Prognostic significance of C1–C2 facet malalignment after surgical decompression in adult Chiari malformation type I: a pilot study based on the Chicago Chiari Outcome Scale

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OBJECTIVE The authors assessed the prognostic significance of various clinical and radiographic characteristics, including C1–C2 facet malalignment, in terms of surgical outcomes after foramen magnum decompression of adult Chiari malformation type I.

METHODS The electronic medical records of 273 symptomatic patients with Chiari malformation type I who were treated with foramen magnum decompression, C1 laminectomy, and duraplasty at Mayo Clinic were retrospectively reviewed. Preoperative and postoperative Neurological Scoring System scores were compared using the Friedman test. Bivariate analysis was conducted to identify the preoperative variables that correlated with the patient Chicago Chiari Outcome Scale (CCOS) scores. Multiple linear regression analysis was subsequently performed using the variables with p < 0.05 on the bivariate analysis to check for independent associations with the outcome measures. Statistical software SPSS version 25.0 was used for the data analysis. Significance was defined as p < 0.05 for all analyses.

RESULTS Fifty-two adult patients with preoperative clinical and radiological data and a minimum follow-up of 12 months were included. Motor deficits, syrinx, and C1–C2 facet malalignment were found to have significant negative associations with the CCOS score at the 1- to 3-month follow-up (p < 0.05), while at the 9- to 12-month follow-up only swallowing function and C1–C2 facet malalignment were significantly associated with the CCOS score (p < 0.05). Multivariate analysis showed that syrinx presence and C1–C2 facet malalignment were independently associated with the CCOS score at the 1- to 3-month follow-up. Swallowing function and C1–C2 facet malalignment were found to be independently associated with the CCOS score at the 9- to 12-month follow-up.

CONCLUSIONS The observed results in this pilot study suggest a significant negative correlation between C1–C2 facet malalignment and clinical outcomes evaluated by the CCOS score at 1–3 months and 9–12 months postoperatively. Prospective studies are needed to further validate the prognostic value of C1–C2 facet malalignment and the potential role of atlantoaxial fixation as part of the treatment.

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KEYWORDS facet malalignment; outcomes; Chiari malformation; foramen magnum decompression; syringomyelia; Chicago Chiari Outcome Scale; cervical

Chiari malformations are rare hindbrain anomalies with reported prevalence rates between 0.1% and 1%.1 To date, no consensus exists regarding the etiology or pathophysiological mechanism of Chiari malformation,2 although several theories have been proposed for its pathogenesis, including the instability theory.3–7 The most common subtype, Chiari malformation type I (CMI), is often identified during adulthood, and surgical treatment consists of foramen magnum decompression (FMD) with or without duraplasty to restore normal CSF flow.8
Various anomalies are frequently associated with this entity, including atlantoaxial instability, adding to the complexity in management and variability in surgical outcomes. Recently, atlantoaxial instability has been proposed as the triggering factor leading to tonsillar herniation, acting as a compensatory protective phenomenon to prevent spinal cord injury.

To effectively evaluate the postoperative outcomes, several outcome scales have been proposed in the literature. Among the available scales, the Chicago Chiari Outcome Scale (CCOS) has been objectively validated and has shown high reliability and prognostic value as justification for its use to evaluate the clinical outcomes of CMI patients during the postoperative course. 

C1–C2 cervical fusion, in addition to FMD, has been proposed as a feasible surgical strategy for the management of CMI when atlantoaxial instability is present. C1–C2 facet malalignment is defined as anterior or posterior atlantoaxial facet dislocation in the sagittal plane, and it is considered to be a key radiographic indicator of atlantoaxial instability.

To our knowledge, no previous studies have used a validated multifactorial outcome measurement to assess the relationship and influence of atlantoaxial malalignment on surgical outcomes after FMD. Therefore, the objective of this study was to determine the prognostic significance of various clinical and radiographic characteristics, including C1–C2 facet malalignment, in terms of post-FMD surgical outcomes assessed using the CCOS.

Methods

Patient Population

The Institutional Review Board of the Mayo Clinic approved this retrospective study before initiation (18-003951). The electronic medical records of 273 symptomatic patients with CMI who were treated with FMD, C1 laminectomy, and duraplasty at the Mayo Clinic were retrospectively reviewed. Exclusion criteria included the following: 1) age < 18 years; 2) lack of follow-up at 1–3 and 9–12 months; 3) CMI related to any gross structural brain or spinal malformation, hydrocephalus, infection, tumor, or connective tissue disorder; and 4) previous surgery-associated craniovertebral junction anomalies. In total, 52 consecutive adult patients with adequate preoperative clinical and radiological data and a minimum adequate follow-up of 12 months were included in this retrospective cohort study.

