Cox® Technic Treatment of Neurofibromatosis Type 1 with Cervical and Lumbar Radiculopathy
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ABSTRACT

Introduction: To demonstrate the treatment of a patient with Neurofibromatosis Type 1 with Cox Flexion-Distraction with 5 year follow-up.

Clinical Features: This case involves a 43 year old male with chronic low back pain and sciatic radiculopathy treated with Cox® Technic flexion distraction (CTFD). This case includes 5 year follow-up. Imaging confirmed neurofibromatosis type 1, 2 years after having begun CTFD. His most recent MRI revealed innumerable lesions in the cervical and lumbar regions confirming neurofibromatosis type 1. Treatment for cervical radiculopathy was started recently after symptoms developed. Improvement was measured via VAS and orthopedic examination.

Intervention and Outcome: Treatment consisted of Cox® FD. He initially started at protocol I and reached maximum improvement in approximately 4 weeks. His VAS scores were initially 8/10 and improved to a 0-2/10. Laser therapy was applied to his sacroiliac and coccygeal regions with each treatment.

Conclusion: This case demonstrates successful management and treatment of a patient with Neurofibromatosis Type 1 using Cox® Technic.

Key Words: Neurofibromatosis Type 1, Dural Ectasia, Cox Technic, Flexion/Distraction, Chiropractic, Manual Therapy, Physical Therapy.

INTRODUCTION:
Dural ectasia (dural dysplasia) is defined as a ballooning (dilatation) of the dural sac, bony erosions with spinal canal widening, pedicle erosion, foraminal enlargement and scalloping of the posterior vertebral bodies or sacrum. It is most commonly found in patients with connective tissue disorders such as Marfan syndrome, neurofibromatosis type I (NF1), Ehlers Danlos syndrome (EDS), osteogenesis imperfecta and ankylosing spondylitis. Several studies have shown that dural ectasia may be associated with such conditions as back pain, headaches, radiculopathies, or incontinence. A "classic" picture of dural ectasia may consist of low back pain, headache, proximal leg pain, weakness and numbness above and below the knee, and genital/rectal pain. Symptoms, when present, are typically moderate to severe, occur several times per week (often daily), are commonly exacerbated by upright posture, and are not always relieved by recumbency. MRI and CT are highly specific in diagnosing dural ectasia. Plain film radiography may reveal vertebral scalloping but is not specific.

Neurofibromatosis is an inherited autosomal dominant disorder. It is the most frequent single-gene disorder affecting mankind, with up to 50% of cases due to spontaneous mutation. There are three types: neurofibromatosis type 1 (NF1), neurofibromatosis type 2 (NF2), and schwannomatosis. The cardinal and defining features of NF1 are café au lait macules (6 or more), neurofibromas, skinfold (axillary) freckling, iris Lisch nodules, and characteristic osseous dysplasia. The typical osseous dysplasias are bony erosion to the osteoligamentous canal and within the sacrum, which may lead to
intrapelvic meningoceles. NF1 affects up to 1 in 3000 individuals with an equal prevalence between men and women. Up to 10% of patients present later in life withforme fruste or atypical manifestations. Neurofibromas are benign fibroblastic neoplasms of peripheral nerve sheaths. Malignant degeneration to neurofibrosarcoma occurs in 4-11%. There are three main types of neurofibroma: cutaneous, spinal and plexiform. Intraspinal neurofibromas can provoke cord compression. The most common signs and symptoms are pain and radiculopathy due to compression of the affected nerve root. Regardless of nerve origin, imaging demonstrates greater tumour burden than patient symptoms would suggest. Annual magnetic resonance imaging (MRI) is mandatory because the tumours may suddenly and rapidly start growth cycles or become malignant. Annual neurological examination is recommended and any unexplained neurological signs or symptoms require investigation and referral to a neurologist. Urgent advice is mandatory if individuals experience acute or progressive sensory disturbance, motor deficit and incoordination or sphincter disturbance which could be indicative of an intracranial lesion or spinal cord compression. Headaches with walking, morning vomiting and altered consciousness are suggestive of raised intracranial pressure and constitute a medical emergency. Neurological complications develop from tumours and malformations, including aqueduct stenosis.

