



Jefferson

Philadelphia University +
Thomas Jefferson University

Lower Facial & Neck Dystonias

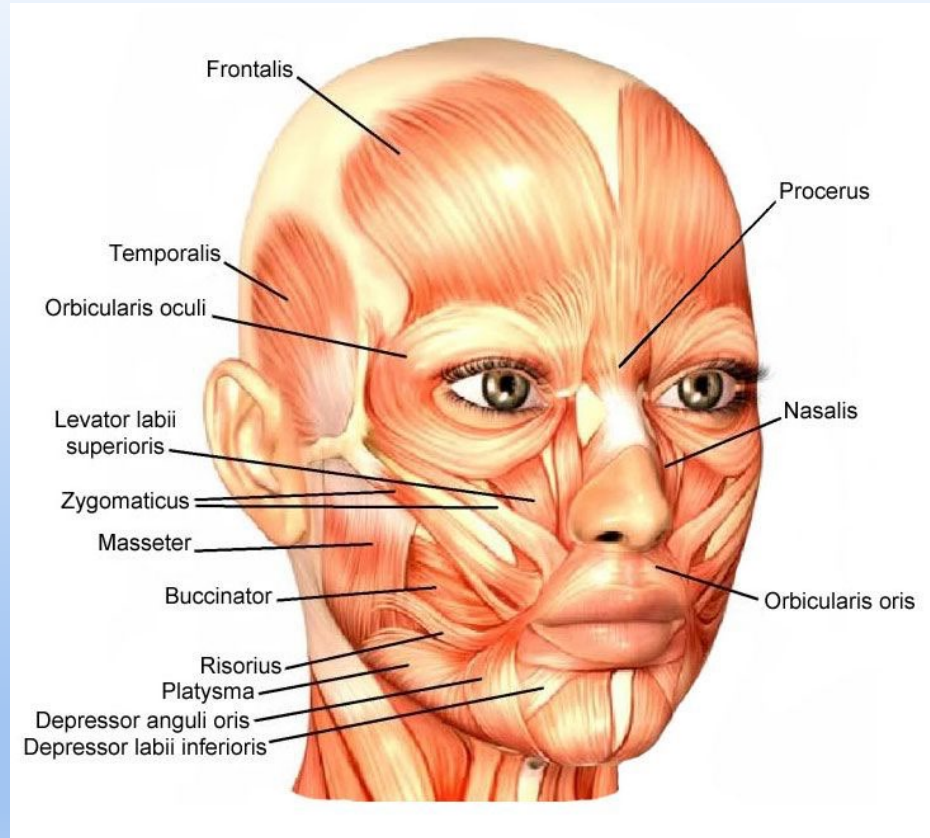
Howard Krein MD, PhD, FACS
Associate Professor
Division of Facial Plastic and Reconstruction
Thomas Jefferson University

Personal Introductions

- **Howard Krein, MD, PhD, FACS**
 - Local born and raised
 - UMDNJ, PhD, Neuroscience
 - Jefferson, MD
 - Jefferson, Otolaryngology- Head and Neck Surgery Residency
 - VCU, Facial Plastic & Reconstructive Surgery Fellowship
 - Facial Plastic and Reconstructive Surgery
 - Head and Neck Microvascular Surgery
 - Jefferson Facial Nerve Center
 - Herbert Kean Center for Facial Aesthetics

