Treating blepharospasm and hemifacial spasm with botulinum toxin

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Objectives

- 1. Learn how to recognize blepharospasm and hemifacial spasm.
- 2. Learn approach to management of these disorders with botulinum toxin.
- 3. Be aware of common pitfalls and how to avoid them.

Blepharospasm definition

- Forceful, uncontrolled contractions of the eyelids
- A focal dystonia

Primary "Benign essential blepharospasm" vs Secondary Causes

- Dry eye
- Allergies
- Blepharitis- inflammation of eyelids
- Blinking Tics
- Parkinson disease
- Tardive dystonia
- Structural lesions- basal ganglia, thalamus, cerebellum, brainstem
- Wilson's disease

- Distinct from eyelid myokymia- common quivering of eyelid

- Meige syndrome: Blepharospasm plus oromandibular dystonia

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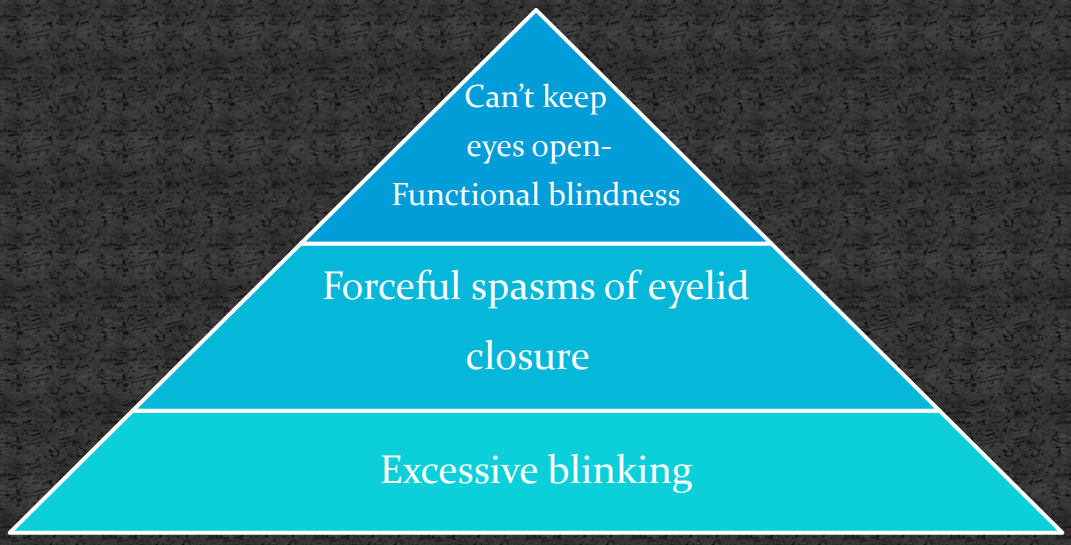
Continuum of severity



Clinical presentation

- May complain of eye irritation
- Frequent blinking
- Spasms shut
- Bright lights, wind, stress, fatigue, and certain activities may be exacerbating factors.
- May have a "geste antagoniste" (sensory trick)- voluntary intervention that may afford temporary improvement

Blepharospasm- Epidemiology

- Estimated 2,000 new diagnoses of BEB/year in the U.S.
- Prevalence of BEB in the general population is approximately 5 per 100,000 individuals.
- 20,000-50,000 individuals in the U.S.
- Usually sporadic, but familiar forms exist
- At least twice as common in women

https://ghr.nlm.nih.gov/condition/benign-essential-blepharospasm#statistics
https://rarediseases.org/rare-diseases/benign-essential-blepharospasm/

Videos: Blepharospasm

Blepharospasm- Treatments

- Chemodenervation- First-line (Botulinum toxin injections)

- Oral medications:
- Anticholinergic medications- trihexyphenidyl, benztropine
- GABA'ergic- benzodiazepines (clonazepam), baclofen
- Dopaminergic medications- levodopa
- Dopamine depleting medication- tetrabenazine

- Surgical options:
- Myectomy
- Deep brain stimulation

- AAN Guidelines (2016)- Level B recommendation ("should be considered") for Ona and Inco; level C ("may be considered") for Abo

