Dystonia



Objectives

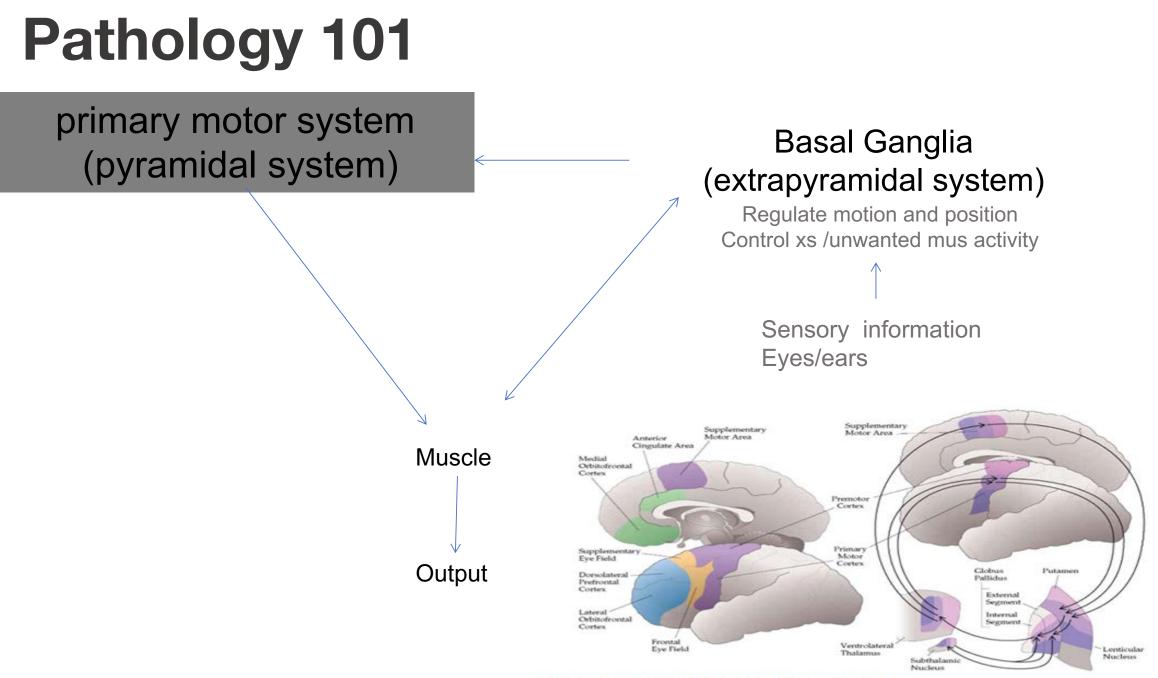
- To have an understanding of the pathology of the origin of Dystonia
- To understand the various classification/locations and potential causes of dystonia
- To have a heightened awareness of the effect of dystonia on a person
- To be aware of treatment principles for the management of dystonia



Dystonia

- Sustained muscle contractions that can cause involuntary twisting and repetitive movements resulting in abnormal postures that sometimes can be painful. The dystonia can be focal (involve only one area of the body) or may include many body parts. Treatments include oral medications, botulinum toxin injections and DBS.
- A state of abnormal muscle tone resulting in muscular spasm and abnormal posture, typically due to neurological disease or a side effect of drug therapy.
- The movements may resemble a tremor.
- Dystonia is often initiated or worsened by voluntary movements, and symptoms may "overflow" into adjacent muscles.





https://www.google.com/images/drprafullkdavemd.com

Dystonia is a brain circuit disorder

- Basal Ganglia ORIGIN
- Cerebellar function, sensory processing, motor inhibition, neuroplasticity and somatotopic cortical organisation but the pathology remain largely unclear
 - Cassidy A pathophysiology of idiopathic focal dystonia ACNR 2010 10 14 18
- Many parts of the BG
- Many ways to arrive at dystonia
- Distribution defines effect
- Inherited dystonias come from changes in basic cell processes



'PAM'

'Our muscles themselves are fine. They're just receiving confusing instructions ("mis-transmissions"), like a mis-coached football team running around in senseless patterns, no offense or defence, as they fail to accomplish coordinated plays let alone score a touchdown! https://dystoniamuse.com/



No inhibition...