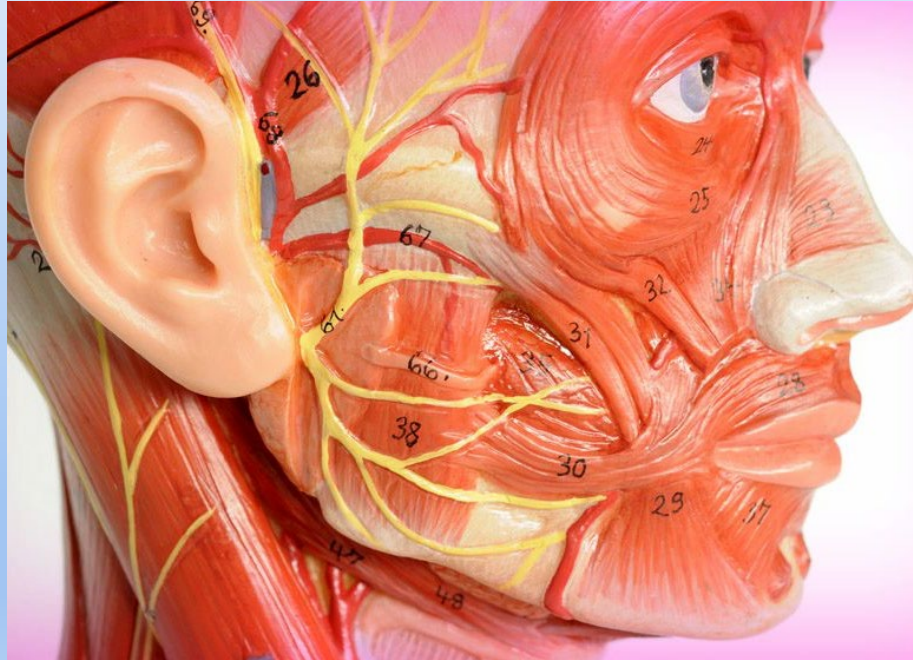
Muscles of Facial Expression

- Innervated by CNVII distal branches in a variably redundant pattern
- Results in non-uniform paresis
- Buccal branching is more redundant than marginal mandibular or frontal branch innervation

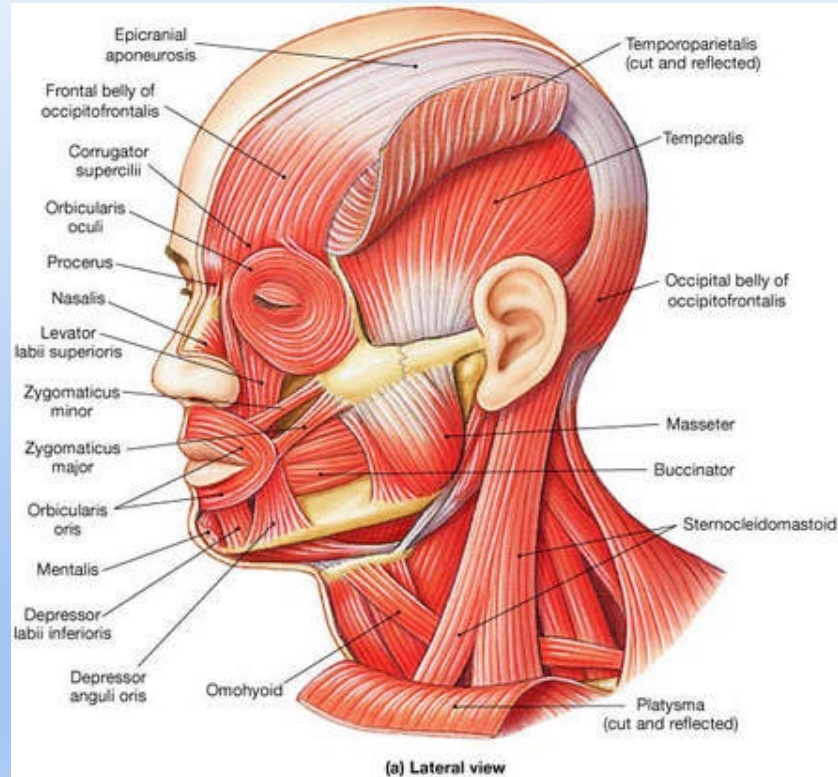


Muscles of Facial Expression

- Innervated by CNVII distal branches in a variably redundant pattern
- Results in non-uniform paresis
- Buccal branching is more redundant than marginal mandibular or frontal branch innervation

