

# Not if you have BLEPHAROSPASM!

---

## What is it?

Blepharospasm, a form of focal dystonia, is the 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 resulting from the misfiring of neurons within the central nervous system. It involves the fifth and seventh cranial nerves.

## 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.

## 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.

## Is it temporary?

Unfortunately, no. Less than 2% of cases dissipate spontaneously. The great majority of patients must deal with it for life.

## Does it progress?

In some instances, 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).

If it progresses, at its worst, the patient cannot willfully open his or her eyes and is functionally 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.

## How is it diagnosed?

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

However, 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 treated?

Since the problem is neurologic in nature, the physician may 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, Xeomin® / incobotulinumtoxinA), Dysport® / abobotulinumtoxinA, Myobloc® / rimabotulinumtoxin B), 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. Allergan and Merz offer patient assistance programs for those who qualify.

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.

## Are there other complications?

In some patients, apraxia of eyelid opening associated with blepharospasm is the most common cause of failure or minimal improvements with botulinum toxin. It is the transient inability to open the eyelids in the absence of spasms.

## How do you live with blepharospasm?

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 offers research grants to investigators and participates in the Dystonia Coalition, a collaboration of medical researchers and patient advocacy groups that is working to advance the pace of clinical and translational research in the dystonias to find better treatments and a cure. It also participates in the Dystonia Brain Collective based at the Harvard Brain and Tissue Resource Center.

## What resources are available to me?

The BEBRF serves as a central clearing house for the distribution of printed educational/medical informational material, CDs and DVDs relating to blepharospasm, Meige and hemifacial spasm as needed by patients and physicians; directs patients to doctors who treat these disorders and offers encouragement through personal contact with the office as well as with a web of District Directors, State Coordinators, Area Representatives and support group meetings. An Internet website provides up-to-the-minute information, links to other related sites and a bulletin board which serves as an international support group. The Foundation also produces a bi-monthly newsletter, which is sent to thousands of patients and medical professionals; and holds symposiums each year in different geographical areas.

Mattie Lou Koster, *BEBRF Founder*  
1912-2001

### MEDICAL ADVISORY BOARD

Mark Hallett, M.D., Chair . . . . . Bethesda, MD  
L. Craig Evinger, Ph.D. . . . . Stony Brook, NY.D.  
Andrew Harrison, M.D. . . . . Minneapolis, MN  
Joseph Jankovic, M.D. . . . . Houston, TX  
Laurie J. Ozelius, Ph.D. . . . . New York, NY  
James R. Patrinely, M.D. . . . . Houston, TX  
and Pensacola, FL  
Peter J. Savino, M.D. . . . . LaJolla, CA  
Alan B. Scott, M.D. . . . . San Francisco, CA  
Mark Stacy, M.D. . . . . Durham, NC  
Edwardo Tolosa, M.D. . . . . Barcelona, Spain

### DISTRICT DIRECTORS & E-MAIL

Peter Bakalor, Western  
AK, AZ, CA, CO, HI, ID, MT, NV,  
NM, OR, UT, WA, WY  
[dir-w@blepharospasm.org](mailto:dir-w@blepharospasm.org)

Barbara Benton, Eastern  
CT, DE, DC, MA, MD, ME, NH, NJ, NY,  
NC, PA, PR, RI, SC, VT, VA, WV  
[dir-e@blepharospasm.org](mailto:dir-e@blepharospasm.org)

Shelley Goddard, North Central  
IL, IN, IA, KY, MI, MN, NE, OH, SD, WI, ND  
[dir-n@blepharospasm.org](mailto:dir-n@blepharospasm.org)

Fran Morgan, Southern  
AL, AR, FL, GA, KS, LA, MS, MO, OK, TN, TX  
[dir-s@blepharospasm.org](mailto:dir-s@blepharospasm.org)

*For further information, contact:*

### BENIGN ESSENTIAL BLEPHAROSPASM RESEARCH FOUNDATION

P. O. Box 12468 • Beaumont, TX 77726-2468, USA  
(409) 832-0788 • Fax (409) 832-0890  
E-Mail: [bebrf@blepharospasm.org](mailto:bebrf@blepharospasm.org)  
WEBSITE: <http://www.blepharospasm.org/>

*BEBRF is an exempt organization under Section 501(C)(3).  
All donations are tax deductible.*

Revised 2013



**BLIND  
BUT WITH  
PERFECT  
VISION**

**Impossible?**

**BENIGN ESSENTIAL  
BLEPHAROSPASM**