

Types of neck dystonia



Torticollis



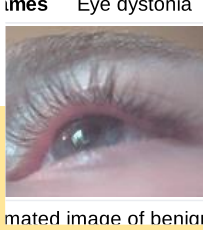
Laterocollis



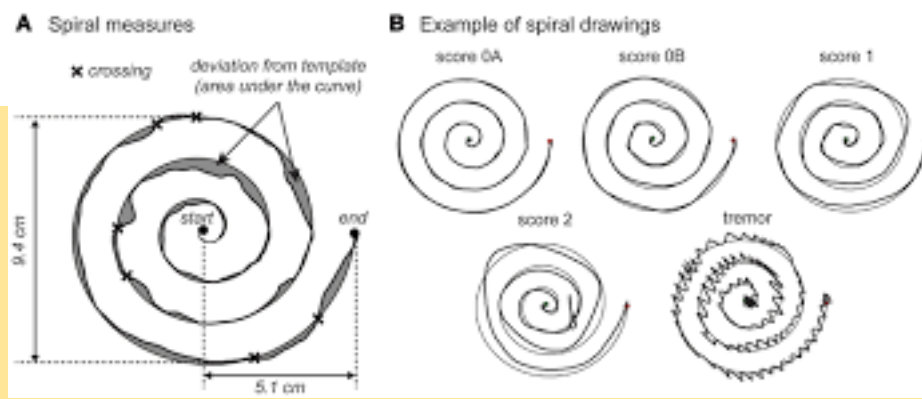
Retrocollis



Antecollis

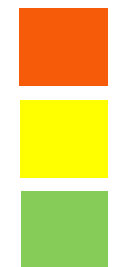


imitated image of benign



Movement Disorders I

Ya Min Kyaw
 Associate Professor
 Senior Consultant Neurologist





Movement Disorders

- **Movement disorders** are clinical syndromes with either an excess of movement or a paucity of voluntary and involuntary movements, **unrelated to weakness or spasticity**



Common Movement Disorders

- two major categories- *hyperkinetic* and *hypokinetic*
- **Hyperkinetic** movement disorders refer to dyskinesia, or excessive, often repetitive, involuntary movements that intrude upon the normal flow of motor activity.



Movement Disorders

- **Hypokinetic** movement disorders refer to
 - akinesia (lack of movement)
 - hypokinesia (reduced amplitude of movements)
 - bradykinesia (slow movement)
 - rigidity

I. Hypokinesias

- | | |
|--|---------------------------|
| A. Akinesia/bradykinesia
(parkinsonism) | F. Freezing
phenomenon |
| B. Apraxia | G. Hesitant gaits |
| C. Blocking (holding) tics | H. Hypothyroid slowness |
| D. Cataplexy and drop attacks | I. Rigidity |
| E. Catatonia, psychomotor
depression, and
obsessional slowness | J. Stiff muscles |

II. Hyperkinesias

- | | |
|-------------------------------|--------------------------------|
| A. Abdominal dyskinesias | M. Moving toes and fingers |
| B. Akathitic movements | N. Myoclonus |
| C. Ataxia/asynergia/dysmetria | O. Myokymia and synkinesis |
| D. Athetosis | P. Myorhythmia |
| E. Ballism | Q. Paroxysmal dyskinesias |
| F. Chorea | R. Periodic movements in sleep |
| G. Dystonia | S. REM sleep behavior disorder |
| H. Hemifacial spasm | T. Restless legs |
| I. Hyperekplexia | U. Stereotypy |
| J. Hypnogenic dyskinesias | V. Tics |
| K. Jumping disorders | W. Tremor |
| L. Jumpy stumps | |

This table and the definitions for each item were developed by S. Fahn, C. D. Marsden, and J. Jankovic and published in Table 1.1 in reference 7.



Common Movement Disorders

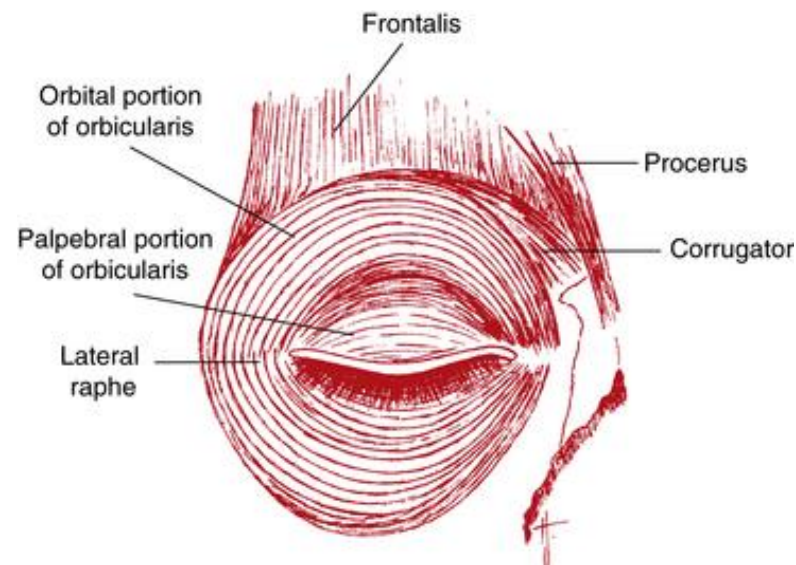
- Blepharospasm
- Hemifacial spasm
- Cervical dystonia
- Benign Essential Tremor



