

Overview of Facial Dystonia

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Disclosure Information

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- I do not intend to discuss an unapproved/investigative use of a commercial product/device in my presentation.

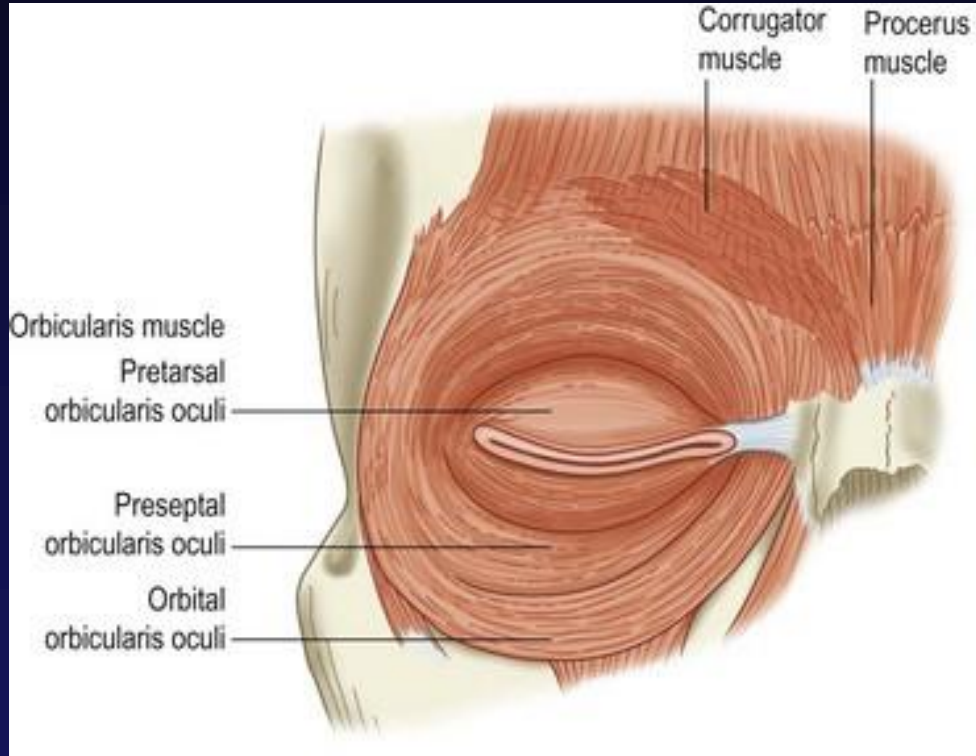
Outline

- Define dystonia
- Types of dystonia
 - Eyelid myokymia
 - Benign essential blepharospasm
 - Orofacial dystonia
 - Meige
 - Brueghel
 - Hemifacial spasm
 - Apraxia of lid opening
 - Aberrant regeneration of facial nerve

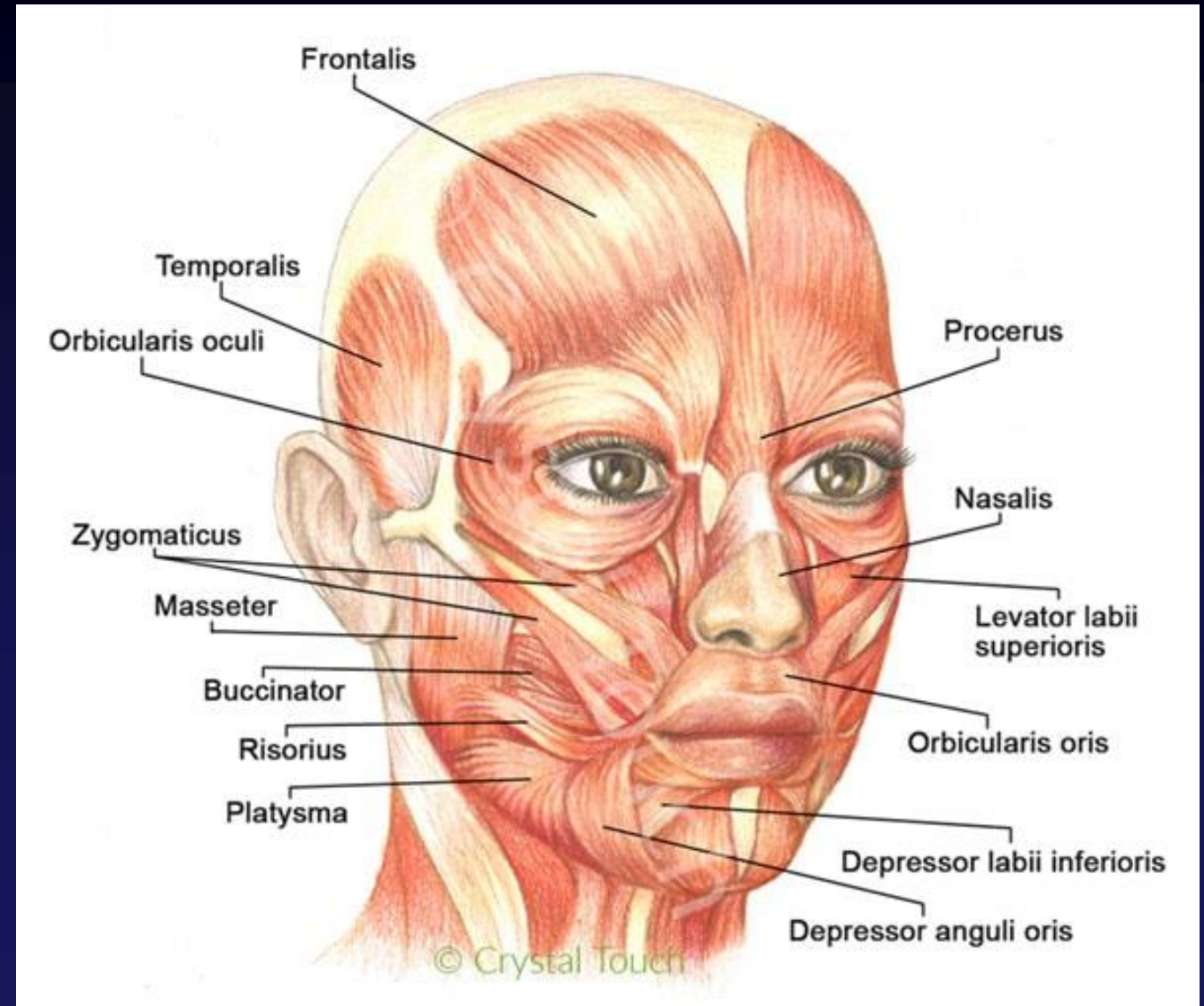
What is dystonia?