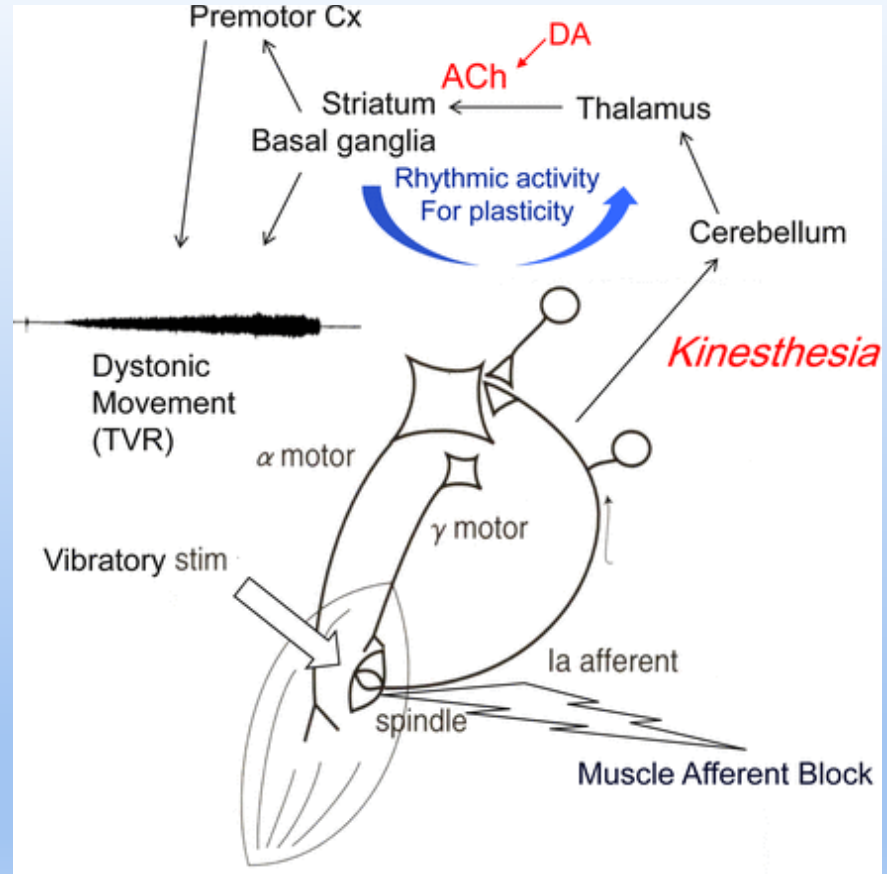


Muscles of Mastication



General Overview

- Highly variable neurological movement disorder characterized by involuntary muscle contractions that are patterned and repetitive
- Exact cause of dystonia is not yet known but may involve alteration in several regions of the brain or the communication between them (abnormal function of the basal ganglia)
- Dystonia may be congenital, acquired, or idiopathic



Overview

- Dystonia is classified by three main factors: age; the areas of the body affected; and the underlying cause
- Age: Childhood onset - 0 to age 12, Adolescent onset - age 13 to 20 Adult onset - older than age 20
- Body Part: Focal, Segmental
- Cause: Primary (idiopathic), Secondary (symptomatic), Heredodegenerative

How are Lower Facial and Neck Dystonias Diagnosed...