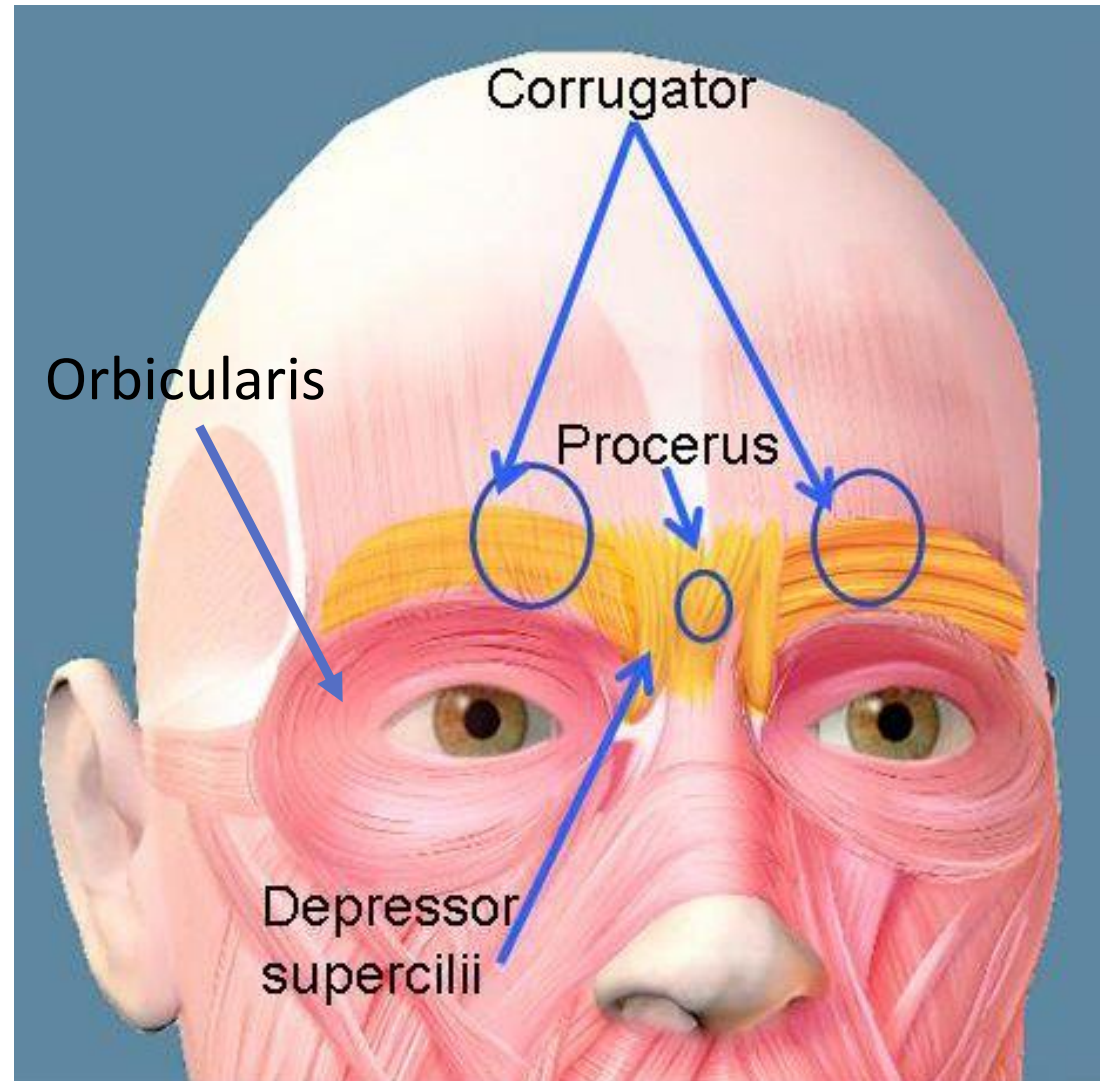
Blepharospasm

Blepharospasm

- Blepharospasm is abnormal contraction of the eyelid muscles.
- often refers to benign essential blepharospasm (BEB)
- a bilateral condition
- a form of focal dystonia leading to episodic closure of the eyelids protractor muscles (orbicularis oculi, procerus, and corrugator superciliaris)



Blepharospasm





Blepharospasm- **Clinical Presentations**

- Age: onset most commonly occurs during years 40-60
- Gender: Female > Male (2-4:1)
- exact cause – unknown
- **Risk Factors**
- Head or facial trauma.
- Family history of dystonia or tremor.
- Reflex blepharospasm
- Stress
- Medications- used to treat Parkinson's disease



Blepharospasm

- **Symptoms and Signs**

- limited to the bilateral eyelids
- begin as mild and infrequent twitches
- progress over time to forceful and frequent spasms of the eyelids, which abate during periods of sleep
- In advanced cases- can cause functional blindness from periodic inability to open the eyes



Blepharospasm

- **Meige's syndrome**- associated with facial grimacing
- **Reflex blepharospasm (Secondary)** - accompanied by photophobia and ocular inflammation
- **Reflex blepharospasm** which is triggered by
 - severely dry eyes and blepharitis
 - intraocular inflammation
 - meningeal irritation
 - light sensitivity



Blepharospasm





Blepharospasm

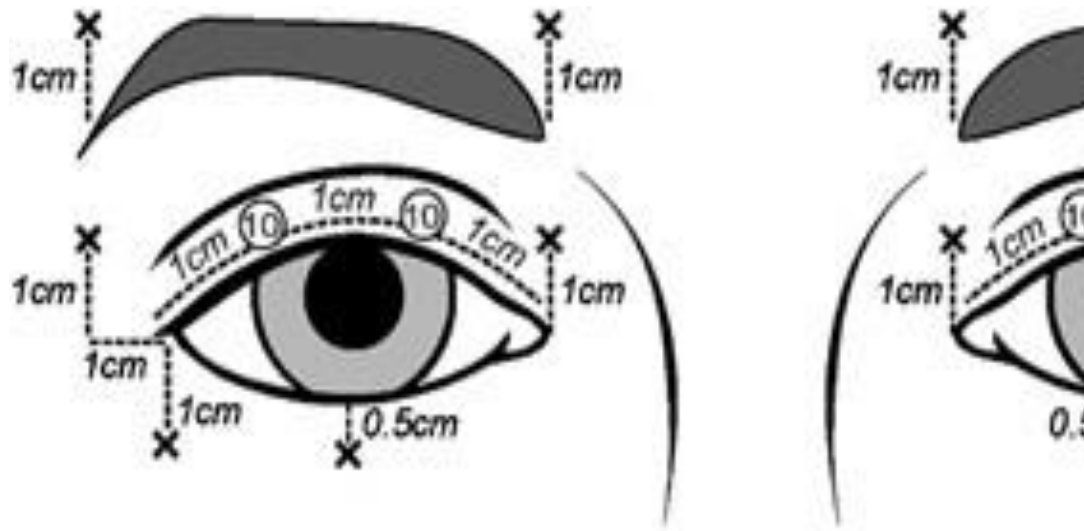
- **Diagnosis**
 - is a clinical diagnosis
 - is made by careful history taking and physical exam
- **Laboratory test and Diagnostic procedures**
 - generally unrevealing
 - rarely indicated in the workup



Blepharospasm

- **Management**
- **First Line**
- Periodic injection of a **botulinum toxin A** -the treatment of choice
- Botulinum toxin A injection is directed into or adjacent to the orbicularis oculi muscle, every 3-4 months.
- Dosage -2.5 to 5 units per injection site and 4 to 8 sites per eye.
- Average onset of action is in 2-3 days
- average peak effect occurs at about 7-10 days,
- duration of 3-4 months