- Involuntary muscle contractions
 - Sustained contraction vs intermittent contraction (tonic vs clonic)
 - → abnormal postures or repetitive movements
- Can affect 1 muscle, groups of muscles, or muscles throughout the body
- Thought to be caused by damage or abnormalities in basal ganglia
- Can be idiopathic, genetic, or acquired

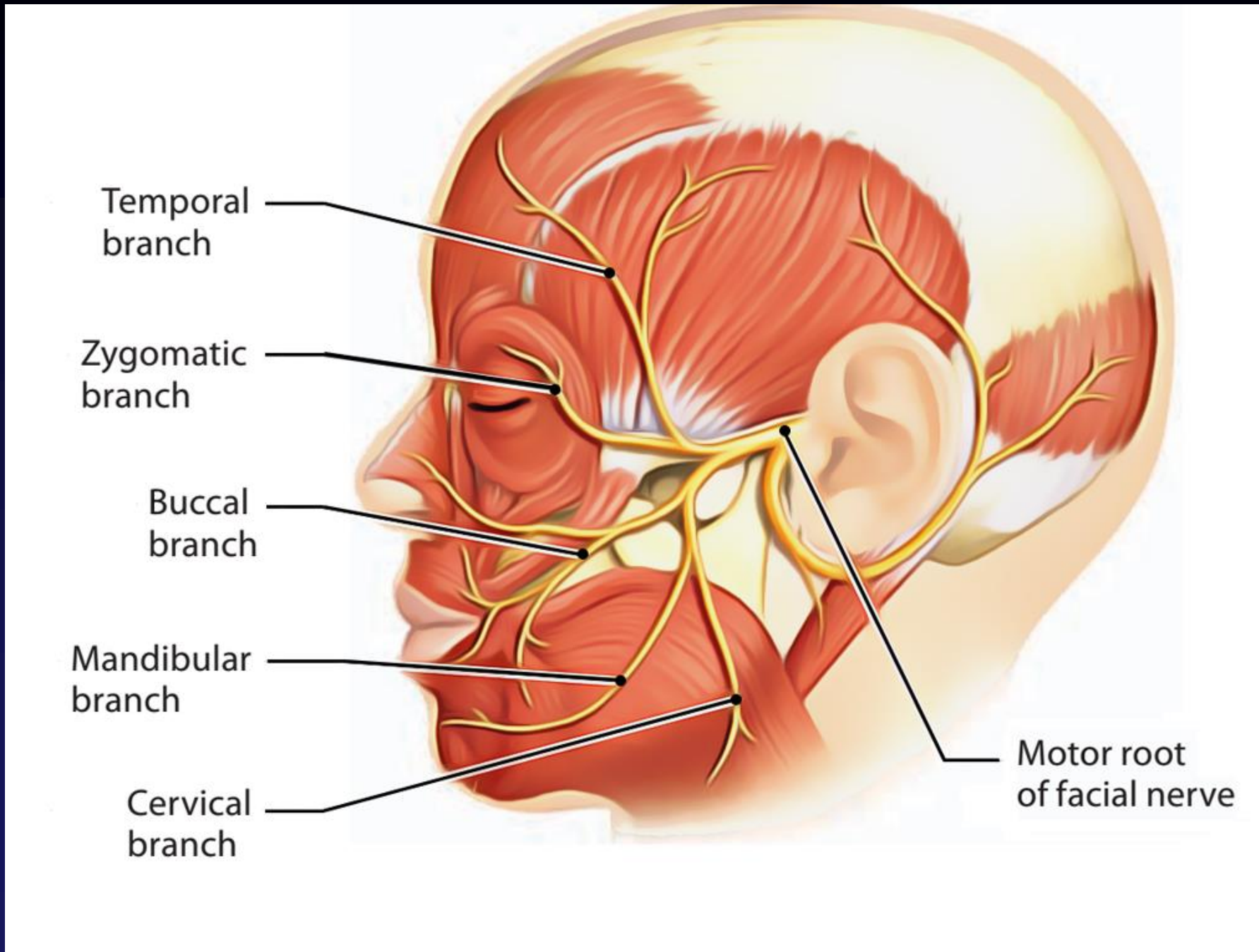
Facial muscle anatomy



<https://clinicalgate.com/eyelid-anatomy-and-function/>



<https://crystal-touch.nl/our-facial-muscles-and-their-functions/>



Types of facial dystonia

- Eyelid myokymia
- Blepharospasm
- Orofacial dystonia
 - Meige
 - Brueghel
- Hemifacial spasm