The purpose of this study is to demonstrate the use of CTFD in a patient with NF1. CTFD is evidenced based spinal manipulation that is well researched, from case studies by certified chiropractors to federally funded biomechanical and high-level randomized clinical trials.

Literature searches (MeSH, Pubmed, ResearchGate, Core) involved the following terms: Neurofibromatosis Type 1, dural ectasia, Cox Technic, flexion/distraction, chiropractic, manual therapy, and physical therapy.

CASE REPORT

History:
A current patient for the past seven years with a history of chronic low back pain presented with pain that radiated into the sacroiliac and coccyx regions bilaterally as well as with bilateral sciatic radiculopathy more pronounced on the right side and into the right foot. He had chronic low back pain that was initially treated with diversified adjusting and laser therapy. After approximately 18 months he could no longer tolerate manipulation. He only received laser therapy for pain management and exercise instruction which kept his pain level at a 1 or 2/10 on the VAS. When he presented with sciatic radiculopathy, his VAS score was 8/10. He complained of a frequent urge to urinate that may have precipitated the recent complaint. He did not complain of recurrent headaches. At the time he had been a delivery driver for a garment and linen company which involved frequent bending and lifting along with jumping in and out of the vehicle.

Examination:
Examination revealed mild left antalgia. Range of motion testing was decreased in extension (10°) and right lateral flexion (15°). Deep tendon reflexes and lower extremity muscle strength testing were unremarkable. He had paresthesia in the L5 dermatome on the right side only. Bilateral SLR was normal. Positive Goldthwaite on the right (50°). Kemp’s was positive bilaterally. Palpable tenderness and hypertonicity of erector spinae muscles from L4 to the sacrum, piriformis, obturator internus, gemelli, hamstring, gastrocnemius and soleus muscles. Sacroiliac joint testing was normal. Heel-toe walk was normal. Romberg’s test was also normal. He uses Celebrex as needed for pain.
**Diagnosis:**
A diagnosis of a central disc protrusion at L5/S1 was made and Cox FD protocol 1 was performed with an L2 contact based on Tolerance Testing. A request was made to his family physician for lumbosacral x-rays and CT/MRI.

**Imaging:**
X-ray imaging revealed limbus vertebrae at L4 and L5 and enlargement of the osteoligamentous canals of the lumbar spine (Figure 1).

![Figure 1. Lumbar Radiographs. Limbus vertebrae and enlarged osteoligamentous canals.](image)

CT imaging of the lumbosacral spine revealed extensive dural ectasia of L3-L5 nerve roots as well as the sacral nerve roots up to S2 level involving dilatation of the neural foramina. No evidence of disc bulge or prolapse in the lumbar spine (Figure 2).
Upon discovering the dural ectasia the patient was evaluated for joint hypermobility using the Beighton and Brighton methods which is used in the evaluation of Marfan Syndrome. He did not display any hypermobility in any joints. A request was made to his urologist to correlate his urinary frequency with his dural ectasia by means of an MRI.
Figure 4. Thoracolumbar MRI. Enlarged nerve root sheaths (white outlined arrows), intrapelvic meningoceles anterior to sacrum and posterior to rectum (solid black arrows), scalloped vertebrae (white arrows).
Figure 3. Cervical MRI. Dumbbell tumor (solid white arrow), intrathecal neurofibromas (white outlined arrows), cord compression by lesion at level of atlas and lower cervical spine (solid black arrows), enlarged nerve root sheaths (black outlined arrows).

