

Symptomatic Improvement of Dysautonomia and Pain in Patients with Craniovertebral Instability via Occipital Cervical Fusion

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Introduction

Joint hypermobility in patients with inherited conditions of connective tissue has recently been shown to cause excess mobility of the movement of the odontoid process in relationship to the skull base. A subset of these patients will develop distortion of the relationship of the odontoid to the brainstem leading to tonsillar herniation, severe headaches and mechanical neck pain. These patients are also known to have associated abnormalities of the function of the autonomic nervous system. The purpose of this study is to assess the effect of reduction of the distortion of the brainstem and fusion of the occiput to the upper cervical spine could have an effect on the debilitating effect of the dysautonomia.

Methods

A prospective outcome study of patients undergoing intraoperative reduction of abnormal occipito-cervical relationships with distortion of the brainstem who presented with severe unremitting headaches and mechanical neck pain were selected for study. All patients had documented type 3 Ehlers Danlos Syndrome (hypermobility). They all had abnormal movement of the occipito-cervical junction, excess movement on flexion-extension MRI studies and improvement with use of

Results

Karnofsky scores improved significantly post operatively ($P < 0.05$). Pain was relieved or substantially improved in 63% of patients, palpitations were improved in 55%, Orthostatic intolerance improved in 52% and disabling chronic fatigue was improved in 48%. In total, 23/27 would definitely have gone through surgery in retrospect. All patients had normalizations of the cranial-axial angle via intraoperative reduction.

Conclusions

In this small series, pain, dysautonomia, and chronic fatigue syndrome responded to occipitocervical reduction and occipito-cervical fusion.

Learning Objectives

1. Describe the role of occipital-cervical stabilization in relieving dysautonomia caused by brainstem compression and strain.

References