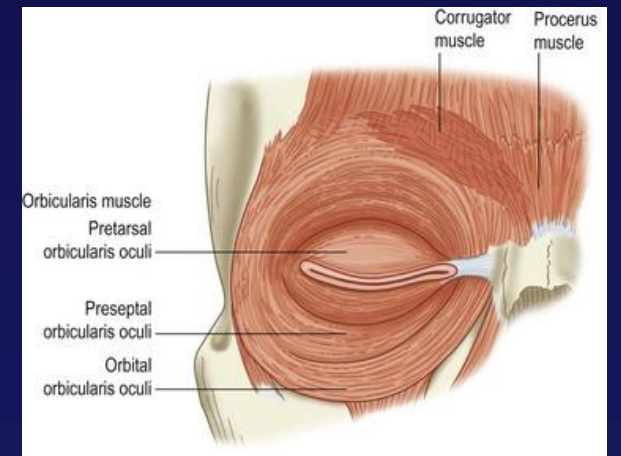
- Apraxia of eyelid opening
- Aberrant regeneration of facial nerve

Eyelid myokymia

- Fascicular contractions within orbicularis muscle
 - Twitching, flickering, rippling movements
- Usually affects one side
- Associated with stress, fatigue, caffeine
 - “Medical student’s disease”
- Treatment: avoid triggers, usually self-resolves
- Very rarely is the first manifestation of BEB or HFS

Benign Essential Blepharospasm

- Involuntary episodic closure of eyelids
 - Contractions of eyelid protractors
 - Tonic – eye is closed for prolonged time
 - Clonic – multiple contractions
- Ranges from mildly increased blink rate to forceful contractions
- Eyelid involvement ONLY
- BOTH sides affected

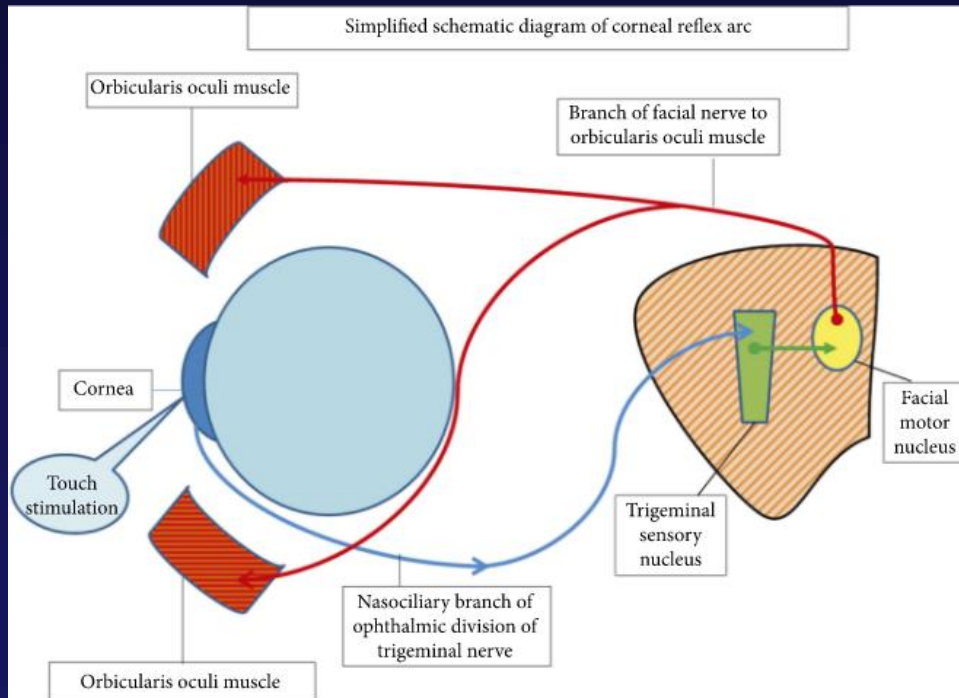


Blepharospasm

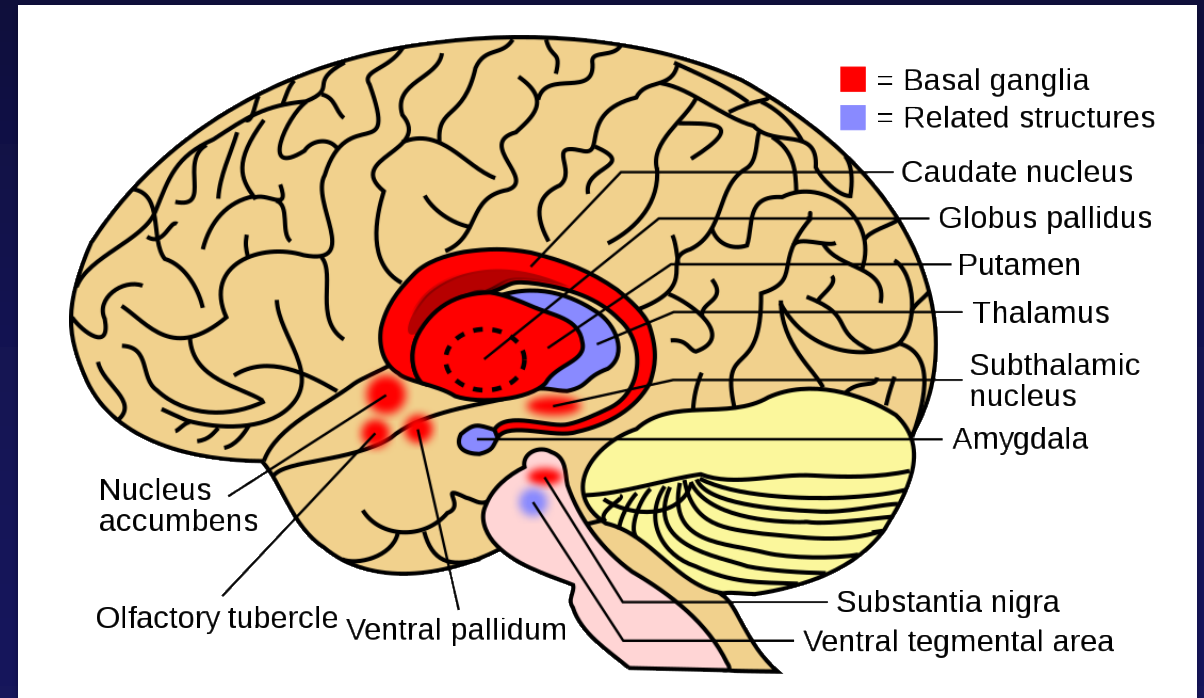
- Population
 - Ages 40-60 years
 - Female > male (2-4:1)
 - Prevalence of 1.6-30/100,000
- 2,000 cases diagnosed annually in the US