Blepharospasm



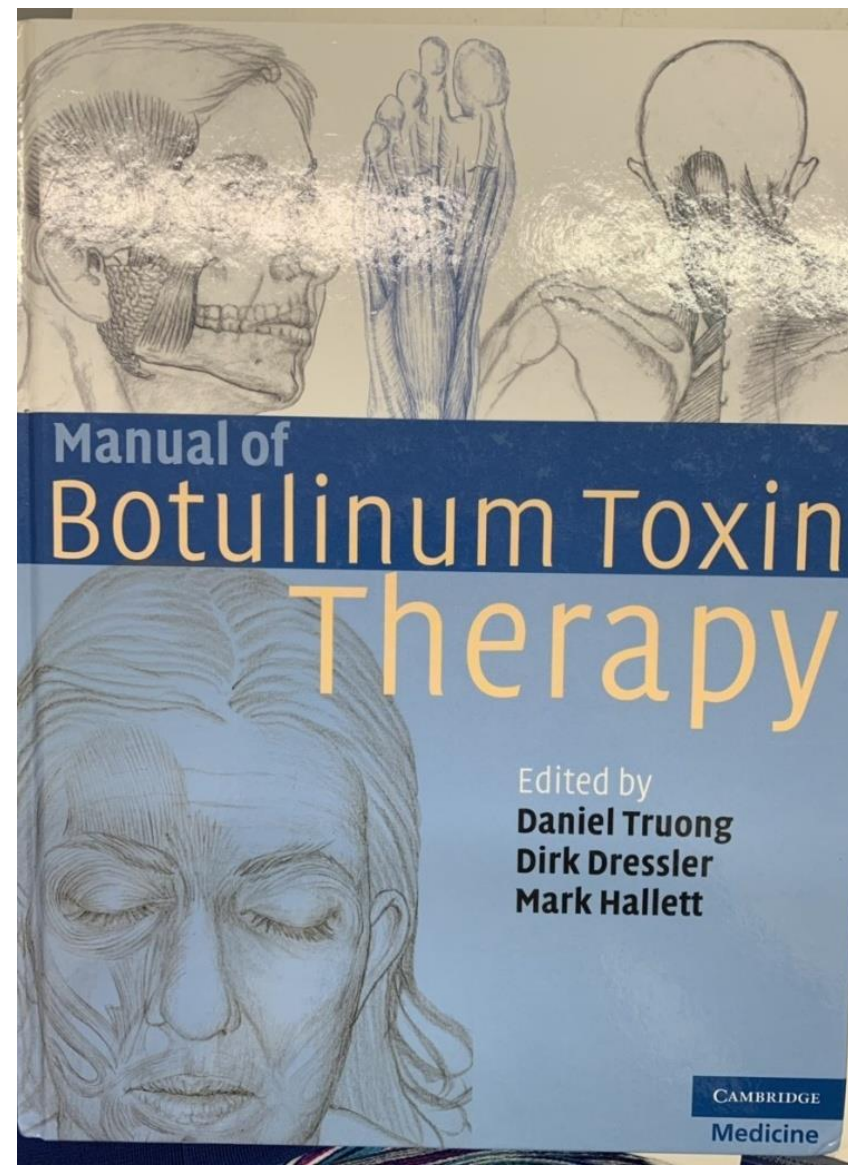
Botox Injection in
Blepharospasm





NHS

Calderdale and Huddersfield
NHS Foundation Trust







Blepharospasm

- **Management**
- **Second line**
- Oral medications such as **muscle relaxants and sedatives** - rarely effective in the treatment of BEB
- In secondary causes - treat the underlying etiology.

Blepharospasm

- **Management**

- Artificial tears should be used to reduce ocular irritation and dryness

- Use of FL-41 tinted lenses is helpful in those patients with photophobia

- Reduce stress



ful in



Hemifacial Spasm





Hemifacial Spasm

- characterized by irregular, involuntary muscle contractions (spasms) on one side (hemi-) of the face (-facial)
- facial muscles are controlled by the facial nerve, which originates at the brainstem and exits the skull below the ear where it separates into five main branches.

Hemifacial Spasm

- both men and women
- middle-aged or elderly women more frequently
- much more common in some Asian populations
- Bilateral- rare
- may be caused by
 - a facial nerve injury
 - a tumour <1%
 - no apparent cause
- compression of the facial nerve by vessels of the posterior circulation (Eg. vertebral artery)

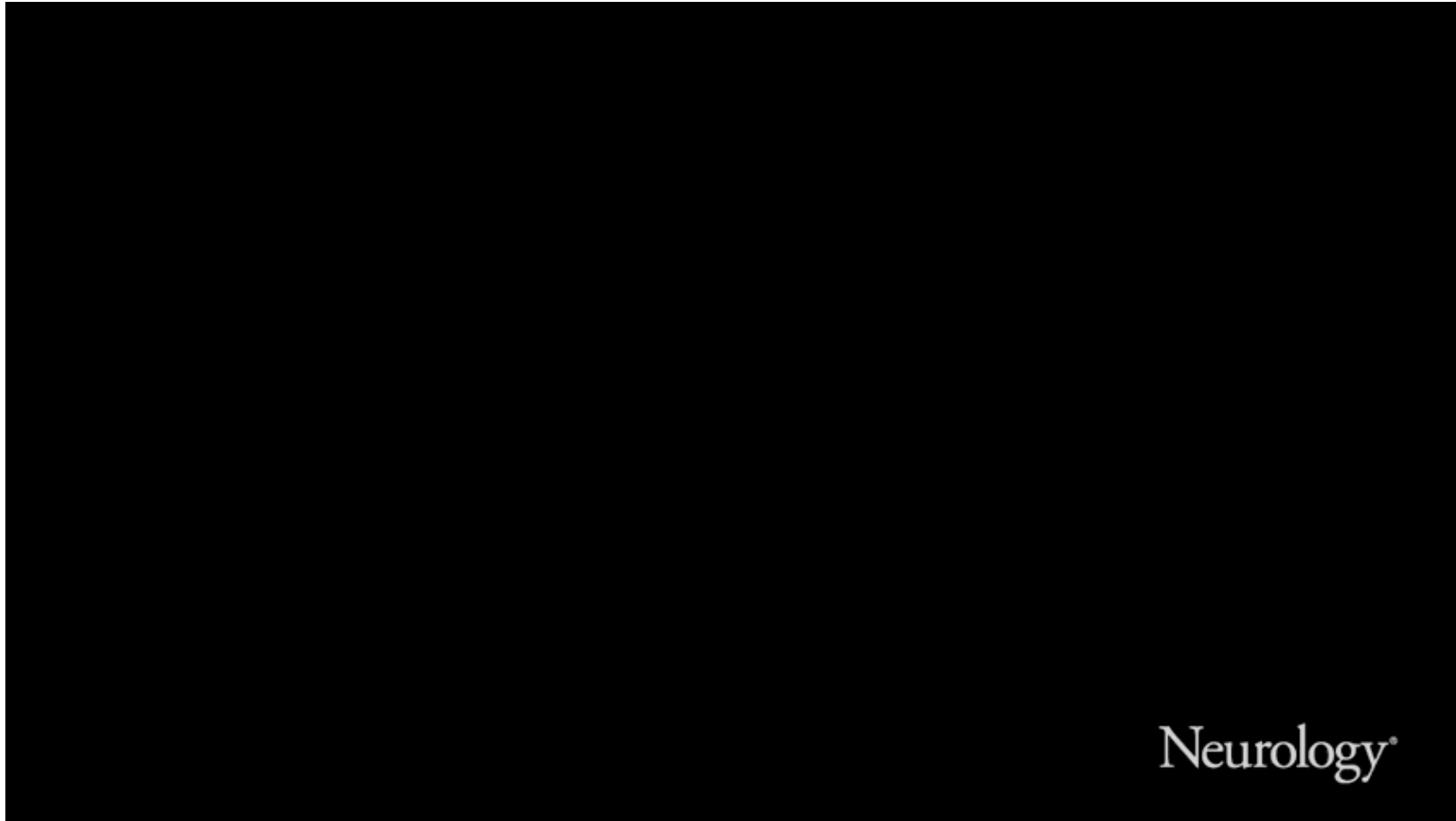


Hemifacial Spasm

- two forms: typical and atypical
- **Typical form**; twitching usually starts in the lower eyelid in orbicularis oculi muscle, it spreads to the whole lid, then to the orbicularis oris muscle around the lips, and buccinator muscle in the cheekbone area
- **Atypical hemifacial spasm**; twitching starts muscle around the lips, and in the cheekbone area in the lower face, then progresses up to the orbicularis oculi muscle in the eyelid as time progresses