- Clinical Diagnosis
- Must rule out other motor/neurological causes
- No validated diagnostic criteria (no specific Laboratory or imaging study)

Meige Syndrome

- Aka Brueghel's syndrome
- Combination of upper face (blepharospasm) and lower face (Oromandibular) dystonias
- First described in 1900s
- Women>men
- Varied age of onset (30-70)

Meige Syndrome -Blepharospasm symptoms

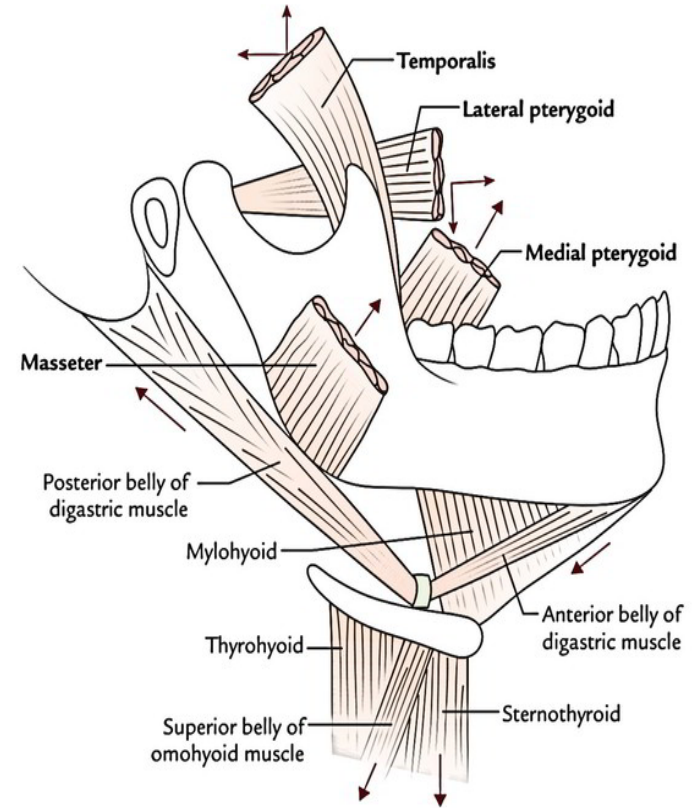
- the first symptom to appear is an increased rate of blinking^[2]
- uncontrollable squinting/closing of eyes
- light sensitivity (photophobia)
- squinting/eyes closing during speech
- uncontrollable eyes closing shut (rare instances completely causing blindness)

Meige Syndrome - Oromandibular Symptoms

- difficulty opening the mouth (trismus)
- clenching or grinding of the teeth (bruxism)
- spasms of jaw opening
- sideways deviation or protrusion of the jaw
- lip tightening and pursing
- drawing back (retraction) of the corners of the mouth
- deviation or protrusion of the tongue.
- jaw pain
- difficulties eating and drinking
- difficulties speaking (dysarthria)

