Thinking Outside the Box for Cervical Dystonia (Torticollis) in Adults:

Role of PT for Prevention, Early Identification and Intervention Strategies

Nancy Byl, PT, MPH, PhD, FAPTA
Professor Emeritus, UCSF, Department of Physical Therapy and Rehabilitation Science, School of Medicine, Center for PT Health and Wellness

Purpose of Lecture I

- Define cervical dystonia/torticollis including clarifying the differences between childhood and adult cases
- Become familiar with different clinical presentations of of adults with cervical dystonia
- Outline potential factors for early identification of CD in patients with mechanical neck pain (MNP)
- Summarize some unique additions to the history and physical examination for patients with CD
- Discuss sensitivity and specificity of making an early identification of CD in patients with MNP

Purpose Lecture II

- Outline the objectives for CD rehabilitation
- Summarize PT interventions for CD (standard versus integrated learning based training)
- Review the effectiveness of PT intervention strategies for CD (meta analysis /case series)
- Summarize patient reported quality of life living with CD

Purpose of Lecture II

- Describe a current, longitudinal clinical trial designed to define PT guidelines for CD (Netherlands)
- Raise questions about a paradigm shift or quackery: videos of response to mouthpiece for TMJ problems in patients with CD
- Summarize key points

Cervical Torticollis/Dystonia: Adults

- Involuntary, focal movement disorder of the neck with *end range posturing in rotation, side bending and excessive extension or flexion (sometimes with oscillations)*

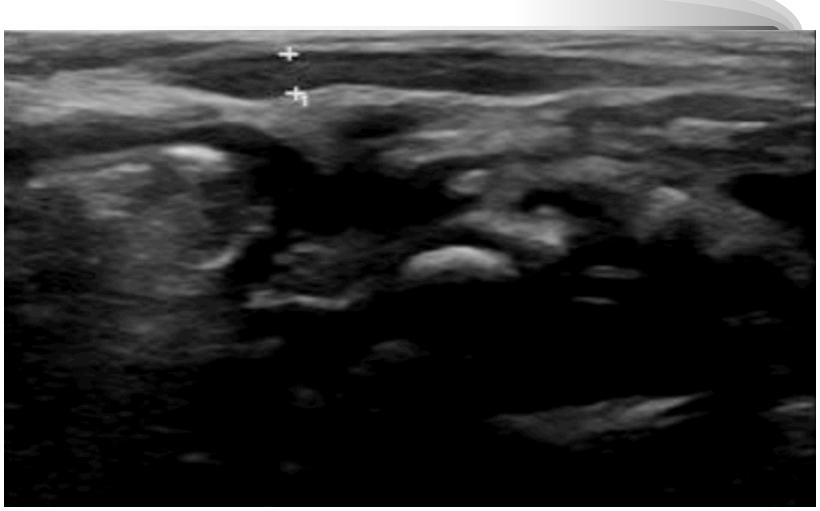
Clinical Presentation of CD in Adults

- Poor inhibition/over excitation
  - co-contractions agonists/antagonists
  - "tonic", "jerking" or tremor oscillations
- Can not only involve the neck but the jaw, eye lids, shoulder, mouth, tongue, eye tracking
- Usually painful (constant muscle contractions) May have degenerative changes (x-ray/MRI)
- ***Sensory tricks decrease involuntary movements***

Cervical Torticollis: Children

- Positional asymmetry of the neck noted at birth or shortly after birth
  - Usually associated with positioning in utero
  - Physical findings
    - Shortening and thickness of neck muscles:one side
    - Muscle spasms, but no dystonic" co contractions of agonists and antagonists
    - Responds well to stretching and retraining with supervised PT and parental collabortion at home

MRI:Cervical Torticollis Children



Examples of Adults with Cervical Dystonia: UTube

Types of Sensory Tricks in CD

- Inversion
- Elevation of arms
- Run fingers in hair
- Chew on toothpick
- Touch side of face (opposite to turning/ side bending
- Go to high altitude
- Rock in rocking chair
- Sail a sailboat
- Lie on back with full extension of OA joint
- Side propping on unaffected side
- Bend down with head between legs and arms up to ears
- Have a mouthpiece molded

Example of "Sensory Tricks"

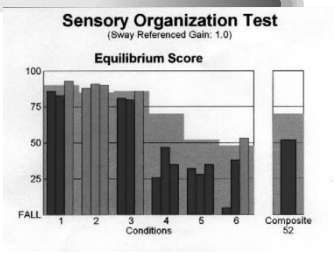
Head between knees

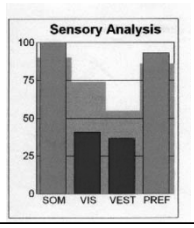
Retrocollis Elevate arm /turn head to stop retrocollis

Clinical Presentation with Extensor Muscle Weakness:Forward Head

Clinical Presentation of Poor Balance: Normal MRI but Abnormal Posturography

- In case series of 5 subjects with CD, all abnormal SOT (Byl , 2010)
- Normal sway in stable conditions
- Increased sway and lost balance in complex sensory challenged conditions
- All four failed (condition 5); 3 also failed condition 6 (visual, vestibular, somatosensory)





Clinical Presentation:Adults with CD

- Asymmetric overactivity
  - Muscle tension in upper trapezius , levator scapulae, splenius capitus
  - Opposite side: overactivity of (SCM)
- Difficult to turn in one direction
- Position of head; side bent with excessive extension or flexion and rotation in opposite direction

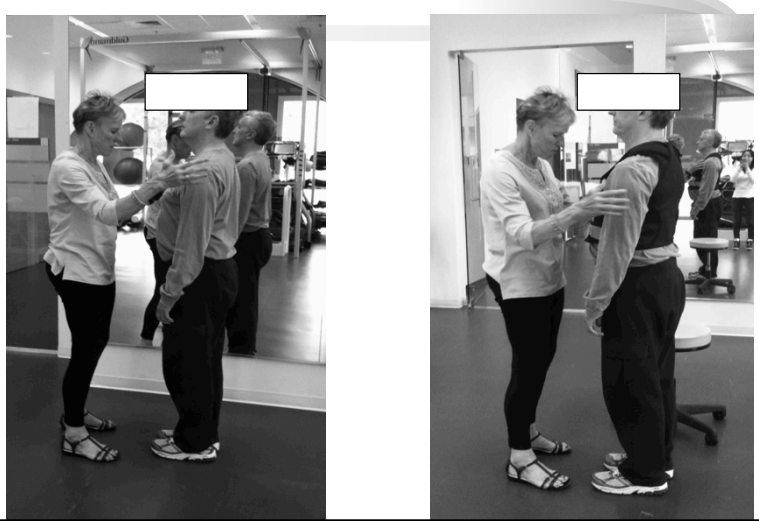
- Symmetric over-activity
  - Hyoids and scalenes
  - Head pulled forward
  - Weak neck extensors
- Under activity of SCM on the side of the bias of the head (tilted or turned)
- Slow onset
- Cramping relaxes with sleep
- Unaware of good alignment

Clinical Presentation of CD: Oscillatory Head Movements

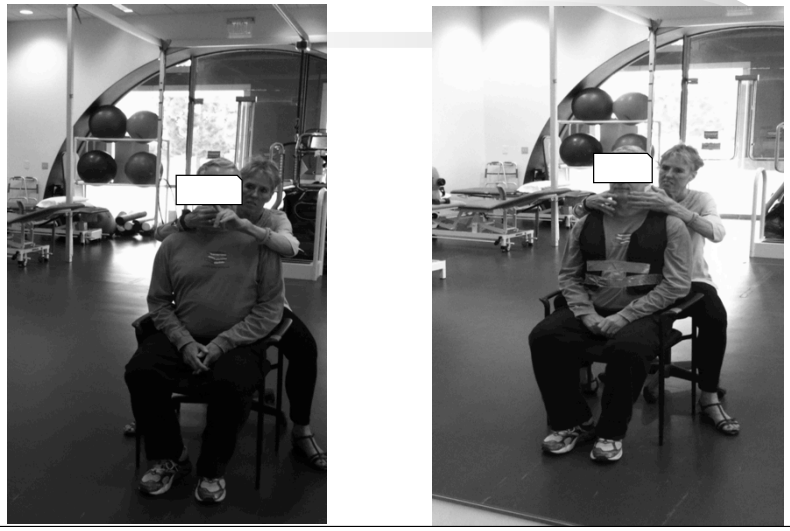
- Oscillations: Defect in central neural integrator
  - _ PD (resting tremor) _ CD (movement tremor)
- Type I oscillations in CD
  - Small oscillation with frequency of 3-9 Hz
  - Some view as similar to essential tremors
- Type II oscillations in CD
  - Irregular, jerky and large amplitude; 0.1 and 0.5 Hz
  - Some suggest the jerky oscillatory movements confirm diagnosis of dystonia (Shaikh et al,2015)

Oscillatory Head Movements

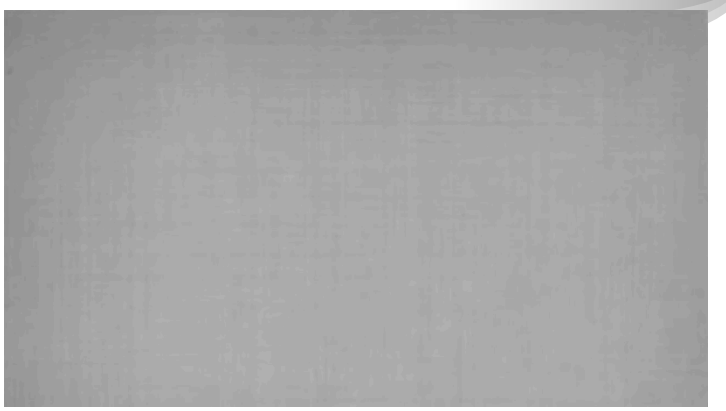
Clinical Presentation: Abnormal Postural Righting



Clinical Presentation with Difficulty with Eye Tracking While Head Kept in Neutral



Are we Missing Cervical Dystonia in Patients with Stiff Neck or Painful Cervical Spams:



