

Chiropractic Care Of A Patient With Crouzon Syndrome And Multiple Congenital Anomalies At The Cervical Spine: A Case Report

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Citation

L Henry. *Chiropractic Care Of A Patient With Crouzon Syndrome And Multiple Congenital Anomalies At The Cervical Spine: A Case Report*. The Internet Journal of Chiropractic. 2013 Volume 3 Number 1.

Abstract

Objective: The objective of this case report is to describe the chiropractic care of a patient with an unusual condition: Crouzon syndrome and fusion from occiput to C3.

Introduction: Crouzon syndrome is a genetic disorder that includes craniosynostosis, the premature closure of the cranial sutures, and affects the shape of the head and face. Cervical spine anomalies are common in patients with Crouzon syndrome. The author was unable to find any previous reports regarding chiropractic care of patients with Crouzon syndrome.

Intervention and Outcome: A 24-year-old male with Crouzon syndrome presented with neck and upper back and shoulder pain, accompanied by frequent numbness at the arms and hands. He had prior history of craniofacial surgery for Crouzon syndrome. He was evaluated by physical examination and radiographs of the cervical spine. He was diagnosed with vertebral subluxation (segmental dysfunction) of the cervical spine producing cervicgia complicated by congenital fusion of the spine from the occiput to C3. Treatment consisted of spinal manipulation, physiotherapy modalities and therapeutic exercises. The patient's spinal pain resolved following chiropractic care. The patient was subsequently referred for neurology consultation and to a surgeon for carpal tunnel syndrome due to work-related repetitive stress injury.

Discussion: The benefits of spinal manipulation for neck pain and back pain are well established, however, there are no published studies on the chiropractic management of patient's with Crouzon syndrome. This patient's care was complicated by congenital fusion from occiput to C3.

Conclusion: In this case, there is an apparent resolution of spinal pain following chiropractic management of a patient with multiple spinal congenital anomalies.

INTRODUCTION

Crouzon syndrome, also known as Crouzon disease and craniofacial dysostosis, is a genetic disorder in which premature closure of the sutures prevents normal cranial development, causing deformity of the head and face. Crouzon syndrome is caused by mutation in the fibroblast growth factor receptor 2 (FGFR2) gene. The mutation is autosomal dominant when inherited or may be due to new mutation. Crouzon syndrome is the most common craniosynostosis syndrome with incidence of 16 per million newborns.¹ Characteristic appearance includes increased intraorbital space and shallow orbits with ocular proptosis, maxillary hypoplasia and beaked nose. Crouzon syndrome may result in dental problems, hearing loss, or poor vision.² Cleft lip and palate may be present. Intelligence is usually not affected. In addition to characteristic craniofacial abnormality, Crouzon syndrome may present with stylohyoid ligament calcification, cervical spine anomalies,

and deformities of the elbows, hands or viscera, or acanthosis nigricans.³

Treatment of craniosynostosis generally consists of surgery to improve facial appearance and alleviate pressure on the brain and cranial nerves. Less severe cases may be managed by cranial molds.⁴

There is an increased incidence of congenital anomalies at the cervical spine in patients with Crouzon syndrome compared with the normal population and some authors have suggested that cervical spine fusions are progressive. Proudman et al found a 40% incidence of cervical spine anomalies in Crouzon syndrome patients.³ Anderson et al found increased incidence of butterfly vertebra and congenital fusions, with an 18% incidence of block vertebra in the children treated. C2-C3 was the most common level affected, with C5-C6 fusion occurring nearly as often. The authors suggested that the incidence might be higher in

adults in cases of progressive cervical fusion. Sequential imaging studies showed progression of cervical fusion in some patients.⁵ Kreiborg et al reported 25% incidence of cervical fusion in Crouzon syndrome and stated that cervical fusion at C2-C3 versus C5-C6 can help distinguish between Crouzon syndrome and the Apert syndrome on radiograph in most cases.⁶ Hemmer et al found spinal fusions in 38% of Crouzon syndrome patients, with upper cervical fusions being most common. The level of fusion was not specific enough for differential diagnosis from other forms of craniosynostosis.⁷

Vertebral anomalies in pediatric patients are not as easy to recognize compared to abnormalities of the head and neck, which may lead to delayed diagnosis. Manaligod and colleagues suggest radiographic imaging, preferably computed tomography, to diagnose spinal anomalies in a timely manner and reduce risk of neurological injury.⁸

A literature search was conducted using the Index to Chiropractic Literature and PubMed. The keywords were “Crouzon syndrome” and “craniosynostosis” for the Index to Chiropractic Literature and “chiropractic Crouzon syndrome,” “spinal manipulation Crouzon syndrome” and “chiropractic craniosynostosis” for PubMed. There was no literature concerning chiropractic care for Crouzon syndrome although there was one relevant peer-reviewed paper: A case report by Alcantara and Doucet⁹ described the chiropractic care of a 3-week-old girl with craniosynostosis with fusion of the posterior fontanelle. The treatment consisted of gentle high-velocity, low amplitude chiropractic adjustments and craniosacral therapy. After six visits the authors reported an increase in cranial diameter and long-term follow up revealed progression in cranial development without need for surgical intervention.

