Cervical Dystonia: Disease Profile and Clinical Management

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Cervical dystonia, the most common focal dystonia, frequently results in cervical pain and disability as well as impairments affecting postural control. The predominant treatment for cervical dystonia is provided by physicians, and treatment can vary from pharmacological to surgical. Little literature examining more conservative approaches, such as physical therapy, exists. This article reviews the etiology and pathophysiology of the disease as well as medical and physical therapist management for people with cervical dystonia.

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Cervical dystonia (CD) is the most frequently occurring focal dystonia, with an incidence of 8.9 per 100,000 people.[1] This condition often results in cervical pain and disability as well as impairments affecting postural control. The purpose of this report is to review the clinical characteristics of CD as well as medical and physical therapist management for patients with CD.

Background and Demographics

Dystonia is a condition that results in sustained, involuntary muscle contractions, which often cause twisting or repetitive movements or abnormal postures.[2] Dystonia can affect any voluntary muscle, and the condition is rare and often misdiagnosed.[3] Dystonia involving the neck muscles is called "cervical dystonia." However, this condition also is commonly referred to as "torticollis" or "spasmodic torticollis." These terms can be misleading, because "torticollis" implies an impairment that is purely rotatory, whereas patients often have combined postures associated with flexion, extension, or side bending. Additionally, the term "spasmodic" is applied to describe movements that are intermittent or clonic and tremulous.[4]

Epidemiological studies have shown ratios of occurrence in men to women of 1:1.4 to 1:2.2 and mean ages of onset of 39.2 years for men and 42.9 years for women.[5,6] The classification of dystonia is based on signs or symptoms (focal versus generalized), age of onset (childhood versus adult), and etiology.[2] Because generalized dystonia has a younger onset and CD typically has an adult onset, classification is based on etiology: primary versus secondary. Patients with primary CD are considered to have an idiopathic cause, with no evidence from history, physical examination, or laboratory tests (excluding dystonia genetic studies) of a secondary cause for their symptoms.[2] Patients with secondary CD may have an abnormal birth or developmental history, exposure to drugs known to cause dystonia (tardive dystonia), or neurological illness.[7]

The progression of CD among people is variable, and the symptoms may vary within an individual. Only 10% to 20% of patients have a period of remission, and these patients often experience a recurrence within several months or years.[8,9] Additionally, symptoms typically worsen over the course of the first 5 years before stabilizing.[9] Pain is the primary cause of disability in patients with CD and occurs in two thirds to three quarters of patients.[10,11] Two thirds of patients require analgesics during their illness.[12] Jahanshahi[13] showed that stress or self-consciousness and walking/fatigue/carrying objects increased symptoms in 80% and 70% of people, respectively.

"Sensory tricks," also known as "geste antagonistique," often can reduce dystonic postures or movements. Patients often can relieve symptoms by touching their chin, the top of the head, or the back of the head. In the study by Jahanshahi,[13] 88.9% of patients were able to maintain a midline cervical posture by using sensory cues. However, no statistical analysis was performed on the data. Some patients have reported reduced dystonic spasms and abnormal head positioning by simply thinking of a sensory trick.[14] Finally, Krack et al[15] fitted patients with a mechanical device that provided constant contact to the occiput and shoulders; all 5 patients reported reduced spasms of posterior cervical muscles (retrocollic posturing/cervical extension) during gait. The physiology of these maneuvers is unknown.[16]

Wissel and colleagues[17] examined the temporal pattern of activation of 6 cervical muscles by using electromyography (EMG) (surface electrodes placed over the sternocleidomastoid [SCM] muscle and needle electrodes placed into the muscle bellies of the splenius capitis muscles bilaterally) in 25 patients with CD. The patients demonstrated a significant reduction in head deviation (scored as described by Tsui et al[18]) while performing an antagonistic gesture. The authors[17] found that 52% of the patients had a reduction in EMG activity during initial arm movement, prior to the fingers touching the facial target area. The remaining 48% of the patients required facial-finger contact for a reduction in muscle activation. However, the EMG data were not statistically analyzed. Altering sensory input and activating other motor programs to adjust for dystonic posturing may have been responsible for the results. The authors[17] suggested that different physiological mechanisms may affect what appear to be clinically indistinguishable sensory maneuvers.

