

Dystonia

A Comprehensive Approach to Care and Treatment

Presenter:

Mindy Kaye Bixby D.O.

Movement Disorder Specialist

Board Certified Neurologist

Talk Overview

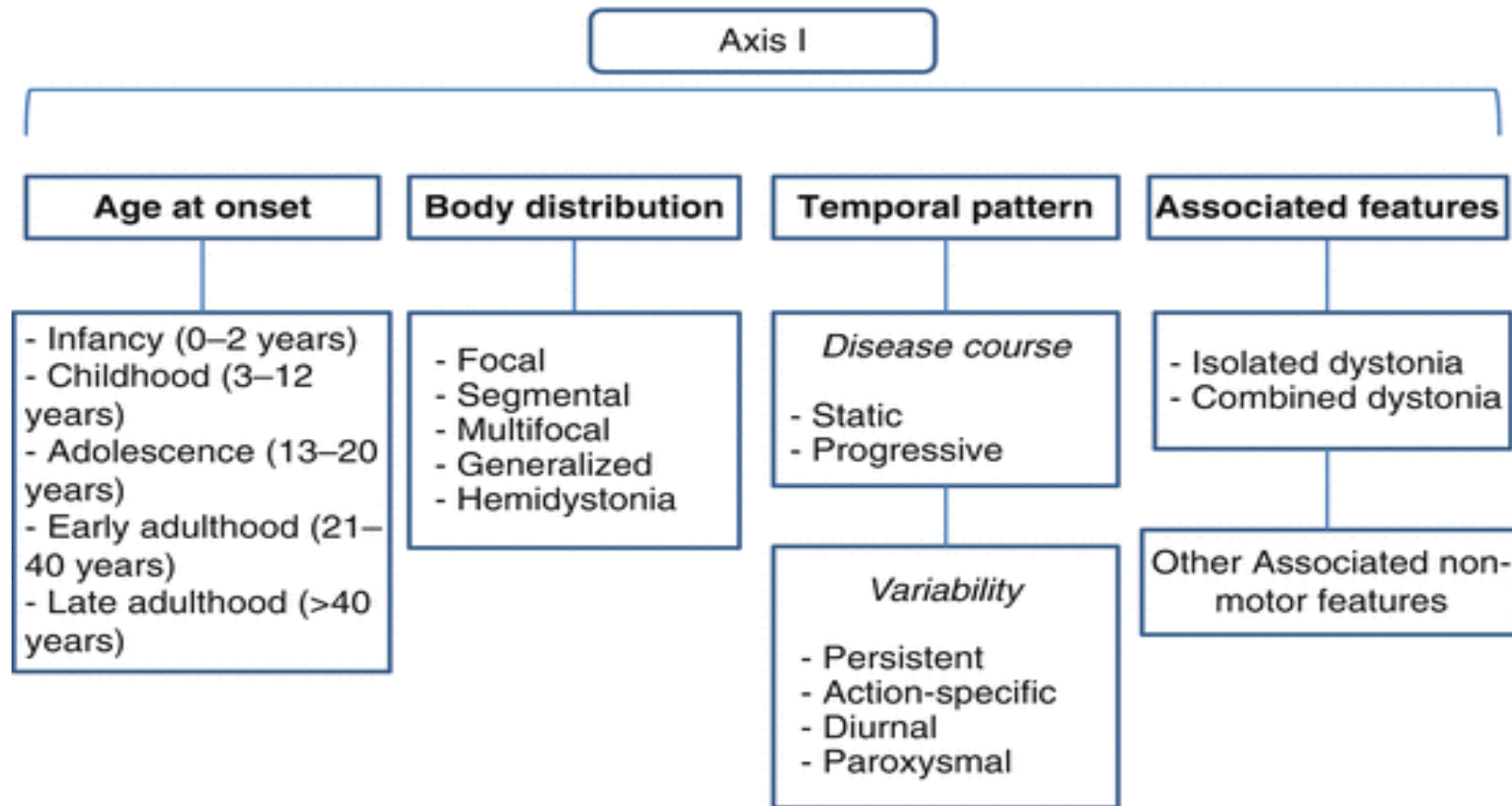
- Definition of Dystonia
- Idiopathic versus Other Etiology
- Head to Toe Discussion of Types of Dystonia
- Approach to Diagnosis
- Management of Dystonia
- Future Research and Treatment Discussed

Definition of Dystonia

- **Dystonia** is a movement disorder in which a person's muscles contract uncontrollably. The contraction causes the affected body part to twist involuntarily, resulting in repetitive movements or abnormal postures.
- **Dystonia** can affect one muscle, a muscle group, or the entire body



Classification of Dystonia



Focal Dystonia

- Focal: one body part
 - ❖ Blepharospasm
 - ❖ Oromandibular
 - ❖ Spasmodic Torticollis
 - ❖ Limb Dystonia



Segmental Dystonia

- Segmental: occurring in 2 contiguous body parts
 - Cranial + Brachial
 - Cranial + Cervical
 - Cranial + Axial