Clinical Presentation: CD versus Cervical Neck Tension with Tinnitus

- ENT examination positive for tinnitus (> 3 months)
- Negative MRI, no history whiplash injury
- No history of cervical surgery or TMJ pain

- CST (37/87 patients)
  - Neck Bournemouth Q>14
  - Absence of trigger points: (sensitivity 82%)
  - Positive manual rotation
  - ↑+ Adapted Spurling Test
  - ↑neck pain with tinnitus
  - Muscular tension with sleep
  - Decreased hearing

- Non CST/?CD (50/87 pts)
  - Neck Bournemouth Q< 14 (sensitivity 80%)
  - No ↑ ↑ neck pain/spasms
  - Positive trigger points
  - Excessive muscle irritability
  - Asymmetric head position
  - Muscle tension ↓sleeping

Cervical Dystonia VS Stiff Neck

Cervical Dystonia	**Wry Neck/Stiff Neck**
• Slow onset; may not be painful; can include jerking	• Acute onset; often after a night's sleep or physical exertion
• Tension in upper trapezius, levator, splenius capitus same side; excessive SCM activity opposite side	• Primary complaint -pain
• Side bending and extension same side; rotation opposite	• Side bent to one side
• Cannot initiate turning in direction opposite to rotation	• Tension upper trap, levator, splenius capitus same side
	• Can side bend in opposite direction but feel stretch pain

Cervical Dystonia VS DDD/Wry Neck

Cervical Dystonia	**Stiff Neck/ Degen Arthritis**
• Quiet when sleeping	• Persistent nightly pain/spasm
• Made worse with postural tasks and/or eye movts	• Most comfortable with head supported (e.g. supine)
• May be irritated with collar	• Relieved a little with cervical collar
• *Sensory tricks* can reduce abnormal movements	• Not altered by eye movements
• *Oscillatory* head movements/tremors in 28-68% of patients	• Not altered by *sensory tricks*
	• No *tremors or oscillatory* head movements

History, Physical Exam and Tests for Mechanical Neck Pain

- History: Onset, psychosocial, work, trauma, family hx
- ROM; symmetry; strength; endurance; movement quality; sensation; pain (nature); postural alignment
- Standardized tests recommended
  - VAS (0-10 with score >2)
  - Asymmetric Manual Rotation Test (passive/active rotation - CROM)
  - Quality of movements C0-C2; C2-C7 (rated 1-3)
  - Hypermobility/normal/hypomobility)- abnormal in 2/3
  - AST: segmental provocation test (extension, lateral flexion and rotation: + and pain > 2

History and Physical Exam for Cervical Spine:Mechanical Neck Pain

- Craniocervical flexion test (CCFT)
  - Patient in supine with hook lying position with neck in neutral, facing horizontal and mid line bisecting neck longitudinally
  - Posterior glide on BP cuff at 20 mm Hg
  - Hold maximum posterior glide at each 2 mm hg to 30 mm hg for 2-3 second without abnormal movement patterns
  - Endurance: Isometric contraction of 10 seconds at 5 pressures: (20,25,30,35,40)

Unique History Questions for CD

- Family Hx: genetics, family members with cervical dystonia,other focal dystonia , neuro-degemerative disease
- Personal and occupational Hx: excessive neck demands
- Previous trauma (e.g. MVA)
- Personality factors: Type A, perfectionist,
- Psychological factors: compulsive, phobic, perseverative
- Behavioral risk factors: stress, depression, anxiety psychopathology
- Disease:DDD, spondylosis or spondylolisthesis
- TMJ problems, grinding, bruxing teeth

Unique Physical Exam for CD

- Evaluate balance and postural righting reactions
  - One foot and tandem romberg
  - Eyes closed; on stable and unstable surface
  - Effect of inversion, weighted vest, ankle/arm weights
- Evaluate visual tracking, saccades, double vision, fixation, nystagmus,VOR/OKN (e.g. check reading ability with head moving/words moving)
- Video tape patient at rest and with activities that stimulate involuntary movements

Additional Neurological Examination for CD

- Check reflexes: arms, legs, abdominal (usually normal
- Check neural tension (often positive)
- Evaluate neurosensitivity
  - Hypersensitive to touch (tactilly defensive)
  - Sharp dull discrimination- normal
  - Skin around neck and shoulders
    - Localization- abnormal
    - Graphesthesia- abnormal
    - Stereognosis- abnormal
    - Vibration may increase or decrease hypersensitivity

Additional Standarized Testing for CD

- Evaluate depression (Beck Depression Scale)
- Evaluate stress/anxiety (Beck Anxiety Scale)
- Evaluate fatigue (Fatigue Inventory)
- Assess sleep (Sleep Questionnaire)
- Evaluate involuntary movements
  - Toronto Western Twister Scale
  - Comprehensive Cervical Dystonia Scale

Standardized Testing for Functional Outcomes in CD

- Severity of CD
  - Tsui Scale (different aspects of abnormal posture: maximum 25 points)
  - Clinical Global Impressions-Severity of Illness Scale (CGI-S)
- Disability
  - TWSTRS Toronto Western Spasmodic Torticollis Rating Scale disability section 7 point likert scale
  - Functional Disability Questionnaire (FDQ) 27 items measuring impact of CD on function

Standardized Tests for Outcomes in CD

- Active ROM (measure with the Cervical Range of Motion Meter frame with 3 inclinometers – [CROM])
- Neck strength (extension, flexion, rotation in both directions)–weak on one side/direction
- Quality of Life
  - Craniocervical Dystonia Questionnaire (CDQ-24)
  - High correlation with SF 36

Physical Findings: Risk Factors for Cervical Dystonia

- History of neck trauma
- Parent has CD, PD or tremor
- Ashkanzi Jewish ethnicity
- Personality: Type A, perfectionist, phobic
- Normal reflexes
- Poor balance/postural righting
- Asymmetric head position at end range in one direction and difficulty turning in opposite direction
- Jerky neck movements
- Difficult moving slowly
- Excessive muscle tension during day -quiet at night
- Sensory tricks:change position
- Poor sensory discrimination

"Ruling in a CD Diagnosis"

Compared to patient with Orthopedic Neck Problems	Yes =1	No = 0	(comment)
1. Normal Neurological Exam except task specific involuntary movements and maybe twitching beyond neck	X		
2. Uncontrollable asymmetiric muscle tension	X		
3. Difficulty turning in one direction	X		
4. Muscle tension relaxes with sleep	X		
5. Slow onset of problem	X		
6. Uses sensory tricks to stop or enhance movement	X		
7. May or may not have pain	X		
8. Type A Personality(pefectionist, impatient, compulsive	X		
9. Under stress	X		
10. Poor postural righting	X		
11. Oculomotor control problems (VOR, OKN, diplopia)	X		
12. TMJ problems; grinding teeth	X		
13. Ashkanzi Jewish ethnicity	X		
14. . Sensory discrimination problems (graphesthesia, stereognosis	X X		

Total >12 High Probability of CD

Benefit of Diagnostic Criteria

LR+=.86= 5.4
.16

LR-= .14=.166
.84

	Has FHD	Does Not have FHd	
Score >12	25	5	Sensitivity 25/29=86%
Score <12	4	26<	Specificity 26/31=84%

False negative 5/30 (17% have a positive test and no disease
False positive 4/29 (13.7% have negative test, but have disease

LR+= sensitivity/1-specificity (high) LR- 1-sensitivity/specificiey (low)
ROC plots sensitivity by 1-specificity (want combination high sens and low 1-spec)

Summary

- It often takes years before the diagnosis of focal dystonia is confirmed
- Early identification of risk factors for CD lends itself to preventive activities
- Prevention of CD is easier to address than treatment

Lecture II: Physical Therapy Intervention Strategies to Supplement Botulinum Toxin Injections

Objectives for Physical Therapy

- Work as part of a team (including the patient and the family) to educate about CD/rehabilitation strategies
  - Establish positive health behaviors (including regular exercise, sleep, nutrition, healthy life style, stress management, counseling)
  - Stop abnormal movements
  - Maintain normal range of motion and good postural alignment
  - Strengthen neck stabilization and weak, corrective muscles

Objectives for Physical Therapy

- Decrease pain and muscle tension (massage,STM, jt mobs)
- Reduce neural tension in the upper limb
- Stretch short muscles
- Improve integrative balance
  - Integrate normal balance/ postural righting responses into daily activities
  - Facilitate good biomechanics of neck movements
- Learn to separate eye and head movements
- Find postures and sensory tricks to allow normal movement without cramping

Objectives of Physical Therapy

- Inhibit dystonic muscles (e.g. fatigue, biofeedback, electrical stimulation or active/resistive contractions)
- Reinforce voluntary contractions of corrective muscles (e.g. moving head to opposite side of torticollis attitude)
- Reduce neural and sensory hypersensitivity (relaxation techniques/imagery)
- Decrease TMJ problems /grinding of teeth

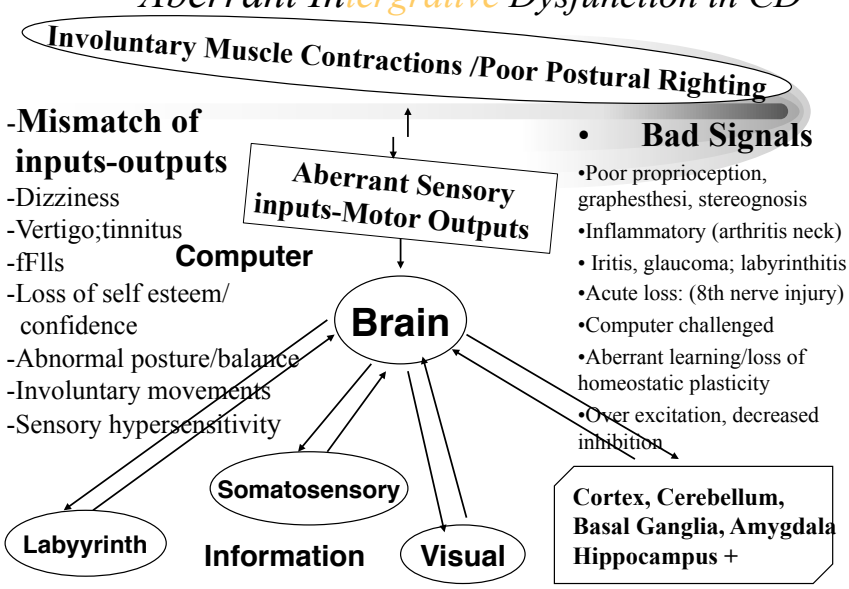
General Physical Therapy for CD

- There are no" standardized" programs for treating patients with all forms of CD
- Traditional, standard programs include:
  - Range of motion exercises (active /passive)
  - Muscle elongation and posture exercises
  - EMG biofeedback training (turning muscles off/on)
  - Electrotherapy-stimulate weak corrective muscles
- Standard programs decrease pain and spasm but do not necessarily improve functional ADLs