- Usually, when a movement is made, excitation and inhibition work in harmony so contraction of the agonist muscles is coordinated with relaxation of the antagonist muscles.
- In patients with dystonia there is deficient inhibition of the antagonist muscles. This can result in **co-contraction**, where the agonist and antagonist muscles contract together.
- Research has identified that the major role of the basal ganglia is to balance excitation and inhibition (just like a pair of scales). However, in dystonia this delicate balance is not attained. It is not yet certain whether the problem is with the direct pathway, the indirect pathway or both. However, as dystonia appears to result from insufficient inhibition in the muscles, it may be that the indirect pathway is failing, resulting in impaired suppression of muscle activity. The lack of inhibition of antagonist or surrounding muscles ultimately causes the cocontraction or overflow phenomena seen in dystonia.
- One area of focus has been to look at whether dystonia is caused by a shortage of the inhibitory neurotransmitter GABA. It seems plausible that a shortage of an inhibitory neurotransmitter such as GABA may play a role, since dystonia seems to be a failure of inhibition.
- At present this is a theory that remains unproven; however, one of the treatments for dystonia, which helps in some cases, is to prescribe medications that increase the quantity of GABA such as benzodiazepines, gabapentin or baclofen.
- Courtesy of <u>http://www.dystonia.org.uk/</u>



QOL

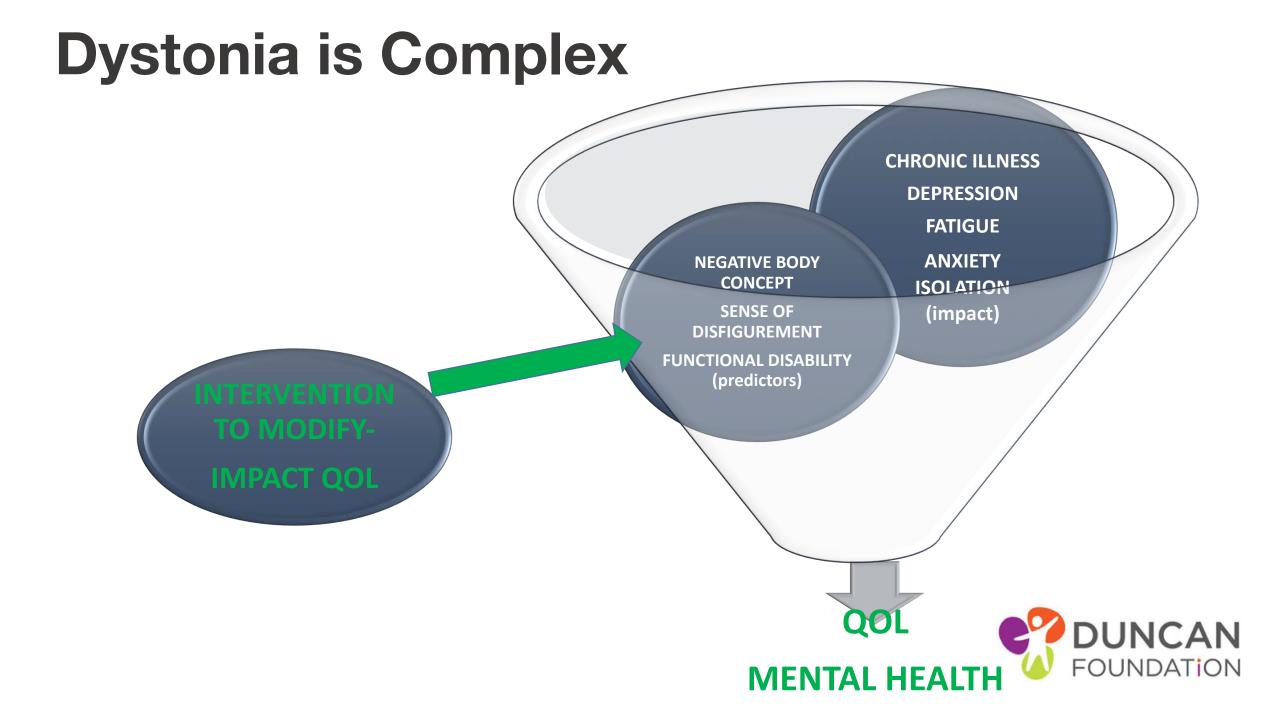
- Mobility
- Daily activities
- Pain uncommon in early stages
 - cervical dystonia 75%
- Mental Health 65% mod or s. anxiety or depression.