Foramen Magnum Decompression

All patients underwent suboccipital craniectomy with inclusion of the foramen magnum rim, C1 laminectomy, duraplasty, and reduction of the cerebellar tonsils using microsurgical techniques. Resection of the cerebellar tonsil tips was performed with a subpial technique or resection by coagulation, or a combination of both, based on the extent required to accomplish patency and complete decompression of the fourth ventricle. In some cases, patients presented with arachnoid scarring and adhesions that required a sharp dissection. Duraplasty was performed in all cases with pericranium or an artificial dural substitute.

Statistical Analysis

One neurosurgeon (M.L.L.) assessed patient records and imaging. Values for continuous variables are expressed as means with standard deviations and ranges, and frequency distributions are used to describe the categorical

Outcome Assessment

Preoperative and postoperative clinical symptoms were assessed for each patient using the NSS. The NSS score ranges from 0 to 5, with 5 representing normal function. Postoperative patient outcomes at the 1- to 3-month and 9- to 12-month follow-ups were also evaluated with the CCOS. 

The CCOS is a validated scoring system specific for CMI and grades postoperative outcome into 4 main components. Written records were converted into numerical values from 1 to 4 for each component and were summed to yield a total score of 4 to 16. The lowest score (4) represented complete postsurgical incapacitation and the highest score (16) represented excellent recovery.

Statistical Analysis

One neurosurgeon (M.L.L.) assessed patient records and imaging. Values for continuous variables are expressed as means with standard deviations and ranges, and frequency distributions are used to describe the categorical

Radiographic Characteristics

A uniform MRI protocol was followed for all CMI patients, with the patient’s neck kept in a neutral position. The radiological variables recorded on MRI included the length of tonsillar herniation, atlantodental interval (ADI), and presence of basilar invagination (BI), syrinx, and C1–C2 facet malalignment. Length of tonsillar herniation was defined by the perpendicular distance between the tip of the cerebellar tonsil and McRae’s line (basion-opisthion line). The ADI is a small slitlike space between the posterior aspect of the anterior atlas ring and the anterior aspect of the odontoid process. BI was defined as the odontoid crossing the Chamberlain line between the hard palate and posterior rim of the foramen magnum for ≥ 2.5 mm. Although atlantoaxial instability is presumed to be present in cases of normal C1–C2 facet alignments, no imaging studies, including dynamic studies, have been able to confirm this. For this reason, in this study C1–C2 facet malalignment was classified only into anterior and posterior dislocation or normal alignment (Fig. 1). Furthermore, shift of both facets to the same side, or shift of one facet without compensatory shift of the other joint to the contralateral side, was required to be considered facet malalignment in order to account for the shift of normal head rotation.

Preoperative Demographics and Clinical Presentations

Sex, age, and clinical symptom type and duration were extracted for the 52 included patients. Preoperative clinical symptoms were graded using the Neurological Scoring System (NSS) for the following symptom categories: 1) occipital headache, 2) neck pain, 3) motor deficits, 4) sensory deficits, 5) gait instability, 6) bowel/bladder weakness, and 7) swallowing function deficit. Patients were subsequently categorized into two groups according to the presence or absence of symptoms.
variables. The preoperative and postoperative NSS scores were compared using the Friedman test. Because CCOS scores were normally distributed, bivariate analysis (using the Spearman correlation test or Pearson correlation, depending on distribution) was conducted to identify the preoperative variables that correlated with the outcome measures (CCOS scores). For the variables with p values < 0.05 on the bivariate analysis, multiple linear regression analysis was subsequently performed to check for independent associations with the outcome measures. Statistical software SPSS version 25.0 (IBM Corp.) was used for the data analysis. Significance was defined as p < 0.05 for all analyses.

Results

Preoperative Demographics and Clinical Presentations

Fifty-two adult patients with preoperative clinical and radiological data and a minimum follow-up of 12 months were included. Demographic data and clinical presentation are presented in Table 1. The cohort consisted of 6 male and 46 female patients with a mean age of 40.8 ± 14.1 years (range 19–76 years). The median duration of symptoms was 36 months, with an IQR of 23–75 months and a range of 6–480 months. The most predominant preoperative complaint was headache (90.4%), followed by neck pain (86.5%) and gait instability (53.8%).

