
NEUROLARYNGOLOGY:
LARYNGOLOGIC MANIFESTATIONS OF
NEUROPATHOLOGY
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OBJECTIVES

- UNDERSTAND THE METHODS OF CLASSIFICATION FOR THESE DISORDERS
- RECOGNIZE SIGNS/SYMPTOMS OF NEUROLARYNGEAL DISORDERS
- CONSIDER THE LOCATION/ETIOLOGY OF THE LESION
- IDENTIFY AND DESCRIBE NEUROLOGIC DISORDERS THAT AFFECT THE LARYNX
- DEFINE WORK-UP FOR THESE DISORDERS
- DESCRIBE TREATMENT OPTIONS

NEUROLARYNGOLOGY

- PERIPHERAL: PARESIS/PARALYSIS
- MOVEMENT DISORDERS
- PYRAMIDAL DISEASE
- NEURODEGENERATIVE/NEUROMUSCULAR



Appleby AL, Herman AG, Smith ID, Aboukhalil M, Adkins RW, Sulica L. *Subglottic cysts: Common movement-related disorders affecting the larynx: a report from the neurolaryngology committee of the AAO-HNS. Otolaryngol Head Neck Surg.* 2020;133(5):634-643.

SIGNS SUGGESTIVE OF A NEUROGENIC VOICE DISORDER

VOCAL FATIGUE
 VOCAL TREMOR
 WEAK OR BREATHY VOICE
 VOCAL STRAIN OR STOPPAGE
 ALTERED RESONANCE
 ACQUIRED DYSPHAGIA
 ACCOMPANYING DYSPHAGIA

Waxman, L. "Management of Laryngeal Lesions of the Voice." *Am J Otol Rhinol Laryngol*. 2016;37(10):1131-7.

Where is the lesion?	
Cortex	Aphasia Aphonia Dysarthria Dysphonia Stridor
Extrapyramidal system	Vocal strain Pitch breaks Tremor Spasmodic movements Focal, regional, generalized dystonias
Cerebellum	Ataxia Dysmetria Tremor Discoordination
Brainstem	Flaccid paralysis Usually involves >1 cranial nerve

NEUROLARYNGEAL WORK-UP

- HISTORY
- PHYSICAL EXAM
- NEURO-LARYNGEAL EXAMINATION
- VIDEOSTROBOSCOPY
- DIAGNOSTIC TESTS
 - EMG
 - FEEST
 - RADIOGRAPHIC
 - LABORATORY
- NEUROLOGY REFERRAL

HISTORY

- WHAT IS THE PROBLEM?
- ONSET?
- WHAT MAKES IT WORSE/BETTER?
- ASSOCIATED SYMPTOMS

EXAM

- VOICE
 - QUALITY: ROUGH, STRAINED, BREATHY
 - FLOW: DECREASED BREATH SUPPORT, PROJECTION OR VOLUME
 - CONTROL: PITCH
 - FATIGUE, DECREASED RANGE, VOICE BREAKS
- NEURO
 - STARTS WHEN THE PATIENT WALKS IN
 - OBSERVE WRITING, ARCHIMEDES
 - NEURO EXAM: STRENGTH, SENSATION, DISDIADYMNESIS/CB
- NEUROLARYNGEAL EXAM
 - USE FLEXIBLE SCOPE
 - ASSESS MOBILITY OF VOCAL FOLDS
 - SPORE
 - DIFFERENTIATE NEUROLOGIC FROM JOINT HYPO-MOBILITY. EVALUATE TONE, LOOK FOR MOVEMENT DISORDERS
 - IS THERE ACTIVITY AT REST?
 - ARE ACCESSORY MUSCLES IN USE?
 - IS THE MOTION CONSISTENT?
 - ISOLATION: ADDUCTOR, ABDUCTOR, TENSOR
 - AGILITY
 - COORDINATION

ELECTROMYOGAPHY: USES

- CONFIRM PARALYSIS AND PROGNOSIS
- DIFFERENTIATE LARYNGEAL PARALYSIS FROM CA JOINT ANKYLOSES
- DIAGNOSIS OF: MG, ALS, MYOPATHY
- DETERMINING SITE OF LESION: RLN, SLN, HIGH VAGAL
- EVALUATING LARYNGEAL SYNKINESIS
- DELIVERY OF BOTOX IN HYPERFUNCTIONAL DISORDERS



Meyer TJ, Hagi AD, Bitzer A. Electromyography of laryngeal and pharyngeal muscles. *Neurologic disorders of the larynx*. Thame; 2009.