Blepharospasm

- Can be isolated or caused by other conditions
- Cause: ?defective circuit



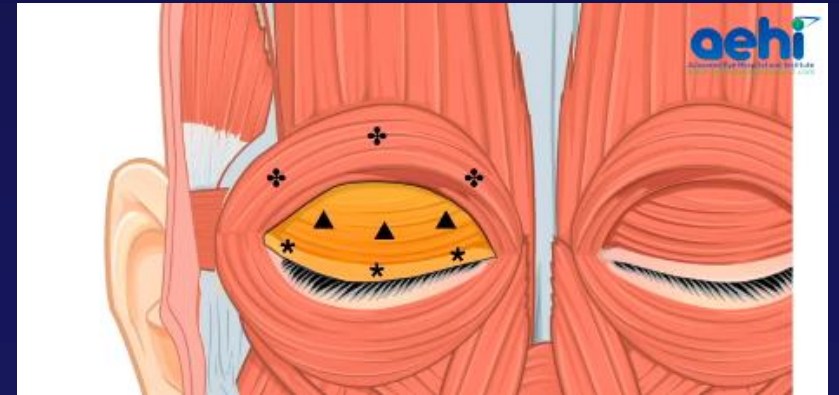
<https://www.hindawi.com/journals/prm/2021/6664736/>



