

## Review on Blepharospasm

B.Sowmya,Ch.Sharon Roja,Dr.Brahma reddy, K.Sravanthi.

Nalanda Institute of Pharmaceutical sciences, Kantepudi (V), Sattenapalli (M), Guntur dist, A.P.

### ABSTRACT

Blepharospasm is any abnormal contraction or twitch of the eyelid. The Condition should be distinguished from the more common, and milder, Involuntary quivering of an eyelid, known as myokymia. Benign essential blepharospasm (BEB) is a craniofacial dyskinesia characterized by repeated, involuntary contractions of the orbicularis oculi, procerus, and corrugator muscles. The mechanism underlying benign essential blepharospasm is not yet clearly understood but is thought to be multifactorial, with a likely genetic component and environmental trigger. It has been suggested that BEB may be partly due to abnormalities in the basal ganglia and dopaminergic system involving a subclinical loss of striatal dopamine.

### Key words:

Blepharospasm,  
Benign essential blepharospasm (BEB),  
orbicularis oculi, dystonia, dry eye, facial trauma.

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### \*Corresponding Author

Name: B.Sowmya

Email: sowmyabhavanasi20@gmail.com

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## INTRODUCTION

Blepharospasm the word is taken from Greek: βλέφαρον / blepharon- eyelid, and σπασμός / spasmus-spasm, an uncontrolled muscle contraction<sup>1</sup>. Blepharospasm is any abnormal contraction or twitch of the eyelid. The Condition should be distinguished from the more common, and milder, Involuntary quivering of an eyelid, known as myokymia. The person's eyelids feel like they are clamping shut and will not open without great effort. People have normal eyes, but for periods of time are effectively blind due to their inability to open their eyelids.

It is of two types: essential and reflex blepharospasm. The benign essential blepharospasm is a focal dystonia-a neurological movement disorder involving involuntary and sustained contractions of the muscles around the eyes. The term essential indicates that the cause is unknown. Reflex blepharospasm is a type of secondary blepharospasm triggered by dry eyes, blepharitis, intraocular inflammation, meningeal irritation, and light sensitivity<sup>2</sup>.

## EPIDEMIOLOGY

The first record of blepharospasm and lower facial spasm was found in the 16th century in a painting titled De Gaper. At that time, and for several ensuing centuries, patients with such spasms were regarded as being mentally unstable and often were institutionalized in insane asylums. Little progress was made in the diagnosis or treatment of blepharospasm until the early 20th century, when Henry Meige (pronounced "mehzh"), a French neurologist, described a patient with eyelid and midface spasms, spasm facial median, a disorder now known as Meige syndrome<sup>3</sup>. Benign essential blepharospasm (BEB) is a craniofacial dyskinesia characterized by repeated, involuntary contractions of the orbicularis oculi, procerus, and corrugator muscles. These spasms cause involuntary blinking that can become severe and lead to functional blindness. BEB has a prevalence of 5 to 13 per 100,000 individuals, is almost 3

times more common in females, and has a median age of diagnosis at 53 years of age<sup>4</sup>.