64%

- exacerbate symptoms.
- Employment status.
 - 69% reduced productivity,
 - 31% reduced hours or responsibilities
 - 19% reported unemployment as a result of their condition (Molho 2009)
- Gudex 1998
- Social life 75%
- Self-conscious 80% (Dystonia Society 2011)







Classification

- Areas affected
 - generalised, focal, multi focal, segmental, hemi-dystonia
- Origin
 - primary idiopathic out of the blue 50%, inherited ?precise cause, further classified by the gene
 - Secondary acquired disease, environmental, infection, drug, trauma damage to BG
- And location
 - Cervical = neck, blepharospasm = eyes, writers cramp = hand



Diagnosis

- History BI (anoxia, trauma, CVA) psychiatric (anti psychotic drugs)
- No blood test or lab test as chemical/functional
- Not visible on CT/MRI
- Exclusion



Cervical Dystonia

- Patterned, repetitive, and spasmodic or sustained muscle contractions resulting in abnormal movements and postures of the head and neck may affect any combination of neck muscles result in jerky head movements or periodic or sustained unnatural positioning of the head (dystonic posturing).
- Extrapyramidal system fine tunes to maintain 'set point'
 - CD set point altered so overactive contraction of push pull balance into 'new set point'
- Cervical Dystonia >Spasmodic Torticollis
 - as may not be spasmodic and may or may not consist of torticollis (head turning)



• Ostrem J

Cervical Dystonia: Characteristics

- Cause is unknown
- Familial history approx 12%
- Neck trauma
- F>M, 30-50yrs
- Neurologic examination normal
- Sensory tricks-partial, temporary relief



Cervical Dystonia: Characteristics

- Pulling sensation involuntary twisting or jerking
- Worsen gradually
- Plateaus 5 years
- Spontaneous remissions rare
- Each subtype activates **different** pattern of muscles resulting in the abnormal neck/head posture

combination

• Tremor



Anatomy

Splenius capitis Rotation towards Extends head

Levator scapulae Lateral tilt same side

Trapezius Extend head Rotates scapular Sternocleidomastoid Chin forward, rotation opposite small flexion

> Scalene Rotates towards Flexion

Anatomy

Trapezius

Splenius capitis Sternocleidomastoid Rotation towards Chin forward, Extends head rotation opposite small flexion Levator scapulae Lateral tilt same side Extend head Rotates scapular

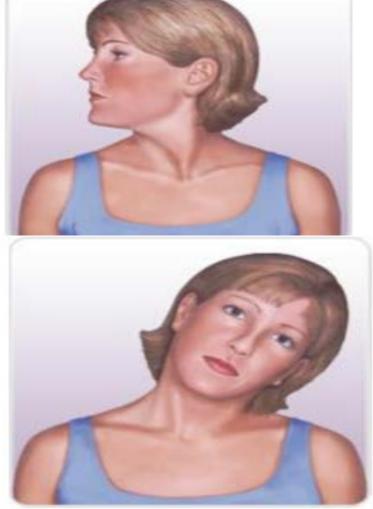
Muscles involved in cervical dystonia

Torticollis

- Contralateral (opposite side)sternocleidomastoid
- Contralateral trapezius
- Ipsilateral (same side)splenius capitis
- Ipsilateral splenius cervicis
- Ipsilateral levator scapulae

Laterocollis

- Sternocleidomastoid
- Ipsilateral splenius capitis
- Ipsilateral scalene complex
- Ipsilateral semispinalis capitis and longissimus
- Ipsilateral levator scapulae
- Trapezius



FUUNDATION

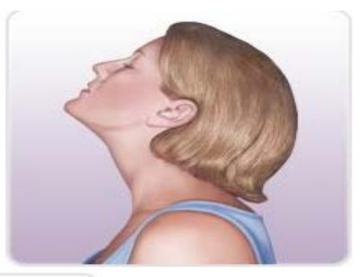
Muscles involved in cervical dystonia

Retrocollis

- Bilateral splenius capitis
- Bilateral levator scapulae
- Posterior vertebral muscles