Beyond Muscle Imbalance and Weakness: An Important Theoretical Construct

- If the underlying etiology of cervical dystonia is multifactorial which drive abnormal sensory and motor processing (involuntary muscle contractions, poor integrative balance, excessive excitation, poor inhibition) …..**then**
- Successful retraining must address the CNS imbalance is to restore normal, homeostatic, sensory-motor learning

Aberrant Intergrative Dysfunction in CD



Integrating Principles of Neural Adaptation in PT Retraining Strategies for CD

- Neural retraining activities must require
  - Attention
  - Repetition of normal more than abnormal movements
  - Feedback to monitor success
  - Progression of difficulty in task practice
  - Daily practice spaced over time
  - Enhanced internal proprioception and postural alignment with gravity
  - New learning everyday and interaction with others

Evidence Based Literature: Match PT to Each Patient Needs

- **Myoclonic form**: quick, involuntary muscle jerking or twitching (myoclonus) that usually affects arms, neck, and trunk.
  - Objective of PT : try to quiet/immobilize head (reduce pathological head movements)
    - Position of eyes: train eyes to follow objects to side opposite torticollis to reduce spasm and correct bias of CD
    - In front of a mirror: achieve controlled alignment and slow movement several times a day using "Thinking Man's Posture" : engage hyoids and semispinalis and slowly move head in neutral

Evidence Based Literature: Match PT to Each Patient Needs

- **Tonic Form CD**: Muscle contractions are sustained, causing abnormal posture of the head and neck
- Objective PT: rehabilitate corrective muscles
  - Place head in opposite position to dystonic posture
  - Contract the corrective muscles while avoiding increased tonic resistance and preventing overflow
  - Focus selectively on the inactive muscles
  - Integrate progressive positioning:
    - Supine position (possibly with torso slightly raised
    - Progress to sitting, then standing and then walking

A Traditional Community Program: Spasmotic Torticollis Recovery Clinic (STRC Inc)

- Management….. not cure
  - Positive health: Walk regularly, eat a balanced diet, consider psychological counseling to reduce stress
  - Decrease muscle tension/pain at work /recreation
    - Try to maintain good posture
    - Get regular massages
    - Actively range neck motion; avoid extension always
    - Wear a cervical collar if helpful
    - Strengthen weak muscles (turn opposite to pull)

Abigail Brown-New Mexico

General Program for CD (Brown STRC Inc)

- Increase ROM of the neck
  - active movements
  - joint mobilization (careful not to increase tone) if indicated due to degenerative structural changes
  - stretch tight muscles (Abegail Brown. New Mexico)
- Improve ergonomics
  - wear cervical collar when driving
  - set up ergonomic work station

Program Elements:STRC

- Gently self massage neck while supine/sitting
  - Consider purchasing a neck massager unit
  - Massage before bed and first thing in AM
- Do gentle traction: manual, mechanical or towel
- Use one good pillow for sleeping
- Eat well balanced meals
- Program Intensity
  - Go for 1 week to train with Abigal ($1000) or
  - Purchase l ($500) and self train at home
- Effectiveness limited**: No brain retraining!!**

Patient Requirements for Remediating CD

- Use all tricks/ strategies to stop abnormal movements
- Think positively re recovery
- Use mental imagery to retrain (e.g. "inner game of tennis"; imagery and self healing ")
- If CNS is hyperexcitable (on autonomic drive of fight/ flight) quiet nervous system (rocking, swinging, relaxation, quiet place, vibration)
- Use mental imagery to achieve normal head control
- Improve balance/postural righting
- Make video of a person moving head normally (e.g.Recognise for hand – noci.com)
  - Watch the video
  - Imagine it is "you"
  - Copy the movements

Patient Requirements for Remediating CD

- Do learning based training at home and at work 10-100x/ day(corrective exercises)
  - Mirror biofeedback or neurobiofeedback
  - Electrical stimulation of corrective muscles
  - Sensory tricks as necessary
- Create physical opportunities to improve postural righting (e.g. unweighting environments)
  - Float in water keeping neck quiet
  - Walk in water up to neck and keep head quiet
  - Walk with unweighting equipment (e.g. AlterG un-weights with air; other equipment unweights with harness
    - Skip - jog un-weighted)

Patient Requirements for Remediating CD

- Learn more about the principles of neuroplasticity
  - Softwired (Merzenich)
  - Brain Rules (Medina),
  - Brain changes itself (Doidge)
- Restore ability to right the head against gravity
  - Start with inversion, supine, propping on side (head quiet)
  - Progress to partial sitting ,full sitting then standing
- Learn to control eye movements :erform following movements with head kept quiet
  - start supine, - then sitting - then standing)

Patient Requirements for Remediating CD

- Learn to come from supine to sit and sit to stand without using anterior neck muscles (scalenes)
- Learn to breath diaphragmatically (do not use scalenes)
- Retrain the ability to voluntarily turn the head in both directions, slowly without jerking (in prone, supine ,sitting) then progress to faster movements
- Restore ability to tuck chin in (open OA jt) and practice turning trunk on head instead of head on trunk

Patient Requirements for Remediating CD

- Manage stress and depression
- Learn to use a computer /keyboard without strain
  - Maintain neutral posture
  - Put screen at eye level
  - All fingers resting down, move from sh and elbow
  - Avoid touch typing
  - Keep wrists in neutral
  - Take frequent breaks
  - Consider sitting on a Swiss ball

Discover Dynamic Activities to Inhibit the Dystonic Retrocollis

Bending over arms over head

Try reading with inversion

Inversion Traction

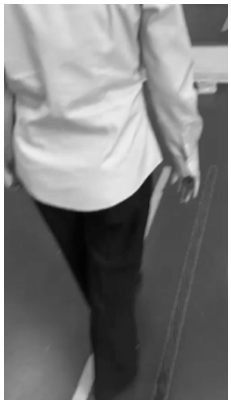
Other Activities

- Sitting and bouncing on ball
- Propping on side
- Prone, head down /up gravity
- BWSTT
- Rolling
- Use lazar pointe on head
- Biofeedback
- Guided imagery
- Follow ball with eyes
- Comfortable cervical support

Example: Stop Abnormal Movements with Active Lazar Training (Pointer on Headband)

- Keep head still with lazar pointer on target for 10-20 seconds (supine, sitting, walking)
- Turn head smoothly in both directions keeping on different targets
- Look up and down smoothly
- Side bend head in both directions
- Squat and lunge while keeping head straight
- Walk forward and backwards and keep head quiet
- Stand on an unstable surface and keep head still

Example of Using Lazar to Improve Alignment



- Take one mirror and have it flat
- Place another mirror at 90 o to the flat mirror
- Place the nose of the patient on the mirror that is 90 o
- The patient must try to keep the head still and not let the face get distorted

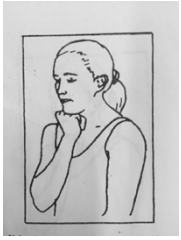
Use mirror imaging to facilitate normal alignmentt

Patient Requirements for Remediating CD;Improve Posture

- Carefully assess response to manual traction; can make things worse or better.
  - Can do home towel traction for relaxation
  - Can try "pumpable collar to distract neck"
- Strengthen lower abdominals
- Do sensory retraining around the neck and shoulders (graphesthesia, stereognosis)

Patient Requirements for Remediating CD:Improve Posture

- Strength core muscles to stabilize neck
  - Tape neck and shoulder (increase sensation)
  - Wear cervical collar when driving or working
- Ask friends, family and co -workers to remind you to stand tall and keep head aligned
- Strengthen the deep neck muscles
  - isometric exercises (rotatories, multifidi)
  - Thinking mans posture
- Strengthen lower abdominals



Patient Requirements for Remediating CD: Improve Balance with Home Practice

- Step high over objects with reciprocal arm movements
- Sit on a large therapy ball
  - Bounce on ball to encourage righting of head and trunk
  - Balance on ball in good posture lifting one leg
  - Balance on ball, good posture, lift one leg and put arms out in front, out to side or throw and catch balls
- Lie on your stomach over a large ball;
  - Balance and lift both legs
  - Balance and lift both arms
  - Balance and bring head to neutral off ball
  - Balance and lift arms, legs and head

Practice Controlled Eye and Head Movements

- While lying supine, have someone move one finger sideways and up/down
  - Follow the finger movement with the eyes
  - When successful add head moving in opposite direction
- Progress from supine to propping on elbows and repeat eye tracking practice
- Progress to crawl position and repeat eye tracking practice

Practices Controlled Eye and Head Movements: VOR and OKN

- Practice reading outloud with the head turning or nodding (slow and fast)
  - Lie on your back under a table with a glass top and book turned down while you read a page
  - Sit and repeat reading with head moving
  - Stand or jog in place and read words on a wall
- Practice reading outloud with the book moving back and forth and up and down (slow and fast)
  - Lie on your back and progress to sitting and standing

Practice Controlled Arm Movements in Sitting Then Standing

- Sitting:
  - Patient should place hands on jaw (Thinking Mans Posture), and repeat eye and head movement tasks that were performed in supine
  - Hold a ball with both hands and circle both arms in large circles first right to left and then left to right while keeping the eyes on the ball
- Then try same tasks standing
  - Stand on a stable surface
  - Stand on unstable surface and repeat eye and head movements

Use Electrical Stimulation to Strengthen Corrective Muscles

- Use ES plus active contraction of muscles for correction ;strengthen weak side
  - Recruit SCM to turn and rotate the neck in the opposite direction of the torticollis
    - While receiving stimulation, patient should actively contract the target muscle
    - When possible, contract and provide isometric resistance while ES is on
    - Recruit neck flexors with ES if have retrocollis; progress resistance

Use Biofeedback fo Retrain Muscles: Inhibition and Contraction

- Rent a biofeedback machine for training (preferably two channel)
  - Put electrodes on the muscles that are over reactive and try to turn them off
  - Put electrodes on muscles that are weak or difficult to turn on and try to turn them on
  - Try to alternate recruiting and turning off muscles
  - Try to reciprocally contract agonist and then release and fire antagonist

Use Gaming Systems (Preferably with a Camera) to Provide Feedback about Movements

- Can perform Wii games for retraining balance
- Look into "EYETOY" programs from Sony or KINECT
  - Watch "Mirror type" feedback from camera during game playing
    - Do functional balance tasks
    - Do activities with hands/arms and keep head quiet
    - Try to move head and neck smoothly keeping it straight while playing the games

How Long Must a Patient Train to Rehabilitate?

