Evaluation of the Need for Upfront Sub-Occipital Craniectomy at the Time of Initial Anterior Decompression in Patients with Platybasia and Concomitant Chiari 1 Malformation

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Introduction

- Disorders of the craniocervical junction (CCJ) are associated with high morbidity and reduced quality of life.[1]
- For irreducible CCJ dysfunction (CCJD) with anterior vectors of compression, the endoscopic endonasasl approach (EEA) has become increasingly utilized for achieving ventral decompression[1,2]
- A certain subset of patients with irreducible CCJD present with a concomitant Chiari Malformation (CM) and posterior vector of compression. Prior studies have suggested that this combination is more common than previously thought in patients identified with CM[3] Additionally, this triad of retroflexed dens, platybasia, and Chiari Malformation may represent its own distinct cluster of CCJ pathology[4]
- The optimal surgical treatment paradigm for patients with CCJD and concomitant CM (CCJD+CM) is unknown and has often been debated whether anterior decompression alone is sufficient or if anterior decompression also requires posterior suboccipital craniectomy[2]

Objectives

- The primary aim of this study is to analyze and characterize those subset of patients who present with both irreducible CCJD and Chiari Malformation.
- The secondary aim of this study is to determine the utility of upfront suboccipital craniectomy (SOC) at the time of index EEA for CCJD.

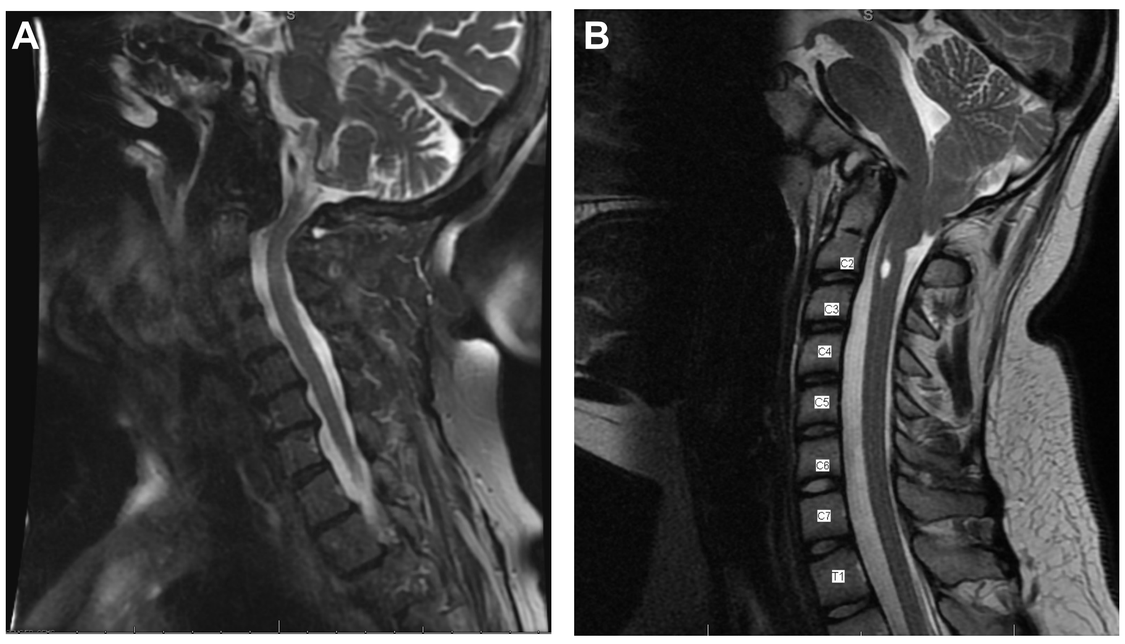
Methods and Materials

- A retrospective review of all patients with irreducible craniocervical junction pathologies from 2008 to 2023 at a single center was performed.
- Radiographic data, outcomes, and surgical approach were obtained.
- When applicable Chi-Square analysis and student's t-test analysis were used to assess for independence.
- Chiari Malformation was defined as tonsillar descent > 1 cm below the foramen magnum