Multifocal Dystonia

- Two or more non-contiguous body parts involved

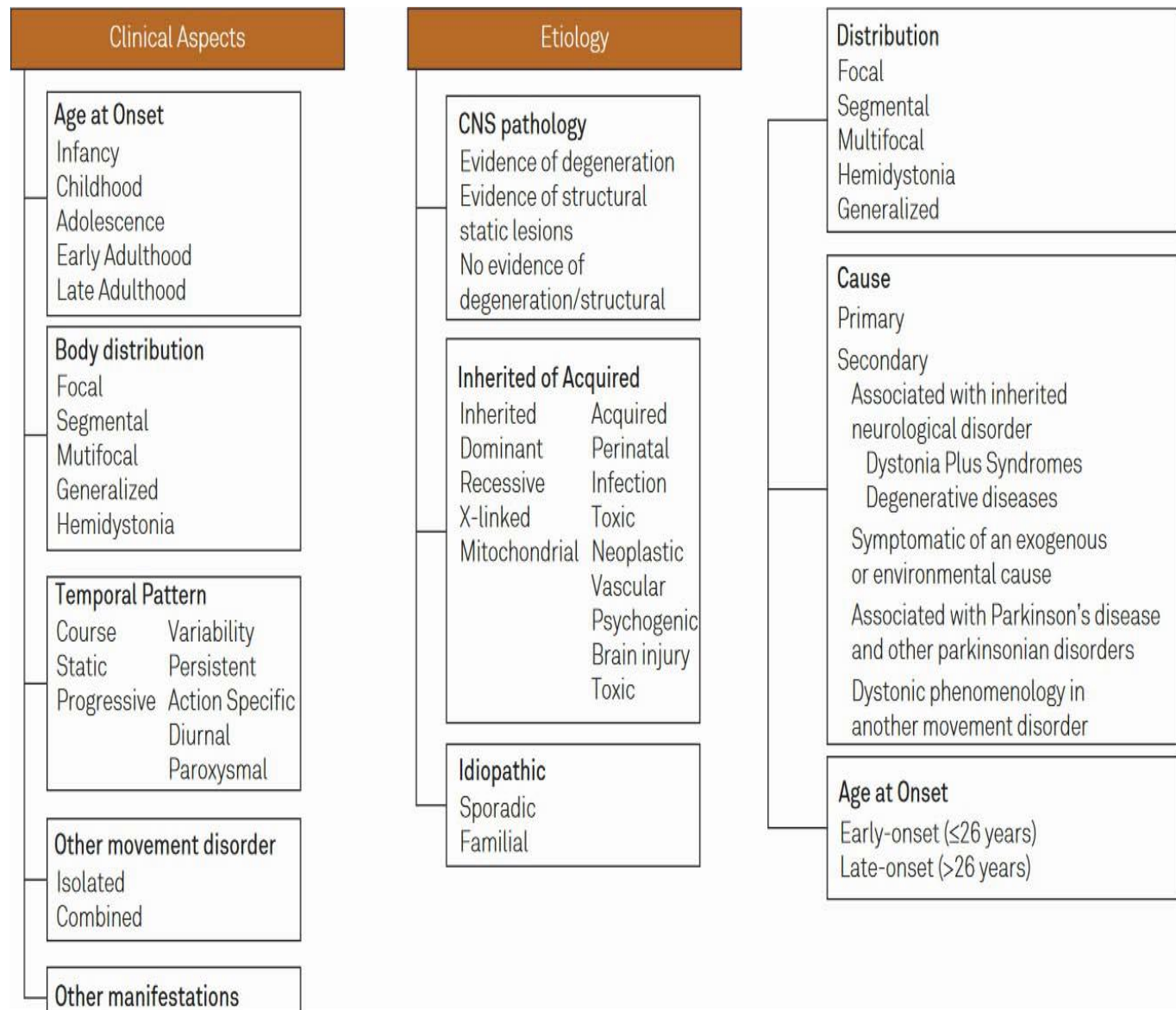


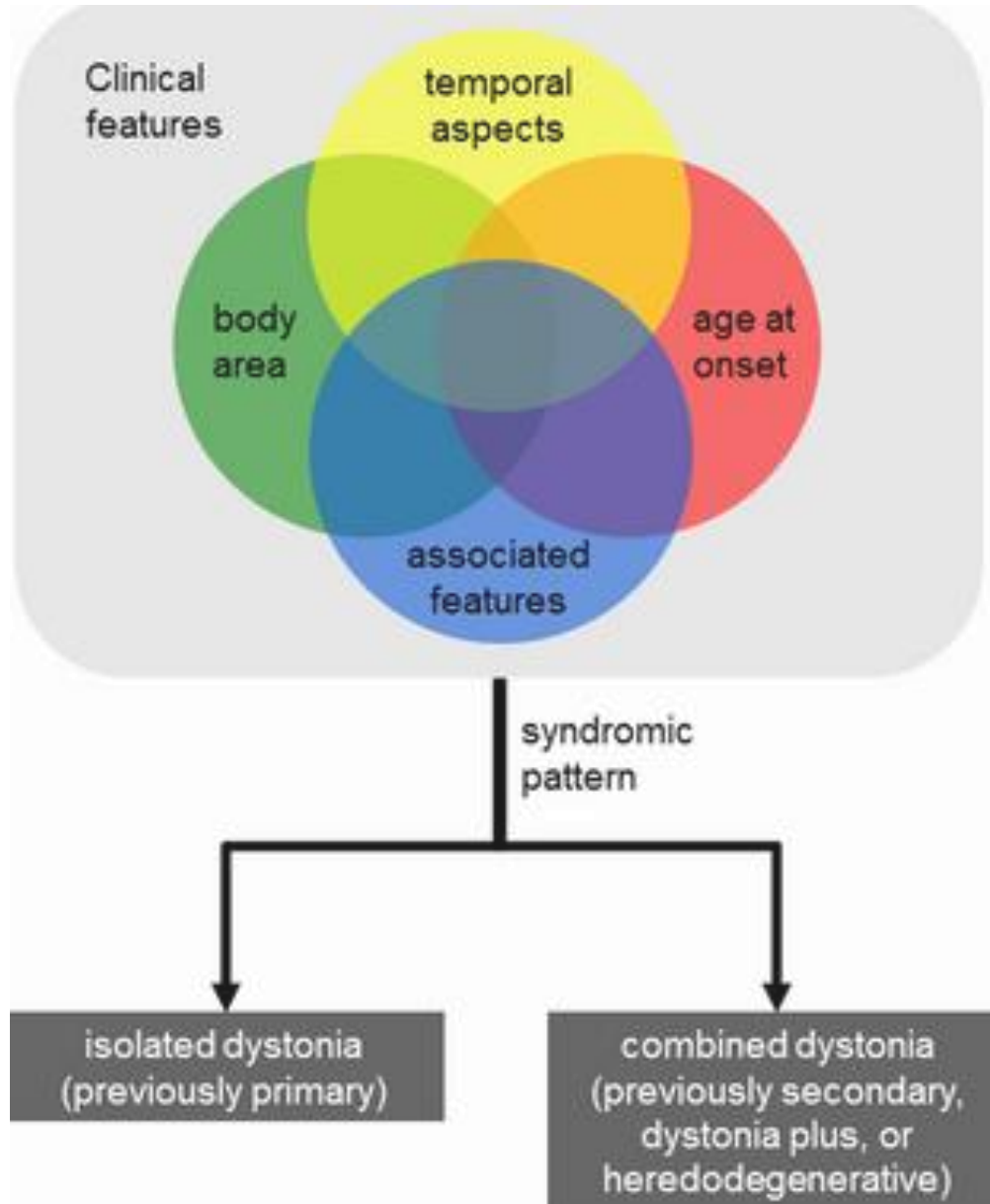
Generalized Dystonia

- Involves Several Areas on Both Sides of the Body
- A combination of Leg Involvement plus of Any Other Area of the Body.