ELECTROMYOGAPHY: TECHNIQUE

- ELECTRODE PLACEMENT: DETECT INSERTIONAL ACTIVITY
- VERIFICATION OF PLACEMENT IN APPROPRIATE MUSCLE
- EVALUATE MUSCLE AT REST: DETECT SPONTANEOUS ACTIVITY
- EXAMINE INDIVIDUAL MOTOR UNITS
- ASSESS RECRUITMENT
- DETECT SYNKINESIS



Meyer TJ, Hagi AD, Bitzer A. Electromyography of laryngeal and pharyngeal muscles. *Neurologic disorders of the larynx*. Thame; 2009.

BOTOX FOR DYSTONIA

- EMG-GUIDED INJECTION OF BOTULINUM A INTO AFFECTED MUSCLES
- DELIVER VOLUME OF ABOUT 0.1 CC INTO EACH VOCAL FOLD
- PATIENTS NEED A VERY WIDE RANGE OF DOSAGES
- USE ANATOMY, INSERTIONAL ACTIVITY, VOLITIONAL VERIFYING ACTIVITY
 - AbSD: TA INJECTION, BILATERAL
 - AbSD: PCA INJECTION, UNILATERAL AND CHECKED WITH FLEXIBLE SCOPE



MOVEMENT DISORDERS

HYPERFUNCTIONAL DISORDERS	
Dystonia	Prolonged muscle contractions resulting in abnormal postures
Tremor	Rhythmic sinusoidal oscillation of a body part or segment
Myoclonus	Sudden brief muscle contractions of a body part or segment
Chorea	Involuntary, irregular, semipurposeful motion flowing from one body part to another
Tics	Abnormal involuntary movement or sound production

- HYPOFUNCTIONAL DISORDERS
- SLOWER
 - STIFFER
 - WEAKER

HYPOFUNCTIONAL DISORDERS	
Central	Medullary disorders Arnold-Chiari malformations Brainstem cerebrovascular accident
Peripheral	Vagal trauma Iatrogenic injury infection Neoplasm
Neuromuscular Junction	Myasthenia gravis Lambert-Eaton syndrome
Myopathies	Myotonic dystrophy Oculopharyngeal muscular dystrophy Myositis Dermatomyositis Fratric thyroid function (dysregulation)

MIXED DISORDERS

Parkinson's disease	
Multiple sclerosis	Manifestations variable: tremor, parkinsonism, dystonia, paroxysmal dyskinesias, ataxia

HYPERFUNCTIONAL DISORDERS

DYSTONIA

- PROLONGED MUSCLE CONTRACTIONS RESULTING IN ABNORMAL POSTURES
- PRIMARY OR SECONDARY, EARLY OR LATE
 - MAY ALSO HAVE TREMOR
- HERITABLE AS AD, AR, OR X-LINKED
- LARYNGEAL DYSTONIA HAS SEVERAL SUBTYPES
- OFTEN ACTION-INDUCED AND ACTION-SPECIFIC
- DYSTONIAS WE SEE ARE USUALLY CRANIAL OR CERVICAL
 - BLEPHAROSPASM, TORTICOLLIS, OMD OR LARYNGEAL
- 6.7% WILL PROGRESS TO GENERALIZED
- GESTE ANTAGONISTE

Shibata A, Kuroki C. The role of genes in causing dystonia. *Eur J Neurol*. 2010; 23(12):1457-65.
 Warrick A, Aminoff E, Edwards J, et al. Whole-genome linkage on Aquapapain family (Dy1) is chromosome 16q24. *Mov Disord*. 2004; 19(12):1457-65.
 Warrick A, Aminoff E, Edwards J, et al. Cervical dystonia in monozygotic twins: case report and review of the literature. *Mov Disord*. 2004; 19(12):1457-65.
 Dalakas G, Gajjar A. The environmental epidemiology of primary dystonia. *Trends Other Hyperkinet Mov*. 2011; 1(1):1-11.