Results

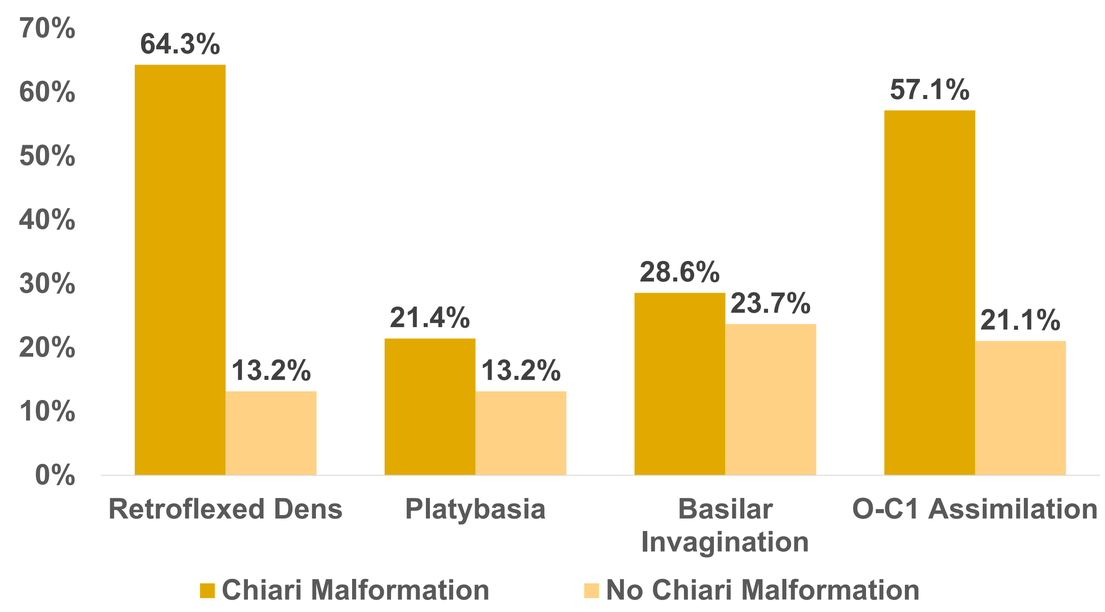
- 52 patients with irreducible CCJD were included in this study, of which 14 patients had CCJD plus concomitant CM (CCJD+CM, Fig. 1). The remaining 38 patients with irreducible CCJD without CM served as controls (Fig. 1)
- CCJD+CM was more common in younger patients (39.7 ± 21.22 v. 60.9 ± 23.9, p=.01).
- The most common presenting symptoms for patients with CCJD+CM were dysphagia (n=10; 71.4%), myelopathy (64.3%; n=9), and chiari-like symptoms (headaches and paresthesias; n=6, 42.9%)

Figure 1. Representative Images of Irreducible CCJD Either Alone or with Concomitant Chiari 1 Malformation



A. CCJD Alone with Rheumatoid Pannus **B.** CCJD+CM with Basilar Invagination, Retroflexed Dens, and Chiari Malformation

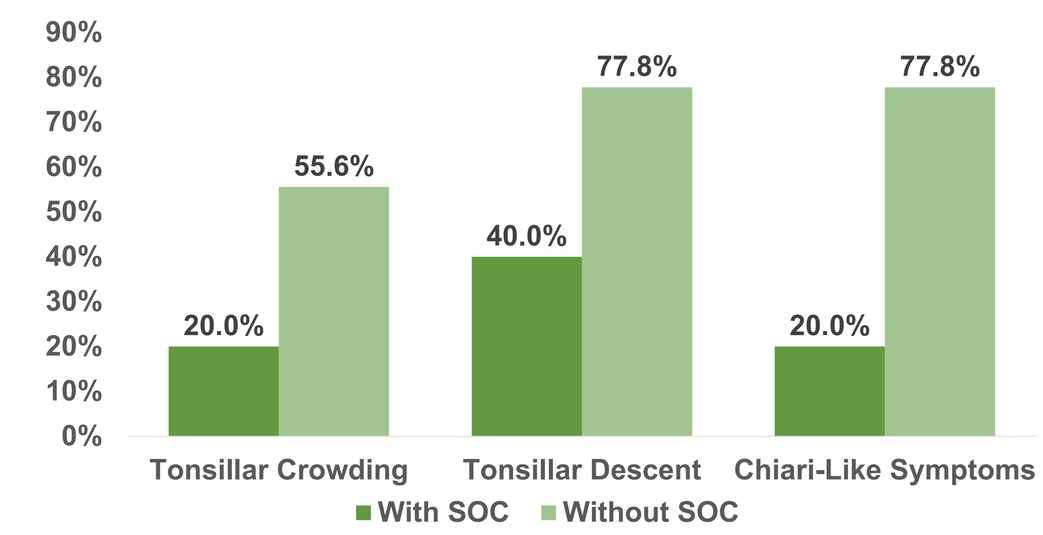
Figure 2. Anatomic Features of CCJ Dysfunction



Results

- Patients w/ irreducible CCJD+CM were significantly more likely to have retroflexed dens (p<.001) and O-C1 assimilation (p=.015)
- Two patients (14.3%) had undergone an SOC prior to presentation and 5 patients (35.7%) had an SOC at the time of their index EEA surgery.
- CCJD+CM patients with SOC either before or during their EEA had 100% rate of fusion including the occiput compared to just 33.3% of those without (p=.015)

Figure 3. Post-Operative Outcomes following EEA with or without Sub-Occipital Craniectomy (SOC)



Results

- Patients with irreducible CCJD+CM who underwent SOC at the time of surgery had lower rates of persistent radiographic tonsillar descent (p=.15), tonsillar crowding (p=.19), and chiari-like symptoms at last follow-up (p=.19) though these did not reach statistical significance
- No patients with CCDJ+CM who did not undergo SOC have been retreated for stenosis though 2 are currently being monitored and considered for SOC decompression

Conclusions

- The presence of irreducible CCJ Dysfunction with concomitant Chiari Malformation (CCJD+CM) appears to be linked to the presence of a retroflexed dens
- CCJD+CM patients who underwent SOC at the time of index surgery had lower rates of persistent radiographic and symptomatic features of CM compared to those who did not, though this did not reach statistical significance.
- All patients with suboccipital decompression at the time of initial surgery required fusion including the occiput
- Further studies are needed to determine whether primary posterior decompression is needed at the time of index ventral surgery for a circumferential decompression in patients presenting with irreducible CCJD+CM.

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