Approach to Diagnosis





Approach to Diagnosis

Sensory Trick



Head to Toe Discussion of Types of Dystonia

- Blepharospasm
- Oromandibular Dystonia (Meige syndrome)
- Spasmodic Dysphonia
- Cervical Dystonia (Spasmodic Torticollis)
- Truncal Dystonia
- Limb Dystonia
- Generalized Dystonia



Blepharospasm

- Involuntary tonic, spasmodic, bilateral eyelid closure
- F>M
- More common in older individuals (60+)
- Causes: idiopathic, Parkinson's disease, post-encephalitic, Tetany, drugs (psychotropics)
- Triggers: bright lights, fatigue, emotional tension
- Treatment: Botulinum toxin injections into the orbicularis oculi, corrugators, procerus muscles



Oromandibular Dystonia (Meige syndrome)

- Focal dystonia characterized by contractions of the face, jaw, and/or tongue causing difficulty in opening/closing the mouth often affecting chewing and speech.
- Meige: Involvement frequent forced spasms of the eye, jaw, tongue and lower facial muscles



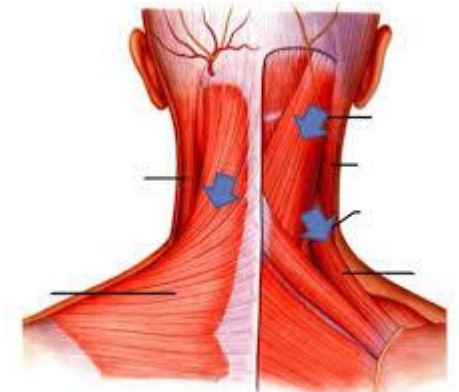
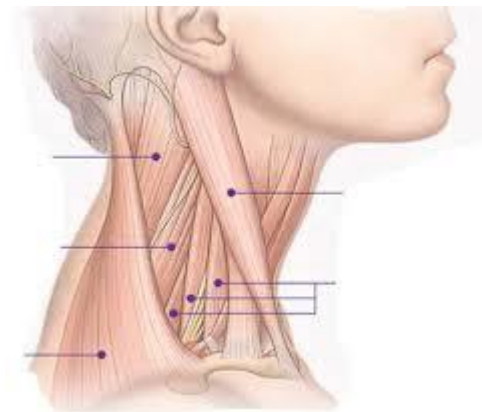
Spasmodic Dysphonia

- Laryngeal Dystonia
- Involuntary spasms of the voice box or larynx
- Dystonic muscles of the vocal cords
- Resulting in tight, strained or strangled sound.
- Causing breaks or interruptions in the voice



Cervical Dystonia (Spasmodic Torticollis)

- Female>Male (2:1)
- CD where neck muscles involuntarily contract into abnormal positions such as: repetitive twisting movements of your head and neck.
- intermittent, in spasms, or constant
- Causes: idiopathic, Parkinson's disease, injury, family history.
- Triggers: stress, fatigue
- Treatment: Botulinum toxin injections to head and neck, Deep Brain Stimulation (DBS) and certain medications.
- Physical therapy



Truncal Dystonia

- Abdominal ticks, truncal posturing
- Female = Male (1:1)
- Posturing of abdominal muscles and/or back spasms that involuntarily contract into abnormal positions such as: repetitive twisting movements of the trunk.
- intermittent, in spasms, or constant
- Causes: idiopathic, Parkinson's disease, injury, family history.
- Triggers: stress, walking, fatigue, eating (full stomach)
- Treatment: Botulinum toxin injections to trunk, Deep Brain Stimulation (DBS), medications discussed later



Limb Dystonia

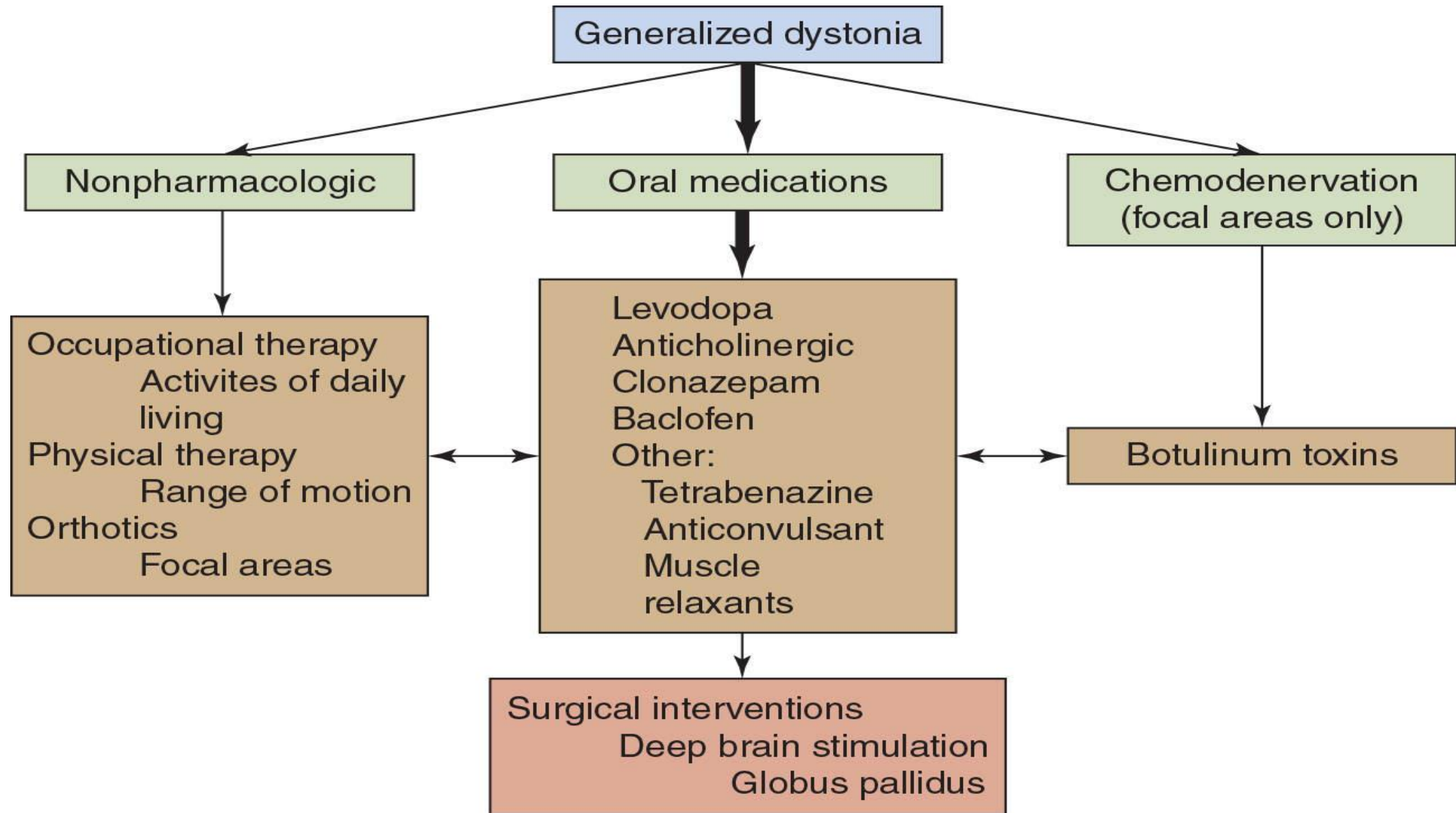
- Female < Male (1:2)
- LD where limbs involuntarily contract into abnormal positions sometimes due to a certain manuevr completed by patient that causes repetitive, posturing and intermittent twisting/flexing movements of the limb.
- intermittent, in spasms, or constant
- Causes: idiopathic, Parkinson's disease, injury, family history. Possible overuse such as musician
- Triggers: stress, fatigue, repetitive movements
- Treatment: Botulinum toxin injections to limbs, Deep Brain Stimulation (DBS), medications