- Most patients have difficulty correcting the abnormal movements on a daily basis
- Retraining takes many repetitions (probably >100,000)
  - Have neck in a normal position more than abnormal positions
  - Use normal head movements and inhibit abnormal movements
  - Integrate normal postural righting activities daily
  - Avoid heavy lifting or other activities that stress the neck
- Patients need to address other life style issues
  - Enroll in a stress management class and maybe counseling
  - Engage in fitness exercises 5-6 hours/week (without increasing dystonia)

Outcomes: PT Effectiveness

Outcomes of Physical Therapy Programs for CD

- There are no randomized controlled trials
- Pre-post trials have small number of subjects
- PT is considered adjunctive to medication or DBS
- Small clinical trials and case reports (Botox and PT) indicate
  - patients may improve head position /decrease pain
  - patients may not gain improved quality of life or ADL…..

Positive Outcomes of PT Intervention:Program Components 3 RCTs

- Program Components
  - Intense motor learning (postural control, balance, strengthening axial musculature and facilitation of voluntary movement) (3) (Tasoreli 2006; El-Baharawy 2012;Queiroz 2012)
  - One on one mobilization techniques of cervical spine and dystonic muscles
- Intervention intensity (impractical?)
  - 40 min/session every other day for 6 weeks or
  - 75 minutes/per session 5 days/week for 5 weeks or
  - 90 min/day, every day for 2 weeks

Suggestions for Improving PT Program to Enhance Effectiveness: Bleton

- Focus on rehabilitation antagonist muscles
- Control of dystonic movements by frequent training in functional context
- Teach exercise in 1-2 PT sessions over a few weeks
- After education, home program

Bleton 2010

- Home program: intensive training required in patient environment (up to +10x a day for at least 10 minutes)
- Patients encouraged to correct dystonic posture during daily life activities by turning their head in the opposite direction of the dystonic posture

What Patients Say About Life with CD (1,071 respondents)

- Mean time since diagnosis: 9.6 years; 1-3 years before diagnosis made
  - 66%: lot of pain
  - 61% experience depression and mood alterations
  - 93 % major impact on life
- Seventy percent satisfied with relationship with MD
- Patient expactations
  - 63% expected freedom from spasms
  - 62 % expected freedom from pain
  - 53% expected to return to normal routine

Life with CD: The Patient Perspective

- Most common treatment was BoNT (86%) followed by oral medications (58%) and physical therapy (35%)
- Among patients on BoNT
  - 5% were fairly/very satisfied
  - 25% fairly/very dissatisfied
  - 20% neither satisfied or unsatisfied with outcomes
- **Conclusions**: Patient's expectations need to be better addressed in therapy

Eligibility for Pilot Study: Sequential Case Series

(Byl , 2010)

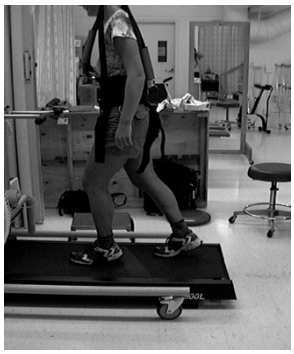
- 5 adults with cervical dystonia
  - Diagnosed by a physician
  - No structural abnormalities of the spine
  - No signs of neurovascular insults, tumors or neurodegenerative disease
  - Had the problem for over a year
  - Tried other interventions- unsuccessful
  - Male and female; 30-65 years of age
  - May or may not be receiving botulinum toxin

Subject Description

- 4 females, one male
- One with family hx of dystonia
- Onset: 3@10 years; 2@3 years
- Stressful jobs: 3 administrators, l musician, l homemaker with 2 year old and husband beginning faculty position)
- Pain scale: 5-6 with muscle spasms
- Posture: Poor dynamic balance; forward head
- All currently receiving botox; one no longer responsive

Supervised Integrative Balance Training

– Walk/jog on a bodyweight supported treadmill with supervision 1x/week while participating in activities that required:
  - Attention
  - Repetition
  - Progression of difficulty
  - Multi-tasking
    – Reading
    – Talking
    – Throwing/catching balls
    – Solving lateral puzzles/math



Pilot Study Sequential Case Series:Procedures

- Additional Intervention
  – Daily home program practicing putting head into neutral position at least 10x/day
  – Aerobic exercise 3-4x/week
  – To guide home training, patients kept notes re activites making them worse and those making them better
- Outcome variables
  – Pain severity (ordinal scale 1-10)
  – Posturography (4 of 5 subjects)
  – Symptoms of dizziness (yes/no)
  – Independence (Café 40)

Results of Case Series

- Compliance
  - All patients kept supervised visits
  - Patients admitted to noncompliance with exercises at home (aerobic exercises 2-3x/week; unable to practice getting into normal position 10x/day)
- With retraining
  - One patient 100% better
  - Three patients improved(head control, posturography, pain, ADL)
  - One patient: nonresponsive to Botox and exercises; scheduled for DBS

Results Case Series

- There were consistencies across subjects:
  - preceding event =stress;
  - type A personalities, high achievers
  - described some early neck trauma (e.g. fall off a bike; MVA; physical struggle in date rape);
  - demanding jobs using poor posture;
  - poor postural righting responses (abnormal SOT)

Post surgical Response DBS:one case

- No measurable effect for first 6 weeks
- Very fatigued post surgery
- 8 -weeks came in for PT and resumed a careful plan for re-learning
- 12- weeks began to be able to keep up upright
- 16- weeks walking with better posture; driving
- 5 years later:replacement electrodes needed
  - doing reasonably well -single mom raising 6 year old
  - still fatigued and cannot work

Current Randomized Clinical Trial of Botox +PT: Netherlands

- Standard/tailored PT (one year)
  - Individualized PT based on evaluation
  - 1-2x/week supervised PT: 4 wks
  - Education provided by PT
  - Functional exercises adopted to daily home situations/practice

    -Muscle stretching -Passive mobilizations -Strengthening
  - Home program with progressive, corrective neural motor training (10x/day until corrected)

Current Randomized Clinical Trial to Create PT Guidelines for CD: Netherlands

- Regular botox injections
  - About every 3-4 months
  - Receiving botox for over a year
- Train PTs in techniques for standardized but tailored PT program
- Random assignment (100 pts)
  - Trained PTs
  - Non trained but licensed PTs
- Training: One year
  - Tailored PT
    - Supervised 2x/week, 4 wks
    - Home Program (daily corrections) + periodic f/u
  - Usual PT
    - PT by licensed PT (usual, standard care)
    - varied frequency as needed

- *Thinking Outside the Box by Using a Mouth Piece for Cervical Dystonia:Paradigm Shift or Quackery?*

Conclusions

- PTs may be able to detect early signs and symptoms of CD in patients with mechanical neck pain
- For effective PT intervention for CD
  - *Collaboration* with an MD team including systemic management, counseling and botox injections is needed
  - *Education about CD and neuroplasticity*
  - *More research on effective PT* needed

Current Evidence: Effectiveness of PT for CD

- Individualized/ supervised exercises for ROM, STM, joint mobilization, strengthening (Biofeedback and ES) and progressive learning based sensory, balance and sensorimotor integrative movements
- **Patient integration of intense home, self directed corrective learning based postural and movement training

Key Points

- Cervical dystonia is a challenging problem to treat
- PT must be individualized for each patient
- Botulinum toxin injections and supplementary PT can reduce pain and disability;may not increase ADLs
- Improvement in quality of life and community participation requires integrative daily, self regulated learning based training at home
- A large RCT in the Netherlands may help us develop better clinical guidelines for PT intervention

THANK YOU

- Panel Discussion

E. Television

0 = No difficulty

1 = Unlimited ability to watch television in normal seated position but bothered by torticollis

2 = Unlimited ability to watch television in normal seated position but requires use of tricks to control torticollis

3 = Unlimited ability to watch television but requires extensive measures to control torticollis **or** is able to view only in nonseated position (e.g., lying down)

4 = Limited ability to watch television because of torticollis

5 = Unable to watch television more than a few minutes because of torticollis

F. Activities Outside the Home *(e.g., shopping, walking about, movies, dining, and other recreational activities)*

0 = No difficulty

1 = Unlimited activities but bothered by torticollis

2 = Unlimited activities but requires simple tricks to accomplish

3 = Accomplishes activities only when accompanied by others because of torticollis

4 = Limited activities outside the home; certain activities impossible or given up because of torticollis

5 = Rarely if ever engages in activities outside the home

III. Pain Scale (MAXIMUM = 20)

A. Severity of Pain Rate the severity of neck pain due to ST during the last week on a scale of 0–10 where a score of 0 represents no pain and 10 represents the most excruciating pain imaginable. Score calculated as: (worst + best + (2*usual))/4

Best ____

Worst ____

Usual ____

B. Duration of Pain

0 = None

1 = Present < 10% of the time

2 = Present 10%–25% of the time

3 = Present 26%–50% of the time

4 = Present 51%–75% of the time

5 = Present > 76% of the time

C. Disability Due to Pain

0 = No limitation or interference from pain

1 = Pain is quite bothersome but not a source of disability

2 = Pain definitely interferes with some tasks but is not a major contributor to disability

3 = Pain accounts for some (less than half) but not all of disability

4 = Pain is a major source of difficulty with activities; separate from this, head pulling is also a source of some (less than half) disability

5 = Pain is the major source of disability; without it most impaired activities could be performed quite satisfactorily despite the head pulling

1. Consky ES, Lang AE. Clinical assessments of patients with cervical dystonia. In: Jankovic J, Hallett M, eds. *Therapy with Botulinum Toxin*. New York, NY: Marcel Dekker, Inc.:1994;211-237.

