Predicting clinical outcomes using morphometric changes in adults with complex Chiari malformation undergoing occipitocervical fusion with or without ventral decompression: patient series

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BACKGROUND The authors assessed the connection between clinical outcomes and morphometrics in patients with complex Chiari malformation (CM) who have undergone posterior fossa decompression (PFD) and subsequent occipitocervical fusion (OCF) with or without ventral decompression (VD).

OBSERVATIONS The authors retrospectively reviewed 33 patients with CM aged over 21 years who underwent PFD and OCF with or without endoscopic endonasal odontoidectomy at the authors’ institution (21 OCF only and 12 OCF + VD). Clivoaxial angle (CXA), pB-C2 (perpendicular line to the line between the basion and C2), atlantodental interval (ADI), basion-dens interval (BDI), basion-axial interval (BAI), and C1 canal diameter were measured on preoperative and approximately 3-month postoperative computed tomography or magnetic resonance imaging scans. Common symptoms included headache, paresthesia, and bulbar symptoms. Clinical improvement after surgery was observed in 78.8% of patients. CXA, ADI, and BDI all significantly increased after surgery, whereas pB-C2 and BAI significantly decreased. OCF + VD had a significantly more acute CXA and longer pB-C2 preoperatively than OCF only. Patients who clinically improved postoperatively showed the same significant morphometric changes, but those who did not improve showed no significant morphometric changes.

LESSONS Patients showing improvement had greater corrections in skull base morphometrics than those who did not. Although there are various mutually nonexclusive reasons why certain patients do not improve after surgery, smaller degrees of morphometric correction could play a role.

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KEYWORDS Chiari malformation; craniocervical instability; occipitocervical fusion; retroflexed odontoid; odontoidectomy

Chiari malformation (CM) type 1 (CM 1) is radiographically characterized by herniation of the cerebellar tonsils below the foramen magnum by at least 5 mm.1 Such tonsillar herniation classically causes dorsal compression and obstruction of cerebrospinal fluid (CSF) flow, requiring posterior fossa decompression (PFD), which is widely accepted as the first-line surgical option.2

However, this dorsal pathophysiology is complicated in a subset of patients with CM 1 with coexisting connective tissue disorders (e.g., Ehlers-Danlos syndrome [EDS]), craniocervical instability (CCI), basilar invagination, and/or retroflexed odontoid.3–5 In addition to many of the classic CM 1 symptoms, such as exertional occipital headache, paresthesia, and ataxia, these complicated patients often present with signs and symptoms of ventral brainstem compression, such as dysphagia, respiratory problems, and dysautonomia.3,5

In such complex CM cases, occipitocervical fusion (OCF) with or without ventral decompression (VD; e.g., endoscopic endonasal

ABBREVIATIONS ADI = atlantodental interval; BAI = basion-axial interval; BDI = basion-dens interval; CCI = craniocervical instability; CM = Chiari malformation; CM 1 = Chiari malformation type 1; CSF = cerebrospinal fluid; CT = computed tomography; CXA = clivoaxial angle; EDS = Ehlers-Danlos syndrome; MRI = magnetic resonance imaging; OCF = occipitocervical fusion; pB-C2 = perpendicular line to the line between the basion and C2; PFD = posterior fossa decompression; POTS = postural orthostatic tachycardia syndrome; VD = ventral decompression.

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Morphometric Measurements

Six morphometric variables were considered and collected on preoperative mid-sagittal imaging (computed tomography [CT] or magnetic resonance imaging [MRI]) closest to the date of surgery and on imaging approximately 3 months after the surgery. These variables were CXA, pB-C2, atlantodental interval (ADI), basion-dens interval (BDI), basion-axial interval (BAI), and canal diameter at the level of C1. CXA is the angle between the slope of the lower clivus and a line along the posterior aspect of C2. pB-C2 is the line drawn perpendicularly to the dura from a line between the posterior aspect of the C2 base to the basion. ADI is the distance between the posterior aspect of the dens and the superior aspect of the dens. BDI is the distance between the inferior aspect of the dens. BAI is the line drawn perpendicularly to the basion from the posterior axial line. For the OCF + VD cohort, pB-C2, ADI, and BDI were collected only preoperatively due to resection of the dens. Measurements were made by a team composed of a neurosurgeon (N.J.M.) and a senior medical student (J.K.C.).

Preoperative Clinical Presentation

Out of 182 total adult Chiari surgical cases at our institution, we identified and reviewed 33 OCF cases, preoperative characteristics of which are summarized in Table 1. An example case is illustrated in Fig. 1. The mean age was 34.6 ± 8.8 years, and 90.9% were female. A formal diagnosis of EDS was carried by 57.6%. Overall, the most common presenting symptoms were headache (100.0%), paresthesia (90.9%), neck/shoulder pain (87.9%), and loss in daily functionality (87.9%). Bulbar symptoms were also prominent, and they included dizziness/vertigo (78.8%), dysphagia (63.6%), respiratory problems (57.6%), and tinnitus (57.6%).

Out of 33 cases, 21 were in the OCF-only cohort, and 12 were in the OCF + VD cohort. Although there were no statistically significant differences in preoperative clinical presentation between these 2 cohorts, the latter trended toward more postural orthostatic tachycardia syndrome (POTS) (41.7% versus 14.3%; p = 0.11) and general dysautonomia (75.0% versus 42.9%; p = 0.15).