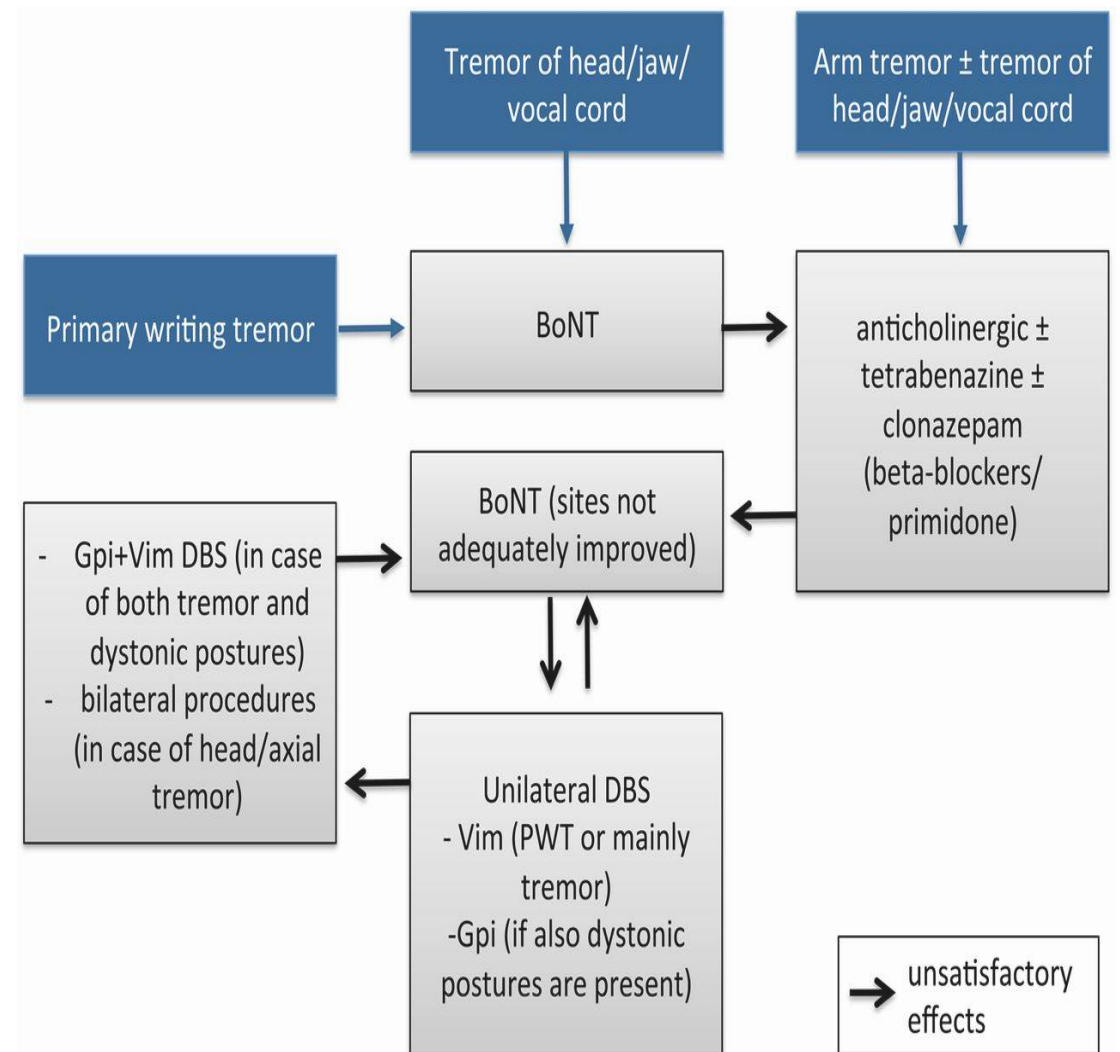
Generalized Dystonia

- Female = Male (1:1)
- Generalized Dystonia occurs throughout the body and is characterized as facial, cervical, truncal and/or limbs involuntarily contract into abnormal positions sometimes due to a certain maneuvers completed by patient that causes repetitive, posturing and intermittent twisting/flexing movements of the limb.
- intermittent, in spasms, or constant
- Causes: idiopathic, neurodegenerative, injury, genetic change to include: stiff person syndrome
- Triggers: stress, fatigue, repetitive movements
- Treatment: Botulinum toxin injections to limbs (however at times not enough benefit throughout the body with a limit of toxin dose due to safety concerns; usually seek a combination of botulinum toxin, Deep Brain Stimulation (DBS) and medications