This rating scale is provided by WE MOVE.
Additional scales and assessment forms are available at **www.wemove.org**
(http://www.wemove.org)

©WE MOVE™

Toronto Western Spasmodic Torticollis Rating Scale (TWSTRS)[1]

PRESENTED BY WE MOVE™ | © 2002

I. Torticollis Severity Scale

A. Maximal Excursion

1. Rotation *(turn: right or left)*

0 = None [0°]

1 = Slight [< 1/4 range, 1°–22°]

2 = Mild [1/4 – 1/2 range, 23°–45°]

3 = Moderate [1/2 – 3/4 range, 46°–67°]

4 = Severe [> 3/4 range, 68°–90°]

2. Laterocollis *(tilt: right or left, exclude shoulder elevation)*

0 = None [0°]

1 = Mild [1°–15°]

2 = Moderate [16°–35°]

3 = Severe [> 35°]

3. Anterocollis/Retrocollis *(a or b)*

a. Anterocollis

0 = None

1 = Mild downward deviation of chin

2 = Moderate downward deviation (approximates 1/2 possible range)

3 = Severe (chin approximates chest)

(continues inside)

b. Retrocollis

0 = None

1 = Mild backward deviation of vertex with upward deviation of chin

2 = Moderate backward deviation (approximates 1/2 possible range

3 = Severe (approximates full range)

4. Lateral shift *(right or left)*

0 = Absent

1 = Present

5. Sagittal shift *(forward or backward)*

0 = Absent

1 = Present

B. Duration Factor *(Weighted x 2)*

0 = None

1 = Occasional deviation (< 25% of the time, most often submaximal)

2 = Occasional deviation (< 25% of the time, often maximal) **or**
Intermittent deviation (25%–50% of the time, most often submaximal)

3 = Intermittent deviation (25%–50% of the time, often maximal) **or**
Frequent deviation (50%–75% of the time, most often submaximal)

4 = Frequent deviation (50%–75% of the time, often maximal) **or**
Constant deviation (>75% of the time, most often submaximal)

5 = Constant deviation (>75% of the time, often maximal)

C. Effect of Sensory Tricks

0 = Complete relief by one or more tricks

1 = Partial or only limited relief by tricks

2 = Little or no benefit from tricks

D. Shoulder Elevation/Anterior Displacement

0 = Absent

1 = Mild (< 1/3 possible range, intermittent or constant)

2 = Moderate (1/3 – 2/3 possible range and constant, > 75% of the time) **or**
Severe (> 2/3 possible range and intermittent)

3 = Severe and constant

E. Range of Motion *(without aid of sensory tricks)*

0 = Able to move to extreme opposite position

1 = Able to move head well past midline but not to extreme opposite position

2 = Able to move head barely past midline

3 = Able to move head toward but not past midline

4 = Barely able to move head beyond abnormal posture

F. Time *(up to 60 seconds) for which patient is able to maintain head within 10° of neutral position without using sensory tricks (mean of two attempts)*

0 = > 60 seconds

1 = 46–60 seconds

2 = 31–45 seconds

3 = 16–30 seconds

4 = < 15 seconds

II. Disability Scale (MAXIMUM = 20)

A. Work *(occupation or housework/home management)*

0 = No difficulty

1 = Normal work expectations with satisfactory performance at usual level of occupation but some interference by torticollis

2 = Most activities unlimited, selected activities very difficult and hampered but still possible with satisfactory performance

3 = Working at lower than usual occupation level; most activities hampered, all possible but with less than satisfactory performance in some activities

4 = Unable to engage in voluntary or gainful employment; still able to perform some domestic responsibilities satisfactorily

5 = Marginal or no ability to perform domestic responsibilities

B. Activities of Daily Living *(e.g., feeding, dressing, or hygiene, including washing, shaving, makeup, etc.)*

0 = No difficulty with any activity

1 = Activities unlimited but some interference by torticollis

2 = Most activities unlimited, selected activities very difficult and hampered but still possible using simple tricks

3 = Most activities hampered or laborious but still possible; may use extreme tricks

4 = All activities impaired; some impossible or require assistance

5 = Dependent on others in most self-care tasks

C. Driving

0 = No difficulty (or has never driven a car)

1 = Unlimited ability to drive but bothered by torticollis

2 = Unlimited ability to drive but requires tricks (including touching or holding face, holding head against head rest) to control torticollis

3 = Can drive only short distances

4 = Usually cannot drive because of torticollis

5 = Unable to drive and cannot ride in a car for long stretches as a passenger because of torticollis

D. Reading

1 = Unlimited ability to read in normal seated position but bothered by torticollis

2 = Unlimited ability to read in normal seated position but requires use of tricks to control torticollis

3 = Unlimited ability to read but requires extensive measures to control torticollis **or**
is able to read only in nonseated position (e.g., lying down)

4 = Limited ability to read because of torticollis despite tricks

5 = Unable to read more than a few sentences because of torticollis

(continues on back)

BMC Neurology

STUDY PROTOCOL Open Access

Cervical dystonia: effectiveness of a standardized physical therapy program; study design and protocol of a single blind randomized controlled trial

Joost van den Dool[1,2], Bart Visser[2], J Hans TM Koelman[3], Raoul HH Engelbert[4,5] and Marina AJ Tijssen[1*]

Abstract

Background: Cervical dystonia is characterized by involuntary muscle contractions of the neck and abnormal head positions that affect daily life activities and social life of patients. Patients are usually treated with botulinum toxin injections into affected neck muscles to relief pain and improve control of head postures. In addition, many patients are referred for physical therapy to improve their ability to perform activities of daily living. A recent review on allied health interventions in cervical dystonia showed a lack of randomized controlled intervention studies regarding the effectiveness of physical therapy interventions.

Methods/design: The (cost-) effectiveness of a standardized physical therapy program compared to regular physical therapy, both as add-on treatment to botulinum toxin injections will be determined in a multi-centre, single blinded randomized controlled trial with 100 cervical dystonia patients. Primary outcomes are disability in daily functioning assessed with the disability subscale of the Toronto Western Spasmodic Torticollis Rating Scale. Secondary outcomes are pain, severity of dystonia, active range of motion of the head, quality of life, anxiety and depression. Data will be collected at baseline, after six months and one year by an independent blind assessor just prior to botulinum toxin injections. For the cost effectiveness, an additional economic evaluation will be performed with the costs per quality adjusted life-year as primary outcome parameter.

Discussion: Our study will provide new evidence regarding the (cost-) effectiveness of a standardized, tailored physical therapy program for patients with cervical dystonia. It is widely felt that allied health interventions, including physical therapy, may offer a valuable supplement to the current therapeutic options. A positive outcome will lead to a greater use of the standardized physical therapy program. For the Dutch situation a positive outcome implies that the standardized physical therapy program forms the basis for a national treatment guideline for cervical dystonia.

Trial registration: Number Dutch Trial registration (Nederlands Trial Register): NTR3437

Keywords: Cervical dystonia, Spasmodic torticollis, Physical therapy, Botulinum toxin, Activities of daily living, Quality of life

* Correspondence: m.a.j.de.koning-tijssen@umcg.nl
[1]Department of Neurology, University Medical Centre Groningen, University of Groningen, Hanzeplein 1, 9700 RB Groningen, The Netherlands
Full list of author information is available at the end of the article

BioMed Central

© 2013 van den Dool et al.; licensee BioMed Central Ltd. This is an Open Access article distributed under the terms of the Creative Commons Attribution License (http://creativecommons.org/licenses/by/2.0), which permits unrestricted use, distribution, and reproduction in any medium, provided the original work is properly cited.

Background

Cervical Dystonia (CD), or torticollis, is a disabling neurological disorder characterized by abnormal positions of the head due to involuntary muscle contractions of the neck [1]. The posture in CD patients can feature one or a combination of postures: rotation (torticollis); lateral tilting (laterocollis); flexion (anterocollis); extension (retrocollis); and lateral shift. With an estimated prevalence of 5.7 patients per 100.000 persons in Western Europe, CD is the most common form of primary adult onset dystonia which usually starts after the age of 30 [2]. Pain is experienced in two-thirds to three-quarters of patients and is a major source of disability, which is strongly associated with the presence of muscle contractions and head deviations [3-6]. Decreased self-efficacy, fatigue, anxiety and depression are other factors associated with disability in cervical dystonia [7]. Research on focal dystonia's, including CD, revealed abnormalities in basal ganglia function, cerebellar function, sensory processing, motor inhibition, neuro-plasticity and somatotopic cortical organisation but the pathophysiology remains largely unclear [8]. Treatment options for CD are mainly symptomatic, aiming to reduce involuntary movements, correct abnormal head positions and reduce pain. Currently, the best evidence based treatment option is to inject the dystonic neck muscles with botulinum toxin (BTX) [9-12]. The effects of BTX fluctuate over time. A peak effect occurs within 2–4 weeks after injections and is followed by a decrease of effect and return of symptoms. On average new injections are given within 12–14 weeks after the previous injections (Figure 1) [13].

