



InSight

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Eyelid Spasms

Part I – Essential Blepharospasm



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INTRODUCTION

There are several different types of eyelid spasms. The three most common are eyelid twitching (myokymia), Essential Blepharospasm, and Hemifacial Spasm. Part I of Eyelid Spasms will focus on Essential Blepharospasm while Part II will look at Myokymia and Hemifacial Spasm.

What is Essential Blepharospasm?

“**Blepharo**” comes from the Greek word for eyelids. **Blepharospasm** is a condition in which there is involuntary blinking of the eyelids that may at times be forceful and sustained (several seconds). Blepharospasm is a type of “**Dystonia**”, a term used to describe “abnormal, involuntary, sustained muscle contractions and spasms”. When the eyelid spasms occur, a temporary inability to see may result due to the forceful involuntary eyelid closure (*Figure 1a and 1b*). Patients with Blepharospasm have normal eyes. The visual disturbance is due solely to the forced closure of the eyelids. In some individuals, other muscles of the face may be involved. With other facial spasms (perioral contractions, lip pursing, chin

thrusting, tongue movements), the patients often have very unusual facial expressions that are uncontrollable (*Figure 2*). Dystonias may also occasionally occur elsewhere including the neck (torticollis, antecollis) or dystonic posturing of the limbs. Involuntary vocalization, such as grunting, frequent throat clearing, and respiratory noises as well as dysphagia and respiratory difficulties may develop. In advanced stages, patients with uncontrolled eyelid and facial spasms may become functionally blind, socially reclusive, and unable to work or care for themselves.

How does Essential Blepharospasm present?

The average age of onset is 56 years with females being more commonly affected than males by 3:1. Blepharospasm usually begins with a gradual increase in the blink rate that eventually progresses to forceful eyelid spasms frequently throughout the day. A fluctuating course characterized by remissions and exacerbations is usual. It may be precipitated or aggravated by bright light, stress, fatigue, driving, reading, watching TV, and a variety of other activities. Sleep, relaxation training, walking and talking may improve the spasms. Some patients learn to avoid situations that aggravate the condition, while others develop certain behavioral techniques that involve using other facial muscles or acts of mental concentration to decrease the frequency and intensity of spasms. Examples include humming, singing, whistling, yawning, coughing, mouth opening, nose pinching, chewing

gum, talking continuously, rubbing the eyelids or applying pressure to other areas of the face, covering one eye, and solving puzzles or problems.

What causes Essential Blepharospasm?

Blepharospasm is thought to be secondary to an abnormal functioning of the “**basal ganglia**,” an area of the brain which plays an important role in the control of fine motor movements. The exact problem in this area is unknown but a neurochemical imbalance is suspected. In some cases it may be a familial disease with more than one family member affected. In other cases, blepharospasm may be secondary to drug therapy for other diseases (ex. medication for Parkinson’s disease, major tranquilizers).

THERAPY

Blepharospasm can be treated with oral medication, injections of Botulinum toxin or surgery.

Drug therapy is effective in fewer than 10% of patients and may only give partial relief. The response is unpredictable and side effects may outweigh the benefits. A variety of medications have been tried including antipsychotics, affective disorder agents, anxiolytic agents, sedatives, drugs for Parkinson’s disease, muscle relaxants, etc.

Botulinum toxin is produced by the bacteria “*Clostridium botulinum*”. Botulinum toxin interferes with acetylcholine release from peripheral motor nerve terminals, resulting in

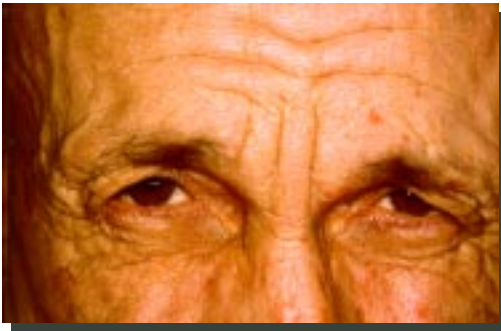


Figure 1A
Pre-spasm,
eyelids open.