Hemifacial Spasm



Neurology®



Hemifacial Spasm- **Diagnosis**

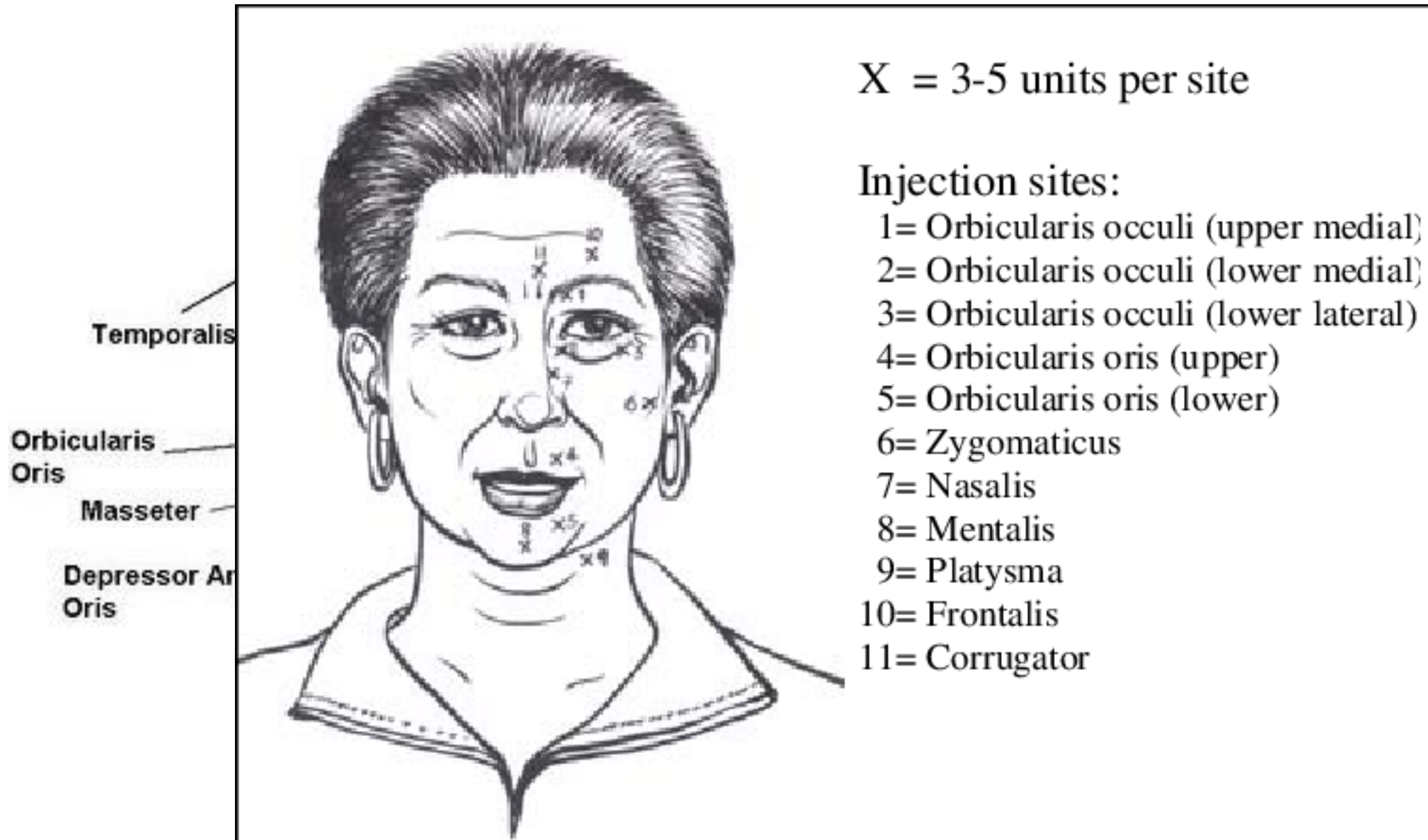
- Mainly clinical
- **Investigations**
 - To detect underlying cause- Eg tumour, vessel
 - MRI
 - CT scan
 - Angiography
 - EMG



Hemifacial Spasm- **Treatment**

- **First line- Botulinum toxin type A**
- Most effective treatment
- Helpful in more than 90% of patients
- Injections – around the eye and cheek, weakens the overactive muscles in the face
- can reduce spasms for three to four months

Hemifacial Spasm- Treatment



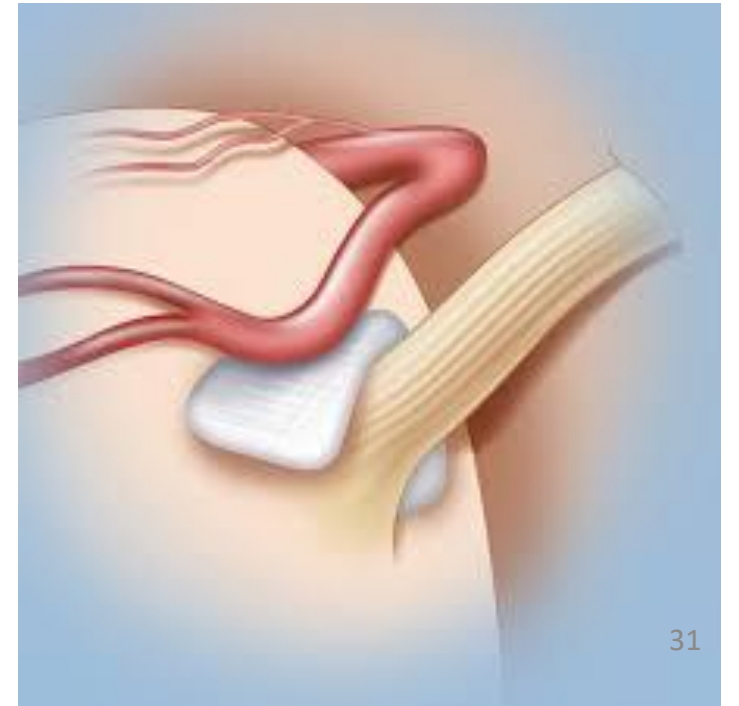


Hemifacial Spasm- **Treatment**

- **Second line- oral muscle relaxants**
 - baclofen
 - Clonazepam
 - carbamazepine
- Less effective

Hemifacial Spasm- **Treatment**

- **Microvascular Decompression**
- most popular surgical treatment
- microvascular decompression relieves pressure on the facial nerve
- a small opening in the skull behind ear
- puts a piece of Teflon padding between the vessels pushing on it.