The role of altered 1a-spindle afferent inputs in modifying the proprioceptive input of arm muscles, proximal shoulder muscles, or both may trigger a reduction in dystonic activity at the spinal, brain stem, or subcortical levels.[19] Investigators[20,21] showed that the application of vibration to the neck muscles to stimulate the Ia-spindle afferent fibers provided temporary relief of dystonic posturing. However, the delay in a change in involuntary head movements in response to a vibration stimulus indicated the involvement of centrally controlled mechanisms in response to altered peripheral proprioceptive inputs.[21]

Patients with CD often demonstrate deficits in postural control. Investigators have reported changes in vestib-

ular function and perception of body orientation, such as asymmetries in the vestibulo-ocular reflex (VOR),[22,23] vestibular hyperreactivity,[24] difficulty recognizing postural and visual vertical,[25,26] and abnormal postural responses to the application of a vibration stimulus to the neck muscles.[27] Moreau and colleagues[28] examined body sway in 10 patients with CD and age-matched control subjects during static conditions with eyes open and closed as well as during dynamic testing of lateral and anterior or posterior displacement with a rocking platform. Dynamic tests were repeated 6 weeks following injection of botulinum toxin A (BtA), when head posture was significantly improved. Patients with CD demonstrated significantly greater deficits than control subjects only in the lateral sway component of dynamic activities before and after injection.[28] Eye closure significantly increased dynamic balance parameters (lateral and anterior or posterior sway) in both groups. However, the effect was greater in the CD group, particularly for lateral sway.[28] The authors[28] concluded that patients with CD had a greater reliance on vision for maintaining postural stability.

Whether these postural control abnormalities are a cause or a consequence of CD is unclear. Colebatch et al[29] found that abnormal vestibulocollic reflexes were more commonly present in patients with CD with a duration of greater than 5 years and concluded that changes were more likely to be compensatory than causal. However, Stell et al[30] reported an asymmetric response of the VOR that persisted after head position improved following botulinum toxin injections in patients with CD. Additional studies[23,31] have determined that abnormalities of the VOR are unrelated to head posture and have suggested primary involvement of the vestibular system. No studies regarding physical therapist examination or treatment of postural control deficits in this patient population have been performed.

Pathophysiology

In most cases, the anatomic origin of symptoms is unknown. Patients with dystonia have persistent co-contraction of agonists and antagonists.[2] Dystonia was initially thought to be a result of impaired reciprocal inhibition, represented by multiple levels of the central nervous system producing inhibition of a muscle when the antagonist is activated.[32,33] Reciprocal inhibition is reduced in patients with dystonia, and patients often have reduced inhibition in asymptomatic limbs, suggesting a more widespread abnormality.[34] Vibration of sensory afferents can produce action dystonia (dystonia that occurs only during voluntary movement),[35] and blockage of gamma and Ia-spindle afferent fibers reduces action dystonia.[34] Feiwell et al[36] demonstrated that patients with cranial dystonia affecting the orbicular muscle of the eye and the oromandibular muscles had abnormal sensory processing in the primary sensorimotor area in response to a vibration stimulus. Abnormalities of the blink reflex recovery curve[37] and Hoffmann reflex recovery curve[38,39] suggested a reduction in spinal cord and brainstem inhibition in patients with torticollis and generalized dystonia.

Using a soleus muscle Hoffmann reflex EMG method, Sabbahi and colleagues[40] noted neurophysiologic differences in people with generalized dystonia, people with CD, and subjects who were healthy. However, studies in which computed tomography and magnetic resonance imaging were used demonstrated involvement of the caudate or putamen in patients with torticollis, hemidystonia, or generalized dystonia.[41,42] Studies in which positron emission tomography was used suggested a possible role of D_1 and D_2 dopamine receptors in the putamen,[43,44] but a subsequent study[45] questioned the role of the receptors in the striatothalamic pathway.

