

# Effect of Underlying Connective Tissue Disorders on Pediatric and Adolescent Chiari I Malformation Neurosurgical Patients: A National Inpatient Sample Analysis

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## BACKGROUND

Chiari malformation type I (CMI) is a relatively common neurosurgical finding in both pediatric and adult populations. However, the pathophysiology of CMI is multi-faceted, complex, and still poorly understood. Furthermore, the role of connective tissue disorders (CTDs) on craniocervical instability and CMI remains poorly characterized. Given the high likelihood of surgical complications commonly seen in children and adolescents with CTDs, the authors assessed CTD patients who underwent neurosurgical treatment of their CMI for demographics, presenting symptoms, comorbidities, and perioperative courses.

## METHODS

Patients with CMI admitted for suboccipital decompression or laminectomy from January 2008 to September 2015 were captured using the National Inpatient Sample (NIS). Information on patient demographics, comorbidities, and perioperative course was collected. Diagnoses and procedures were determined by ICD-9-CM and ICD-9-PCS codes, respectively. Descriptive and regression analyses as well as categorical subgroupings were performed in SPSS version 26 (SPSS Inc., Chicago, IL, USA).

## RESULTS

38,611 CMI patients, 362 of whom with CTD, were identified. CMI patients with CTD were more likely to be female ( $p<0.001$ ) and present during the teenage (ages 13-18;  $p=0.055$ ) or young adult (ages 19-35) years ( $p<0.001$ ). Compared to the CMI without CTD group, far fewer patients within the CTD group were young children (ages 0-12;  $p<0.001$ ), late adult age (36-65 years old), or geriatric age ( $>65$  years old;  $p<0.001$  &  $p=0.022$ , respectively). Despite the predominantly teenage and young adult age of patients with CTD and CMI, they have more chronic issues ( $p<0.001$ ): systemic comorbidities include asthma ( $p=0.006$ ), postural orthostatic tachycardia syndrome, cardiac dysrhythmias, gastroparesis, and elevated white blood cell count (all  $p<0.001$ ). Central nervous system (CNS) comorbidities include migraine, tethered cord, and urinary incontinence (all  $p<0.001$ ). They have increased arthropathy and joint instability, including craniocervical instability (all  $p<0.001$ ), as evidenced by increased concomitant posterior cervical fusion surgeries and application of cervical halo procedures (both  $p<0.001$ ) during the same inpatient stay. Patients with CTD had longer length of stay (6.6 vs. 4.0 days), more procedures during hospitalization, and increased hospitalization charges (\$87,811.2 vs. \$58,497.2; all  $p<0.001$ ).

## CONCLUSIONS

Patients with coexisting CTD and CMI are more likely to present with complex Chiari and associated CCI. Most are female, teenage, rather than young children, and present with systemic, CNS, and joint abnormalities. They have longer, more complicated, and more expensive hospitalizations. Understanding the pediatric population age subsets in which these coexisting conditions present, how to make these diagnoses, and the unique perioperative and post-surgical challenges these patients face allows neurosurgeons to utilize specific guidelines for this patient population to best prepare and achieve optimal outcomes.