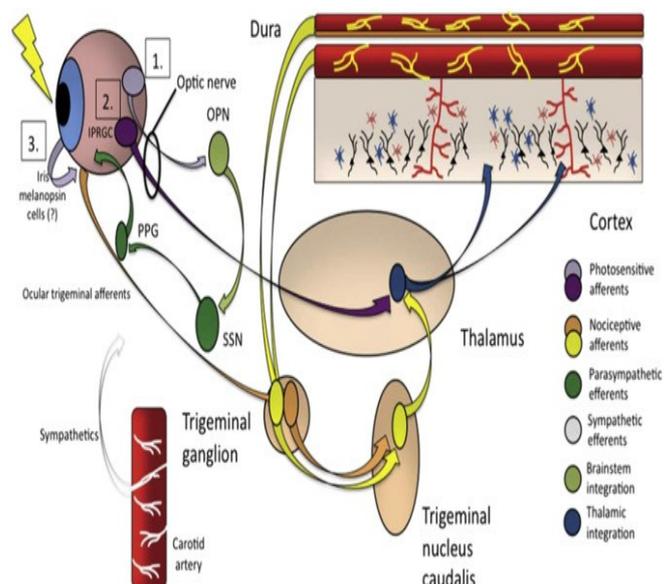
## United States

It is estimated that there are at least 50,000 cases of blepharospasm in the United States, with up to 2000 new cases diagnosed annually. The prevalence of blepharospasm in the general population is approximately 5 in 100,000<sup>5</sup>.

- Age: onset most commonly occurs during years 40-60
- Gender: Female > Male (2-4:1)
- Incidence: 2,000 cases diagnosed annually in the U.S.
- Prevalence: 1.6-30/100,00<sup>6</sup>.

## PATHOPHYSIOLOGY

The mechanism underlying benign essential blepharospasm is not yet clearly understood but is thought to be multifactorial, with a likely genetic component and environmental trigger.



It has been suggested that BEB may be partly due to abnormalities in the basal ganglia and dopaminergic system involving a subclinical loss of striatal dopamine. This loss of dopamine creates avulnerability of the trigeminal blink circuit to excitation by an environmental insult or external trigger. There is also a progressive loss of dopamine in the substantianigra that occurs with aging. Therefore, there is an increased excitability of the trigeminal blink reflex with age, and this association correlates with the fact that BEB is typically diagnosed at a later age<sup>7</sup>.

**CAUSES**

- Some causes of blepharospasm have been identified; however, the causes of many cases of blepharospasm remain unknown, although some educated guesses are being made. Some blepharospasm patients have a history of dry eyes and/or light sensitivity, but others report no previous eye problems before onset of initial symptoms<sup>10</sup>.
- Some patients with blepharospasm report a familial occurrence of the affliction. In families with autosomal dominant familial dystonia, affected members may have a generalized or segmental dystonia, while other members have various focal dystonias, such as isolated blepharospasm<sup>11</sup>.
- Some drugs can induce blepharospasm, such as those used to treat Parkinson's disease, as well as sensitivity to hormone treatments, including estrogen-replacement therapy for women going through menopause.
- Blepharospasm can also be a symptom of acute withdrawal from benzodiazepines<sup>12</sup>

**RISK FACTORS**

Variable risk factors for blepharospasm have been reported:

- Head or facial trauma.
- Family history of dystonia or tremor.
- Reflex blepharospasm is reportedly triggered by severely dry eyes and blepharitis, intraocular inflammation, meningeal irritation, light sensitivity.
- Stress may exacerbate benign essential blepharospasm.
- Medications, such as those used to treat Parkinson's disease, have been associated with blepharospasm.
- Earlier studies suggested that cigarette smoking was a negative risk factor, but more recent studies no longer support this<sup>8</sup>.

**SIGNS AND SYMPTOMS**

- Excessive blinking and spasming of one or both eyes – characterized by uncontrollable eyelid closure of durations longer than the typical blink reflex.
- The spells of spasming may last for minutes or even hours
- Uncontrollable contractions or twitches of the eye muscles and surrounding facial area. Some sufferers have twitching
- symptoms that radiate into the nose, face and sometimes, the neck area
- Dryness of the eyes
- Sensitivity to the sun and bright light<sup>9</sup>.

**DIAGNOSIS**

- The diagnosis of blepharospasm is clinical and is made by careful history taking and physical exam.
- Eyelid myokymia
- Secondary to ocular irritation (ie dry eyes, blepharitis, entropion, intraocular inflammation, photosensitivity)
- Secondary to meningeal irritation (ietemporoparietal strokes)
- Meige syndrome (blepharospasm- oromandibulardystonia)
- A symptom of systemic disease associated with lesions of the brainstem and basal ganglia (Parkinson's, Huntington's, Wilson's, progressive external ophthalmoplegia,progressivesupranuclear palsy.)
- Facial tics (Tourette syndrome)
- Hemifacial spasm (contraction of the entire one side of the face)
- Cerebral palsy
- Functional (Focal brain injury or tumor)<sup>13</sup>.

