

Facial and Eyelid “Twitch” Disorders

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We have been asked to provide a description of disorders which may “look something like” Benign Essential Blepharospasm (BEB). We have listed below such disorders with a brief description of each and the ways in which each of these might differ from BEB and from one another.

Our patients report that the onset of BEB (any of the three forms listed below) may be sudden (overnight) or more insidious (starting as a “twitch” in one eyelid or spasms of both the upper and lower eyelids on one side). This onset variability may sometimes pose considerable confusion early on in the disorder.

Benign Essential Blepharospasm

There are three general types of BEB:

1. Spastic BEB (BEB_S)
Bilateral (both eyes) forcefully close, usually synchronously (at the same time), although one eye may be more affected (squeeze close harder) than the other. The spasms abate during sleep and may be variable in their severity from moment to moment, day to day, and with different activities. Infrequently begins as a twitching in one eyelid or both eyelids (upper and lower eyelids) on the same side of the face.
2. Pseudo-Apraxic BEB (BEB_A)
Bilateral (both eyes) drift closed, usually synchronously (at the same time), although one eye may be more affected (be more difficult to open) than the other.
3. Combined BEB (BEB_C)
Patients affected with combined BEB show features of both BEB_S and BEB_A.

Meige Syndrome

Features of BEB_S, BEB_A, or BEB_C with simultaneous involuntary spasms or writhing movements of the mid-face (cheeks), lower-face (mouth and/or tongue), and possibly extending down into the neck. Some consider the BEB disorders and Meige Syndrome to be different examples of a single set of related disorders known as craniofacial dystonias.

Tardive Dyskinesia

Writhing movements of the tongue, mouth, or lips in isolation. Sometimes seen in association with increased blink frequency. Usually a side effect (sometimes irreversible) of specific medications. Other rare, but sometimes similar appearing disorders may include *Tardive Dystonia*, *Tourette Syndrome*, *Tardive Tourettism*, and *Tardive Myoclonus*.

Benign Eyelid Twitch

(Also called *eyelid myokymia*)

This is a fine fasciculation (tiny muscle contractions) generally affecting one eyelid (more often a lower eyelid, but upper eyelids as well). Affected patients may feel as though their entire eyelid is “jumping wildly,” but observers barely notice the movement. Twitching is episodic, lasting seconds to hours over minutes to months, but always eventually resolves on its own. Associated with stress, fatigue, and caffeine use. Does not involve upper and lower eyelids on the same side or eyelids on

both side of the face at the same time and rarely involves more than one eyelid at different times. “Myokymia” is probably a bad name for this problem, as the term is also commonly used to describe another, very different disorder.

Hemifacial Spasm

Spastic contractions of muscles involving an entire half of the face. Often progresses to show some weakness of the face muscles on the same side, despite frequent spasms. Closes one eye. Most often due to irritation of the *Facial Nerve* near the brainstem, but in some cases may also be associated with decreased hearing on the affected side. Very rarely, may occur bilaterally (on both sides of the face), but when it does, the two sides spasm asynchronously (not together). The hallmark of this disorder is the development of spasms BEFORE any weakness is apparent.

Bells Palsy / Facial Nerve Injury with Aberrant Regeneration

The *Facial Nerve* (also called the 7th Cranial Nerve) may be injured from any number of causes. The term *Bells Palsy*, was originally used to describe spontaneous 7th Nerve palsy (weakness) of unknown cause. Many researchers believe the majority of cases of Bells Palsy are caused by one of the *Herpes Viruses* (that reside in the nervous system of 50-90% of adults across the world, either *Herpes Simplex* or *Herpes Zoster*), although many other known causes of “spontaneous” 7th Nerve weakness may simply not be tested for or recognized. After injury, as this nerve recovers, it may grow back in strange ways (so called *Aberrant Regeneration*), and cause spasms which may look something like Hemifacial Spasm. An injury to the Facial Nerve at the back of the brain may result in something that looks like Hemifacial Spasm in the lower half of one side of the face. Repeated episodes of facial weakness on one side of the face or episodes on both sides of the face may indicate a disorder called *Melkersson-Rosenthal Syndrome*. The hallmark of these Facial Nerve problems is a weakness on the face BEFORE spasms develop.

Oculomotor Nerve (3rd Cranial Nerve) Injuries

The 3rd *Cranial Nerve* is important for upper eyelid opening. An injury to this nerve from any cause (stroke, trauma, tumor, etc) may cause the upper eyelid to stay closed and not open and may look sort of like BEB_A if both sides are involved (as can happen with massive head trauma), but there are other signs that help physicians to easily distinguish between these disorders.

Blepharoptosis

Blepharoptosis is generally the term used to mean a droopy upper eyelid. There are many types and causes. Most commonly this is a problem in one eye, but bilateral (both upper eyelid) involvement is not uncommon, either. Typically, however, causes and effects of a droopy upper eyelid are constant, that is the eyelid(s) are almost always droopy to the same level. BEB_A, on the other hand, often has periods where the eyelids will open to different degrees and then completely close. It can be a little confusing when blepharoptosis and BEB_A coexist, but videos or photos of a person with BEB_A when their eyes are “the best they ever get,” can help a clinician tease these disorders apart. *Myasthenia Gravis* and related disorders may cause variable droop of the upper eyelids and may rarely be confused with BEB_A, but a carefully recounted history by a patient and a thoughtful physical examination should easily distinguish these disorders. Some additional unusual causes of blepharoptosis which may be particularly confusing, especially early in their onset, include variants of *Parkinson's disease*, *Progressive Supranuclear Palsy*, *Periodic Paralysis*, various *myopathies* (including mitochondrial myopathies), and the *Chronic Progressive External Ophthalmoplegias*.

Multiple Sclerosis

Is a relatively rare disorder which may cause “writhing” muscle movements anywhere on the face, often called *mytonia* or *myotonic contractions*. These muscle contractions are sometimes described as appearing “like a worm crawling under the skin” and do not resemble the facial spasms or contractions seen in most of the above described disorders.

Secondary Blepharospasm

Secondary blepharospasms (forceful eyelid closing in response to eye irritation or even some medications) may strongly resemble BEB_s and in fact often co-exist with BEB_s. Any irritant to the eye, such as a tear film abnormality (Dry Eye, Toxic Tear Syndrome, Tear Film Instability, etc) may result in secondary blepharospasm. Placing a couple of drops of topical anesthetic in the eye can often help to identify a role of many types of secondary blepharospasm, although habitual eyelid squeezing may be more difficult to tease out.

Compiling the above list of disorders was a difficult task, and we’ve probably crossed the line many times between giving too much information and not providing enough, and we have certainly erred in mentioning some rare entities and excluding others that are more common, but perhaps more obviously distinct from BEB, and generalities have been made in order to provide some sort of framework for non-physicians. The bottom line, however, is that the diagnosis of BEB can be complicated and difficult, and BEB is best distinguished from many other disorders by an experienced physician well-trained in recognizing the myriad of eyelid movement dysfunctions.