Etiology

The pathogenesis of CD is unclear. Some cases of genetic mutations have been identified,[46,47] but these mutations typically result in generalized dystonia rather than focal CD. In patients with dystonia primarily in the cervical region, the presentation consists of persistent co-contraction of the agonists and antagonists. A history of head or neck trauma is present in 5% to 21% of patients with CD, and the traumatic event may or may not be the "trigger" for a patient's dystonia.[11,48–50] The onset of dystonia may occur immediately following a traumatic event or up to 12 months following an injury.[51] Furthermore, the injury may be mild or attributable to chronic, repetitive stress or to a single traumatic event.[52]

Differential Diagnosis

Most focal dystonias are idiopathic, although there are other secondary, symptomatic focal dystonias.[53] It is important to distinguish between the idiopathic and symptomatic forms to adequately determine prognosis and course of treatment. In patients with adult-onset focal dystonia that is presumed to be idiopathic, laboratory and neuroimaging tests are not useful because a specific cause is rarely found. Details of the onset, distribution, and clinical characteristics are useful.[53] The most common type of symptomatic adult-onset focal dystonia is tardive dystonia, which can be induced by long-term neuroleptic medication administration as well as the use of antiemetic agents or some antivertiginous agents.[54,55] As part of a patient history, patients should be questioned regarding the use of these medications. Patients with tardive dystonia are less responsive to treat-

ment than those with idiopathic dystonia.[56–58]

Patients with idiopathic CD typically report a gradual onset of symptoms. This differs from posttraumatic dystonia, in which there is a history of recent trauma in the same bodily region as the focal dystonia.[59,60] Patients with posttraumatic dystonia differ from those with idiopathic CD, as they have a marked limitation of range of motion, absence of geste antagonistique, and lack of improvement after sleep.[52,61]

In idiopathic dystonia, the only abnormal neurological finding is the presence of dystonic postures and movements. Torticollis can accompany degenerative parkinsonian syndromes.[62] However, patients would likely demonstrate additional neurological signs and symptoms.

If a patient is under 40 years of age, tests for Wilson disease should be performed.[53] This is an inherited disorder that results in copper accumulation in the body. Patients with this disease can have dystonia, and the condition can be treated with medication and diet modification.[63–66]

Examination

An examination by a physician or physical therapist should include palpation and postural evaluation to determine which muscles are overactive and contributing to improper alignment. The patient's responsiveness to geste antagonistique (sensory cues) should be determined. Cervical active and passive range of motion should be assessed prior to and subsequent to the use of sensory cues. A physical therapist examination also should include an assessment of muscle length, strength (force-generating capacity), and preferred movement patterns of the shoulder girdle and thoracic and lumbar spine to determine appropriate patient education and home exercises.

The examination also may include a commonly used outcome measure, the Toronto Western Spasmodic Torticollis Rating Scale (TWSTRS) (Appendix 1).[67] This scale includes an assessment of the dystonic position of the head, neck, and shoulders, the effectiveness of sensory cues, the length of time the patient can keep the head in the midline, and the range of head and neck movements. The scale includes a global outcome score (total score) and scores on subscales for severity (0–35), disability (0–30), and pain (0–20), with higher scores indicating greater impairment. In a study by Comella and colleagues,[68] this scale showed good interrater reliability (Spearman rank coefficient [r_s]) (.76–.98) across all subscales. However, Salvia et al[69] found excellent reliability for the total score (.99) and for the disability and pain subscale scores (.88) but greater variation for the severity subscale score (.37–.98). Additionally, Lindeboom et al[70] reported that the disability subscale may be more sensitive to change than the impairment-related subscales (pain and severity).

The scale described by Tsui et al[18] (Appendix 2) is a commonly used measure of head and shoulder positions, duration of sustained movements, and head tremor in patients with CD. This measure includes scores from 0 to 25, with lower scores indicating a lower degree of severity. Interrater reliability (Pearson *r*) has been demonstrated to be .86[67]; however, no studies have examined the responsiveness or validity of the scale.