ADDUCTOR SPASMODIC DYSPHONIA (ASD)

- PRIMARY DYSTONIA RESULTS IN SPASMODIC CONTRACTION OF THE ADDUCTORS
- VOICE IS INTERMITTENTLY STRAINED/STRANGLER ESPECIALLY WITH VOWELS FOLLOWED BY A GLOTTIC BREAK
- TASK-SPECIFIC (DOES NOT OCCUR WITH YELLING, SINGING, HUMMING, WHISPERING)
- HERITABLE
- CAN DISPLAY MIXED CHARACTERISTICS

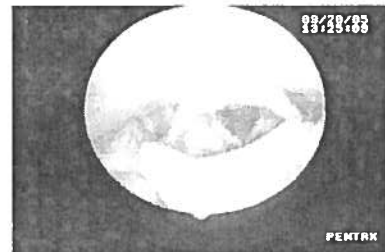
Smith MA, Barabási A, Corrado C, et al. The focal dystonias: Current views and challenges for future research. *Mov Disord*. 2014; 29(14):2153-63.
 Barabási A, Smith MA, Fong J, et al. Neurologic Disorders of the Larynx. In: Aminoff BA, Vekovick M, eds. *Movement Disorders of the Larynx and Neck*. New York, NY: Thieme; 2019. pp.140-155.



ABDUCTOR SPASMODIC DYSPHONIA

- PRIMARY DYSTONIA CAUSED BY SPASMODIC CONTRACTION OF THE ABDUCTORS
- VOICE IS INTERMITTENTLY BREATHY, EFFORTFUL OR APHONIC
 - UNVOICED CONSONANTS FOLLOWED BY VOWELS
- TASK-SPECIFIC
- HERITABLE
- CAN BE MIXED

Shibata A, Aminoff E, Edwards J, et al. Abductor laryngeal dystonia: a series treated with botulinum toxin. *Laryngoscope*. 2007; 117(12):2182-9.
 Aminoff E, Edwards J, Barabási A, et al. Botulinum toxin treatment of laryngeal dystonia. *Laryngoscope*. 2007; 117(12):2182-9.
 Aminoff E, Edwards J, Barabási A, et al. Botulinum toxin treatment of laryngeal dystonia. *Laryngoscope*. 2007; 117(12):2182-9.



TREATMENT

- BEHAVIORAL
 - VOICE THERAPY
 - SENSORY TRICK
- MEDICATIONS
 - MUSCLE-RELAXANTS, ANXIOLYTICS, DOPAMINE RECEPTOR BLOCKERS, XYREM
- BOTULINUM TOXIN
- SURGERY
 - RLN SECTION
 - SELECTIVE LARYNGEAL ADDUCTOR DENERVATION-REINNERVATION (SLAD-R)
 - DEEP BRAIN STIMULATION

Bres JH, Farn S, Musteert C, et al. Localized injections of botulinum toxin for the treatment of focal dystonia and hemifacial spasm. *Ann Disord*. 2017; 237: 24-1187

OROMANDIBULAR DYSTONIA

- FOCAL OR SEGMENTAL DYSTONIA
- INCIDENCE 70 PER MILLION
- USUALLY INVOLVES JAW OPENING OR CLOSING
- TASK-SPECIFIC (SPEAKING VS. EATING)

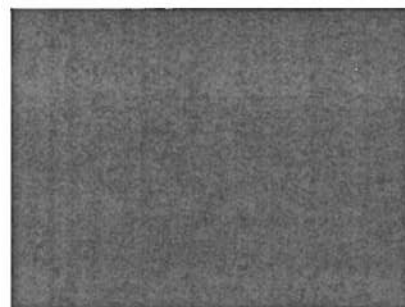
Donofoan, JM, CD Nardien, SA Schneider, KP Bhatia. Nardien's book of Movement Disorders. Oxford: Oxford University Press; 2017



MUSCLE TENSION DYSPHONIA

- DISTURBED VOCAL FOLD BEHAVIOR CAUSED BY INCREASED TENSION OF THE PARALARYNGEAL MUSCULATURE
- FUNCTIONAL DISORDER
- ETIOLOGY
 - PSYCHOLOGICAL OR PERSONALITY FACTORS
 - VOICE ABUSE
 - COMPENSATION FOR UNDERLYING PATHOLOGY
- WHERE WE COME IN
 - RECOGNIZE THE DISORDER
 - REFER TO AN APPROPRIATE SPEECH PATHOLOGIST
 - MEDICALLY/SURGICALLY ADDRESS OTHER PATHOLOGY

Van Houtte E, Van Lierde and S. Claeys. Pathophysiology and treatment of muscle tension dysphonia: A Review of the Current Knowledge. *Journal of Voice*, 2011, 25(2), p. 202-207