Management of Dystonia



Medications for Dystonia

- **Anticholinergics:**

Artane, Cogentin

- **Dopaminergics:**

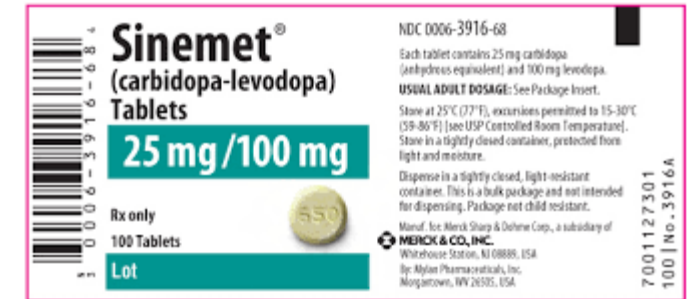
Sinemet, Parlodel, Amantadine

- **GABAergics:**

Valium

- **Anticonvulsant:**

Topamax, Keppra



Botulinum Toxin



XEOMIN



MYOBLOC



BOTOX



DYSPORT

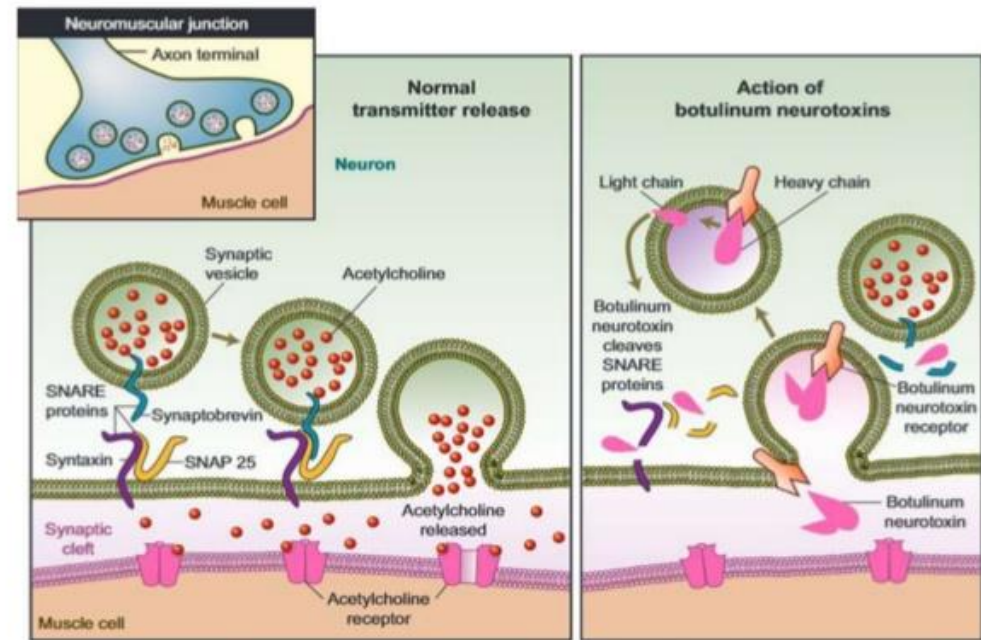


Botulinum toxin

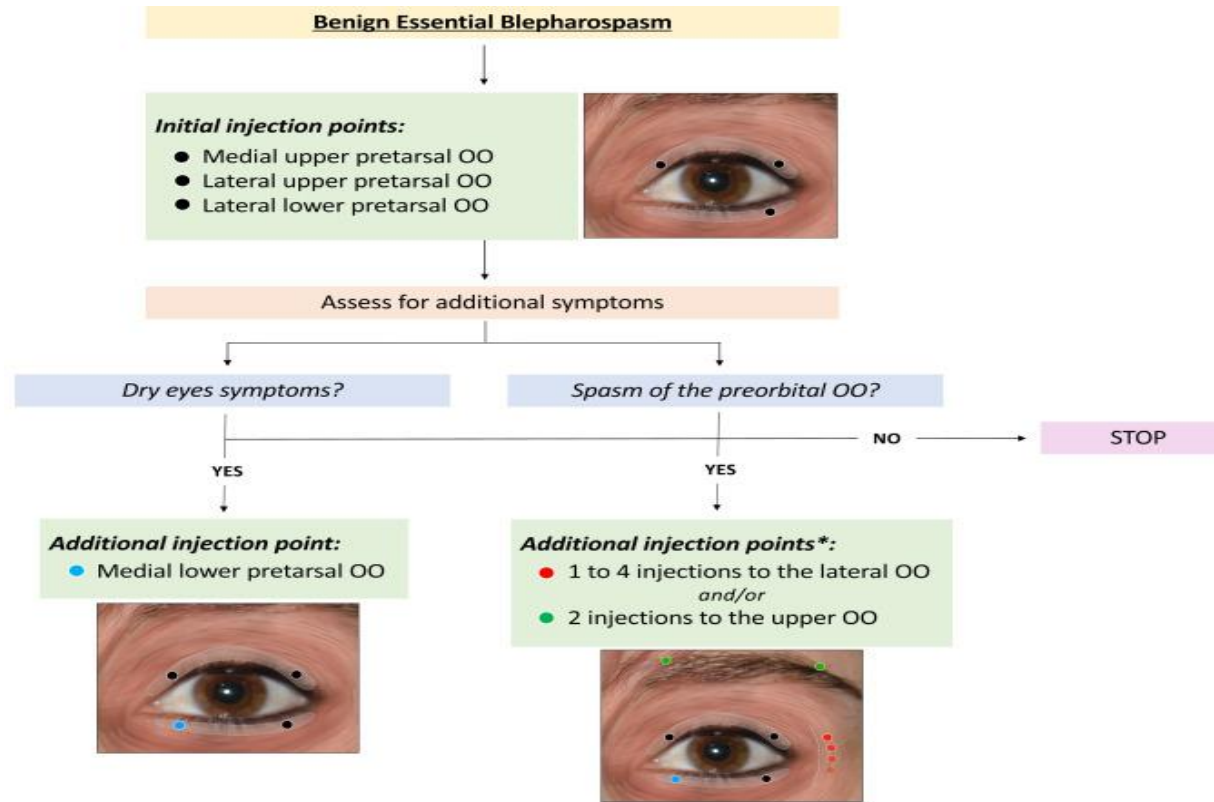
Mechanism of Action:

A neurotoxin that paralyzes muscles by inhibiting release of acetylcholine from presynaptic vesicles at the neuromuscular junction

ACTION OF BOTULINUM TOXIN

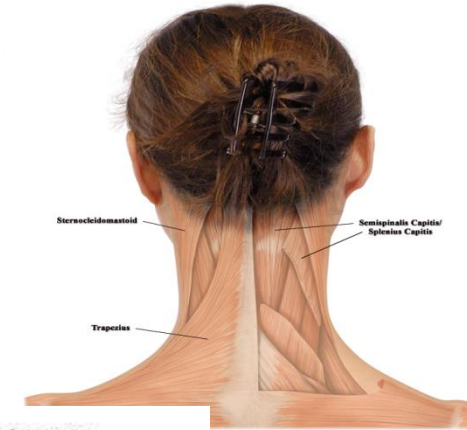
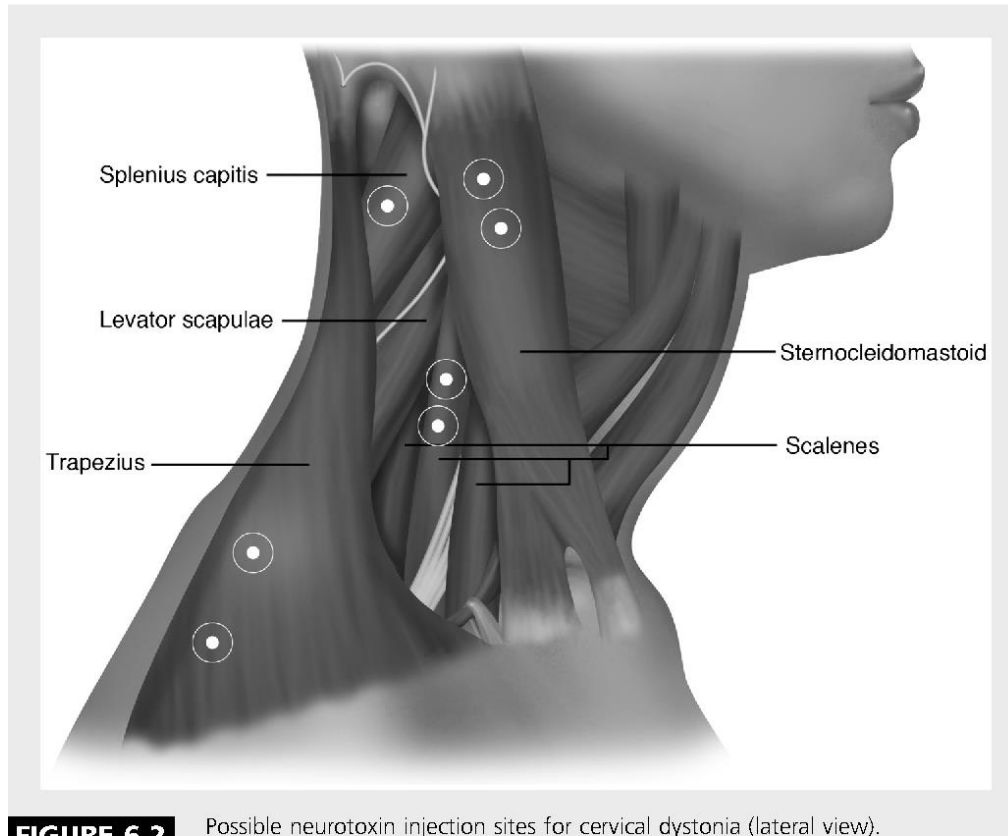