Radiographic Characteristics

Preoperative radiographic characteristics are presented in Table 1. There were 21 (40.4%) patients with BI, 7 (13.5%) with ADI > 3 mm, 34 (65.4%) with C1–C2 facet malalignment, and 15 (28.8%) patients with a syrinx, with a mean tonsillar herniation of 11.16 ± 5.52 mm.

Clinical Presentations at Follow-Up

Analysis of the predominant preoperative clinical features revealed that at last follow-up, occipital headache improved in 38 of 47 patients (81%), neck pain in 35 of 45 (78%), motor deficits in 6 of 11 (55%), sensory deficits in 14 of 24 (58%), gait instability in 20 of 28 (71%), bowel/bladder weakness in 4 of 4 (100%), and swallowing function in 5 of 6 patients (83%). There was a statistically sig-

<table>
<thead>
<tr>
<th>Variable</th>
<th>Value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age, yrs</td>
<td>40.8 ± 14.1</td>
</tr>
<tr>
<td>Sex</td>
<td></td>
</tr>
<tr>
<td>Male</td>
<td>6 (11.5)</td>
</tr>
<tr>
<td>Female</td>
<td>46 (88.5)</td>
</tr>
<tr>
<td>Symptom duration, mos, median (IQR)</td>
<td>36 (23–75)</td>
</tr>
<tr>
<td>Occipital headache</td>
<td>47 (90.4)</td>
</tr>
<tr>
<td>Neck pain</td>
<td>45 (86.5)</td>
</tr>
<tr>
<td>Motor deficits</td>
<td>11 (21.2)</td>
</tr>
<tr>
<td>Sensory deficits</td>
<td>24 (46.2)</td>
</tr>
<tr>
<td>Gait instability</td>
<td>28 (53.8)</td>
</tr>
<tr>
<td>Bowel/bladder weakness</td>
<td>4 (7.7)</td>
</tr>
<tr>
<td>Swallowing function</td>
<td>6 (11.5)</td>
</tr>
<tr>
<td>Tonsillar herniation, mm</td>
<td>11.16 ± 5.5</td>
</tr>
<tr>
<td>ADI, mm</td>
<td>2.32 ± 1.02</td>
</tr>
<tr>
<td>&lt;3</td>
<td>45 (86.0)</td>
</tr>
<tr>
<td>&gt;3</td>
<td>7 (13.5)</td>
</tr>
<tr>
<td>BI*</td>
<td></td>
</tr>
<tr>
<td>Absent</td>
<td>31 (59.6)</td>
</tr>
<tr>
<td>Present</td>
<td>21 (40.4)</td>
</tr>
<tr>
<td>Syrinx</td>
<td>15 (28.8)</td>
</tr>
<tr>
<td>C1–C2 facet malalignment (anterior &amp; posterior)</td>
<td>34 (65.4)</td>
</tr>
<tr>
<td>Anterior</td>
<td>2 (3.8)</td>
</tr>
<tr>
<td>Posterior</td>
<td>32 (61.6)</td>
</tr>
</tbody>
</table>

Values are expressed as the number of patients (%) unless indicated otherwise; mean values are given ± SD.

* BI was present when the tip of the odontoid process was ≥ 2.5 mm above Chamberlain’s line.
significant improvement in the NSS score for the majority of symptoms, with the exceptions of swallowing and bowel/bladder function (Table 2).

**Bivariate Analysis**

The results of bivariate analysis among the various clinical and radiological variables tested for associations with CCOS score are shown in Table 3. We found that motor deficits, syrinx presence, and C1–C2 facet malalignment had a significant (p < 0.05) negative association with CCOS scores at the 1- to 3-month follow-up, while the CCOS scores at the 9- to 12-month follow-up were negatively associated only with swallowing function and C1–C2 facet malalignment (p < 0.05). Based on these results, the other nonsignificant variables were excluded from further analyses, and motor deficits, syrinx presence, C1–C2 facet malalignment, and swallowing function were included in the multiple linear regression analysis.

**Multiple Linear Regression Analysis**

Multivariate analysis showed that syrinx presence and C1–C2 facet malalignment were independently associated with CCOS score at the 1- to 3-month follow-up, and swallowing function and C1–C2 facet malalignment were independently associated with CCOS score at the 9- to 12-month follow-up (Table 4). The ANOVA results and goodness of fit of the regression model showed significant simultaneous influences between syrinx presence, motor deficit, and C1–C2 facet malalignment and CCOS score at the 1- to 3-month follow-up (p = 0.003), as well as swallowing function and C1–C2 facet malalignment and CCOS score at the 9- to 12-month follow-up (p = 0.000) (Table 5).

