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
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