TREMOR

- INVOLUNTARY OSCILLATORY MOTOR ACTIVITY WITH CONSTANT PERIODICITY AND AMPLITUDE
- RANGES IN FREQUENCY, AMPLITUDE AND EXACERBATING/ATTENUATING FACTORS
- FREQUENTLY A COMPONENT OF ESSENTIAL TREMOR
- VOICE IS TREMULOUS OR QUAVERING, VOICE BREAKS OR PITCH/LOUDNESS CHANGES
- TREMOR IS PRESENT AT RELATIVE REST AND ACTIVITY; OFTEN PRONOUNCED WITH ACTIVITY
- TREATMENT
 - VOICE THERAPY
 - MEDICATIONS: ORAL ANTIHYPERTENSIVES, ANTICONVULSANTS, CCB, BZD

ICD-9-CM	ICD-10-CM	ICD-10-CM
34.00	G25.00	Essential tremor
34.01	G25.01	Essential tremor of the head
34.02	G25.02	Essential tremor of the voice
34.03	G25.03	Essential tremor of the hand
34.04	G25.04	Essential tremor of the arm
34.05	G25.05	Essential tremor of the leg
34.06	G25.06	Essential tremor of the foot
34.07	G25.07	Essential tremor of the face
34.08	G25.08	Essential tremor of the trunk
34.09	G25.09	Essential tremor of the neck
34.10	G25.10	Essential tremor of the jaw
34.11	G25.11	Essential tremor of the larynx
34.12	G25.12	Essential tremor of the pharynx
34.13	G25.13	Essential tremor of the esophagus
34.14	G25.14	Essential tremor of the stomach
34.15	G25.15	Essential tremor of the intestine
34.16	G25.16	Essential tremor of the rectum
34.17	G25.17	Essential tremor of the bladder
34.18	G25.18	Essential tremor of the uterus
34.19	G25.19	Essential tremor of the vagina
34.20	G25.20	Essential tremor of the penis
34.21	G25.21	Essential tremor of the testis
34.22	G25.22	Essential tremor of the epididymis
34.23	G25.23	Essential tremor of the vas deferens
34.24	G25.24	Essential tremor of the ureter
34.25	G25.25	Essential tremor of the bladder neck
34.26	G25.26	Essential tremor of the urethra
34.27	G25.27	Essential tremor of the penis
34.28	G25.28	Essential tremor of the testis
34.29	G25.29	Essential tremor of the epididymis
34.30	G25.30	Essential tremor of the vas deferens
34.31	G25.31	Essential tremor of the ureter
34.32	G25.32	Essential tremor of the bladder neck
34.33	G25.33	Essential tremor of the urethra
34.34	G25.34	Essential tremor of the penis
34.35	G25.35	Essential tremor of the testis
34.36	G25.36	Essential tremor of the epididymis
34.37	G25.37	Essential tremor of the vas deferens
34.38	G25.38	Essential tremor of the ureter
34.39	G25.39	Essential tremor of the bladder neck
34.40	G25.40	Essential tremor of the urethra

• **BOIX INJECTION, INJECTION AUGMENTATION**
Boix J, et al. J Voice. 2013;28(1):1-10. doi:10.1016/j.jvoice.2012.07.001. Epub 2012 Oct 11. PMID: 23011111



CHOREA

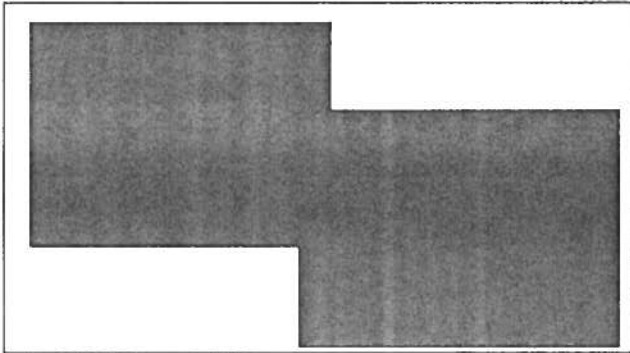
- INVOLUNTARY NONREPEITIVE, RANDOM MUSCLE CONTRACTIONS
- DISORDERS WITH CHOREA
 - HD, SPONGIFORM ENCEPHALOPATHIES, NEUROACANTHOSIS, WILSON'S DISEASE, CHOREA GRAVIDUM
- VOCAL DYSFUNCTION RESULTS FROM CHOREIC AND DYSTONIC MOTION IN HEAD AND NECK
- VOCAL RESULTS
 - DECREASED MPT
 - INCREASED NOISE-TO-HARMONIC RATIO
 - IRREGULAR PITCH FLUCTUATION
 - SUDDEN PHONATORY INTERRUPTION
 - MISPLACEMENT OF ARTICULATORS