Cervical Dystonia



Rotated
(Torticollis)



To the side
(Laterocollis)



Backward
(Retrocollis)



Forward
(Anterocollis)



Cervical Dystonia

- is a condition in which neck muscles involuntarily contract into abnormal positions
- It causes repetitive twisting movements of head and neck
- can be intermittent, in spasms, or constant
- painful and disabling in some cases



Cervical Dystonia

- Women- twice as often as men
- people between ages 40 and 60
- In most cases-unknown. **Possible causes** include:
 - medication that blocks dopamine, such as some antipsychotics
 - injury to the head, neck, or shoulders
 - a genetic mutation- 10-25% have a family history of the disease
 - a psychological problem
 - neurological disorders, such as Parkinson's, Wilson's

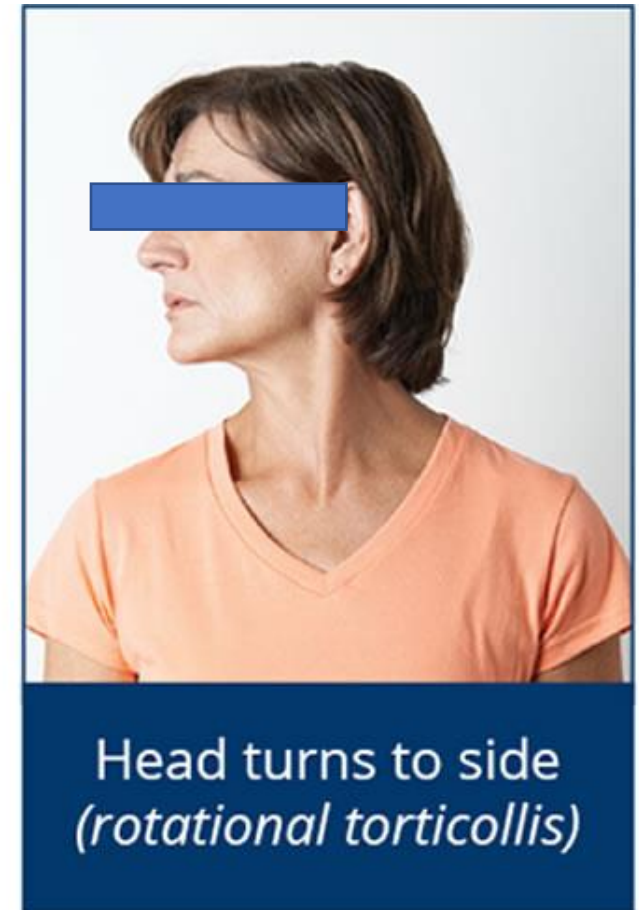


Cervical Dystonia

- most common abnormal movement – **tortillis**
- **anterocollis**
- **retrocollis**
- **laterocollis**
- Some may have a combination of these movements.

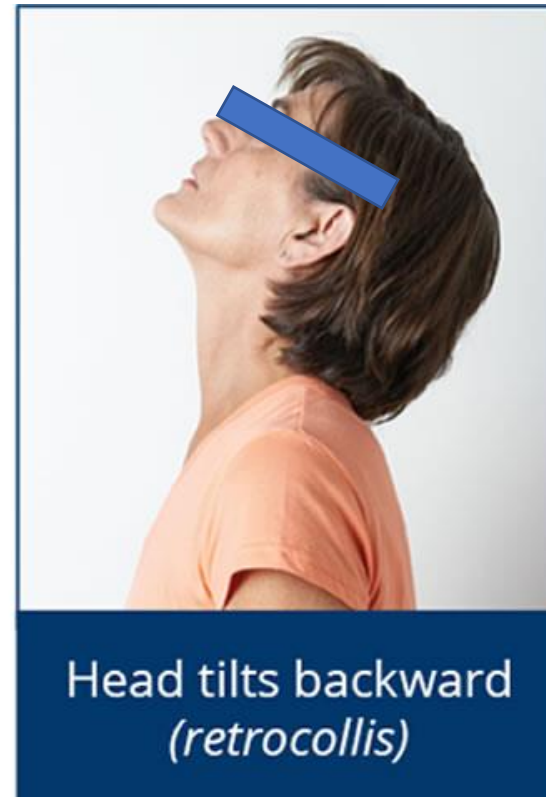
Cervical Dystonia

- most common abnormal movement - a twisting of the head and chin sideways, toward the shoulder, called **tortillis**.
- When sustained, the abnormal posture is referred to as tonic
- Spasmodic torticollis- jerky head movement or more rhythmic dystonic head tremor



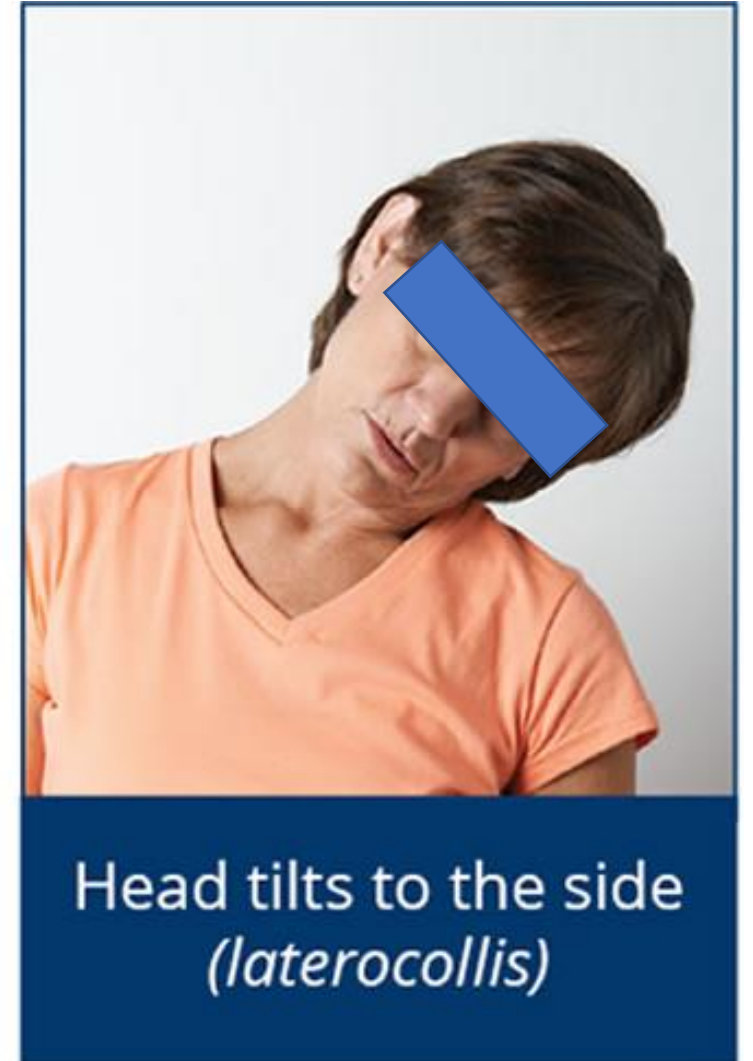
Cervical Dystonia

- tipping forward, chin downward, known as **anterocollis**
- tilting backward, chin upward, called **retrocollis**



Cervical Dystonia

- tilting sideways, ear to shoulder, known as **laterocollis**
- Associated with elevation of shoulder



Common postures involved in cervical dystonia

According to 1 study of 300 patients¹:

82%
of patients



Torticollis
(rotated)

42%
of patients



Laterocollis
(to the side)

25%
of patients



Anterocollis
(forward)

29%
of patients



Retrocollis
(backward)

66% of cervical dystonia patients present with a combination of postures

Cervical Dystonia- **Clinical Presentations**

- **Symptoms**
- begin gradually → may get worse and then reach a plateau
- severity – varies
- neck pain that radiates to the shoulders -75%
- a raised shoulder, headache
- Tremors – hand, head tremor
- enlargement of the neck muscle, - 75 %
- Stress, excitement, some physical positions - aggravate symptoms

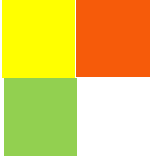
Cervical Dystonia- Clinical Presentations (Video)



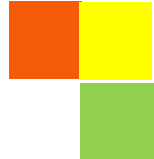
Cervical Dystonia- **Clinical Presentations**

- **Sensory Trick**

- The “**sensory trick**” or “**geste antagoniste**” is a characteristic and unique feature and may serve as a diagnostic clue to the diagnosis of focal **dystonia**.
- simple sensory tricks - can help stop a spasm
 - lightly touching the side of face, chin, cheek, or the back of head.



