients must deal with it for life.

Are there other complications?

Apraxia of lid opening associated with blepharospasm is the transient inability to open the eyelids in the absence of spasms.

If only one side of the face is affected, then it is almost certainly not BEB or Meige, but it could be a completely different syndrome called hemifacial spasm. The cause of hemifacial spasm differs from BEB, as does the approach to therapy.

How is it diagnosed?

Diagnosis is often difficult. Because these conditions are fairly rare, many physicians are unacquainted with them. Referrals to many specialists are common. Incorrect diagnosis sometimes delays proper treatment.

What causes it?

The best research to date shows that abnormal metabolic activity in the area of the brain called the basal ganglia causes excess electrical activity in the seventh facial nerve. The cause for the abnormal brain activity is, as yet, unknown.

How is it treated?

Since the problem is neurologic in nature, the physician will often try a variety of anticholinergic drugs.

Some patients report modest relief with drug therapy, but many of them abandon the pills because of bothersome side effects. The most common and effective current treatment involves injections of small amounts of a highly purified and diluted form of botulinum toxin (BOTOX® / onabotulinumtoxinA, Myobloc® / rimabotulinumtoxinB, Dysport® / abobotulinumtoxinA, Xeomin® / incobotulinumtoxinA) into the eye-closing muscles. This reduces the transmission of electrical impulses between the nerve endings and the muscles, relieving much of the spasms. Though this is an office procedure that takes only a few minutes, it is expensive and must be repeated every few months. These injections are usually administered by a neurologist, an ophthalmologist or a neuro-ophthalmologist. Medicare and private insurance should cover the injections in part. Patient assistance programs are for those who do not have Medicare coverage.

Those patients who are not helped by injections or drugs may explore surgery. A partial, limited or full myectomy removes part of the muscles that cause the eyelids to close. The result is that the overactive nerve endings no longer have the muscles to stimulate. The majority of patients who have this operation are pleased with the results. Sometimes used with, or instead of myectomy, the frontalis sling is an implanted elastic aid to help lift the upper lid.

Hemifacial spasm – How is it treated?

For treatment of hemifacial spasm, botulinum toxin can also be used, but the operation of microvascular decompression, where a small sponge or pad is placed to separate the seventh nerve from a blood vessel that presses on it, may well correct the problem

How do you live with it?

Most patients have to learn to adjust their lives to accept some limitations. With injections or surgery, many patients are able to continue to work. Driving becomes a problem for some people and they have to rely on others for chauffeuring or on public transportation.

What is being done about it?

The Benign Essential Blepharospasm Research Foundation (BEBRF) raises funds through public and private contributions for the research necessary to find the cause and a cure for blepharospasm and Meige. It provides encouragement to patients through educational materials and direct personal contact. Through a web of District Directors, State Coordinators, Area Representatives and Support Groups, patients are directed to medical centers where these conditions are treated. Also, the Foundation provides a central clearing house for the distribution of printed informational material, CDs and DVDs relating to blepharospasm, Meige and hemifacial spasm as needed by patients and physicians. An Internet Web site provides up-to-the-minute information, links to other related sites and a bulletin board which serves as an international support group. A bi-monthly newsletter is sent to thousands of patients and medical professionals; BEBRF support group meetings are held each year in different geographical areas.

Mattie Lou Koster, *BEBRF Founder*
1912-2001

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