https://en.wikipedia.org/wiki/Basal_ganglia

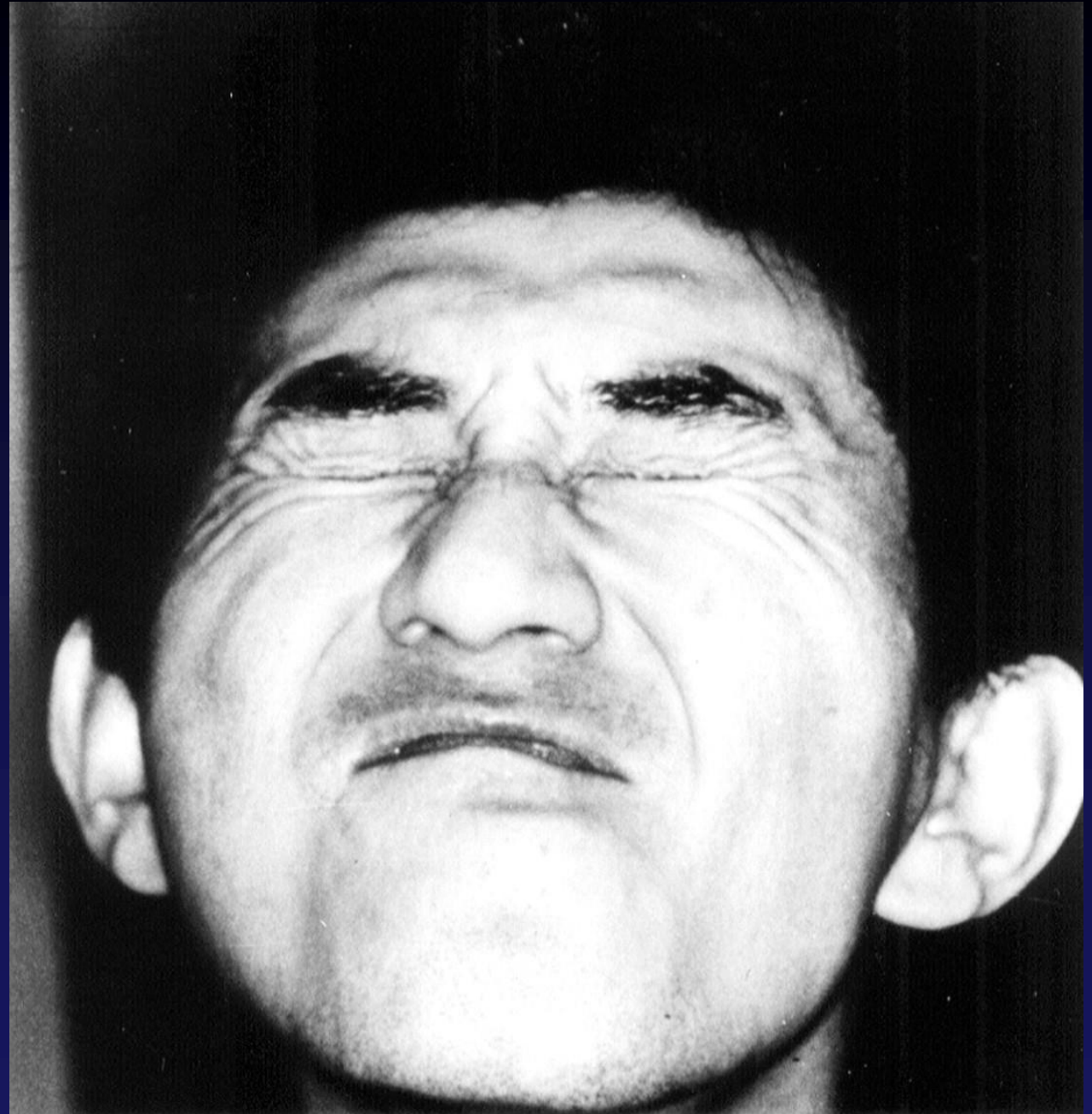
Blepharospasm

- Treatment:
 - Artificial tears, lid scrubs, tinted lenses
 - Botulinum toxin injections
 - Consider oral medications
 - Consider excision of eyelid muscles (myectomy)



Meige Syndrome

- Blepharospasm + oromandibular dystonia (lower facial spasm)
- BOTH sides affected



Meige Syndrome

- Population
 - 30-70 years old, mean age 55 years old
 - Female > males (2:1)
 - 5/100,000
- Can start as blepharospasm and then spread to involve lower face
 - Spread is more common early in disease

Meige syndrome

- Cause: dopaminergic and cholinergic hyperactivity
 - Decreased functioning of inhibitory neurons
 - Abnormal sensorimotor processing
 - Abnormal control of CN nuclei in brainstem by basal ganglia
- Can be primary or secondary

Meige Syndrome

- Treatment:
 - “Sensory tricks”
 - 1st line systemic medication
 - 2nd line botulinum toxin
 - Reserved for blepharospasm component
 - Deep brain stimulation

Brueghel Syndrome

- Jaw opening dystonia
- +/- eyelid involvement



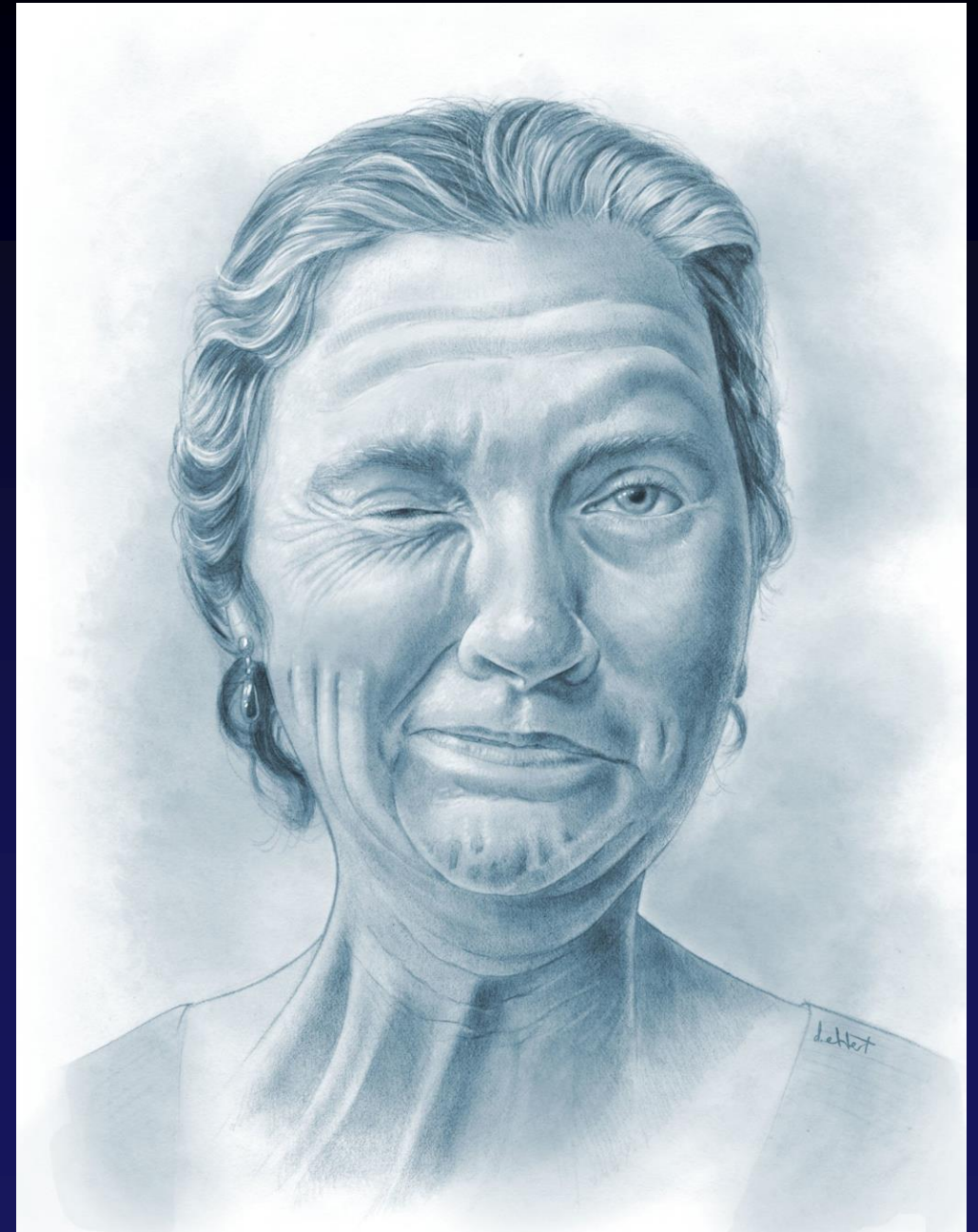
<https://jnnp.bmj.com/content/jnnp/39/12/1204.full.pdf>