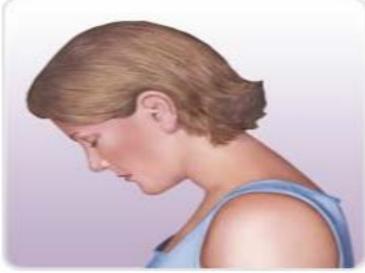
(semispinalis capitis and longissimus)

• Upper trapezius



Anterocollis

- Bilateral sternocleidomastoid
- Scalene complex





Cervical Dystonia

- VIDEOs
- <u>https://www.youtube.com/watch?v=mSwo28t5t3k</u>
- https://www.youtube.com/watch?v=ZcFPI
- Includes sensory trick



Treatment

- Pain
- Depression
- Local injection Botulinum Toxin
- Surgery
- DBS
- Therapy input upper cervical/ chiropractic scoliosis management



Treatment theory

Muscular contraction is abnormal - excessive co-contraction, misfiring



- Reduces the speed and force of the movement
- Weakens antagonists

AIM - rebalancing dystonic muscles and under performing antagonistic muscles

National Institute of Neurological Disorders and Stroke



Treatment opts

- Pain relief
- Maint ROM
 - Gentle joint mobilisation
 - Postural alignment
 - stretches
- Strengthening antagonist
- Voluntary automatic control of head position
- Decreasing intensity dystonic
 - Relaxation direct effect or coping?
- Simple steps in order to re-educate movement patterns.
- Proprioception
 - Adapted to tilt



Treatment challenges

Rare - Evidence is limited on effects of PT

Lack of clinically experienced therapists

The PT = intense motor learning exc (postural control, balance, strengthening axial musc and facilitation of voluntary movement) and mobilisation tech of Cx spine and dystonic mus.

Studies compare Botulinum Toxin (BTX) + PT vs BTX alone Study intensity -PT = 40 min / session every 2nd day for 6 weeks , 75 mins sessions 5 days/wk for 5 wks, up to 90 min/day for 2 weeks = not feasible



Jean Pierre Bleton

Physiotherapist of the Parkinson's Unit in the Neurology Department at the Rothschild Foundation in France – the Guru!!!

Intervention Goals

- maintain the **flexibility** of the spinal column and the cervical muscles.
- diminish the intensity of the spasms
- achieve voluntary control, then automatic control of the correct head position
- and, if you have pain, to alleviate pain

Requires

- learning to **contract** the muscles that correct your CD.
- repeating the contraction of these muscles
- learning how to hold your head in a straight and stable position voluntarily
- to succeed in maintaining a correct head position in everyday activities



PT intervention

Maintain ROM of the cervical column gentle, manual positioning – to stretch each of the muscles causing CD

Stimulate and guide the contraction of the muscles that correct the position of the head

HEP technique

Modify the program

Identify appropriate position for relaxation of the muscles

Relax localized muscular tension muscle relaxing rolling movements and gentle manual traction

Correct compensatory curvatures of the vertebrae that may exist at the thoracic or lumbar level

Home exercise programme

Effectiveness requires

- Quality slow performance correcting positions held for 6 to 8 secs
- Quantity several times a day 10 to 15 minutes.
- Between each repetition there must be a period of rest as least as long as the period of exercise.
- Mirror feedback to achieve the best possible correction.



ROTATIONAL TORTICOLLIS

Rotation with hands behind your head;

This exercise consists of turning your head completely in the direction **opposite** your CD, fingers crossed behind your head at the occipital bone. Throughout the duration of this exercise, the bust and arms remain immobile. Only your head turns.

Exercise:RecommendedNot recommendedRotation to be made to the:RightLeft





resting position execution of the exercise

Cx Rotation with extension

Raising your arms above your head, your hands holding a light weight about 1kg while at the same time turning your head in the in the correcting direction.