Expert panel reviewed published evidence

- **Classification of recommendations**
- A: Established as effective, ineffective, or harmful for the given condition in the specified population (Level A rating requires at least two consistent Class I studies)
- B: Probably effective, ineffective, or harmful for the given condition in the specified population (Level B rating requires at least one Class I study or at least two consistent Class II studies)
- C: Possibly effective, ineffective, or harmful for the given condition in the specified population (Level C rating requires at least one Class II study or two consistent Class III studies)
- U: Data inadequate or conflicting; given current knowledge, treatment is unproven

Expert panel reviewed literature

- For the treatment of blepharospasm, the evidence supported a Level A recommendation for BoNT-A, A/Ona, and A/Inco; a Level B recommendation for A/Abo; and a Level U recommendation for B/Rima.
- For hemifacial spasm, the evidence supported a Level B recommendation for BoNT-A and A/Ona, a Level C recommendation for A/Abo, and a Level U recommendation for A/Inco and B/Rima.

Labeled Doses for botulinum toxin for blepharospasm

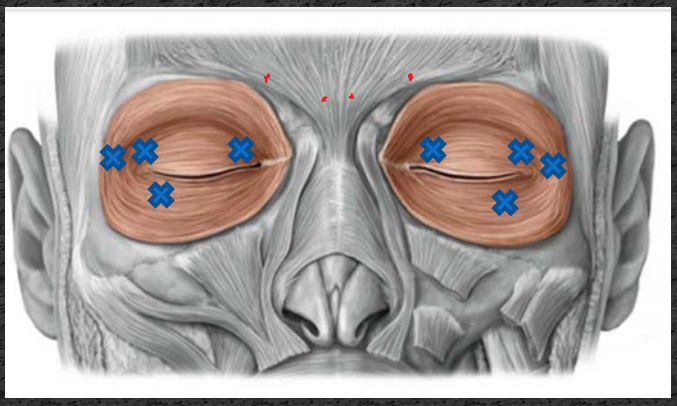
- Onabotulinum toxin A (BOTOX)- 1.5-2.5 units per site (Max 200units/30 days)
- * Also approved in pediatric patients 12yo and up
- Incobotulinum toxin A (Xeomin)- 1.5-2.5 units per site (Max 35units/eyelid)

- Not FDA approved for:
- Abobotulinum toxin A (Dysport)
- Rimabotulinum toxin B (Myobloc)

Blepharospasm- Muscles commonly injected

- Orbicularis oculi
- Procerus
- Corrugator

BLEPHAROSPASM TREATMENT



1.25-2.5 units/site
→ up to 12.5/site

Ona- and Inco-botulinum toxin

https://www.studyblue.com/notes/note/n/practical-3-muscles/deck/13180988

** Be aware of entry point versus where needle ends up, and where toxin will end up.*

Practical aspects

- - Parallel to eyelid
- - Retract extra tissue as needed
- - You will see a bleb form.
- - Caution not to advance the needle when you advance the plunger
- - Can dilute 100units/1cc or 2cc, as is your preference.
- - Ona comes in 100unit or 200unit quantities (medical); Inca is available in 50unit or 100 unit quantities.