**Complications**

There were 11 postoperative complications identified during follow-up: 4 (7%) cases with wound dehiscence and 5 (9%) cases with pseudomeningocele that resolved with conservative treatment. Only 2 (3.8%) patients developed persistent pseudomeningocele that required surgical repair.

**Discussion**

FMD with or without duraplasty is the most common procedure for the management of symptomatic CMI with the goal of restoring normal CSF flow. Despite vast surgical experience, outcomes after surgical intervention remain highly variable. According to the available literature, although approximately 98% of the patients report some symptomatic improvement 6 months after FMD, around 78% of patients still have persistent symptoms at 3 months, 71% at 1 year, and 68% at ≥ 2 years. Multiple factors contribute to the variability in clinical outcomes, especially the numerous anomalies associated with this disease, including atlantoaxial malalignment and instability. Consequently, failure to assess C1–C2 instability may...
result in persistence or recurrence of symptoms after surgical decompression.\textsuperscript{21,25} Here, we report the findings of a retrospective cohort study in which we used the CCOS score to evaluate the prognostic significance of both clinical and radiographic characteristics of patients with CMI after an FMD procedure at our institution.

Preoperative Demographics and Clinical Presentations

Chiari malformation is still categorized as a rare disease by the Office of Rare Diseases of the National Institutes of Health, and epidemiological data for this condition are limited.\textsuperscript{2} In our study, the mean patient age was 40.75 ± 14.08 years, which is within the age range reported in previous studies.\textsuperscript{14} In regard to clinical presentation, occipital headache (90.4%) was the most common presenting symptom, followed by neck pain (86.5%) and gait instability (53.8%) (Table 1), rates similar to those previously reported in the literature.\textsuperscript{21} Less frequent symptoms included motor deficits, sensory deficits, bowel/bladder dysfunction, and swallowing difficulty.

Radiographic Characteristics

Historically, multiple radiographic anomalies have been frequently described in conjunction with CMI, the most common being syringomyelia, scoliosis, BI, and atlantoaxial malalignment and C1–C2 instability.\textsuperscript{8} Incidence rates of BI vary in the literature from 9.2% to 84%.\textsuperscript{10,13,22,23} We identified BI in 21 (40.40%) of all patients presenting with CMI (Table 1). Multiple factors have been associated with the variability of incidence of BI, such as nutritional factors, childbirth-related trauma due to poor delivery practices,\textsuperscript{12} and the lack of strict diagnosis criteria for BI.\textsuperscript{24} C1–C2 facet malalignment is a strong radiographic indicator of atlantoaxial instability and has been associated with preoperative myelopathy, as well as development of postoperative myelopathy if the instability is not addressed.\textsuperscript{15,25,26} In our study, the prevalence of ADI < 3 mm was 86%, and posterior C1–C2 facet malalignment was dominant, affecting 61.56% of cases. These findings result from posterior facet malalignment coupled with dislocation of the facet of the atlas posterior to the facet of the axis.\textsuperscript{15}

Postoperative Follow-Up

Previously reported FMD failure rates range from 20% to 50%,\textsuperscript{20,27} while rates of persistent syringomyelia following FMD range from 0% to 40%.\textsuperscript{23} FMD failures may be due to the absence of a duraplasty or the presence of extensive scarring, sutures involving the obex, arachnoid web or residual C1 lamina, atlantoaxial instability, or myelopathy.\textsuperscript{16} Additionally, 20%–30% of patients do not attain a significant degree of neurological improvement after surgical decompression.\textsuperscript{15}