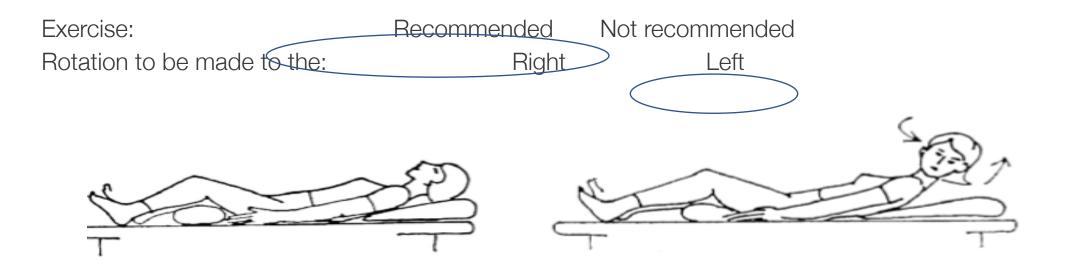
Goal = extension of the whole spinal column, accompanied by a correction of CD.





Flexion with Cx rotation

GOAL; strengthen the sternocleidomastoid muscle (SCM) that corrects your CD. Bring the chin towards the sternum avoiding rotation, then turn your head in the direction away from your CD.



NOTE can be done with El Stimulation

• Electrical Stim on the



Left



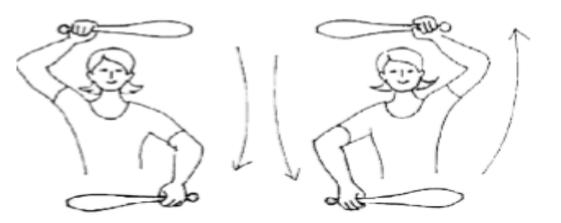








Sensory trick carryover training



Arms moving head still

Progression into coordination of head stability with whole body activities – marching/walking



Electrical muscle stimulators

- Low frequency current.
- Contraction 6 to 8 seconds ; relaxation is approx. double.
- Slow ramp up and down
- Repeat 2 to 3 times, 15 to 20 mins/day.

Generally used on the sternocleidomastoid.

Bleton



Biofeedback

Muscle activity reproduced in beeps or in a visual signal

change with contraction of the muscles.

Feedback to train the contractions in e.g. SCM agonist and antagonist

Mirror feedback another option

Biofeedback for cervical dystonia:

- case series (80 adults, 69 with spasmodic cervical dystonia and 11 with focal dystonia) examining auditory and visual EMG biofeedback. It found that clinically significant improvement of dystonia was achieved by 45/80 (56%) of people at 8 to 12 weeks with biofeedback.
 - While it seems a reasonable approach, it needs to be tested using well-designed RCT



Visual biofeedback

- Vestibular function and altered perception of body orientation
 - reliance on vision for head control and maintaining postural stability.
- portable head-lamp synchronize gaze and head movements and to provide a visual feedback of head position to enhance central sensorimotor integration processes in CD.
- Finding improves quality of life in cervical dystonia by reducing dystonic posture and neck pain. And that
 - Further studies in larger series are needed to assess long-term practicability and efficacy and to identify whether the efficacy of visual biofeedback varies among certain subtypes of CD (e.g. tonic vs. phasic CD, presence or absence of neck pain).
 - J. Mueller



Dos...

- 30 min +/day
- Relax -reduce any stress, depression or anxiety
- Perform active corrective exercises
- Keep your head in the correct alignment TV/radio -giving your body lots of support in sitting
- Arrange your work area so correcting position
- Hold the telephone on the corrective side
- Position so talking to people to the corrective side
- Perform retraining exercises in the swimming pool
- Length not stretch



Try to avoid...

- adopting the posture of torticollis
- holding your head with your hand
- shoulder strap bag
- isolation

ineffective or even harmful.

- massage of spasmodic muscles
- mechanical traction or manipulation
- wearing a cervical collar
- analgesic therapies such as infra-red or lasers



The link

http://dystonia.tripod.com/bleton.pdf



Other emerging

- Auditory stim different in each ear
- Optokinetic stim lateral
- Laser post it notes for training



Outcome measures

- Cervical Dystonia Severity Scale (CDSS)
 - method utilizes a protractor and wall chart to grade the
 - severity of the patient's head deviation from neutral in eachof 3 planes of motion (axial, coronal, and sagittal).
 - http://www.wemove.org
- Columbia Torticollis Rating Scale
- Unified Dystonia Rating Scale (UDRS) focal
- The Fahn–Marsden dystonia scale generalized