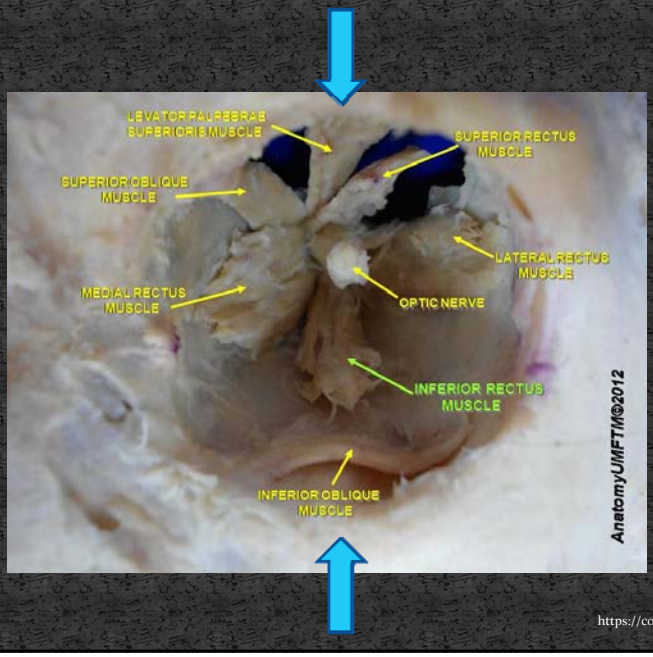
Pitfalls and Potential side effects

- Ptosis→ Avoid levator palpebrae superioris.
- Diplopia→ Avoid medial lower lid area (inferior rectus).
- Bruising
- Dry eye

- Recognize that frontalis activation may be compensatory.

- Be prepared for blinking, twitching, jerking away from you! Stabilize your hand.

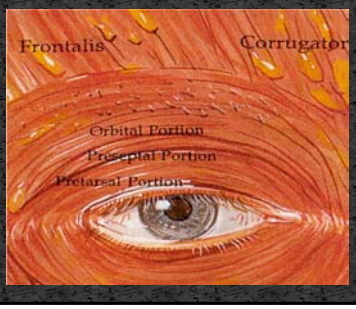
https://www.studyblue.com/notes/note/n/ap-lab-final/deck149354/3



https://commons.wikimedia.org/wiki/File:Slide12uu.JPG

Apraxia of eyelid opening

- Temporary loss of voluntary reopening
- Pretarsal motor persistence
- Levator inhibition (analogous to freezing of the levator palpebrae)
- Seen in 1/3 of patients with blepharospasm
- Patient requires to activate the frontalis to open the eyes
- Also seen in Parkinsonian syndromes



What to expect

- Benefit within 1-2 weeks (usually 3-8 days)
- Reduced blinking and spasms→ Improved ability to keep eye open
- Reemergence of the symptoms when the therapy wears off
- Patients may continue to have the injections as long as they wish to.
- May require dose titration initially, or with time.

Part 2

Hemifacial spasm- Definition

- Involuntary contraction of the facial muscles
- "Synchronous tonic and/or clonic contraction of facial muscles, caused by dysfunction of the ipsilateral facial nerve"
- Typically unilateral, though rare bilateral (asynchronous) cases occur.

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Hemifacial spasm- clinical features

- Often starts around orbicularis oculi
- Twitching and closing of the eye
- Pulling of the face
- May result in facial asymmetry
- Often worse with talking and activities, stress, and fatigue

Hemifacial spasm- Epidemiology

- Prevalence of HFS in the general population is approximately 11 per 100,000 individuals
- More common in women and in Asian populations.
  - Females 14.5 per 100,000
  - Males 7.4 per 100,000

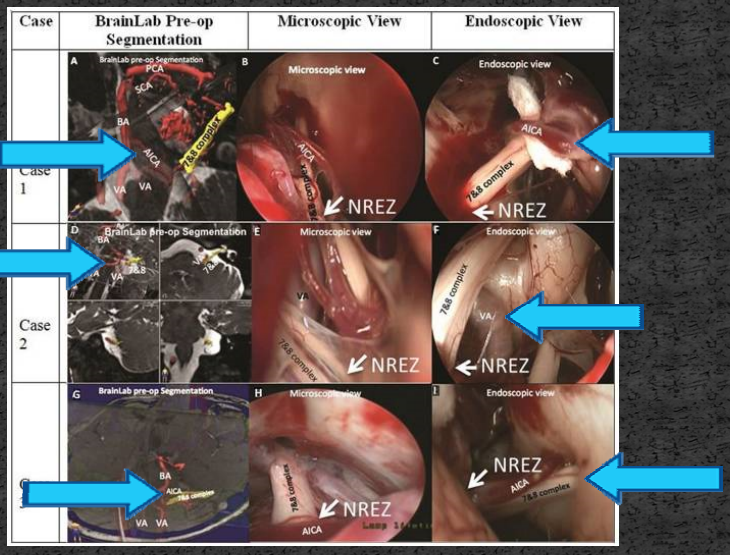
Auger RG, Whisnant JP. Hemifacial spasm in Rochester and Olmsted County, Minnesota, 1960 to 1984. Arch Neurol. 1990;47:1233-1234.