**Treatment Plan:**
He was treated three times a week for 4 weeks and returned to his previous pain level. Initially he reported significant improvement in pain and radiculopathy within 8 visits. After the fourth week, he had an exacerbation while working due to reaching and lifting bags of linen. He continued treatment at the same frequency for 4 more weeks to return to maximum improvement. For the past two years his pain level has been kept between 0-2/10 on VAS. Recent MRI revealed multiple “innumerable” lesions in the cervical and lumbar regions with sparing of the thoracic spine. A diagnosis of NF1 was made by a neurologist.

**Outcome:**
Two months ago he complained of numbness and tingling in his hands at night and occasionally when driving. He did not complain of neck pain or headache. Surface electromyography of the wrists showed bilateral neurofibromas of the median nerves. He was treated with foramen magnum pump alongside protocol II for the lumbar spine. His hand and wrist symptoms diminished with one treatment but have
not resolved. He currently receives protocol II treatment to the full spine with foramen magnum pump, five minutes of autodistraction with the cervical and ankle harnesses, and a locked headpiece. He continues to receive laser treatment bilaterally at the fifth lumbar and sacroiliac joints at his request. He has had to make a change in his employment as he can no longer do work that involves lifting.

**DISCUSSION:**
The current study is similar to what most chiropractors experience with low back pain and neck pain patients in the general population. The treatment provided was based on the patient’s history, clinical examination and tolerance to treatment, both within and between sessions. Imaging was not a significant factor in determining his course of treatment. In both episodes of sciatic radiculopathy, the patient improved to pre-symptomatic pain levels within 8 treatments over four weeks. His cervical pain and radiculopathy improved to pre-symptomatic levels with just two treatments.

There is no consistent or widely accepted pattern of care for treating NF1 and reference to similar studies is limited. Kitchen et al employed spinal manipulation and postural counselling on a 31 year old male and an 8 year old male with NF1. Gajeski et al demonstrated a marked reduction in pain and paresthesia with improved function using lumbosacral manipulation, interferential therapy, heat and rehabilitative exercise in a 45 year old woman with nine months of treatment. Lennington et al discussed an adverse reaction to manipulation of the cervical spine in a patient with NF1 but the authors were speculative as to the actual mechanism of injury claimed by the patient. Colloca and Polkinghorn reported a reduction in pain medication use with chiropractic manipulation in two patients with EDS. Morley and Perrealt also showed a reduction in pain levels over 10 months of treatment for hypermobile joint pain in a 20 year old woman with EDS. Within thirteen weeks, Helmers and Irwin used a combination of traction, distraction, ultrasound, interferential current, massage, and exercise and postural training in a 17 year old female with a sixteen year history of cervical pain and headache. In this study, manual cervical traction was considered as conservative to manipulation.

Federally funded research has shown that CTFD applied to the spine:
1. Decreases intradiscal pressure in the lumbar spine up to -192mmHg.
2. Decreases intradiscal pressure in the cervical spine up to -502mmHg.
3. Increases intervertebral disc height.
4. Increases intervertebral foraminal area up to 28%.

CTFD is an evidence based non-surgical, chiropractic spinal manipulation procedure that is applied using the Cox® Table by a certified practitioner. Tolerance testing is both mandated and expected with treatment. Tolerance testing provides a tailored approach for every treatment session and acts as a gauge for the patient’s sensitivity to treatment at every visit. Recent studies have shown that Cox® Technic certified doctors are able to deliver treatment within the prescribed traction force range with and without visible aids.

**STUDY LIMITATIONS:**
Case studies are typically limited by bias. However, where randomized controlled trials reduce subjectivity, case studies incorporate subjectivity and provide a holistic account of the phenomenon. One drawback of case studies is a lack of empirical materials. The current study only uses VAS as a measured outcome. However, this is balanced by the complexity of NF1. The limitation of objectivity in this study is balanced by the qualitative application of treatment, specifically CTFD and it’s protocols. As a case study, the reader decides what can apply to his or her context.
CONCLUSION:
The current study demonstrates that CTFD can be used to treat NF1 patients with low back and neck pain, and associated radiculopathy.

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References: