Cervical Dystonia

One type of dystonia – Julie Rope
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

Chan J
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
Sternocleidomastoid
Chin forward, rotation opposite

Scalene
Rotates towards

Trapezius
Extend head
Rotates scapular

Splenius capitis
Rotation towards
Extends head

Levator scapulae
Lateral tilt same side

Sternocleidomastoid
Chin forward, rotation opposite
small flexion

Scalene
Rotates towards
Flexion
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
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
  • https://www.youtube.com/watch?v=mSwo28t5t3k
  • https://www.youtube.com/watch?v=ZcFPK

• Includes sensory trick
Treatment

• Pain
• Depression
• Local injection – Botulinum Toxin
• Surgery
• DBS
• Therapy input – upper cervical/ chiropractic – scoliosis management
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
- Studies compare Botulinum Toxin (BTX) + PT vs BTX alone

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.

Study intensity - PT = 40 min/session every 2nd day for 6 weeks, 75 mins/session 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
- 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

HEP technique
- Modify the program
- Identify appropriate position for relaxation of the muscles
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: Recommended Not recommended
Rotation to be made to the: Right Left

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 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.

Exercise: Recommended  Not recommended
Rotation to be made to the: Right  Left

NOTE can be done with El Stimulation
• Electrical Stim on the Right  Left
Sensory trick carryover training

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

Arms moving head still
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
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 each of 3 planes of motion (axial, coronal, and sagittal).
  • [http://www.wemove.org](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
• Effectiveness of a **standardised physical therapy program**: study design and protocol of a single blind randomised controlled trial. *BMC Neurology* 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 1 very detailed – refer to article.
<table>
<thead>
<tr>
<th>Principle</th>
<th>Explanation</th>
<th>Application in standardized PT program</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. Passive mobilisation of the neck</td>
<td>Passive mobilization techniques of the neck create stress relaxation in the collagen fibers of the cervical facet joints. This helps to increase ROM.</td>
<td>Passive mobilisation techniques are applied by PT’s.</td>
</tr>
<tr>
<td>2. Muscle stretching for relaxation</td>
<td>Stretching elongates the dystonic muscle and helps to relax it due to the Golgi tendon reflex.</td>
<td>Passive stretching of dystonic muscles.</td>
</tr>
</tbody>
</table>

**Motor learning principles (Kleim & Jones[20])**

<table>
<thead>
<tr>
<th>Principle</th>
<th>Explanation</th>
<th>Application in standardized PT program</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. Use it or lose it</td>
<td>Failure to drive specific brain functions can lead to functional degradation.</td>
<td>Activation of antagonists.</td>
</tr>
<tr>
<td>2. Use it and improve it</td>
<td>Training that drives a specific brain function can lead to an enhancement of that function.</td>
<td>Training of antagonists in order to improve voluntary movement of the head.</td>
</tr>
<tr>
<td>3. Specificity</td>
<td>The nature of the training experience dictates the nature of the plasticity.</td>
<td>Functional training of activities of daily living tailored to the patients needs.</td>
</tr>
<tr>
<td>4. Repetition matters</td>
<td>Induction of plasticity requires sufficient repetition.</td>
<td>Exercise of newly gained tasks 5–10 times a day for 10–15 minutes.</td>
</tr>
<tr>
<td>5. Intensity matters</td>
<td>Induction of plasticity requires sufficient training intensity.</td>
<td>Training intensity is tailored for the individual and monitored over time.</td>
</tr>
<tr>
<td>6. Time matters</td>
<td>Different forms of plasticity occur at different times during training.</td>
<td>1 year of therapy is divided in 3 stages according the 3 stages model of Fitts &amp; Posner[45].</td>
</tr>
</tbody>
</table>
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
Online resources

- [www.dystoniasociety.com](http://www.dystoniasociety.com)
- Dystonia Medical Research Foundation (DMRF)
  - [www.dystonia-foundation.org](http://www.dystonia-foundation.org) info@dystonia-foundation.org
- The Dystonia Society
  - [http://www.dystonia.org.uk/](http://www.dystonia.org.uk/) info@dystonia.org.uk
- National Spasmodic Dysphonia Association (NSDA)
  - [www.dysphonia.org](http://www.dysphonia.org) nsda@dysphonia.org
- Benign Essential Blepharospasm Foundation (BEBRF)
  - [http://www.blepharospasm.org/](http://www.blepharospasm.org/) bebrf@sbcglobal.net
- WeMove (Movement disorders)
  - [http://www.wemove.org](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 measure 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 planes. The 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
References


Cassidy A pathophysiology of idiopathic focal dystonia ACNR 2010 10 14 – 18


Chronicles of a dystonia muse _https://dystoniamuse.com/


Dystonia foundation organisation – living with dystonia physical therapy resource

Joost van den Dool, et al; Effectiveness of a standardised physical therapy program: study design and protocol of a single blind randomised controlled trial. BMC Neurology December 2013, 13:85

Mueller J, Wissel J Visual biofeedback treatment improves cervical dystonia Department of Neurology, Vivantes Hospital Berlin Spandau, Academic teaching Hospital of the Charité, Berlin

Ostrem Jill L., MD Diagnostic criteria for cervical dystonia: Can botulinum neurotoxin manage, as well as, cure the problem? Professor of Neurology UCSF Department of Neurology Movement Disorder and Neuromodulation Center Bachmann Strauss Dystonia and Parkinson’s Disease Center of Excellence Dec 2015


National Institute of Neurological Disorders and Stroke