<https://n.neurology.org/content/46/6/1767>

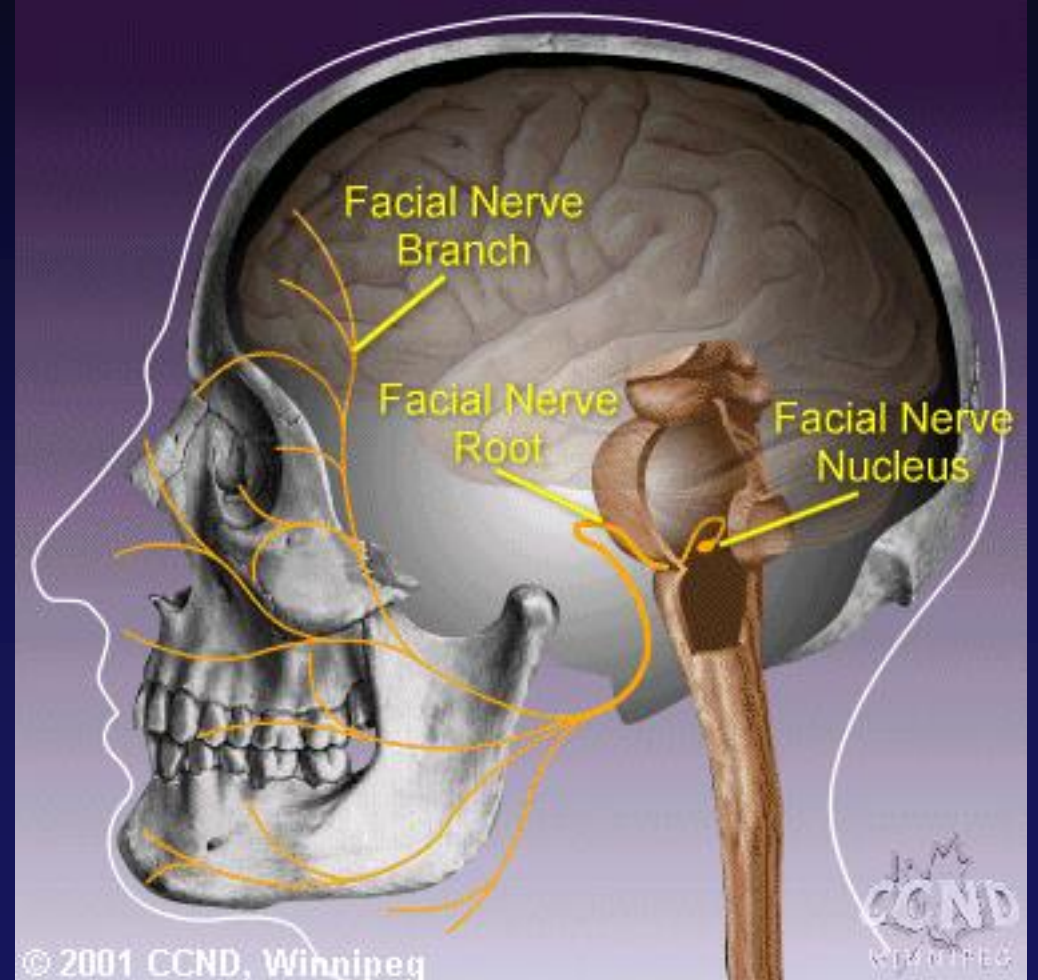
Hemifacial Spasm

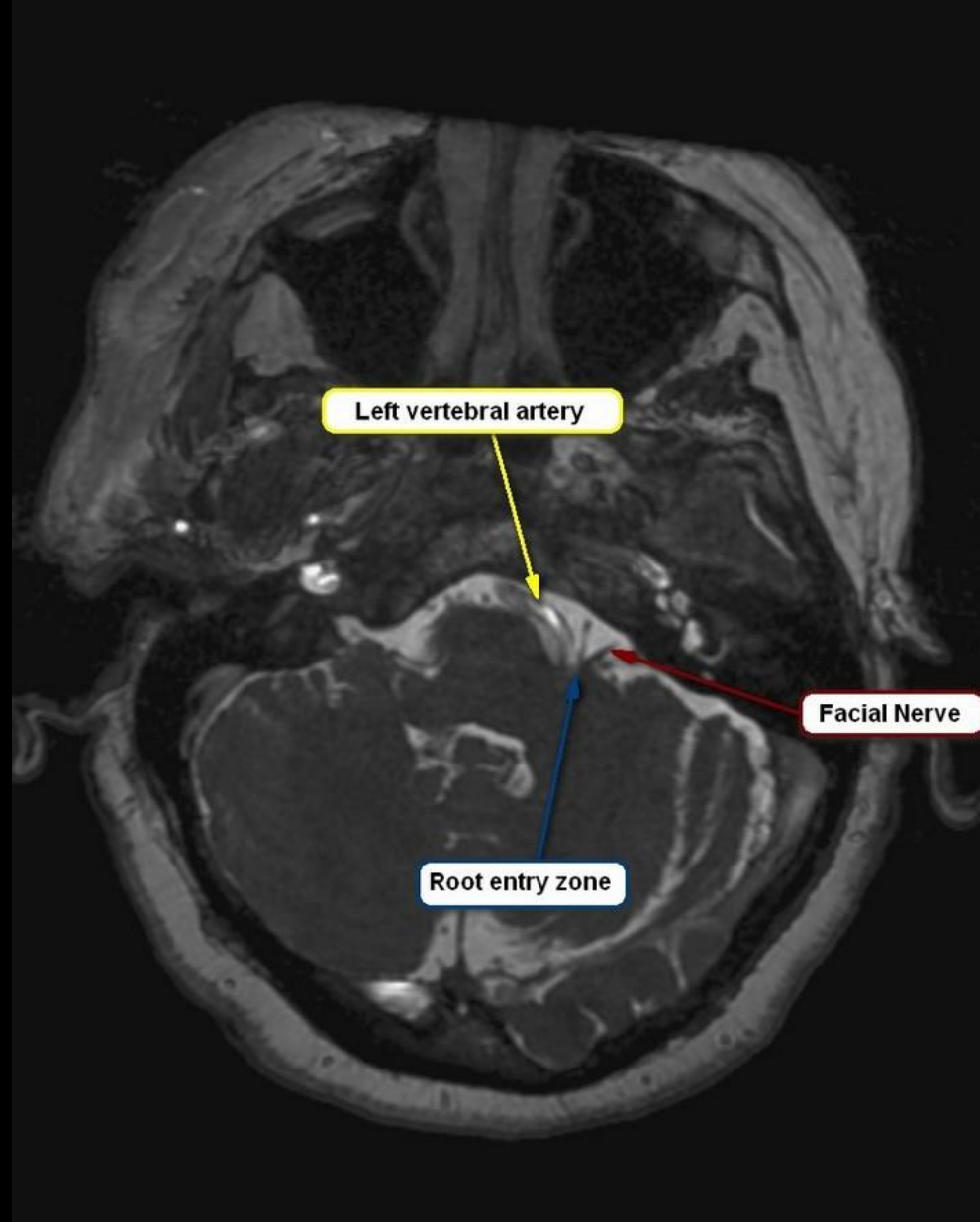
- Involuntary upper and lower facial contractions
- Usually affects one side only
- Population
 - Average age 50-60 years old
 - Women > men (2:1)
 - 8-15/100,000 in US



Hemifacial Spasm

- Causes