Cervical Dystonia- **Clinical Presentations**





Cervical Dystonia- **Clinical Presentations**

- Possible **complications** include:
 - the spread of involuntary motions to other parts of your body
 - bone spurs in the spine
 - cervical spine arthritis
 - also have a higher risk of depression and anxiety



Cervical Dystonia-Diagnosis

- Mainly clinical, exclude differential and secondary causes
- Investigations for differentials
- MRI, CT, ceruloplasmin level and genetic testing
- genetically determined forms of dystonia -DYT6, DYT&,DYT13
- Congenital or acquired disorders of cervical spine
- Tumors of posterior fossa, foramen magnum and spinal cord



Cervical Dystonia- **Management**

- Cervical dystonia is a serious neurological disorder
- a mix of treatments, including:
 - botulinum toxin
 - physical therapy
 - counseling
 - Treatment of complications



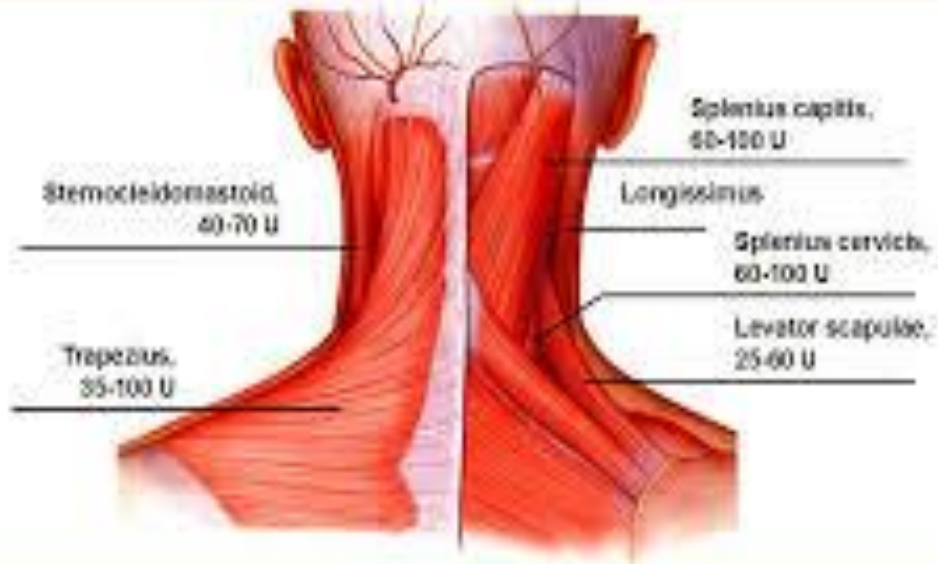
Cervical Dystonia- **Management**

- **First line- Botulinum toxin injections**
- primary treatment for pain relief
- every 12- 14 weeks
- relieve pain and other symptoms in 75%
- EMG guidance experienced a prolonged benefit, a lower incidence of dysphagia and higher incidence of discomfort

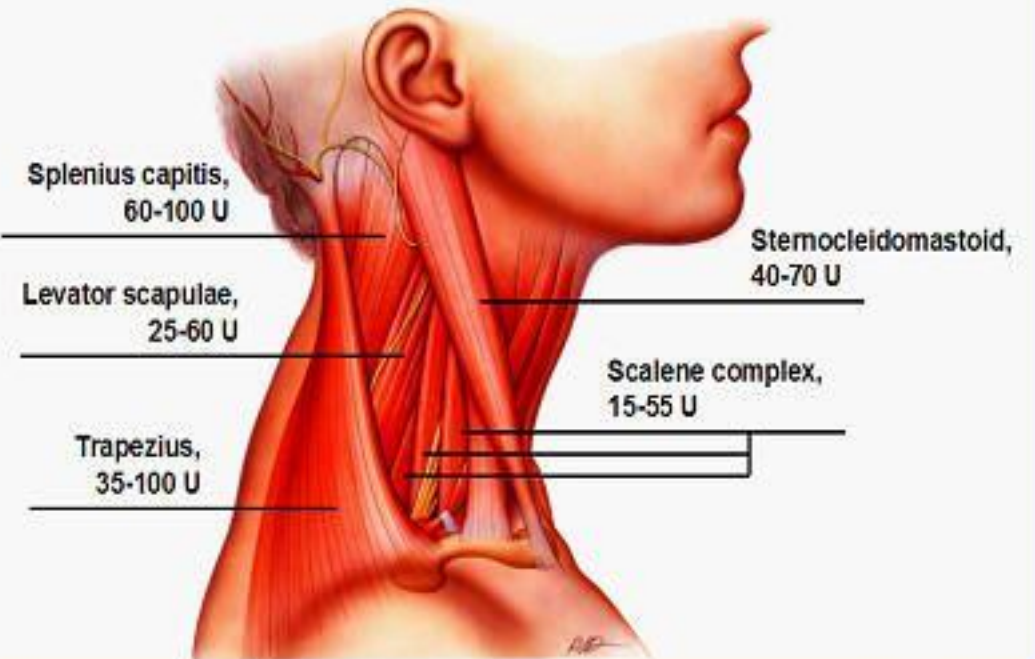
Cervical Dystonia- Management

- **First line- Botulinum toxin injections**

Neck and Shoulder Muscles Affected in Cervical Dystonia



Neck and Shoulder Muscles Affected in Cervical Dystonia





Cervical Dystonia- **Management**

- **Medications**

- anticholinergics, such as trihexyphenidyl and benztropine which block the neurotransmitter **acetylcholine**
- dopaminergics, such as levodopa, bromocriptine, and amantadine which target the neurotransmitter **dopamine**
- GABAergics, such as diazepam, which target the neurotransmitter **GABA-A**
- **anticonvulsants**, such as topiramate- has reported successful for symptoms