In addition to BTX treatment, many CD patients in the Netherlands are referred for physical therapy (PT). However, due to the rarity of CD, experience among Dutch PT's is lacking. Besides, the evidence for the effects of PT on the ability to perform activities of daily living in CD is very limited. [14] Only two small Randomized Controlled Trials (RCT) and one open controlled study investigated the effects of a PT program on CD [15-17]. All studies compared BTX treatment in combination with a PT program versus BTX treatment alone. All studies showed significant better scores on pain and disability in the groups receiving BTX treatment with an additional PT program. The PT programs in all three studies consisted of intense motor learning exercises (postural control, balance, strengthening axial musculature and facilitation of voluntary movement), and mobilization techniques of the cervical spine and dystonic muscles. PT programs varied from 40 minutes per session every other day for six weeks [16], 75 minutes per session 5 days a week for five weeks [17] up to 90 minutes a day for 2 weeks [15]. Although the results of these PT treatments were positive, it is difficult to implement them to current regular care of chronic diseases provided by physiotherapists and exercise therapists. For most patients and therapists it will not be feasible to combine such an intensive program with their daily lives and practice.

One approach towards the treatment of CD was suggested by the French physiotherapist J.P. Bleton [18]. The main goals of this program are the rehabilitation of the antagonist muscles and the control of the dystonic movements by frequent training in a functional context. Exercises are taught during one or two PT sessions a week. After teaching the patients, intensive training is required in the patient's environment (up to 10 times a day for 10 minutes). In addition, patients are encouraged to correct the dystonic posture during their daily live activities by turning their head in the opposite direction of the dystonic posture. Eventually, patients should be able to control the dystonic movements independently. This approach with increasing control of the patients and decreasing therapist involvement seems more applicable than the intensive programs by Tassoreli, El Bahwrady and Queiroz [15-17]. The effect of the Bleton treatment has never been investigated in a large randomized controlled trial. Not only for practical reasons, but also based on the pathophysiological knowledge of CD, a

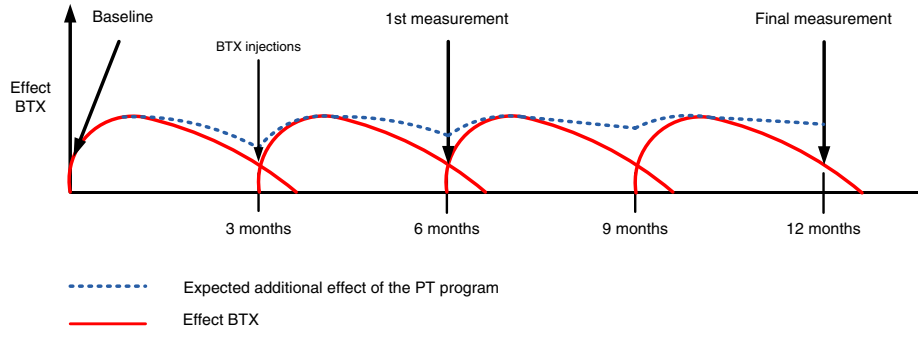


Figure 1 Effect of BTX and expected additional effect of PT. Effect of BTX. Increasing lines indicate a better effect of BTX and less severity of CD, pain and disability to perform daily life tasks. Red lines illustrate the normal effect of BTX, blue dotted lines illustrate the expected additional effect of the PT program.

longer treatment period with a more functional approach and independent continuation of the treatment program seems more appropriate for the long term benefits of PT [15].

In the current study we developed a PT program with elements of the approach suggested by Bleton. We added current knowledge of motor (re) learning principles, coaching and principles of providing feedback in a standardized PT treatment program (Table 1). This resulted in a standardized, tailored PT program comprising of a one year training that aims to improve the ability to perform daily life tasks by emphasizing independent training in the patient's own environment [18-20]. The standardized PT treatment program was developed according the AGREE standards [21] in corporation with the Royal Dutch Society for Physiotherapy (KNGF), the Society for Cesar- and Mensendieck Exercise Therapy (VvOCM) and the Amsterdam School of Health Professions (ASHP). The standardized PT program was developed within the DystonieNet, a national collaboration between neurologists and allied health professionals in research, education and treatment of CD, which was initiated by the neurology departments of four university hospitals in the Netherlands [22]. The standardized PT program aims to relearn or adopt alternative/new movement strategies to improve activities in daily life situations.

The primary objective of this study is to evaluate the effectiveness of the standardized PT program on improving the ability to perform daily life activities in CD patients compared to usual PT that is given in Dutch private practices. Both PT programs are add-on treatment to BTX-injections. Measurement will take place just prior to the BTX injections as it is hypothesized that the effects of the PT program will mainly occur in the period between the injections when the BTX wears off and symptoms return (Figure 1). Secondary objectives are to evaluate the effects on severity of CD pain, quality of life, anxiety and depression.

In addition, cost effectiveness will be detremined by comparing the costs and health utility of the new standardized PT program with the care as usual PT treatment. It is hypothesized that the standardized PT program will be more cost effective and more effective in improving the ability to perform daily life tasks of CD patients than regular PT.

A positive outcome of this study will lead to the development of a national treatment guideline which will be implemented via the Dutch DystonieNet.

Methods/design

Study design

The study will be conducted as a multi-center single blind randomized controlled trial in three Dutch university hospitals. Patients will be randomly assigned to the experimental group or control group using a computerized randomization protocol. Patients in the experimental group will be referred to specialized PT's who are trained prior to the study to perform the standardized PT program. Patients in the control group will be referred to regular PT's and receive care as usual. All data will be collected at baseline, after six months and after one year. In order to determine the additional effects of a PT program, measurements will be performed briefly before the BTX injections at the outpatient clinics of the hospitals. This implies that we measure the effect of the PT program in a period that BTX has the least effect on the symptoms of dystonia (Figure 1). Measurements will be performed by a blind and independent assessor since it is impossible to blind the therapists and patients for treatment allocation.

Participants

The study aims to include 100 patients with primary CD of 30 years and older, stable on BTX treatment for more than one year. Exclusion criteria are secondary or hereditary forms of dystonia, dystonia in other bodyparts than the neck and patients who had surgery for the treatment of dystonia.

Interventions

Standardized, tailored PT program

Subjects in this group receive a one year PT program according the standardized PT program in combination with BTX injections. The PT program will start two weeks after the injections. The emphasis of the PT program is on the 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 [18-20]. A summary of the theoretical basis with respect to *'muscle stretching and mobilizations', 'motor (re)learning', 'transference and generalization', 'feedback' and 'self management',* is displayed in Table 1.

Regular PT

Subjects randomized in this group will receive BTX injections and regular PT once a week for a period of one year. In contrast to the standardized PT program, interventions are not given by specialized therapists. Due to the rarity of CD, the average therapist in the Netherlands has little knowledge about CD. It is likely that common interventions like massage, stretching and exercise of the dystonic muscles are used. Specific information of the weekly sessions and treatment will be retrieved from the local PT's after the patients finished the study period of one year.

Table 1 Theoretical background of the standardized PT program

Muscle stretching /relaxation and mobilisations (de Morree [43], Fung [44])		
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]
7. Salience matters	The training experience must be sufficiently salient to induce plasticity.	Functional training of activities of daily living tailored to the individual needs of the patient
8. Age matters	Training-induced plasticity occurs more readily in younger brains.	
9. Transference	Plasticity in response to one training experience can enhance the acquisition of similar behaviors.	Functional training of activities of daily living tailored to the patients needs and variation and random practice
10. Interference	Plasticity in response to one experience can interfere with the acquisition of other behaviors.	
Transference and generalization (Shea & Morgan [45], Schmidt & Lee [46])		
Principle	*Explanation*	*Application in standardized PT program*
1. Random practice	Enhances the transference and generalization of a task	Tasks or exercises are given in a random order
2. Variation of practice	Enhances the transference and generalization of a task	Specific tasks or exercises are performed in different contexts
Feedback (Shea et al. [47], Schmidt & Lee [46])		
Principle	*Explanation*	*Application in standardized PT program*
1. Summary Knowledge of Results	Feedback is essential for learning to take place. Summary KR is that KR is given after an entire set of trials during an exercise instead of after each individual trial. It is the most effective form for the retention and transference of a task.	Feedback is given after each set of trials of a task. Each task is performed at least 5 times after feedback is provided
Self management (Fitts & Posner three-stage model [48])		
Principle	*Explanation*	*Application in standardized PT program*
1. Cognitive phase	The learner is concerned with understanding a task and developing strategies to perform a task and how the task can be evaluated. These efforts require a high degree of cognitive activity	During the first month patients receive 2 PT sessions a week to (re)learn and understand movement strategies. Movement strategies will be practiced at home 5–10 times a day for 10–15 minutes
2. Associative phase	The learner has selected the best strategy for a task and starts to refine it. This stage requires less cognitive activity	During this stage patients receive 1 PT session. Movement strategies from the first stage will be increased in difficulty. Movement strategies will be practiced at home 5–10 times day for 10–15 minutes

Table 1 Theoretical background of the standardized PT program *(Continued)*

3. Autonomous phase	The learner is able to perform a skill automatically. A low degree of attention is required.	During the last (autonomous) stage, patients are encouraged to perform the learned tasks independently and to improve and maintain their (re)gained abilities themselves. Therapists will have a coaching role. Patients receive one PT session a month for additional advice and motivation.

Outcome variables

Disability

Disability as measured with the disability subscale of the Toronto Western Spasmodic Torticollis Rating Scale (TWSTRS) is the primary outcome of this study. The TWSTRS scale is a widely used scale in research and is a valid and reliable tool to measure severity, disability and pain in CD (Kendall Tau = 0.85, $p < 0.01$) [23,24]. The disability section is a six point Likert scale which consists of six items like driving a car, reading and performing ADL activities (max 30 points). Lower scores indicate less disability.

Disability will also be measured with the Functional Disability Questionnaire (FDQ). The FDQ is a 27 item scale to measures the impact of CD on daily functioning. Questions are asked about the extent to which CD affects the engagement in and performance of a sample of activities at the present time. Each item is rated on a 5-point scale (maximal score 68 points) The FDQ has a high reliability (r=.93, P<0.001) [25].