A more recent outcome measure, the Cervical Dystonia Impact Profile (CDIP-58),[71] was developed in 2004 and found to yield reliable and valid data. It is a 58-item measure of the health impact of CD and has 8 subscales (head and neck symptoms, pain and discomfort, upper-limb activities, walking, sleep, annoyance, mood, and psychosocial functioning). Cano et al[72] examined the responsiveness of this scale in patients with CD following botulinum toxin injections. The CDIP-58 was found to be more responsive in detecting statistical and clinical changes than the Medical Outcomes 36-Item Health Survey Questionnaire, the Functional Disability Questionnaire, and the pain and disability subscales of the TWSTRS. However, this scale has not been widely used as an outcome measure, as indicated in the current literature. Additionally, the authors[72] suggested further studies examining the responsiveness of the CDIP-58 as well as refinement of the walking subscale. Prior to the development of the CDIP-58, Cano et al[72] recommended the TWSTRS as the outcome measure of choice.[73]

Treatment

Current treatment for CD is provided predominantly by physicians, and treatment can vary from pharmacological to surgical. There is a paucity of literature examining more conservative approaches, such as physical therapy. The medical (pharmacological and surgical) and physical therapy management of CD is discussed below.

Medical Management—Pharmacologic

Oral medications. Oral medications provide only modest symptomatic benefit in the treatment of dystonia.[74] For cases of tardive CD, caused by exposure to dopamine receptor antagonist medications, the treatment is the avoidance of the causative agents.[7] Medications used in low doses, such as benzodiazepines, baclofen, or anticholinergic agents, may be useful in the early stages of CD.[27,75] Greene and colleagues[75] reported better outcomes

in patients receiving anticholinergic agents (50% reporting a good response) than in those receiving clonazepam (21%), baclofen (11%), or benzodiazepines (13%). However, higher doses of these medications in later stages often cannot be tolerated because of side effects (dry mouth, cognitive disturbance, drowsiness, diplopia, glaucoma, and urinary retention). Initial studies may support the use of tetrabenazine, a presynaptic catecholamine-depleting agent, in combination with lithium.[76] The use of clozapine may be beneficial in treating tardive dystonia but has not been effective in treating idiopathic dystonia.[77,78]

Botulinum toxin. The most commonly used pharmacotherapy is BtA injections into overactive, dystonic cervical musculature.[79] Botulinum toxin A is currently thought to be the most effective medical treatment for CD—more effective than oral medications.[80] Botulinum toxin A provides graded, reversible denervation of the neuromuscular junction by preventing the release of acetylcholine from the presynaptic axon of the motor end plate. Two preparations of BtA (Dysport* and Botox†) are available. Three units of Dysport is approximately equal to 1 unit of Botox.[81] Physicians performing injections must provide a dose of sufficient quantity to weaken the dystonic muscles but minimize diffusion into adjacent, uninvolved muscles. Injections should be performed with EMG guidance to improve precision and to ensure the use of the lowest dose for the longest dosing interval.[82] An optimal initiation dose of 500 units (Dysport) is recommended for patients with CD.[83] The average duration of benefit from botulinum toxin injections is 12 to 16 weeks.[7,84]

The most common side effects include injection site pain (5% to 28%), dysphagia because of spread to adjacent muscles (11% to 40%), dry mouth (3% to 33%), excessive weakness of injected or adjacent muscles (0% to 56%), and fatigue (3% to 17%).[83,85–87] Less common adverse events include generalized weakness without objective signs of weakness, malaise, and headache.[7]

A recent randomized, double-blind, placebo-controlled trial[88] was performed with 80 patients receiving 500 units of Dysport versus a placebo. Patients were observed from baseline to 20 weeks postinjection. Truong et al[88] found that Dysport was significantly more effective than the placebo at weeks 4, 9, and 12, as demonstrated by improvements in TWSTRS scores. Two studies examining safety following repeated injections showed that patients continued to demonstrate a statistically significant benefit after receiving repeated injections for up to 10 years.[89,90]

Botulinum toxin B (BtB) often is used when patients become resistant to BtA. Multiple studies[88,91,92] have shown the efficacy of BtA or BtB, as measured by patient self-report rating scales, a reduction in TWSTRS scores, patient self-report, or a combination of these factors (Appendix 3). A recent systematic review of the literature on BtB via a meta-analysis indicated that injections were safe and efficacious in single doses but that more studies pertaining to long-term follow-up are needed.[84] No studies have compared physical therapy intervention with the use of botulinum toxin, nor has physical therapy been studied as an adjunct to injections.