In the present study, we report postoperative improvements in occipital headaches (80.85%) and neck pain (77.78%), but as in previous studies, a large number of our patients had no improvement/changes in motor deficits (45.45%), gait instability (28.57%), and sensory deficits (no improvement in 37.50% and worsening in 4.17%) (Table 2). The lack of change in these deficits may have resulted from the presence of myelopathy, which can develop before or after surgery and is the factor considered to have the strongest impact on patient-perceived outcomes, along with headache type and syrinx size.\textsuperscript{24}

\begin{table}[ht]
\centering
\begin{tabular}{lccc}
\hline
Variable & CCOS Score (1–3 mos) & CCOS Score (9–12 mos) \\
& B & SE & p Value & B & SE & p Value \\
\hline
Motor deficits & 0.095 & 0.265 & 0.722 & NA & NA & NA \\
Swallowing function & NA & NA & NA & 1.100 & 0.435 & 0.015 \\
Syrinx & -1.023 & 0.482 & 0.039 & NA & NA & NA \\
C1–C2 facet malalignment & -1.231 & 0.406 & 0.004 & -1.784 & 0.509 & 0.001 \\
\hline
\end{tabular}
\caption{Multiple linear regression analysis with CCOS score}
\end{table}

\begin{table}[ht]
\centering
\begin{tabular}{lcccc}
\hline
Variable & CCOS Score (1–3 mos) & CCOS Score (9–12 mos) \\
& Sum of Squares & df & Mean Square & F & p Value & R² & Sum of Squares & df & Mean Square & F & p Value & R² \\
\hline
Regression & 31.309 & 3 & 10.436 & 5.401 & 0.003* & 0.252 & 58.690 & 2 & 29.345 & 9.647 & 0.000† & 0.283 \\
Residual & 92.749 & 48 & 1.932 & 1.932 & & & 149.060 & 49 & 3.042 & & & \\
Total & 124.058 & 51 & 2.075 & 1.932 & & & 207.500 & 51 & & & \\
\hline
\end{tabular}
\caption{ANOVA and goodness-of-fit analysis with respect to CCOS score}
\end{table}

B = unstandardized coefficient; NA = not applicable.

Boldface type indicates statistical significance (p < 0.05).

\* Predictors: motor deficits, syrinx, C1–C2 facet malalignment.

† Predictors: swallowing function, C1–C2 facet malalignment.
Bivariate Multiple Linear Regression Analyses

We found that in the bivariate analysis, preoperative features of motor deficits, syrinx, and C1–C2 facet malalignment have significant correlations with postoperative CCOS score at the 1- to 3-month follow-up; however, in the multivariate regression analysis motor deficits were not significantly associated with the CCOS score at the 1- to 3-month follow-up (p = 0.722) (Table 4). Furthermore, C1–C2 facet malalignment was the only variable to show a significant correlation to the CCOS score at both the 1- to 3-month and 9- to 12-month follow-ups in both bivariate and multivariate analyses (Table 4). Consequently, we recommend that the presence of atlantoaxial instability be assessed by looking for C1–C2 facet malalignment during the perioperative period in adult patients undergoing surgical decompression for CMI. The finding of atlantoaxial instability, in turn, may lead to further surgical considerations, including C1–C2 fusion, when atlantoaxial instability is identified as previously proposed by Goel.13

Limitations

This study has the inherent limitations of a retrospective analysis and is prone to errors due to inconsistent or inaccurate medical records. Additionally, only one unblinded author assessed patient records and imaging. Also, the variability between surgeons based on experience, skills, and preferred techniques should be accounted for, such as the size of craniotomy and type of duraplasty. The algorithm that we provided is based on the physician’s interpretations rather than the patient’s perception of improvement. While the CCOS score is a validated, multifactorial outcome measure for CMI, it has the drawback of being a retrospective, provider-based scoring system.10,11

Conclusions

The observed results in this pilot study suggest a significant negative correlation between C1–C2 facet malalignment and clinical outcomes evaluated by using the CCOS at 1–3 and 9–12 months postoperatively. Prospective studies are needed to further validate the prognostic value of C1–C2 facet malalignment and the potential role of atlantoaxial fixation as part of the treatment.

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References


**Disclosures**

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

**Author Contributions**

Conception and design: Loe, Vivas-Buitrago. Acquisition of data: Loe, Vivas-Buitrago. Analysis and interpretation of data: Loe, Vivas-Buitrago, Heemskerk, Tripathi. Drafting the article: Loe, Vivas-Buitrago, Domingo. Critically revising the article: Quinones-Hinojosa, Domingo, Heemskerk, Bendok, Bydon, Abode-Iyamah. Reviewed submitted version of manuscript: Quinones-Hinojosa, Domingo, Bendok, Bydon, Abode-Iyamah. Approved the final version of the manuscript on behalf of all authors: Quinones-Hinojosa. Statistical analysis: Heemskerk, Tripathi. Study supervision: Quinones-Hinojosa, Bendok, Bydon, Abode-Iyamah.

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