Treatment of Blepharospasm



*These preorbital injections can be omitted at the initial treatment and done as retouching if the periorbital muscle is still spastic at 2 weeks of follow up

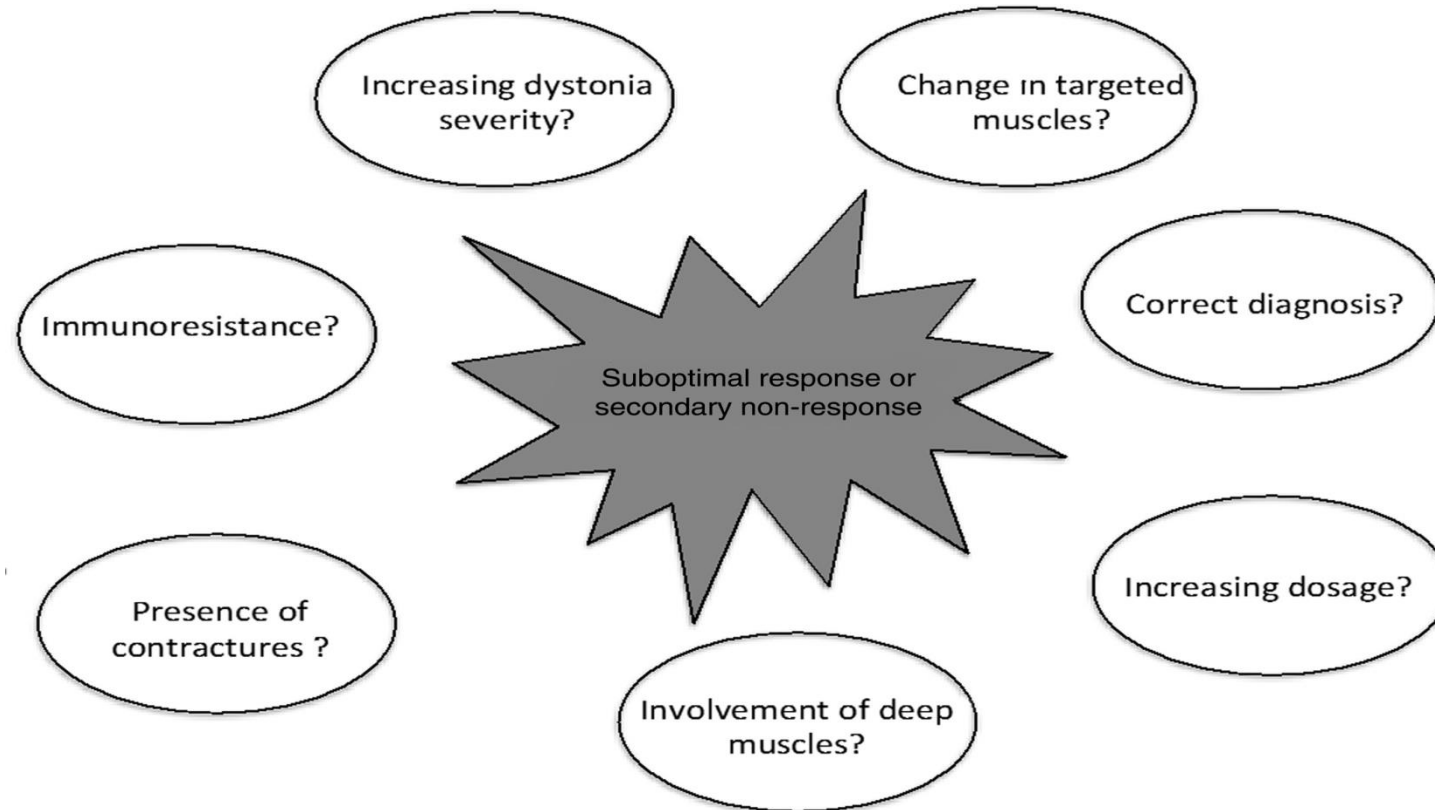
Treatment of Cervical Dystonia



Types of neck dystonia

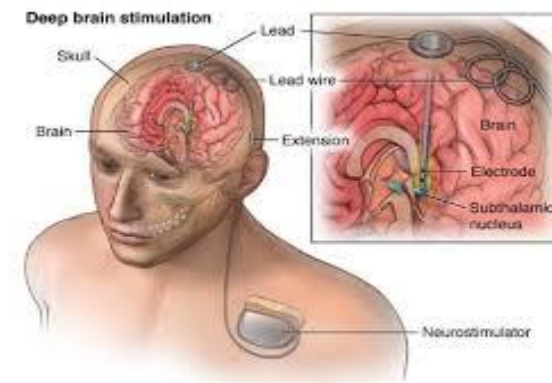
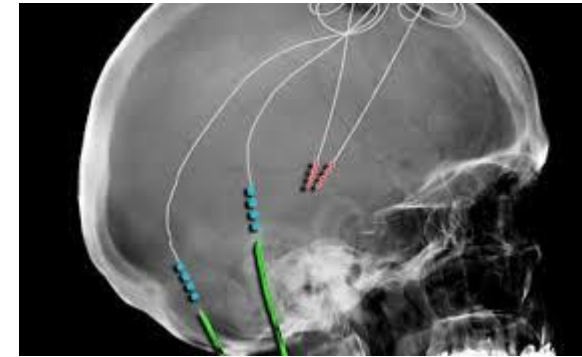


What happens if botulinum toxin is not helping your Dystonia?



Deep Brain Stimulation for Dystonia

- A brain surgery that places a lead in a location where dystonia is most pronounced
- With programming of the lead some have significant improvement of dystonia.