Hemifacial spasm- Causes

- Some cases are due to compression of the facial nerve by an ectatic vessel
- Cases have been reported secondary to many insults along the facial nerve course- trauma, demyelinating, infection-related, tumor, etc.
- Could be that compression leads to ephatic transmission from the injured site, or that there is central hyperexcitation
- Idiopathic cases

One group looked at high resolution imaging for causes and compared with surgical findings in refractory cases. .

Pre-Operative Image-based Segmentation of the Cranial Nerves and Blood Vessels in Microvascular Decompression: Can we Prevent Unnecessary Explorations?



MRI with 7th nerve protocol

Hemifacial spasm- Treatments

- Chemodenervation- First-line (Botulinum toxin injections)

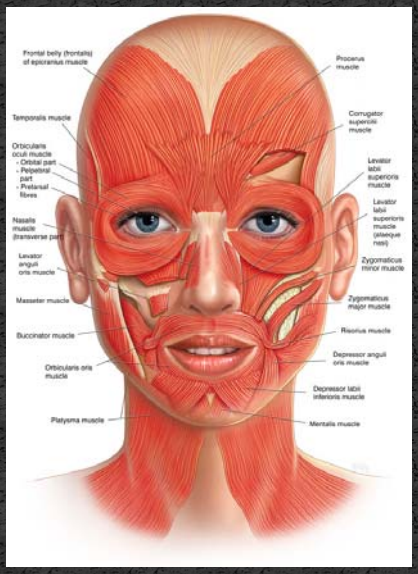
- Oral medications:
- GABA'ergic- benzodiazepines (clonazepam), baclofen
- Anticonvulsants- carbamazepine, gabapentin, levetiracetam, zonisamide

- Surgery- Microvascular decompression may be considered in refractory cases, weighing risks against potential benefits.

Videos: Hemifacial spasm

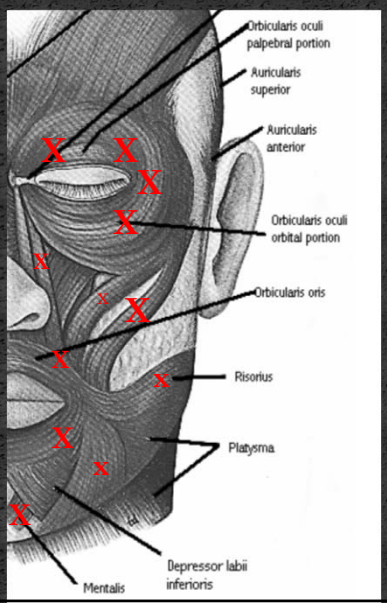
HEMIFACIAL SPASM (Off-label)- Muscles

- Similar to blepharospasm, but may add:
- Zygomaticus
- Levator labii alequae nasi
- Risorius
- Orbicularis oris
- Buccinator
- Depressor anguli oris
- Mentalis
- Platysma
- Etc.
- Generally 2.5-10 units per location (referring to onabotulinum or incobotulinum toxin)



http://slarts.weebly.com/archimbaldo.html

Hemifacial spasm



Individualize the pattern to the patient.

Hemifacial spasm- Potential pitfalls

- Same risks as discussed in blepharospasm apply.
- Treatment with botulinum toxin injections may aggravate facial asymmetry.
- Caution with zygomaticus → May paralyze action in smile.
  → May impair ability to retract the lips
- Caution with orbicularis oris → May impair ability to use straw
  → May cause droop of the mouth
- * Sometimes injecting only the upper face is adequate.
- * At times may consider injecting a small amount into the unaffected side to improve symmetry.

With good technique, you will dramatically reduce the impact on quality of life impairment from these conditions!

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Thank you!

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