- CD Impact Profile, CDIP-58
- CDQ-24
- Short Form-36 Health Survey (SF-36)
- Tsui score- severity and pain low correlation with QOL



Joost van der Dool et al

- Effectiveness of a standardised physical therapy program: study design and protocol of a single blind randomised controlled trial. <u>BMC</u> <u>Neurology</u> December 2013, 13:85
 - Bleton
 - motor relearning principles
 - coaching
 - feedback
- modern principles to enhance neuroplastic changes
- tailored, evidence based intervention more effective



Standarised program

- Botox 2 wks for 1 year standardised
 - functional performance of the exercises adapted to daily life situations, muscle stretching, passive mobilization of the neck and training principles which have found to be relevant for neural rehabilitation and motor learning and will be performed by trained physical therapists.
- A summary of the theoretical basis is displayed in Table <u>1</u> very detailed refer to article..



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Principle	Explanation	Application in standardized PT program
1. Passive mobilisation of the neck	Passive mobilization techniques of the neck create stress relaxation in the collagen fibers of the cervical facet joints. This helps to increase ROM	Passive mobilisation techniques are applied by PT's
2. Muscle stretching for relaxation	Stretching elongates the dystonic muscle and helps to relax it due to the Golgi tendon reflex.	Passive stretching of dystonic muscles
Motor learning principles (Kleim & Jones[20])		
Principle	Explanation	Application in standardized PT program
1. Use it or lose it	Failure to drive specific brain functions can lead to functional degradation.	Activation of antagonists
2. Use it and improve it	Training that drives a specific brain function can lead to an enhancement of that function.	Training of antagonists in order to improve voluntary movement of the head
3. Specificity	The nature of the training experience dictates the nature of the plasticity.	Functional training of activities of daily living tailored to the patients needs
4. Repetition matters	Induction of plasticity requires sufficient repetition.	Exercise of newly gained tasks 5–10 times a day for 10–15 minutes
5. Intensity matters	Induction of plasticity requires sufficient training intensity.	Training intensity is tailored for the individual and monitored over time
6. Time matters	Different forms of plasticity occur at different times during training.	1 year of therapy is divided in 3 stages according the 3 stages model of Fitts & Postner [45]

Hydrotherapy

- Bouyancy + resistance
- Improvement in flexibility and strength through the use of a supportive medium.
- Hard work out with the added resistance and turbulence of the water.
- Reduced delayed muscle soreness compared with gym.
- On a air mattress or buoyancy aids
- Reverse fixed points ie turn body not head
- Retrocollis swim on back if on front increase ext
- Side stroke pick correct side



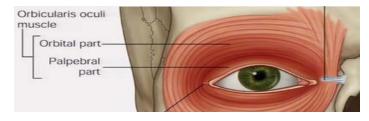
Not just the neck...

- Same principles for treatment management
- Different
- Combinations
- May be task specific ie when writing but not other dexterity



Types

- Blepharospasm
 - eye muscle, squinting (1 but usu. 2), affect vision,
 - inc. sunlight, wind, stress, running/ decr. Humming singing, pinching neck
 - F>M, 50y +
 - Artificial tears don't help
 - Botox to the orbicularis Oculi
- Oromandibular dystonia
 - Jaw musc can't open, clench, grinding
 - Pain, hard to eat, drink, speak
 - ?after dental procedure/trauma
- Meige syndrome
 - facial mus (pharynx, throat, speech swallow)
 - Blepharospasm + Oromandibular dystonia
 - Inc. sTress/ decr. Sing/hum





Types

• Spasmodic dysphoria

- Invol. movements of voice box Add/Abduction
- Can't talk loud, irregular choppy speech, silent at times
- CAN scream, laugh without spasm, whisper and sing high pitched
- Does affect airflow
- Can have mixed F>M, 30 50 yrs onset
- Diag. local anaesthetic to vocal cord speech normal
- Botox alternating one cord at a time
- Writers cramp
 - Focal dominant hand, ache after a page then onset earlier, may develop into complex and associated with CD/ oromandibular
 - Simple writing only Complex all dexterity tasks (buttons, utensils)
 - Usu. Person swaps hand 50% dev in non dom also
- Occupational dystonia
 - Job-musician spasms/feel heavy and pain
 - Rest and Botox but then weakness and in coordination balance but pard if