Severity of CD

Severity of CD will be measured with the Tsui scale [26]. The Tsui scale measures different aspects of abnormal posture and movements in CD patients. It has a maximal score of 25 points. The Tsui-scale is a widely used, standardized and reliable scale (ICC=.86) to measure the severity of CD [26] Lower scores indicate less severity of dystonia.

Severity of CD will also be measured with the Clinical Global Impressions-Severity of Illness Scale (CGI-S) and the Clinical Global Impression - Improvement scale (CGI-I). Both scales are observer- or patient- rated scale that measure illness severity and global improvement on a 7-point scale. Both scales are widely used and reliable and validated for a numerous of disorders (r=0.41 to 0.77, p=0,05 for self perceived measures and r=0.36 to 0.84, p=0.05 of clinician administered measures of anxiety, depression, impairment and quality of life) [27,28].

Active range of motion

To determine the changes in the ability to perform voluntary movements, active range of motion (AROM) will be measured 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. First the resting position of the head will be measured and subsequently the AROM of flexion, extension, lateral flexion and rotation. Although 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 (intratester reliability ranged .63-.93 intertester reliability ranged .74 - .87) [29]. To determine the additional effects of PT on pain, patient are asked to rate their pain on a Numeric Rating Scale (NRS). A score of 0 means no pain and a score of 10 means the worst pain imaginable. The NRS is a validated and reliable tool for the assessment of pain (Spearman r =.94 between VAS and NRS, test-retest reliability ICC = .90) [30,31].

Quality of life

Quality of Life (QoL) will be measured with the Craniocervical Dystonia Questionnaire (CDQ-24) and Short Form 36 (SF-36) [32,33]. The CDQ-24 is a validated and disease specific, self reporting questionnaire to evaluate quality of life of patients with cervical dystonia on a five point likert scale [32]. The CDQ-24 subscales showed moderate to high correlations with those SF-36 subscales measuring similar aspects (Pearson's correlation r = 0.50–0.73; P<0.001, each).The score ranges from 0 to 96 points where lower scores indicate a better QoL. The SF-36 is a validated generic measure for QoL containing 36 items measuring eight dimensions of health [33]. Scores of the different dimensions can range from 0 (worst) to 100 (best).

Anxiety and depression

Since 25 to 59% of the CD patients suffer from anxiety disorders or depression [25,34], the effects of PT are determined with the Beck's Anxiety Index [35] and Beck's Depression Index [36,37]. Both instruments are validated and reliable tools and are rated on a 21 item 4 point Likert scale (BDI: r = .73 with Hamilton Psychiatric Rating Scale for Depression. BAI: test-retest reliability coefficient of .67, R=.54, p=0.05 with anxiety).

Cost effectiveness

To determine the cost effectiveness of both physical therapy programs, the costs per quality adjusted life year (QALY) will be calculated. In addition, cost-effectiveness

related to the clinical outcome will be calculated, with the costs per unit on the TWSTRS-disability scale as the outcome measure. Costs which are associated with loss of productivity due to disability or inability to work will be registered in the subgroup of patients below the age of 65 with the Productivity Costs Questionnaire (PCQ) and EuroQoL-5D (EQ-5D). The PCQ is a 22 item generic questionnaire used to measure absence of work due to health problems and is advised as standard instrument for use in economic evaluations of Dutch healthcare [38,39]. The EQ-5D is a six item, standardized measure of health status in order to provide a simple, generic measure of health for clinical and economic appraisal [39,40].

Sample size

The power calculation is based on the study by Brans et al. investigating the long term effect of BTX on disability and functional health [41]. This study showed an average improvement of 7.1 out of 30 points on the disability subscale of the TWSTRS after 1 year of BTX treatment in CD patients. It is estimated that the additional effect of the PT program according the treatment guideline will be at least half the effect caused by BTX. The cut off for the success of the PT program is therefore, set on an average improvement of 3.5 out of 30 points on the TWSTRS disability scale which is clinically relevant according Brans et al. [42]. With a power of 0.80 and an alpha of 0.05, each group will need 44 subjects. With a loss of 10% taken into account, 50 subjects in each group are required.

Analysis

Differences in all outcome measures, with exception of the measures for cost effectiveness, will be determined with a mixed between-within (repeated measures) analysis of variance for both treatment arms, across three time periods (baseline, after six months and one year). All analyses will be performed under the intention to treat principle in SPSS 20.0. Differences will be considered significant at p-value < 0.05.

The cost effectiveness will be determined by a cost-utility analysis from a societal perspective with a time horizon of one year. Cost-utility analysis facilitates the comparative assessment of health care innovations across different types of interventions, disease areas and health care settings. Incremental cost-utility and cost-effectiveness ratios for the add-on standardized PT program versus add-on regular PT will be calculated as the extra costs per QALY gained and the extra costs per unit decrease in TWSTRS-disability score. The cost effectiveness will be calculated according the most recent guidelines for unit costing in healthcare research [40]. The friction cost method will be applied to calculate the costs of production loss as measured with the PCQ, EQ-5D. Unit costs of production loss will be based on the most recent national guidelines for unit costing in healthcare research [40]. The base year for unit costing will be 2013.

Ethical considerations

In accordance with the local medical ethics committee (MEC) guidelines, written informed consent is required from participants who fulfil the selection criteria. The study has been approved by the Medical Ethics Committee of the Academic Medical Center, Amsterdam (MEC 2012_048). This study is registered under Trial registration number NTR3437 of the Dutch trial registration (Nederlands Trial Register).

Discussion

In our study we aim to fill the gap in evidence based medicine to treat CD patients with PT by performing a large RCT towards the (cost) effectiveness of a standardized, tailored PT program. There are several differences of this study compared to studies reported in the current available literature.

Although other studies have showed added value of short, high intensity PT program on pain and disability in combination with BTX treatment, follow-up measurements were not performed and therefore it is not known if a wash out of treatment effects will occur [15-17]. Since CD is a life lasting disorder, a longer treatment period seems more appropriate to establish lasting changes. We therefore choose for a treatment period of one year in contrary to the other studies which lasted five weeks maximal [15-17]. Another difference with other studies is that the standardized PT program tends to teach patients themselves, how to improve their ability to perform daily life tasks and to manage their symptoms in their own environment. To establish lasting changes and the ability of patients to manage their symptoms in their own environment, we have chosen for a treatment period of one year. The standardized treatment program itself is based on modern principles about motor learning, transference and generalization of learned tasks to enhance lasting (neuroplastic) changes (Table 1) [20,43-48]. Based on these principles, we aimed for a tailored, evidence based intervention that is thought to be more effective than regular interventions.

It is hypothesized that the overall added effect of the standardized PT program on the BTX treatment lies between the periods that the BTX is starting to wear off and the BTX is starting to work again after new injections (Figure 1). Other studies performed measurements in the periods when the peak effect of BTX occurred (2–4 weeks after injections) which make it impossible to determine the additional effects of a PT program on CD

[15-17]. We therefore choose to measure the effects of PT just prior to the BTX injections when the interference of BTX effects are minimal.

Another goal of the standardized treatment program is to make patients less dependent of healthcare providers and to decrease the healthcare costs for this patient group.

In the Netherlands many CD patients are referred for physical therapy. Since CD is a chronic indication for PT, patients receive (except for the first 20 treatments) unlimited reimbursement for PT which results in long lasting use of healthcare in the current, regular situation. We therefore added an economic evaluation to compare the cost effectiveness of the standardized PT program with physical therapy care as usual.

Future implications

In the case of a positive outcome of this study, the standardized PT program will be used as a basis for a national treatment guideline which will be implemented via the Dutch DystonieNet.

Abbreviations

CD: Cervical Dystonia; BTX: Botulinum toxin; PT: Physical therapy; RCT: Randomised Controlled Trial; KNGF: Royal Dutch Society for Physiotherapy; VvOCM: Society for Cesar and Mensendieck therapy; ASHP: Amsterdam School for Health Professions; ROM: Range of Motion; KR: Knowledge of Results; FDQ: Functional Disability Questionnaire; CGI-S: Clinical Global Impression Severity scale; CGS-I: Clinical Global Impression Improvement scale; AROM: Active range of motion; CROM: Cervical range of motion; QoL: Quality of life; BDI: Beck depression index; BAI: Beck anxiety index; SF-36: Short form 36; PCQ: Productivity cost questionnaire; EQ-5D: EuroQoL 5D.

Competing interests

JHTM Koelman and MAJ Tijssen received an unrestricted research grant from Ipsen Pharmaceutical and Allergan Inc. for studies and teaching workshops on dystonia and from Ipsen to finance a specialized dystonia nurse. Ipsen and Allergan have no role in study design, collection, analysis, interpretation of data, in the writing of the report and in the decision to submit the paper for publication. The other authors declare that they have no competing interests.

Authors' contributions

JD wrote the first draft of the manuscript and BV, RE, HK and MKT contributed to the completion of the manuscript. All have made substantial contributions to conception and design of the study. All authors read and approved the final manuscript.

Acknowledgments

The authors wish to thank all the people who have contributed to the development of the standardized PT program and everyone who participates in the study.

Funding

This research is supported by the scientific fund of the Dutch dystonia patient association, Nuts Ohra Fund and the Jacques and Gloria Gossweiler Foundation.

Author details

[1]Department of Neurology, University Medical Centre Groningen, University of Groningen, Hanzeplein 1, 9700 RB Groningen, The Netherlands. [2]Department Exercise Therapy, Amsterdam School of Health Professions, Tafelbergweg 51,1105 BD Amsterdam, The Netherlands. [3]Department of Neurology, Academic Medical Centre Amsterdam, Meibergdreef 9, 1100 DD Amsterdam, The Netherlands. [4]Department of Physiotherapy, Amsterdam School of Health Professions, Tafelbergweg 51, 1105 BD Amsterdam, The Netherlands. [5]Department of Rehabilitation, Academic Medical Center, Meibergdreef 9, 1100 DD Amsterdam, The Netherlands.