Intrathecal baclofen. The final pharmacological intervention involves the use of intrathecal baclofen (ITB) at a high cervical level. Intrathecal baclofen has been used most commonly for the treatment of hypertonicity in various clinical populations, in whom it has shown good success.[93–96] Albright et al[97] demonstrated that ITB was effective in treating generalized dystonia, particularly when the catheter was placed above T4. Seventy-seven subjects (mean age=14 years, range=3–42) received ITB pumps. Subjects had significantly lower dystonia scores (Barry-Albright Dystonia Scale[98]) at 3, 6, 12, and 24 months.[97] Eighty-six percent of subjects reported improved quality of life and ease of care, and 92% retained benefit at the median follow-up period of 29 months; however, the authors[97] did not statistically analyze these 2 variables. Only one study[99] has described the use of ITB in patients with CD via 2 case reports. Dykstra and colleagues[99] reported a reduction in TWSTRS scores and improved 3-dimensional electromagnetically measured cervical range of motion following continuous infusions of ITB at high cervical (C1–C3) levels.

Medical Management—Surgical

Surgical intervention is used when a patient has not responded to pharmacological or other interventions.[74] Although a variety of surgical interventions have been studied, the most commonly used approaches at this time are selective peripheral denervation and deep-brain stimulation (DBS).

Selective peripheral denervation. This surgical approach denervates muscles responsible for abnormal movements and preserves innervation to muscles that are noncontributory. The surgery was pioneered in 1891 by Keen and was further refined by Bertrand in 1993. Bertrand assessed the surgical outcomes of 260 subjects and reported an 88% success rate based on a 4-point scale (poor, fair, very good, or excellent) evaluating the presence or absence

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of detectable abnormal movements.[100] However, no statistical analysis of the data was performed, nor was any information on the reliability and validity of the 4-point classification scale provided. Muscles selected for denervation should be chosen on the basis of an examination of abnormal movements and confirmation with EMG recording. For patients with laterocollis, denervation of the ipsilateral posterior cervical paraspinal, splenius capitis, and SCM muscles is performed. For patients with rotatory torticollis, the same procedure is performed, but denervation involves the contralateral SCM muscle.[101] Clinical symptoms should be stable for at least 1 year before surgery is considered. Furthermore, patients with pure rotatory torticollis with mild extension show the best postoperative results, whereas patients with preexisting fibrosis or severe arthrosis are more likely to show poor results.[102] Adverse effects associated with the surgery may include wound infection (3%), transient balance problems (9%), transient dysesthesia or sensory loss in denervated posterior cervical segments (21%), and dysphagia (36%).[103]

Prospective and retrospective studies have demonstrated a reduction in TWSTRS scores, a reduction in pain, an improvement in head position, an improvement in quality of life, or a combination of these effects.[101,103] In a retrospective study, Cohen-Gadol and colleagues[101] found that head position and pain improved in 77% and 81% of 162 patients, respectively, at 3 months. Long-term follow-up was performed for 130 patients at a mean duration of 3.4 years. Seventy percent of these patients continued to report improved head position and pain. However, the outcome measures used were subjective and have not been validated, and an adequate statistical analysis was not performed. A prospective study by Munchau et al[103] demonstrated a significant reduction in total TWSTRS scores at 6 and 12 months in 40 patients with CD. Subscores for severity, disability, and pain were significantly reduced by 20%, 30%, and 40%, respectively, at 6 months and by 20%, 40%, and 30%, respectively, at 12 months. In both of these studies, physical therapy was initiated while the patients were hospitalized and continued on an outpatient basis for 2 to 3 months to "strengthen and/or retrain the muscles uninvolved in dystonia that are necessary for full neck movement."[101(p1250)] However, no specific information detailing the exercises typically prescribed was provided.

DBS. Before DBS surgery became available, thalamotomy and pallidotomy surgeries were often performed.[104,105] Thalamotomy is now rarely performed because of the potential for serious adverse effects. Most patients with CD required bilateral surgery, raising the risk of speech and swallowing problems.[7] Providing localized brain stimulation to the basal ganglia and thalamus resulted in a reduction in tremor in patients with Parkinson disease.[106,107] As a result, stereotactic surgery was used to treat dystonia, although the precise mechanism of action of DBS remains uncertain.