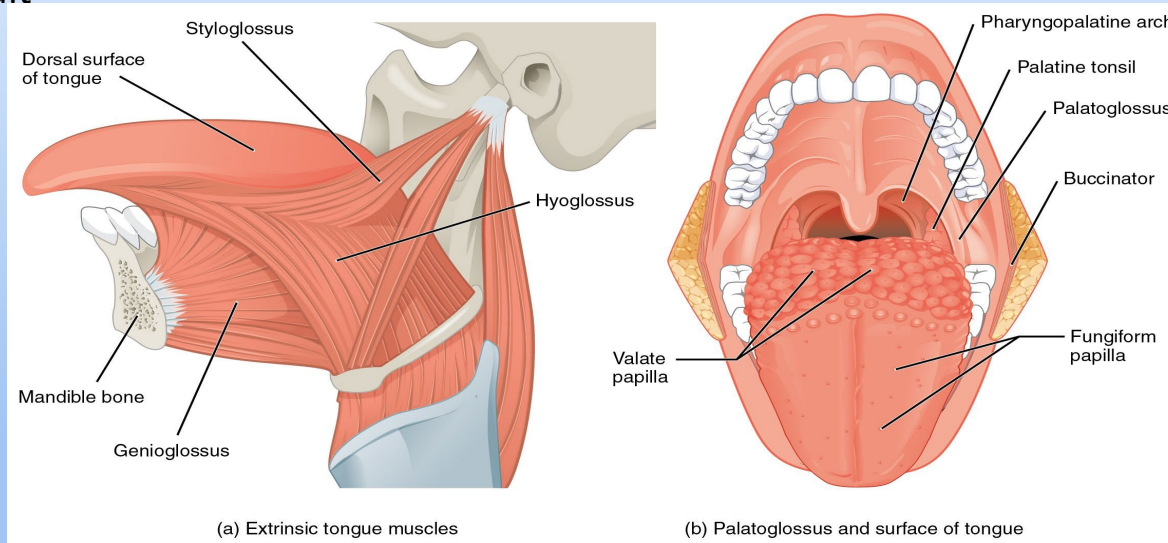
Oromandibular dystonia

- Masseter (closes jaw by elevating the mandible)
- Temporalis (closes jaw, retract mandible)
- Medial Pterygoid (elevation and protrudes mandible)
- Lateral Pterygoid (opens mouth, protrudes the jaw)
- Digastric (opens jaw, elevates the hyoid bone)
- Geniohyoid (opens jaw, elevates and draws hyoid forward)
- Mylohyoid (opens jaw, raises the floor of mouth)



Orofacial and Lingual

- Mainly the zygomaticus major and minor, the orbicularis oris and the buccinators
- In lingual dystonia especially in patients with severe tongue protrusion treatment can be difficult