Received: 23 April 2013 Accepted: 20 June 2013
Published: 15 July 2013

References

1. Fahn S, Marsden CD, Calne DB: **Classification and investigation of dystonia.** *Mov Disord* 1987, **2:**332–358.
2. Epidemiologic Study of Dystonia in Europe (ESDE) CollaborativeGroup: **A prevalence study of primary dystonia in eight European countries.** *J Neurol* 2000, **247:**787–792.
3. Lowenstein DH, Aminoff MJ: **The clinical course of spasmodic torticollis.** *Neurology* 1988, **38:**530–532.
4. Chan J, Brin M, Fahn S: **Idiopathic cervical dystonia: clinical characteristics.** *Mov Disord* 1991, **6:**119–126.
5. Jankovic J, Leder S, Warner D, *et al*: **Cervical dystonia: clinical findings and associated movement disorders.** *Neurology* 1991, **41:**1088–1091.
6. Kutvonen O, Dastidar P, Nurmikko T: **Pain in spasmodic torticollis.** *Pain* 1997, **69:**286.
7. Zetterberg L, Lindmark B, Soderlund A, Asenlof P: **Self-Perceived non-motor aspects of cervical dystonia and their association with disability.** *J Rehabil Med* 2012, **44:**950–954.
8. Cassidy A: **Pathophysiology of Idiopathic Focal Dystonia.** *ACNR* 2010, **10:**14–18.
9. Jankovic J, Brin M: **Therapeutic uses of botulinum toxin.** *N Engl J Med* 1991, **324:**1186–1194.
10. Brashear A, Bergan K, Wojcieszek J, *et al*: **Patients' perception of stopping or continuing treatment of cervical dystonia with botulinum toxin type A.** *Mov Disord* 2000, **15:**150–153.
11. Burgen ASV, Dickens F, Zatman LJ: **The action of botulinum toxin on the neuromuscular junction.** *J Physiol* 1994, **109:**10–24.
12. Truong D, Duane DD, Jankovic J: **Efficacy and safety of botulinum type A toxin (Dysport) in cervical dystonia: results of the first US randomised double-blind placebo-controlled study.** *Mov Disord* 2005, **20:**783–791.
13. Costa J, Espirito-Santo C, Borges A, Ferreira JJ, Coelho M, Moore P, *et al*: **Botulinum toxin type A therapy for cervical dystonia.** *Cochrane Database Syst Rev* 2005, **25**(1):CD003633.
14. Delnooz C, Horstink MWIM, Tijssen MA: **Warrenburg van der BPC: Paramedical Treatment in Primary Dystonia: A Systematic Review.** *Mov Disord* 2009, **24:**2187–2198.
15. Tassoreli C, Mancini F, Balloni L, Pacchetti C, Sandrini G, Nappi G, *et al*: **Botulinum toxin and neuromotor rehabilitation: an integrated approach to idiopathic cervical dystonia rehabilitation: an integrated approach to idiopathic cervical dystonia.** *Mov Disord* 2006, **21:**2240–2243.
16. El-Bahrawy MN, El-Tamawy MS, Shalaby NM, Abdel-Alim AM: **Cervical dystonia: Abnormal head posture and its relation to hand function.** *Egypt J Neurol, Psychiatr Neurosurg* 2009, **46:**203–208.
17. Queiroz MA, Chien HF, Sekeff-Sallem FA, Barbosa ER: **Physical therapy program for cervical dystonia:a study of 20 cases.** *Funct Neurol* 2012, **27:**187–192.
18. Bleton J-P: **Physiotherapy of focal dystonia: A physiotherapist's personal experience.** *Eur J Neurol* 2010, **17**(SUPPL 1):107–112.
19. Shumway-Cook A, Woollacott MH: *Motor control, translating research into clinical practice.* 4th edition. Wolters Kluwer Lippincott Williams & Wilkins; 2010.
20. Kleim JA, Jones TA: **Principles of expierence dependent neural plasticity: Implications for rehabilitation after brain damage.** *J Speech Lang Hear Res* 2008, **51:**s225–s239.
21. AGREE next step consortium: **AGREE II.** *Appraisal of Guidelines for Research & Evaluation (AGREE) Instrument* 2001. www.agreetrust.org.
22. Tijssen MAJ, Dool Van Den J, Hillten Van JJ, Warrenburg Van Der BPC: *DystonieNet.* http://dystonienet.nl.
23. Consky ES, Lang AE: **Clinical assessments of patients with cervical dystonia**. In *Therapy with botulinum toxin.* Edited by Jankovic J, Hallett M. New York: Marcel Dekker; 1994:pp. 211–37.
24. Comella CL, Stebbins GT, Goetz CG, Chmura TA, Bressman SB, Lang AE: **Teaching tape for the motor section of the Toronto Western Spasmodic Torticollis Scale.** *Mov Disord* 1997, **12:**570–575.

25. Jahanshahi M, Marsden CD: **Body concept, disability and depression in patients with spasmodic torticollis.** *Behav Neurol* 1990, **3:**117–131.
26. Tsui JK, Stoessl AJ, Eisen E, Calne S: **Double blind study of botulin toxin in spasmodic torticollis.** *Lancet* 1986, **2**(8501):245–246.
27. Guy W: **Clinical global impression**. In *ECDEU Assessment Manual for Psychopharmacology, revised.* Edited by Guy W. Rockville: MI: National Insitute of Mental Health; 1976:217–222.
28. Zaider TI, Heimberg RG, Fresco DM, Schneier FR, Liebowitz MR: **Evaluation of the clinical global impression scale among individuals with social anxiety disorder.** *Psychol Med* 2003, **33:**611–622.
29. Capuano-Pucci D, Rheault W, Aukai J: **Intratester and intertester reliability of the cervical range of motion device.** *Arch Phys Med Rehab* 1991, **72:**340.
30. Breivik EK, Bjornsson GA, Skovlund E: **A comparison of pain rating scales by sampling from clinical trial data.** *Clin J Pain* 2000, **16:**22–28.
31. Lundeberg T, Lund I, Dahlin L, Borg E, Gustafsson C, Sanda L, *et al*: **Reliability and responsiveness of three different pain assesments.** *In J Rehabil Med* 2001, **33:**279–283.
32. Muller J, Wissel J, Kemmler G, Voller B: **Craniocervical dystonia questionnaire (CDQ-24):development and validation of a disease-specific quality of life instrument.** *J Neurol Neurosurg Psychiatry* 2004, **75:**749–753.
33. Brazier JE, Harper R, Jones NMB, O'Cathain AThomas KJ, Usherwood T, Westlake L: **Validating the SF-36 health survey questionnaire: new outcome measure for primary care.** *BMJ* 1992, **305:**160–164.
34. Müller J, Kemmler G, Wissel J: **The impact of blepharospasm and cervical dystonia on health-related quality of life and depression.** *J Neurol* 2002, **249:**842–846.
35. Beck TA, Epstein N, Brown G, Steer RA: **An inventory to measure clinical anxiety: psychometric proportions.** *J Consult Clin Psychol* 1988, **56:**893–897.
36. Beck TA, Steer RA, Garbin MG: **Psychometric proportions of the Beck Depression Inventory: twenty five years of evaluation.** *Clin Psychol Rev* 1988, **8:**77–100.
37. Fydrich T, Dowdall D, Chambless DL: **Reliability and validity of the Beck Anxiety Inventory.** *J Anxiety Disord* 1992, **6:**55–61.
38. Bouwmans C, Hakkaart-van Roijen L, Koopmanschap M, Krol M, Severens H, Brouwer W: *Productivity Cost Questionnaire.* Erasmus University Rotterdam: Institute for Medical Technology Assesment; 2013.
39. Hakkaart-van Roijen L, Tan SS, Bouwmans CAM: *Handleiding voor kostenonderzoek: Methoden en standaard kostprijzen voor economische evaluaties in de gezondheidszorg College voor zorgverzekeringen.* Erasmus University Rotterdam: Institute for Medical Technology Assesment; 2010.
40. EuroQoL WorkGroup: **EuroQol–a new facility for the measurement of healthrelated quality of life.** *Health Policy* 1990, **16:**199–208.
41. Brans JWM, Lindeboom R, Aramideh M, Speelman JD: **Longterm effects of botulinum toxin on impairment and functional health in cervical dystonia.** *Neurology* 1998, **50:**1461–1463.
42. Brans JW, Lindeboom R, Snoek JW, Zwarts MJ, van Weerden TW, Brunt ER, *et al*: **Botulinum toxin versus trihexyphenidyl in cervical dystonia: a prospective, randomized, double-blind controlled trial.** *Neurology* 1996, **46:**1066–1072.
43. de Morree JJ: *Dynamiek van het menselijk bindweefsel: functie beschadiging en herstel.* Houten: Bohn Stafleu Van Loghum; 2001.
44. Fung YC: *Biomechanics. Mechanical properties of living tissues.* New York: Springer Verlag; 1993.
45. Shea JB, Morgan RL: **Contextual interference effects on the acquisition, retention and transfer of a motor skill.** *In J Exp Psychol* 1979, **5:**179–187.
46. Schmidt RA, Lee TD: *Motor control and learning: a behavioral emphasis.* Champaign, IL: Human Kinetics; 2005.
47. Shea CH, Shebilske WL, Worchel S: *Motor learning and control.* Englewood Cliffs, NJ: Prentice-Hall; 1993.
48. Fitts PM, Posner MI: *Human performance.* Belmont CA: Brooks/Cole; 1967.

doi:10.1186/1471-2377-13-85
Cite this article as: van den Dool *et al.*: Cervical dystonia: effectiveness of a standardized physical therapy program; study design and protocol of a single blind randomized controlled trial. *BMC Neurology* 2013 **13**:85.

Submit your next manuscript to BioMed Central and take full advantage of:

- **Convenient online submission**
- **Thorough peer review**
- **No space constraints or color figure charges**
- **Immediate publication on acceptance**
- **Inclusion in PubMed, CAS, Scopus and Google Scholar**
- **Research which is freely available for redistribution**

Submit your manuscript at
www.biomedcentral.com/submit

BioMed Central