More recently, the use of DBS of the globus pallidus internus (GPi) or the subthalamic nucleus (STN) was implemented in patients with intractable CD.[108] Surgical intervention involving the GPi began in the 1990s and is more common than stimulation of the STN in this population.[104] Most studies reported surgery involving the GPi, and DBS involving the STN was limited to case reports.[109] The surgery involved the placement of microelectrodes into the GPi, typically bilaterally, with identification of the GPi and guidance of the microelectrode placement by microstimulation. In some studies,[109–111] microelectric recording and magnetic resonance imaging were used to guide electrode placement. Once the surgery was performed, multiple visits were required to properly program the settings for the stimulator. The advantages of DBS include the reversibility of the procedure, the ability to adjust the stimulation parameters, and continued access to the therapeutic target. Krauss and colleagues[110] performed DBS with microstimulation guidance on 8 patients and reported the following adverse events: infection (1 subject), lead fractures (3 events in 2 subjects), battery failure (2 subjects), and perioral tightness during DBS adjustment visits (6 subjects).

Prospective studies have shown a reduction in TWSTRS scores[108,110,112] and a reduction in the use of oral medications[110] up to the mean study duration of 20 months to 2 years. Yianni et al[112] reported a 59.5% improvement in the TWSTRS global score and in all subscale scores (severity=63.8%, disability=60.0%, and pain=60.3%) in 6 patients observed for up to 24 months; however, the results were not analyzed for statistical significance. Stimulation was applied to the GPi. However, no details were given regarding the specific surgical procedure. Similarly, Krauss and colleagues[110] reported a statistically significant improvement in TWSTRS severity, disability, and pain subscale scores in 8 patients with CD at 20 months following surgery. No studies have examined the use of physical therapy as an adjunct to DBS surgery.

Physical Therapy Intervention

Very few investigators have examined the effects of physical therapy intervention for the treatment of adult CD. No randomized controlled trials have examined physical therapy intervention, and the studies that have been done were poorly controlled and had small sample sizes. An extensive review of the literature

revealed that few studies have examined "traditional" physical therapy approaches.

Although a few uncontrolled studies demonstrated the use of EMG biofeedback,[113–116] Smania et al[117] carried out the most controlled study, despite examining only 4 subjects. They examined the effects of 2 physical therapy programs on 4 patients with CD. They compared EMG biofeedback with a "novel" physical therapy program. This program included postural reeducation exercises to increase the voluntary control of head posture and to induce the correct perception of head-trunk alignment and passive elongation of myofascial structures to reduce soft-tissue contractures of the cervical spine. Patients received 15 sessions of EMG biofeedback and 15 sessions of the novel physical therapy program. Two subjects received EMG biofeedback first, and 2 subjects received the novel physical therapy program first. Treatment sessions lasted 60 minutes and were performed daily (Monday–Friday) for 6 weeks. The authors[117] found comparable results for EMG biofeedback and the novel physical therapy program in terms of reductions in disability (questionnaire) and in subjects' reports of pain on a visual analog scale, and these results persisted at 3 and 9 months. However, the physical therapy intervention pertaining to the exercises was not well described, nor is it clear that this intervention was applied systematically.

No physical therapy studies examining the implementation of *specific* exercises to isolate and increase activity in antagonistic muscles during dystonic posturing have been performed. Additionally, no research has assessed shoulder and spinal alignment in people with CD or prescribed exercises to improve scapular, glenohumeral, and spinal posture and associated muscle length and strength.

Transcutaneous electrical nerve stimulation has been assessed in only a single report involving a case of post-whiplash dystonia.[118] The case report by Foley-Nolan and colleagues[118] described the use of transcutaneous electrical nerve stimulation with 4 electrodes over C3 to C5 (100 Hz, 100 microseconds) in a 28-year-old patient. Transcutaneous electrical nerve stimulation was gradually decreased from 12 to 3 hours per day over the next 4 weeks. The authors[118] reported that the patient had returned to work at his 6-month follow-up. The patient's history and presentation were poorly described, and no quantitative measures were used to assess outcome.