Deep Brain Stimulation (DBS)

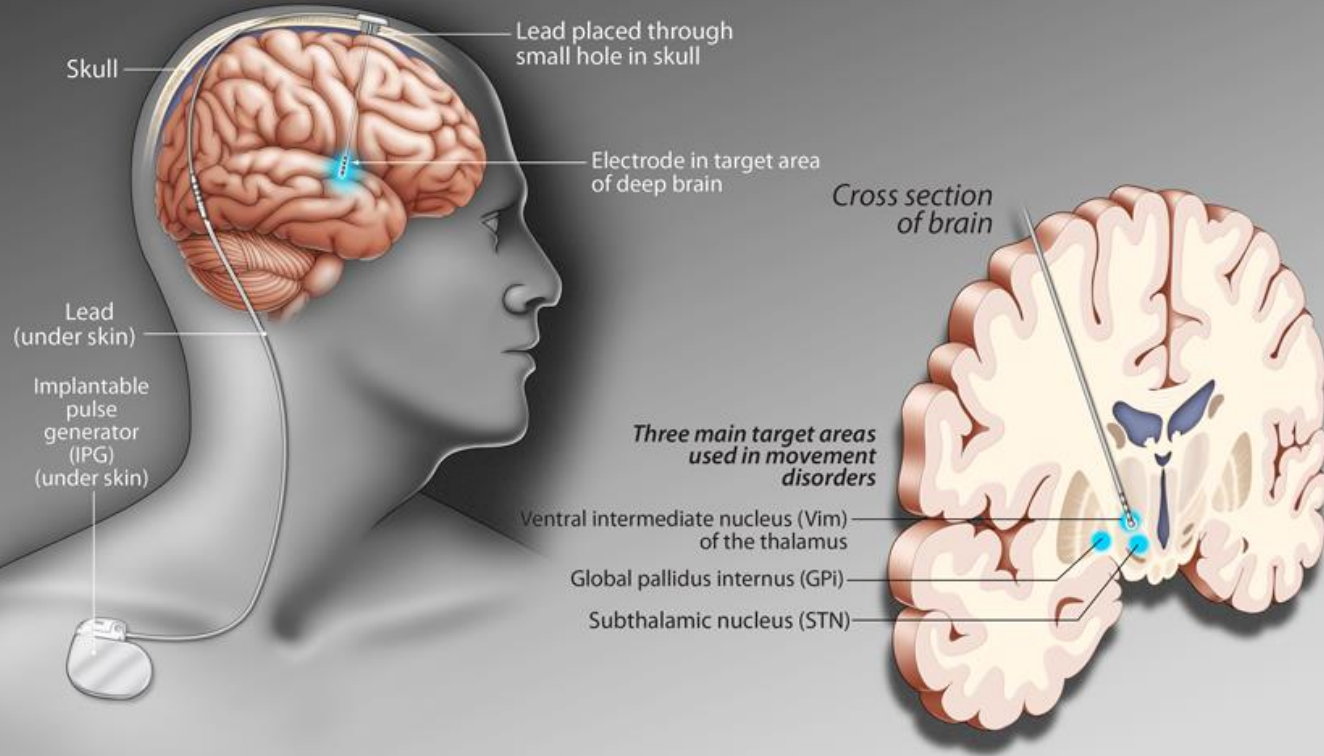


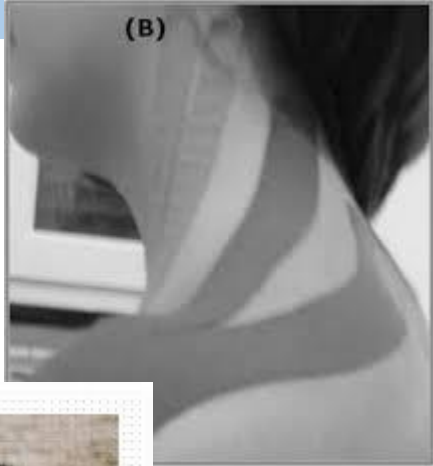
Figure 1. DBS system and the most common targets used for movement disorders. Note that Vim is not an anatomical target but rather a cluster of neurons within the motor thalamus receiving projections from the cerebellum.

Deep Brain Stimulation for Dystonia

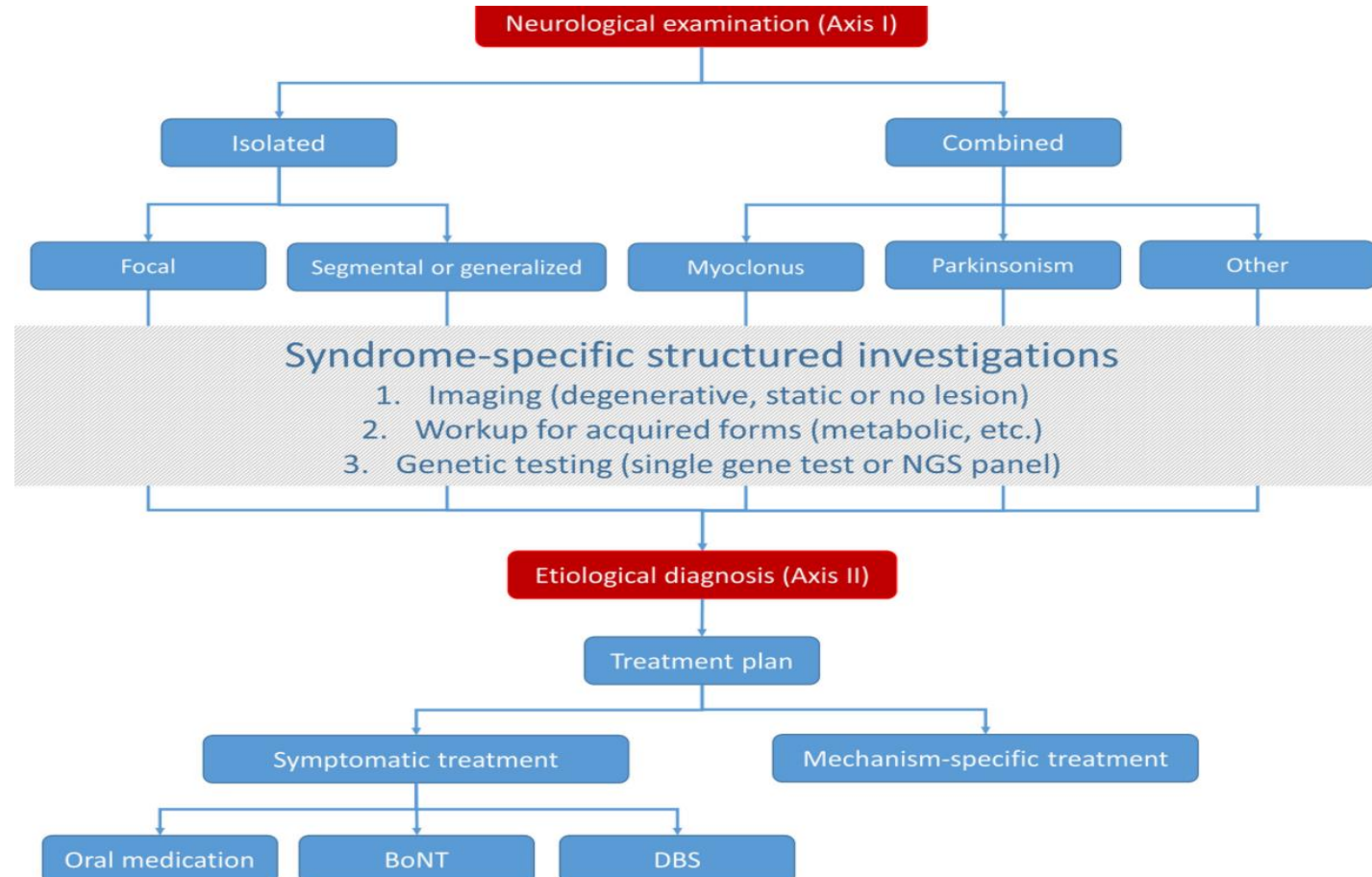
- DBS a pacemaker for Dystonia to modulate the impulses in the brain causing dystonia
- Goal: to improve patient's life through modulating the circuitry causes dystonic posturing.