 - Compression of facial nerve
 - Most common = abnormal artery compressing at exit point
 - Idiopathic





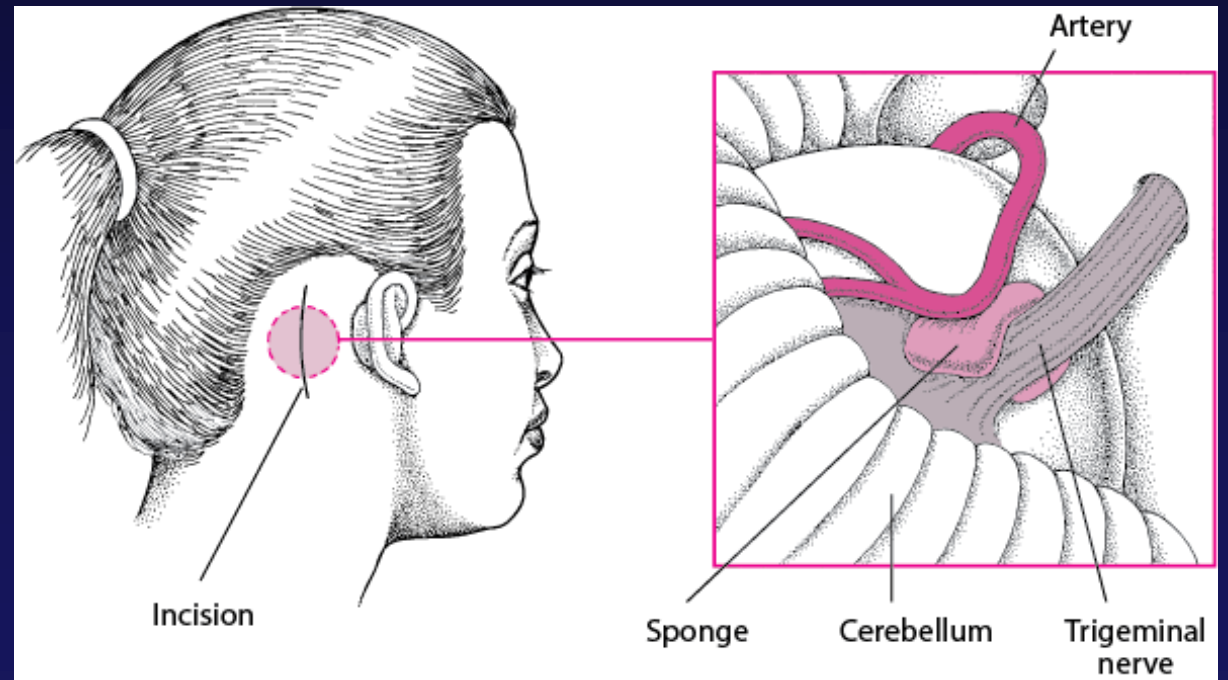
Case courtesy of Assoc Prof Frank Gaillard, Radiopaedia.org,
rID: 2617

Hemifacial Spasm

- Treatment:
 - Botulinum toxin injections
 - Neurovascular decompression surgery (Janetta procedure)