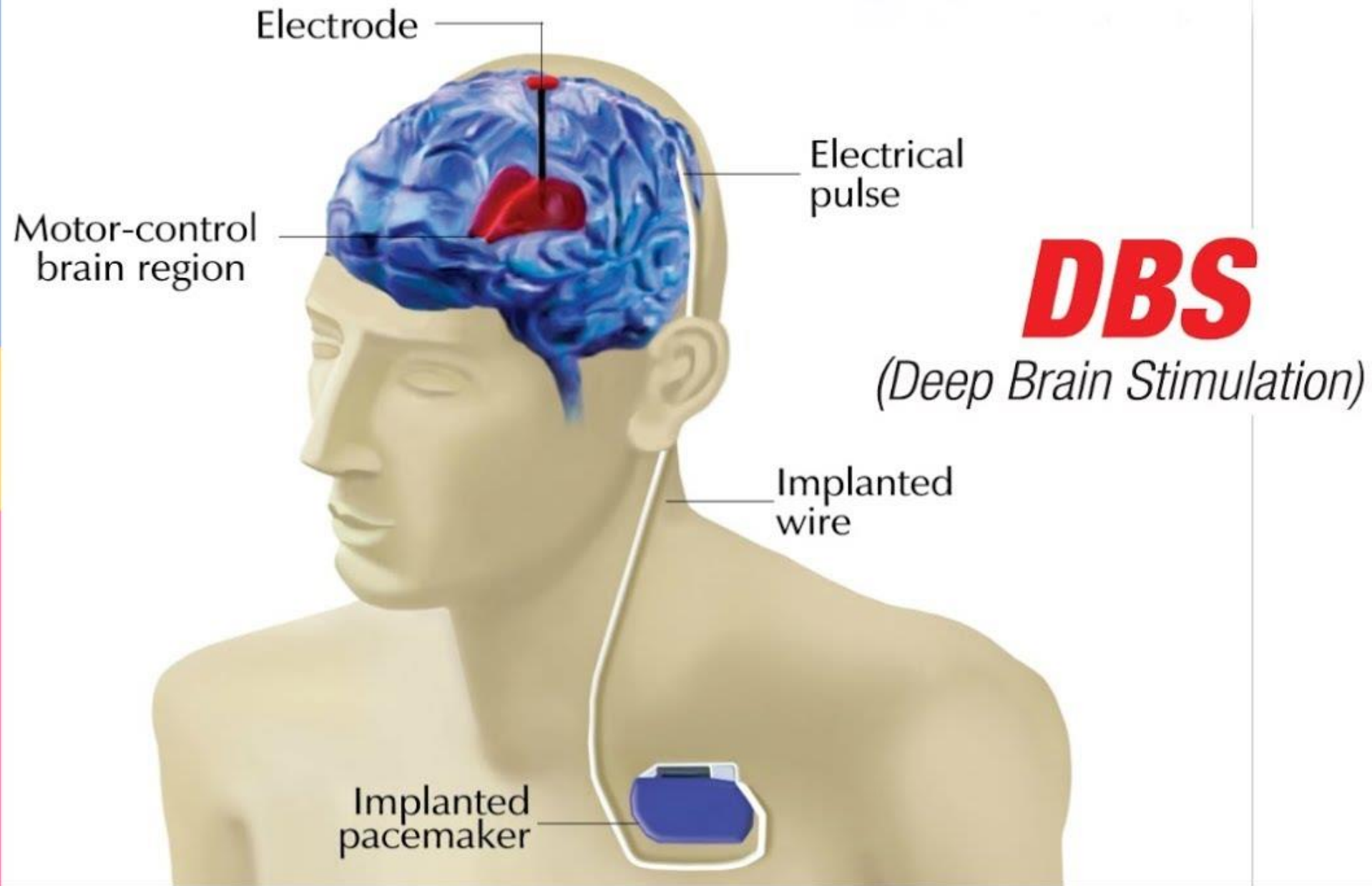
Cervical Dystonia- **Management**

- **Physical therapy**

- includes heat to relax neck and shoulders
- targeted stretching and strengthening exercises.

- **Exercises**

- to relieve symptoms and strengthen muscles.
- Sometimes simple sensory tricks can help stop a spasm.
- lightly touching side of face, chin, cheek, or the back of head
- the effectiveness may lessen in time.

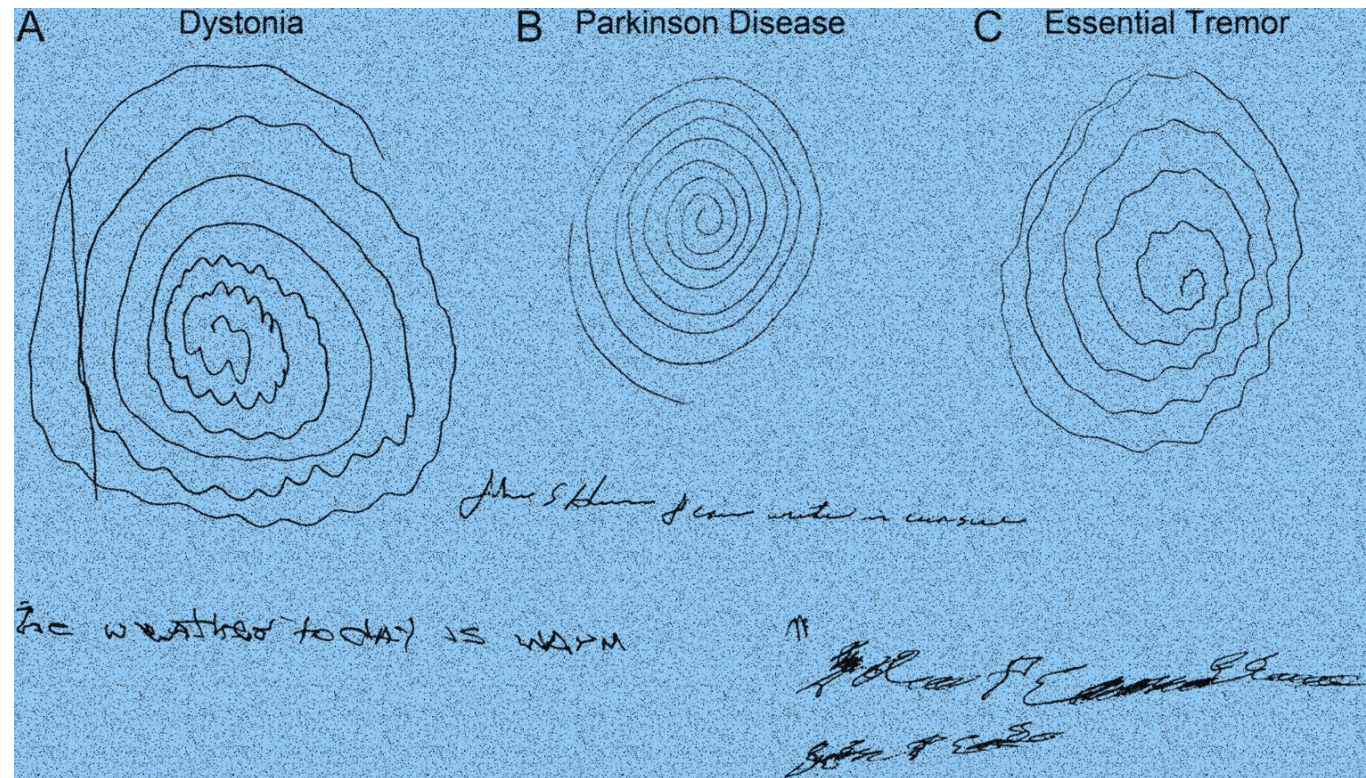


Cervical Dystonia- Management



- to help them more able to manage their motions

Benign Essential Tremor (BET)





Benign Essential Tremor (BET)

- also called **benign tremor, familial tremor, and idiopathic tremor**
- a progressive neurological disorder and the **most common** movement disorder
- characterized by involuntary rhythmic contractions and relaxations (oscillations or twitching movements) of certain muscle groups in one or more body parts



Benign Essential Tremor (BET)

- Large amplitude kinetic tremors
- onset -usually after age 40, but it can occur at any age
- unknown cause but many cases seem to be familial (AD)
- Family history – 50% of cases

Benign Essential Tremor (BET)

- **Clinical Presentations**

- symmetrical, and affects the arms, hands, or fingers;
- sometimes involves the head, vocal cords, or other body parts
- a rhythmic tremor (4–12 Hz)
- either an ***action (intention)*** tremor- during voluntary movements such as eating and writing (goal-directed movements)
- or it is a ***postural tremor***
- not a resting tremor