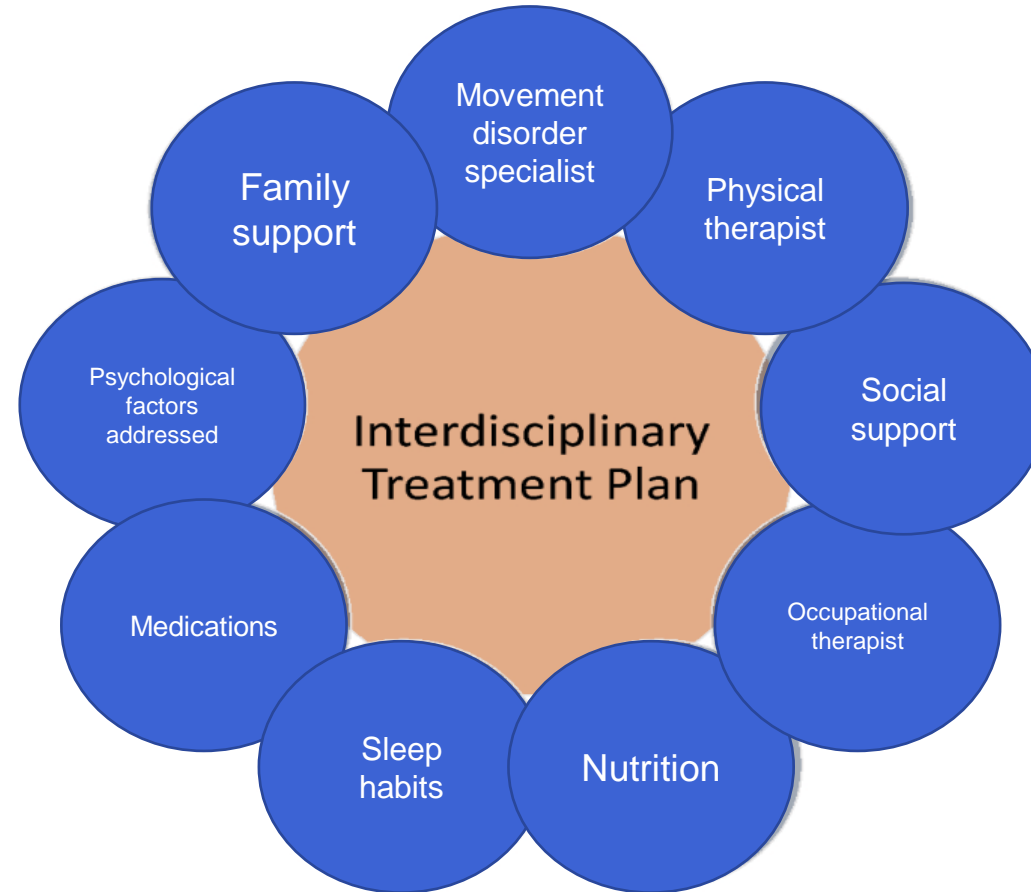
Physical Therapy for Dystonia



TREATMENT PARADIGM

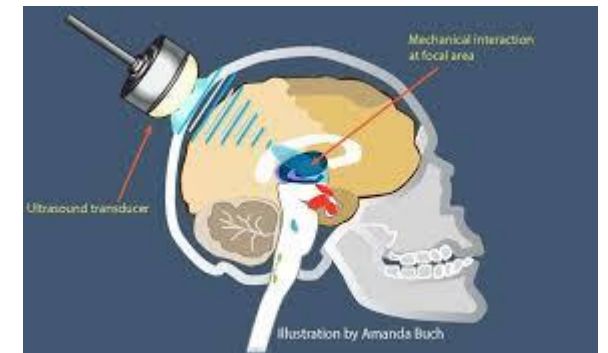
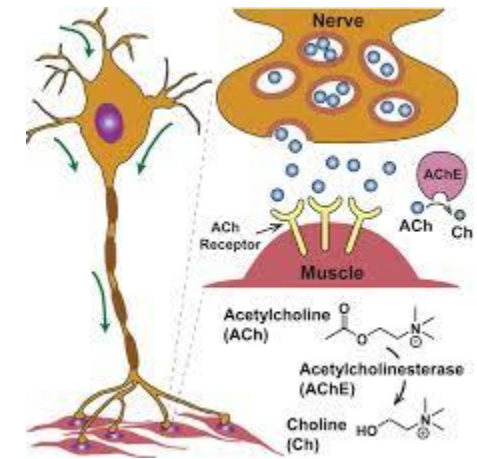
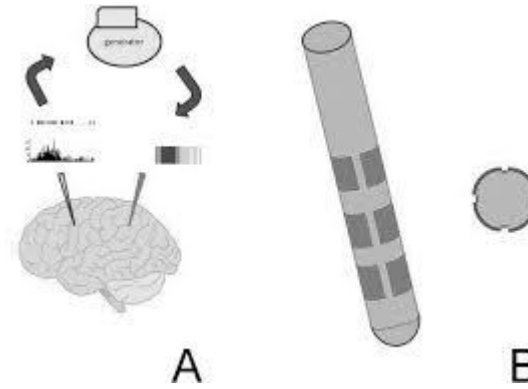


Multifactorial Management of Dystonia



Future Research Targets and Treatments

- **Acetylcholine** is a neurotransmitter of interest because some dystonia patients improve when taking medications that alter levels of acetylcholine.
- **New Approach to DBS:** Sensing capabilities to give stimulus when you need it
- **Focused Ultrasound Therapy:** noninvasive, therapeutic technology with the potential to improve the quality of life and decrease the cost of care for patients with dystonia. This novel technology focuses beams of ultrasound energy precisely and accurately on targets deep in the brain without damaging surrounding normal tissue.
- **Genetic modifiers:** Using genetic biomarkers to prevent or improve gene derived Dystonia (DY) through modifications with gene splicing.





**KEEP
CALM**

AND

SUPPORT

DYSTONIA AWARENESS