In 1996, Davis et al[119] retrospectively attempted to contact and examine 223 patients with torticollis who were treated with iontophoresis between 1958 and 1972. Only 56 people were located and responded to a survey. Of the 56 respondents, 50% reported that they had noted initial improvement, and 51% of these people noted improvement through the time of the survey, although no statistical analysis was performed. However, 5 respondents also had received botulinum toxin injections or peripheral denervation surgery. The authors[119] concluded that the effect of iontophoresis treatment was minimal. However, the measures used for assessing improvement were poorly described and defined.

For patients without CD, little consensus exists on the treatment of chronic cervical pain. The results of 2 recent systematic reviews[120,121] indicated that there may be some benefit from exercise as well as mobilization but that 1 treatment is not superior to the other. Greater benefit may be obtained from multimodal care (exercise plus mobilization), and future studies need to be performed to examine the relative benefit of each specific mode of treatment.[120,121]

Implications for Physical Therapists

On the basis of the available literature, there are several important factors that a physical therapist should consider when examining and treating a patient with CD. First, the examination should include outcome measures that are specific to this patient population (the scale described by Tsui et al,[18] TWSTRS, and CDIP-58) and that would provide information related to a patient's impairments and function. The scale described by Tsui et al would be useful for measuring head position, whereas the TWSTRS and CDIP-58 are excellent standardized measures of pain and disability.

Second, the use of sensory tricks in physical therapy, either as patient education or as an intervention, has not been studied, despite the fact that sensory stimuli have been shown to reduce dystonic spasms and abnormal head posturing in patients with CD.[13–15,17] Physical therapists should examine the responsiveness of patients to sensory cues as well as the timing and segment of the cues that are found to be most effective. This approach would enhance patient education in terms of the techniques that a patient could use to reduce dystonic posturing.

Third, a thorough history of falls or balance deficits and examination of a patient's postural control should be included. Although no studies have identified appropriate interventions for these deficits in patients with CD, they are important factors that can substantially affect a patient's function and are important areas for future study.

A thorough examination of the patient's head position, postural alignment, muscle length and strength throughout the spine and shoulder region, and cervical range of motion is recommended. Specific exercises can be prescribed to improve impairments that exist, but no studies have been performed to assess their effectiveness in reducing pain or disability.

Although mobilization has been effective in treating cervical pain in patients without CD, it is unclear how effective this intervention would be in patients who are unable to relax or "turn off" muscles in the cervical region during a mobilization technique. Because these patients typically experience pain when lifting and carrying objects, the examination could focus on methods that patients use to perform these tasks as well as education in how to modify the tasks to reduce pain.

Summary

The current intervention for people with CD is predominantly medical or surgical. For most other neuromuscular conditions, physical therapy intervention is an adjunct to these 2 approaches. The lack of high-quality research related to physical therapy interventions for the symptoms and disability resulting from CD highlights the need and opportunity for physical therapists to contribute to the management of CD. Physical therapists commonly treat the manifestations of hypertonicity in other neuromuscular conditions and are trained to treat muscle imbalances that contribute to musculoskeletal pain. Further study is needed to investigate the effectiveness of specific therapeutic exercises, patient education, and posture reeducation approaches for people with CD.

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Appendix 1.

Toronto Western Spasmodic Torticollis Rating Scale[a]

I. Torticollis Severity Scale (maximum=35; sum of A through F)

A. Maximal Excursion: Rate the maximum amplitude of excursion by asking the patient not to oppose the abnormal movement; the examiner may use distracting or aggravating maneuvers. When the degree of deviation is between scores, choose the higher of the two.

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)

   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: Provide an overall score estimated through the course of the standardized examination after estimating the maximal excursion (exclusive of asking the patient to allow the head to deviate maximally) *(weighted × 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)

(Continued)

Appendix 1.
Continued

C. Effect of Sensory Tricks
- 0=Complete or partial 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) and 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 the aid of sensory tricks)*. If limitation occurs in more than one plane of motion, use the individual score that is highest.
- 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 (the 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=30; sum of A through F)

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 *(eg, 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

(Continued)

Appendix 1.
Continued

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 headrest) 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
0=No difficulty
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 (eg, lying down)
4=Limited ability to read because of torticollis despite tricks
5=Unable to read more than a few sentences because of torticollis

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 (eg, 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 *(eg, 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; sum of A through C)

A. Severity of Pain: Rate the severity of neck pain due to torticollis 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_____

(Continued)

Appendix 1.
Continued

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 >75% 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 the 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 head pulling

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Appendix 2.