Benign Essential Tremor (BET)

- **Clinical Presentations**

- Tremor can worsen in response to
 - fatigue
 - strong emotions
 - Low blood sugar
 - cold and heat
 - caffeine
 - Medications – Antidepressants, Bronchodilators



Benign Essential Tremor (BET)

- **Clinical Presentations**

- In disabling cases, ET **can interfere** with a person's activities of daily life including feeding, dressing, and taking care of personal hygiene.
- Small amounts of **alcohol – tremor relief** in some, but effect is relatively brief with **rebound increase** in tremor

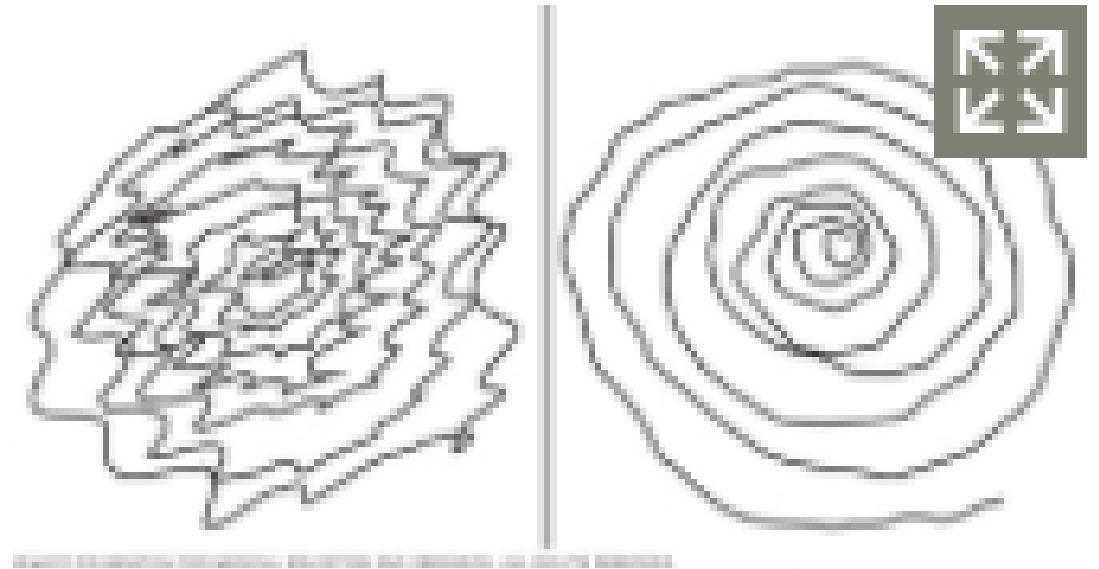


Benign Essential Tremor -**Diagnosis**

- by observing the typical pattern of the tremor (performance test)
- exclusion of known causes of such a tremor - Thyroid disease, metabolic problems, drug side effects, **Parkinsonism**
- **Laboratory test and Diagnostic procedures**
- no medical tests to diagnose essential tremor
- Only for exclusion of others- TFT, level of medications

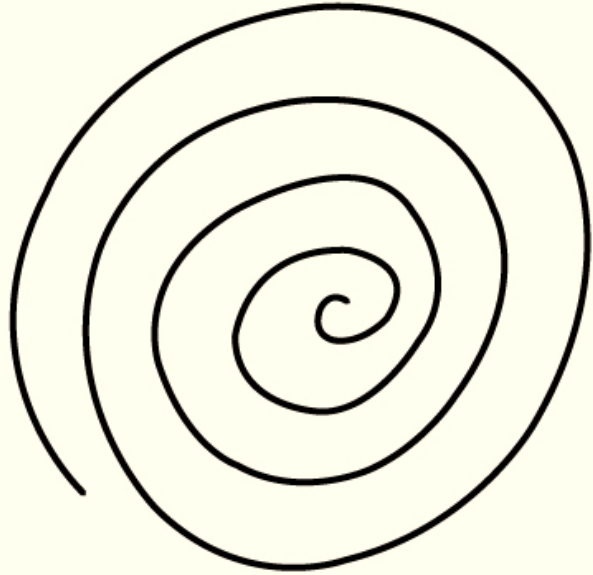
Benign Essential Tremor

- Performance tests
- drink from a glass
- Hold arms outstretched
- Write
- Draw a spiral



Essential tremor test

Benign Essential Tremor



NO TREMOR



PARKINSON'S DISEASE



ESSENTIAL TREMOR



Benign Essential Tremor





Benign Essential Tremor (BET)- Treatment

- **Oral medications**

- **First-line**

- Beta blockers such as propranolol or nadolol and timolol
- anti-epileptic primidone
- have tremor-reducing effects on about **half** of ET patients
- Contraindications- Asthma, peripheral vascular disease, certain heart problems
- Side effects- beta blockers- fatigue, lightheadedness or heart problems, primidone- drowsiness and nausea



Benign Essential Tremor (BET)- Treatment

- **Oral medications**

- **Second-line**

- Anti-epileptics topiramate, gabapentin, levetiracetam
- Side effects- drowsiness and nausea

- **Third-line**

- clonazepam and mirtazapine
- to treat people for whom tension or anxiety worsens tremors.
- Side effects- fatigue or mild sedation.



Benign Essential Tremor (BET)- **Treatment**

- **Botulinumtoxin A (Botox) injections**
- might be useful in treating some types of tremors, especially head and voice tremors
- Side Effects
- Treatment of hand tremors- can cause weakness in fingers.
- Treatment of voice tremors- can cause a hoarse voice and difficulty swallowing

Benign Essential Tremor (BET)- Treatment



- **Deep brain stimulation**
- electrical probe into the (thalamus) that causes tremors
- connect to a pacemaker-like device (neurostimulator) implanted in chest

Benign

Focus

- focus
- The
- of th
- can i
- Side
- walking



area

Benign Essential Tremor (BET)- **Treatment**



- **Avoid caffeine**
- Caffeine and other stimulants can increase tremors.

- **Learn to relax**
- Stress and anxiety tend to make tremors worse
- being relaxed may improve tremors

- **Make lifestyle changes**
- Use the hand less affected by tremor more often.
- Find ways to avoid writing - online banking

Benign Essential Tremor (BET)- Treatment

- **Use alcohol sparingly, if at all**
- Some - notice that tremors improve slightly after drinking alcohol
- Tremors tend to worsen once the effects of alcohol wear off.
- Increasing amounts of alcohol eventually are needed to relieve tremors, which can lead to alcoholism.





Common Movement Disorders I - Conclusion

- Movement disorders are not uncommon
- We can see in our daily clinical practice as well as in our social life
- May be hyperkinetic or hypokinetic
- Increasing awareness can lead to diagnosis



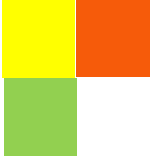
Common Movement Disorders I - Conclusion

- For diagnosis
- Step I : Decide the dominant type of movement disorder(mainly clinical)
- Step II : Make differential diagnosis of the particular disorder
- Step III: Confirm the diagnosis by lab tests and imaging if necessary



Common Movement Disorders I - Conclusion

- Some are difficult to treat but others are manageable even with oral medications
- Botulinum toxin treatment plays an important role in some cases



Thank You