



Clinical Manifestations of Blepharospasm and  
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# Dystonia Defined

- A disorder characterized by involuntary muscle contractions that cause slow repetitive movements or abnormal postures
- May or may not be painful
- May affect only one muscle, groups of muscles, or muscles throughout the body
- The cause for the majority of cases of dystonia is not known

# Common Forms of Focal Dystonia

- Cervical dystonia, also called spasmodic torticollis or torticollis, is the most common of the focal dystonias
- Benign Essential Blepharospasm
- Cranio-facial dystonia
- Meige syndrome
- Oromandibular dystonia

# Other Forms of Dystonia

- Spasmodic dysphonia
- Task-specific dystonias
  - Writer's cramp
  - Musician's dystonia
- Generalized dystonia (affects most or all of the body)
- Multifocal dystonia (involves two or more unrelated body parts)
- Hemidystonia (involves the arm and leg on the same side of the body)

# Benign Essential Blepharospasm (BEB)

- Disorder of progressive involuntary spasms of the eyelid protractors (orbicularis oculi, corrugator, and procerus muscles) resulting in eyelid closure
- Local eye disorders such as dry eye, iritis, or blepharitis can cause eyelid spasm that is not BEB



# BEB Demographics

- Male: Female ratio about 1:3
- Incidence 10 per 100,000
- Onset age usually 40-60 years
- Some familial tendency

# BEB Onset

- Increased blink rate
  - Both eyes
    - At onset, 20% of patients have asymmetric and asynchronous involvement.
- Progression over a few years followed by stability
  - Remission is rare
- Diagnosis is commonly either missed or delayed by 4 to 10 years

# BEB Symptoms

- Forced lid closure
  - Sometimes brow spasm
  - Sometimes midface spasm
- Light sensitivity
- Eye irritation (gritty, dry)
- Functional blindness
- Stress seems to make it worse



# Blepharospasm video



## **Benign Essential Blepharospasm**

# Meige Syndrome

- Combination of two forms of dystonia, blepharospasm and oromandibular dystonia (OMD)
- Also called Brueghel's syndrome and oral facial dystonia

# Apraxia of Eyelid Opening (AEO)

- Most often associated with extra-pyramidal disorders (progressive supranuclear palsy)
- AEO associated with blepharospasm is not actually an apraxia but rather a focal dystonia of the eyelids – pretarsal blepharospasm

# Apraxia of Eyelid Opening in BEB

- difficulty with voluntary eyelid opening
- there is reduced blinking
- when the eyes are open, no overt orbicularis oculi spasm, and no photophobia
- there is frontalis muscle overaction and secondary elevation of the brows

# Hemifacial Spasm

- Synchronous twitching of most/all of the muscles on one side of the face
- NOT a dystonia – it is believed to be caused by a blood vessel that compresses the facial nerve as it exits the brain
- Treated with botulinum toxin or surgical neurovascular decompression

# Diagnosis of BEB

- Diagnosis of benign essential blepharospasm is made clinically; it is a diagnosis of exclusion, having ruled out the presence of associated conditions. Neuroimaging is generally unrevealing and of limited use in the workup



# Diagnosis of BEB

- History
  - Head injury, Parkinsons disease or PSP, schizoaffective disorders
  - Family history of dystonia
  - Medications associated with dry eye, tardive dyskinesia
  - Other disorders

# Diagnosis of BEB

- Examination
  - Intermittent forceful eyelid closure
  - Apraxia
  - Look for local eye causes of lid spasm (iritis, dry eye, blepharitis, trichiasis, etc)
- Lab testing is usually not necessary
- Imaging – CT or MRI usually not necessary

# Treatment of BEB

- Botulinum Toxin
- Surgery
- Oral medication
- Treat associated conditions
  - Dry eye
  - Blepharitis
  - Photophobia
- Sensory tricks

# Sensory Tricks

- Looking down, lifting a trigger point such as an eyebrow, whistling, singing, humming, chewing gum, playing a musical instrument

# Conclusion

- BEB is a dystonic disorder of progressive involuntary spasms of the eyelid protractors (orbicularis oculi, corrugator, and procerus muscles) resulting in eyelid closure
- It may be associated with oromandibular dystonia and is then usually call Meige Syndrome