Figure 1B
Profound spasm,
of eyelids
(Blepharospasm).

temporary paralysis of the injected muscles. Minute doses of botulinum toxin are injected around the eyelid and facial area with a fine needle. The effect usually occurs within the first few days and lasts an average of 3 to 4 months, at which time another injection is given. Muscle relaxants (orphenadrine, baclofen) have been useful in several patients as the effects of botulinum toxin start wearing off.

Long term follow up studies have shown botulinum toxin injection to be a very safe and effective treatment with as many as 90% of patients obtaining significant relief of their spasms. Repeated treatment generally remains effective over a prolonged period in most patients. However, in some individuals, botulinum toxin injections may become less effective over time, possibly because of a re-sprouting of motor endplate receptors or the development of an antitoxin.



Figure 2
Profound eyelid spasms, facial spasms
and neck spasms (torticollis).

Side effects are infrequent and transient. They include ptosis, blurred vision, double vision, ocular irritation and tearing. These side effects resolve spontaneously in a matter of days to weeks.

SUPPORTIVE THERAPY

The psychologic impact of Essential Blepharospasm can be tremendous. Patients often feel their physicians do not believe their symptoms are uncontrolled and therefore may see several physicians before the proper diagnosis is made. It is not unusual for a patient to be initially seen by a physician who is unaware of this disease and refers them for psychiatric evaluation. They are generally greatly relieved when they are finally told they have a real disease and that some treatment is available. It is extremely important for patients to understand their disease process. Patients generally learn their own coping techniques which can be shared with others at support group meetings. A monthly newsletter and information on support groups can be obtained through the Benign Essential Blepharospasm Research Foundation, address:

BEBRF, Inc.,
P.O. Box 12468,
Beaumont, Texas - 7726-2468

At present there is no cure for Essential Blepharospasm and related dystonias. Systemic medications help some patients. Debilitating spasms, however, continue in most cases, rendering patients functionally blind. Botulinum A toxin injection is the best temporary therapy and myectomy is the best long term therapy.

Apraxia of the Eyelids and Blepharospasm?

Apraxia of eyelid opening is a condition that affects about 7% of Blepharospasm patients and rarely occurs on its own. The

Blepharospasm patient with apraxia of lid opening will typically have lid spasms, squeezing the eyelids shut and then for several seconds to minutes after the spasms stop, the patient is unable to open the lids. The eyelids may then come open almost normally for a period of time and then without warning, slowly drop shut again, or be drawn shut by spasms. The apraxia of lid opening patient can be seen raising their brows and trying to open their eyelids as strongly as possible without any elevation of the eyelids. Apraxia is thought to be due to a problem in the neuro-circuitry for opening the eyelids, much like blepharospasm is a problem in the neuro-circuitry causing squeezing of the lids.

To make a diagnosis of apraxia of lid opening, all Blepharospasm or squeezing of the eyelids must be relieved. Botulinum toxin, myectomy or a combination thereof must completely relieve muscle squeezing before a diagnosis of apraxia of the opening can be confirmed.

Blepharospasm patients with apraxia are treated by a limited myectomy in conjunction with tightening of the levator tendon (aponeurotic ptosis repair). By tightening the tendon of the muscle that raises the eyelids, patients can more effectively open their lids. By removing the squeezing muscles in the upper eyelids (limited myectomy), any residual squeezing that is not completely relieved by botulinum toxin is improved. Most patients with apraxia of lid opening can be improved with a combination of limited myectomy, ptosis repair and botulinum toxin. Unfortunately drugs have provided little or no improvement for this disorder.

Upcoming Issues

- ★ Myokymia ★ Hemifacial Spasm
- ★ Trichiasis ★ Dry Eyes