<https://danishbhatti.com/review-of-hemifacial-spasm-differentials/>



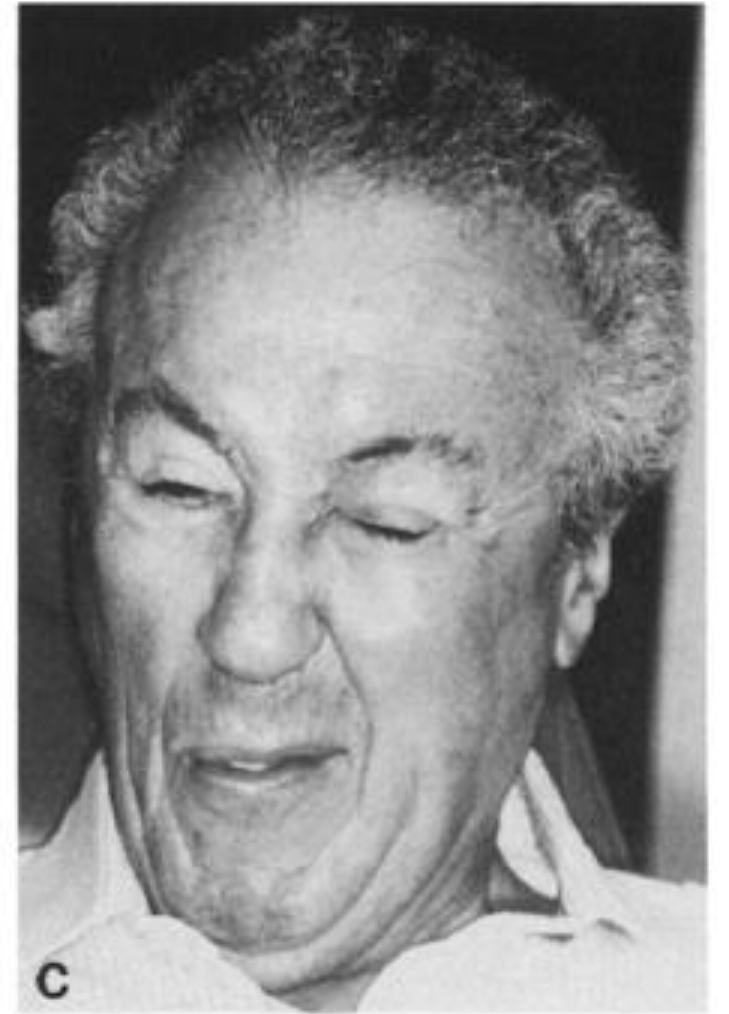
<https://www.merckmanuals.com/home/brain,-spinal-cord,-and-nerve-disorders/cranial-nerve-disorders/hemifacial-spasm>



Blepharospasm



Orofacial spasm



Hemifacial spasm

Apraxia of Eyelid Opening

- Inability to open eyes voluntarily after closing eyelids, in absence of squeezing
 - BOTH sides affected
- Commonly associated with blepharospasm but can be isolated
- Cause: poorly understood
- Treatment: botulinum toxin
 - Also: myectomy, eyelid crutches, ptosis surgery, frontalis sling

Apraxia of Eyelid Opening

> [Ophthalmic Surg.](#) 1990 May;21(5):331-4.

Apraxia of lid opening in blepharospasm

[D R Jordan](#)¹, [R L Anderson](#), [K B Digre](#)

Affiliations + expand

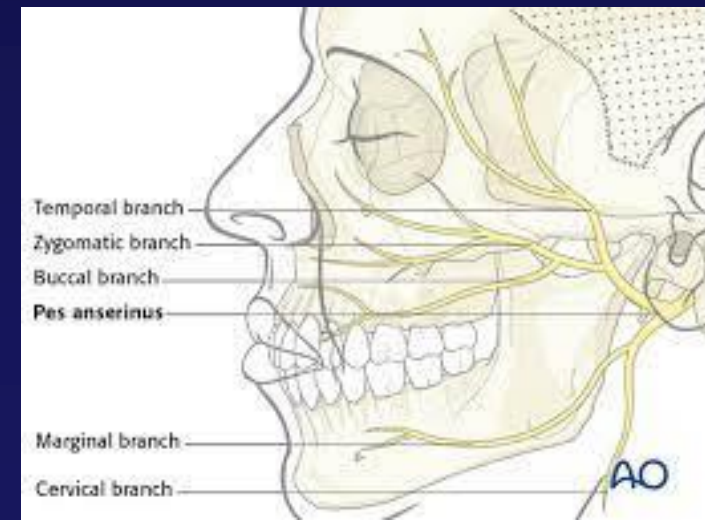
PMID: 2381654

Abstract

Apraxia of lid opening is a nonparalytic motor abnormality characterized by difficulty in initiating the act of lid elevation. It has been reported with extrapyramidal disorders, including Parkinson's disease, Huntington's chorea, progressive supranuclear palsy, and Shy-Drager syndrome. We found seven cases (7%) of functionally disabling apraxia of lid opening in 100 consecutive blepharospasm patients studied. It is important for physicians treating blepharospasm to be aware of the association between these two visually debilitating disorders.

Aberrant regeneration

- Involuntary facial movements accompanying voluntary movements
- Cause: nerve regeneration after CN VII palsy resulting in abnormal connections between peripheral branches of facial nerve
- Types
 - Ocular-oral synkinesis
 - Crocodile tears
 - Hypertonicity



	Blepharospasm	Orofacial spasm	Hemifacial spasm	Apraxia of eyelid opening
Affected sites	Eyelids	Eyelids Lower face	Eyelids Lower face	Eyelids
Laterality	Both sides	Both sides	One side	Both sides
Cause	Unknown	Unknown	Compression of CN VII(vessel) Can be idiopathic	Unknown
Treatment	Botox Myectomy	Oral medications Botox	Botox Decompression of CN VII	Botox Myectomy Ptosis surgery Frontalis sling

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