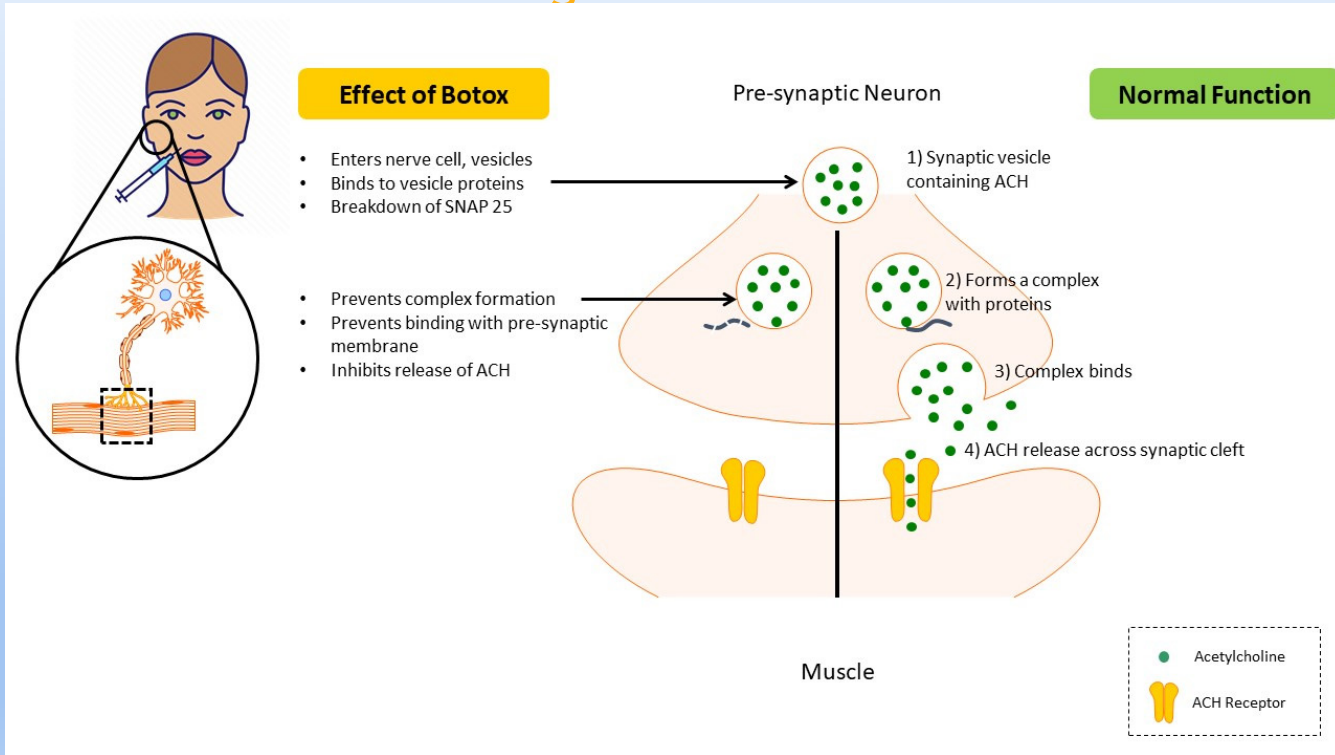
Cervical Dystonia

- A focal dystonia arising from involuntary activation of muscles in the neck and shoulders causing turning, tilting, flexion or extension movements of the head, sometimes combined with elevation or anterior shifting of the shoulders
- Patterned
- Repetitive
- Abnormal head position
- Most common focal dystonia
- Head tremor 60-70%
- Neck Pain 70-80%

Treatments

- Medications
- Clonazepam (Klonopin®)
- Trihexyphenidyl (Artane®)
- Diazepam (Valium®)
- Baclofen (Lioresal®).
- Success with oral medications is limited, however.

Treatment- Botulinum Toxin is the first-line treatment per AAN guidelines



Botulinum Toxin Injection

- Begins with analysis
- Facial muscles involved
- Abnormal movements ie
 - Antero-collis
 - Postero-collis
 - Latero-collis
 - Torti-collis

Botulinum Toxin Injection

- 4 Brands approved for Cervical Dystonias
 - OnabotulinumtoxinA (Botox)
 - IncobotulinumtoxinA (Xeomin)
 - AbobotulinumtoxinA (Dysport)
 - RinabotulinumtoxinB (Myobloc)
- No definitive difference in results
- > 70% of patients benefit on some level
- Timed results, onset and offset
- Can have significant side effects if not done properly

Botulinum Toxin Injection Tools

- EMG - Electromyogram
- Ultrasound

Table 2. Common Muscles and Dosing for Blepharospasm and Oromandibular Dystonia.

Focal Dystonia	Common Muscles Injected	Common Starting Doses (U) (onaBoNT/A, incoBoNT/A) *
Eye Closure	Orbicularis Oculi	20 to 25
Expression	Corrugator	10
	Procerus	5
	Frontalis	20
Jaw Closure	Masseter	50
	Temporalis	40
	Medial Pterygoid	20
Jaw Opening	Lateral Pterygoid	20
	Anterior Belly of Digastric	5 (per belly)
Jaw Deviation	Contralateral lateral pterygoid	20
	Ipsilateral temporalis	40
Tongue Protrusion	Genioglossus	20 (10 each side)

* IncoBoNT/A has been studied and shown efficacious for blepharospasm but data is limited for use in OMD. Dosing data derived from multiple sources [28,33–35].

Deep Brain Stimulation

- Indicated for patients refractory to Botulinumtoxin
- Microelectrodes placed in the BG
- Pulse Generator

Possible Benefits

- Physical Therapy
- Speech and Swallow Therapy
- Acupuncture
- Chiropractic
- Stress Reduction
- Diet
- Exercise

Head and Neck Physical Therapy

- **Stacey Baer, MS, OTR/L, CLT**
 - Graduated Thomas Jefferson University December 2009
 - TJUH (2010 to 2012)
 - NYU Langone Medical Center, Rusk Rehabilitation (2012-2016)
 - New York Presbyterian Columbia University (2016-2018)
 - Facial Nerve Rehab trainings at Mass Eye and Ear (2017)
 - Certification in Lymphedema Therapy (2017)
 - Private practice facial nerve rehab specialist (2019)
 - Co-founder, Facial Nerve Rehab Specialist @ Jefferson Facial Nerve Center (2020-present)