Torticollis Rating Scale of Tsui et al[a]

I. Amplitude of sustained movements (0–9):
   A. Rotation
      0=Absent
      1=<15°
      2=15°–30°
      3=>30°
   B. Tilt
      0=Absent
      1=<15°
      2=15°–30°
      3=>30°
   C. Antero/Retrocollis
      0=Absent
      1=Mild
      2=Moderate
      3=Severe
II. Duration of sustained movements (1–2):
      1=Intermittent
      2=Constant
III. Shoulder elevation (0–3):
      0=Absent
      1=Mild and intermittent
      2=Mild and constant or severe and intermittent
      3=Severe and constant
IV. Head tremor (1–4; severity × duration):
   A. Severity
      1=Mild
      2=Severe
   B. Duration
      1=Occasional
      2=Continuous

[a] Total torticollis score=[I×II]+III+IV=1–25. Adapted with permission from Tsui JK, Eisen A, Stoessl AJ, et al. Double-blind study of botulinum toxin in spasmodic torticollis. *Lancet*. 1986;2:245–247. Copyright 1986, Elsevier.

Appendix 3.

Summary of Efficacy of Botulinum Toxin A (BtA) and Botulinum Toxin B (BtB)[a]

Category	Truong et al[88]	Lew et al[91]	Factor et al[92]
Purpose	To examine the safety and efficacy of a fixed dose of BtA (Dysport) in a multicenter, randomized, placebo-controlled trial	To compare the results of 3 recent multicenter, randomized, placebo-controlled trials examining the efficacy of BtB	To determine the effectiveness of repeated BtB injections in patients who are BtA-resistant
Subjects	n=80; mean age=53 y	Study 1: n=122 (BtA responsive and resistant); mean age=55 y Study 2: n=109 (BtA responsive); mean age=56 y Study 3: n=77 (BtA resistant); mean age=54 y	n=34 (15 BtA resistant, 19 not resistant); mean age=54 y
Injection protocol	Randomized to receive a single injection of BtA (Dysport; 500 U) or placebo; numbers of injection sites and muscles injected were individually determined by the investigators	Study 1: placebo vs BtB (NeuroBloc; 2,500, 5,000, or 10,000 U) Study 2: placebo vs BtB (5,000 or 10,000 U) Study 3: placebo vs BtB (10,000 U); injections in 2–4 muscles determined by investigators	Subjects received up to 10 repeated injections of BtB (NeuroBloc; started at 10,000 U and increased by 5,000 U as needed, with a maximum dose of 25,000 U); numbers of injection sites and muscles injected were not reported
Outcome measures	TWSTRS, pain rating (self-report on visual analog scale), and patient and investigator symptom change ratings (visual analog scale)	TWSTRS, patient analog pain assessment, and patient's and physician's global assessments of change	TWSTRS
Duration of follow-up	Baseline and 2, 4, 8, 12, 16, and 20 wk	Baseline and 4, 8, 12, and 16 wk	Baseline and 4 wk after each injection
Results	Dysport-treated subjects had significantly reduced TWSTRS scores at 4, 8, and 12 wk; positive responses seen in 38% of Dysport-treated subjects compared with 16% of placebo-treated subjects; median duration of response to Dysport=18.5 wk	In all 3 studies, BtB-treated subjects had a significant reduction in the total TWSTRS score compared with placebo-treated subjects; a significant dose response was found, favoring the subjects receiving 10,000 U; median duration of effect was 12–16 wk	TWSTRS scores were significantly decreased at 4 wk, but response magnitude decreased over time; no difference in response between subjects who were BtA-resistant and those who were not BtA-resistant

[a] TWSTRS=Toronto Western Spasmodic Torticollis Rating Scale.