Types

- Generalised dystonia
 - Inherited, rare, local then progresses limbs, torso and facial, may occur at rest
 - Stop during sleep and reduces with rest generally
 - Earlier onset of focal more likely to progress to general
 - If dopamine responsive levodopa, baclofen, intrathecal pump, surgery BG, DBS
- Paroxysmal dystonia Episodic, inherited
 - Kinesogenic, nonkinesogenic, exertion induced and hypnogenic during sleep



Botox

- Botulinum toxin injections for dystonia Botulinum toxin injections (Botox®, Xeomin®, Myobloc®, Dysport®)
- Temporary dennervation
- No signal to contract loose/slackened
- Which muscle? feel muscle EMG to guide
- What dose? Depends...
- 3 days begin to see effect $-\frac{3}{4}$ weeks max effect
- 10 weeks repeat dose
- May develop resistance antibodies
- In conjunction with physiotherapy for antagonist muscle groups and progress midline orientation gait pattern upper limb function - Splinting - DMO
- Serial Casting splint applied/casted/remove 5-7 days later and reapplied continue until reduction in joint range is achieved



Focal hand dystonia

- Important to assess scapular stabilisation
- Somatosensory impairment proprio, putty, coordination exercises
- Kinematics angle changes of the platform writing on changes pressure and angles and improves symptom
- Constraint 4 weeks
- Large hand writing cursive
- Outcome measures
 - time to onset, time 10 consecutive looped L's
 - Writers Cramp Rating Scale movement and speed
 - Arm Dystonia Disability Scale quantify disability



Treatment aims – address intrinsic biomechanical issues and explore ways of creating new "neurological pathways" e.g. for playing the piano.

extensive myofascial work (releasing "trigger points" in tight muscles) of the upper limbs and shoulder girdles and acupuncture treatments together with postural correction work at the piano.

Attention to general health and fitness included institution of a cardiovascular exercise programme (swimming and cycling).

exploring new ways of playing that might use different neurological pathways, (rather than the old ones which had become "smudged").

Altering the height of the piano stool (sometimes quite drastically- the position of the music on the music stand- posture at the piano – sometimes in quite extreme ways, if only to demonstrate that using new ways of playing could bypass many of the dystonic movements

Altering sensory input: Playing with rubber gloves - Covering the ivories of the Steinway with Elastoplast tape- Rubbing the finger tips on a rough surface to "sensitise" them before playing - Playing with white noise through headphones - Using mirrors

Sensory retraining away from the piano to "redefine" the sensori-motor areas of the brain for each finger eg learning Braille, blind fingering of dominoes and other objects

Motor retraining using splints a Type of "constraint induced therapy" where the finger that compensates for the dystonic finger is splinted prior to short periods of practice), and even a short period of cast immobilisation

modifying technique to create a new way of playing (ie using new neurological pathways and patterns



Dopamine Responsive Dystonia

- involuntary muscle contractions, tremors, and other uncontrolled movements (dystonia). from mild to severe but typically improve with sustained use of a L-Dopa.
- usually appear during childhood, most commonly around age 6. The first signs typically the development of inward- and upward-turning feet (<u>clubfeet</u>) and dystonia in the lower limbs. The dystonia spreads to the upper limbs over time;
- Beginning in adolescence, the whole body is typically involved.
- Affected individuals may have unusual limb positioning and a lack of coordination when walking or running. Some people with this condition have sleep problems or episodes of depression more frequently than would normally be expected.
- Over time, parkinsonism. including unusually slow movement (bradykinesia), muscle rigidity, tremors, and an inability to hold the body upright and balanced (postural instability).
- The movement difficulties usually worsen with age but stabilize around age 30.
- A characteristic feature of dopa-responsive dystonia is worsening of movement problems later in the day and an improvement of symptoms in the morning, after sleep (diurnal fluctuation).
- Rarely, adult-onset cases, parkinsonism usually develops before dystonia, and movement problems are slow to worsen and do not show diurnal fluctuations.



Geste Antagonsite

- Indep find, temporary effect, location and pressure differ
- most preferred an ipsilateral light touch on the lower face, chin, or posterior neck - forceful ipsilateral touch, light or forceful contralateral touch, or bilateral manoeuvres
- "supports emerging theories that dystonia is a disorder of sensorimotor integration."
- sensory trick "produces its effect by inhibiting the over-activity of the motor cortex."