**LABORATORY TEST**

- Laboratory tests have no utility in the diagnosis and workup of blepharospasm.

**TREATMENT**

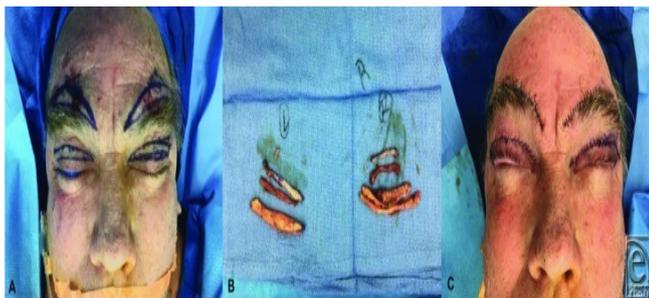
- Drug therapy for blepharospasm has proved generally unpredictable and short-termed. Anticholinergics, tranquilizing drugs and botulinum toxin are the mostly used therapeutic options. However serious side effects can be observed as well as failure of therapy. It is therefore not surprising that new therapies are constantly being tested. In this backdrop new evidence shows Mosapride can be a safe and affordable therapeutic option for blepharospasm<sup>14</sup>.
- Botulinum toxin and myectomy help to controlblepharospasm but may not cure it.
- Botulinum toxin A is a safe, long-term treatment for patients with benign essential blepharospasm, but sustained treatment efficacy may require higher doses in later stages of the disease<sup>15</sup>.

**DRUG CHART<sup>16</sup>:**

Generic Name- Brand Name	Dose for BEB	Typical Dilution
Abobotulinumtoxin A Disport™	40-60 U/eye	Reconstitute 500-U vial in 1ml of 0.9% saline
Incobotulinumtoxin A Xeomin <sup>R</sup>	20-25 U/eye	100 U/1 mL of 0.9% saline
Onabotulinumtoxin A Botox <sup>R</sup>	20-25 U/eye	100 U/1 mL of 0.9% saline
Rimabotulinumtoxin B Myobloc <sup>R</sup>	1250 U/eye	Available as solution of 5000 U/1 mL

## SURGERY

- Surgery is reserved for patients who are poorly responsive to botulinum (/Botulinum\_Toxin\_Use\_In\_Oculoplastics) therapy and are disabled by their symptoms. Patients with apraxia of eyelid opening may often proceed to surgery as chemodenervation with botulinum toxin is less effective. Surgical myectomy of the orbital and palpebralorbicularis muscle in the upper (and sometimes lower) eyelids as well as surgical ablation of the facial nerve are effective in treating BEB. However, the latter procedure has been mostly abandoned owing to high recurrence rates incidence of hemifacial paralysis.
- Surgery in patients with apraxia of eyelid opening may require a frontalis sling rather than a levator repair to help elevated the eyelids<sup>17</sup>.



(a) Preoperative markings for bilateral brow lift and blepharoplasty.  
 (b) Skin and muscle resection.  
 (c) Immediate postoperative result.

## CONCLUSION

Blepharospasm should be considered a form of adult onset focal dystonia. Although prolonged spasms of the orbicularis oculi muscles remain the clinical hallmark of blepharospasm, patient with blepharospasm may be characterised by various types of involuntary activation of periorcular muscles. Although blepharospasm and hemifacial spasm and eyelid disorders with different etiologies. The spasms of lid closure and the disruption of sensorimotor processing of both diseases are remarkably similar. Symptoms of blepharospasm can progress to render a person incapable of activity of daily living, such as reading, and driving and cause an unwanted cosmetic effect. The physiological distress is a real part of the disease and patient education, counselling and support is a valuable as medical treatment.

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