Furl J, et al. J Voice. 2013;28(1):1-10. doi:10.1016/j.jvoice.2012.07.001. Epub 2012 Oct 11. PMID: 23011111

MYOCLONUS

- INVOLUNTARY MUSCLE CONTRACTION/RELAXATION RESULTING IN BRIEF IRREGULAR JERKS
- DRIVER MAY BE CORTICAL, SUBCORTICAL, OR SPINAL SEGMENTAL
- ISOLATED LARYNGEAL MYOCLONUS ARISES WITH INFERIOR OLIVARY NUCLEI, PONTINE TEGMENTAL TRACT OR CEREBELLAR DENTATE NUCLEUS DAMAGE
- SEEN IN ASSOCIATION WITH DYSTONIA, TARDIVE CONDITIONS, PARANEOPlastic SYNDROMES, NEUROBLASTOMA
- CAN PRODUCE BROKEN SPEECH, VOCAL STRAIN, LACK OF COORDINATION
- OFTEN SUPPRESSIBLE WITH SPEECH OR SWALLOW

Cheng VC, Frucht SJ. Myoclonus. Curr Treat Options Neurol. 10:223-9. 2008. PMID: 18411111



HYPOFUNCTIONAL DISORDERS

HYPOFUNCTIONAL DISORDERS

- CENTRAL
 - MEDULLARY
 - ARNOLD-CHIARI
- UMN AND LMN
 - ALS
- LMN
 - CN X INJURY
 - POLIO
- NMJ
 - MYASTHENIA GRAVIS
 - LAMBERT-EATON SYNDROME
- MYOPATHIES
 - OPMD
 - MYOTONIC DYSTROPHY
 - DERMATOMYOSITIS

ALS

- PRESENT WITH FLACCID-SPASTIC MIXED DYSPHAGIA 2/2 TONGUE FASCICULATIONS
- PROGRESSIVE WEAKNESS AFFECTS BREATHING, VOICING AND SWALLOWING



MIXED DISORDERS

- PARKINSON'S DISEASE
- MULTIPLE SCLEROSIS

PARKINSONISM (PARKINSON'S DISEASE)

- SIX CARDINAL SYMPTOMS
 - TREMOR AT REST, BRADYKINIA, RIGIDITY, FLEXED POSTURE, LOSS OF CARDINAL REFLEXES, FREEZING PHENOMENON
- 75% WITH VOICE AND ARTICULATION DEFICITS
 - POOR BREATH SUPPORT
 - POOR BREATH AND VOICING COORDINATION
 - POOR VOCAL FOLD CLOSURE
 - IMPAIRED SENSORY FEEDBACK
- TREATMENT
 - IA FRAMEWORK SURGERY
 - SPEECH THERAPY: LEE SILVERMAN VOICE TREATMENT

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MULTIPLE SCLEROSIS

- DIFFUSE, CENTRAL DEMYELINATION DISEASE
- DIAGNOSIS PROVEN BY CNS PLAQUES SEEN ON MRI AND OLIGOCLONAL BANDS IN CSF
- US INCIDENCE 3/100,000 ANNUALLY
- SPASTIC/SLURRED SPEECH, DYSPHAGIA

CONCLUSIONS

- OTOLARYNGOLOGISTS MAY HELP DIAGNOSE/TREAT NEUROLOGIC DISORDERS
- LARYNGEAL MANIFESTATIONS: HYPERFUNCTIONAL VS. HYPOFUNCTIONAL
- SYMPTOMS AND SIGNS CAN LOCALIZE THE LESION
- MANY OF THESE DISORDERS ARE NOT CURABLE
- SPASMODIC DYSPHONIA IS EFFECTIVELY TREATED WITH BOTOX
- DIFFERENTIATION OF NEUROGENIC AND NONORGANIC PATHOLOGY IS A CHALLENGE
- MAKE FRIENDS WITH A NEUROLOGIST

THANK YOU!

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