DMO

- embraces the neurophysiological approach, where proxima' key points of control, reciprocal inhibition and stimulation of the sensorimotor system are used.
- biomechanical approach of maintaining muscle length, strength and joint alignment.
- To trial effect where you put your hands to have an effect
- omotrain -in theory
- a lumbar belt to load the pelvis then to mimic the rotational panelling use an elastic bandage –pull diagonally up toward the hip to encourage external rotation.
- K taping to facilitate sensory feedback effect





Imbalance

Vestibular Visual and Somatosensory input from receptors in muscles, joints and the skin

Stability of vision, posture and perceived body orientation.

Dystonia - somatosensory input and potentially vision

Cervical pathology - alter cervical muscle function and joint mechanics.

The expected and actual afferent inputs do not match. This can lead to secondary disturbances in the Vestibular and visual systems due to plastic changes in the CNS.

May present as dizziness

• (Brandt 2001)



Imbalance

- Falls
- Community complex environment
- Inactivity



Exercise!!!

let pain be your indicator to modify an exercise. No pain no gain – **not for you**!

Sensible aerobic exercise - cardiovascular system and maintaining optimal health.

It is ok to feel an ache of opposing muscles being worked & normal to feel muscle ache when working a new muscle group.



Neuro pilates

A form of dynamic stabilisation retraining that reconditions the body from the central core to prevent the recurrence of and treat a range of postural, Msk and neurological conditions.

- both avoid the area of Dystonia and go right into it.
- work around an ailment...in the effort to make the 99% of the body that is healthy even healthier so it can support the 1% that is struggling.
- Pilates classes from an instructor with neurological experience.
- four people per class. -specialist attention to every person at the class.



Massage? debatable..

- Cutaneous stimulation of the dystonic muscles reinforces the pathologic activity
- Mixed results
- Acupressure the most economical effective pain relief with measureable low risk
- Traction shown to help
- Direct pressure over SCM ineffective and may injury



Being healthy

- Healthy people- become more active, energetic and healthier versions of themselves.
- Even with dystonia it is important to reach optimal health, becoming proactive about general health.
- Dystonia is sensitive to any kind of stress including fatigue to managing your time to managing your dystonia fight fatigue



Online resources

- <u>www.dystoniasociety.com</u>
- Dystonia Medical Research Foundation (DMRF)
- <u>www.dystonia-foundation.org</u>

info@dystonia-foundation.org

- The Dystonia Society
- http://www.dystonia.org.uk/

info@dystonia.org.uk

nsda@dysphonia.org

bebrf@sbcglobal.net

- National Spasmodic Dysphonia Association (NSDA)
- www.dysphonia.org
- Benign Essential Blepharospasm Foundation (BEBRF)
- <u>http://www.blepharospasm.org/</u>
- WeMove (Movement disorders)
- http://www.wemove.org



Measures

- QUALITY OF LIFE
 - Cranio-cervical Dystonia Questionnaire (CDQ-24) and Short Form 36 (SF-36)
- Disability
 - Toronto Western Spasmodic Torticollis Rating Scale (TWSTRS)
 - Functional Disability Questionnaire (FDQ). The FDQ is a 27 item scale to measures the impact of CD on daily functioning
- RANGE OF MOVEMENT
 - ability to perform voluntary movements, with a cervical range of motion meter (CROM) [29]. The CROM is a frame that will be placed on the head with
 three separate inclinometers to measure AROM in the sagittal, coronal and horizontal planesthe psychometric properties of the CROM in patients with CD
 are unknown, in a healthy population the CROM is a reliable instrument to measure cervical ROM
- PAIN
 - To determine the additional effects of PT on pain, patient are asked to rate their pain on a Numeric Rating Scale (NRS).
- DEPRESSION
 - Beck's Anxiety Index and Beck's Depression Index
- Severity
 - Tsui scale measures different aspects of abnormal posture and movements in CD patients
 - Clinical Global Impressions-Severity of Illness Scale (CGI-S) and the Clinical Global Impression Improvement scale (CGI-I).
 - ADDS arm dystonia scale



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THANK YOU

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