INTERVENTION AND OUTCOME

An athletic 24-year-old male with Crouzon syndrome presented with neck and upper back and shoulder pain, accompanied by frequent numbness at the arms and hands. He reported an insidious onset one-year prior and stated that the symptoms had gradually worsened over the past 6 months. He denied history of trauma. He described moderate intermittent neck pain and tension, rating the pain 6/10 VAS, where 10 represents the most pain. He reported pain at the right elbow and numbness in both arms and hands. He reported that pain interfered with sleep and caused difficulty with his job working as a pizza restaurant manager, which required repetitive motions of kneading and slapping dough

and cutting.

Past medical history consisted of a motor vehicle collision 5 years prior. He also had a history of craniofacial surgery for Crouzon syndrome and, history of migraines with treatment by a neurologist who prescribed topiramate. Relevant family history indicated that his father had Crouzon syndrome. He reported a history of smoking and weight training five days per week.

The patient was 66 inches tall, weighed 128 pounds and appeared athletic. Blood pressure was 116/79, pulse 54 BPM, and respiration was 20 per minute. Cervical range of motion was restricted in extension and right rotation with pain and lateral flexion to the left and right elicited pain. Foraminal compression test was negative for radiating symptoms to the upper extremities but increased local pain at the neck. Shoulder depressor test was painful on both sides. Deep tendon reflexes and dermatomes were normal for the upper extremities. There was weakness at the right triceps (grade 4/5); upper extremity motor was otherwise normal. Eden’s and Wright’s tests were positive bilaterally, indicating thoracic outlet syndrome. Phalen’s and Tinel’s tests were negative. There was restricted motion at the right proximal ulna with tenderness at the medial elbow. Restricted motion was palpable at the right sternoclavicular joint and at the carpal joints on the right. Spasm was present at the scalenes, SCM and trapezius muscles bilaterally. The pectoral muscles were hypertonic. There was tenderness and hypertonicity at C3-C7, with asymmetry and restriction at C5 and C7-T1 and the first rib on the right. No motion was palpable at the upper cervical spine.

AP, lateral, flexion and extension radiographs were taken of the cervical spine. There was a decrease of the cervical lordosis and a right listing at the cervical spine. Basilar impression was present with fusion of the posterior elements at C2-C3 and the appearance of partial fusion from occiput to C3. Flexion and extension views were taken to rule out instability and confirmed that there was no motion from occiput to C3. There was moderate degenerative joint disease at C7-T1.

The patient was diagnosed with vertebral subluxation (segmental dysfunction) of the cervical spine producing cervicalgia, complicated by congenital fusion of the spine.

Intervention consisted of chiropractic adjustments using Diversified technique to correct cervical joint dysfunction, along with electrical stimulation, superficial heat,

intersegmental traction, and home exercises. Myofascial Disruption Technique was used to reduce myofascial trigger points. The patient was counseled on home exercises to stretch the pectoral muscles and postural exercises for the neck. He was seen three times per week for four weeks. At a re-evaluation at four weeks, the patient's neck and upper back pain improved but numbness at the hands persisted in the lateral three digits on both sides, especially on the right side. Chiropractic care continued for an additional three weeks. The care was followed by a resolution of pain at the neck, shoulders and upper back. Upper extremity manipulation resulted in temporary improvement in hand numbness. The patient's hand symptoms worsened with continued repetitive stress at job, which required kneading and slapping pizza dough and cutting pizzas. He was advised to reduce repetitive stress on the hands and wrists. He was counseled to apply ice and instructed on therapeutic exercises for carpal tunnel syndrome. When hand numbness failed to improve the patient was referred to a neurologist for electrodiagnostic testing. Nerve conduction velocity testing demonstrated evidence of bilateral distal median neuropathy, worse on the right and the patient was referred for surgical consultation for carpal tunnel syndrome.

DISCUSSION

The benefits of spinal manipulation for neck pain and back pain are well established. However, there apparently are no published studies on the chiropractic management of patients with Crouzon syndrome. The presence of multiple congenital fusions at the cervical spine produced altered biomechanics and likely predisposed this patient to premature degenerative joint disease and subsequent neck pain. Chiropractic management consisting of spinal manipulation, physiotherapy and exercise was followed by a resolution of the spinal pain.

CONCLUSION

In this case, there is an apparent resolution of spinal pain

following chiropractic care despite complication due to multiple spinal congenital anomalies. Radiographic evaluation of the spine prior to chiropractic care is recommended for patients with Crouzon syndrome due to the high incidence of vertebral anomalies.

ACKNOWLEDGEMENTS

Thank you to John Hart, DC, MHSc, Assistant Director of Research at Sherman College of Chiropractic, for his help revising this manuscript.

FUNDING SOURCES AND CONFLICTS OF INTEREST

There were no funding sources or conflicts of interest.

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