Clinical Outcomes

The mean follow-up duration after surgery was 18.6 months (range 3.3–66.0 months), and postoperative changes in symptomatology are summarized in Table 2. Six patients had complete resolution of preoperative symptoms, 20 had improvement without complete resolution, 6 had relatively unchanged clinical outcomes (including those with transient improvement after surgery followed by return to baseline with symptom recurrence), and 1 had exacerbation of symptoms. Thus, overall, 78.8% (26 of 33) had favorable clinical outcomes. There were no significant differences in patterns of symptom changes between the OCF-only and OCF + VD cohorts. Of note, 73.7% of patients with EDS and 85.7% of patients without EDS in our cohort showed complete or partial resolution after surgery, but this difference was not statistically significant (p = 0.67), likely due to low power.

Complications

In the OCF-only cohort, 4 patients experienced postoperative wound infections requiring surgical washout, and 1 required an elective but same-hospital-stay surgical revision to address postoperative concerns of dysphagia. Out of 12 OCF + VD cases, 1 had
a surgical washout for wound infection, and another had screw mal-
placement requiring reoperation. The overall rates of postoperative
wound infection and revision were 15.2% and 6.1%, respectively.
Of note, all cases except for 1 requiring surgical washout had EDS.

Preoperative and Postoperative Morphometrics

Morphometric measurements are summarized in Table 3. Over-
all, the mean CXA, ADI, and BDI significantly increased after sur-
gery from 134.9° to 142.0° \((p = 0.002)\), 1.5 mm to 2.2 mm \((p =
0.002)\), and 4.7 mm to 6.2 mm \((p = 0.02)\), respectively. The mean
pB-C2 and BAI significantly decreased after surgery from 6.6 mm
to 5.1 mm \((p = 0.01)\) and 8.8 mm to 6.9 mm \((p = 0.003)\), re-
spectively. The mean canal diameter remained stable.

Between the OCF-only and OCF + VD cohorts, the latter preop-
eratively had a more acute mean CXA \((122.6° \text{ versus } 141.9°; p <
0.001)\) and a longer mean pB-C2 \((7.8 \text{ mm versus } 5.9 \text{ mm}; p =
0.01)\). Postoperatively, the mean CXA increased significantly in both cohorts
\((141.9° \text{ to } 146.8°; p = 0.044 \text{ in OCF only}; \text{ and } 122.6° \text{ to } 133.3°; p =
0.007 \text{ in OCF + VD})\). The mean BAI decreased in both cohorts, but
only the decrease in the OCF + VD cohort was statistically significant
\((10.1 \text{ mm to } 6.5 \text{ mm}; p = 0.01)\).

Associations Between Morphometrics and Clinical
Outcomes

In order to clinically contextualize these morphometric changes,
we further stratified the morphometric results into 2 groups based
on their overall clinical improvement (Table 4). The mean follow-up
periods were 18.5 months for the improved group and 19.2 months
for the nonimproved group, respectively. The previously noted mor-
phometric changes (increases in CXA, ADI, and BDI and decreases
in pB-C2 and BAI) were all present and statistically signi-
ficant \((N = 26)\). However, in the nonimproved group
\((N = 7)\), these postoperative morphometric parameters were rela-
tively unchanged.

When the morphometrics of the improved and nonimproved
groups were directly compared, the former was found, preopera-
tively, to have a significantly longer mean pB-C2 \((7.0 \text{ mm versus
5.1 mm}; p = 0.014)\) and BAI \((9.3 \text{ mm versus } 7.0 \text{ mm}; p = 0.048)\).

### TABLE 1. Preoperative characteristics

| Characteristics                              | All Adults N = 33 | OCF Without VD n = 21 | OCF With VD n = 12 | p Value |
|---------------------------------------------|-------------------|-----------------------|-------------------|---------|
| Mean age (± SD), yrs                        | 34.6 (8.8)        | 35.2 (8.6)            | 33.6 (9.5)        | 0.62    |
| F, n (%)                                    | 30 (90.9)         | 20 (95.2)             | 10 (83.3)         | 0.54    |
| Radiographic CM type, n (%)                 | 1                  | 1                     | 1                 | 1       |
| 1                                           | 30 (90.9)         | 19 (90.5)             | 11 (91.7)         |         |
| 1.5                                         | 3 (9.1)           | 2 (9.5)               | 1 (8.3)           |         |
| Associated conditions, n (%)                |                   |                       |                   |         |
| Syrinx                                      | 6 (18.2)          | 4 (19.0)              | 2 (16.7)          | 1       |
| EDS                                         | 19 (57.6)         | 14 (66.7)             | 5 (41.7)          | 0.27    |
| POTS                                        | 8 (24.2)          | 3 (14.3)              | 5 (41.7)          | 0.11    |
| Prior history of CM surgery, n (%)          | 18 (54.5)         | 12 (57.1)             | 6 (50.0)          | 0.73    |
| Preoperative symptoms, n (%)                |                   |                       |                   |         |
| Headache                                    | 33 (100.0)        | 21 (100.0)            | 12 (100.0)        | —       |
| Neck/shoulder pain                          | 29 (87.9)         | 19 (90.5)             | 10 (83.3)         | 0.61    |
| Back pain                                   | 16 (48.5)         | 9 (42.9)              | 7 (58.3)          | 0.48    |
| Extremity pain                              | 12 (36.4)         | 8 (38.1)              | 4 (33.3)          | 1       |
| Dysphagia                                   | 21 (63.6)         | 13 (61.9)             | 8 (66.7)          | 1       |
| Respiratory problems                        | 19 (57.6)         | 11 (52.4)             | 8 (66.7)          | 0.49    |
| Balance instability                         | 13 (39.4)         | 7 (33.3)              | 6 (50.0)          | 0.47    |
| Dizziness/vertigo                           | 26 (78.8)         | 17 (81.0)             | 9 (75.0)          | 0.69    |
| Muscle weakness                             | 17 (51.5)         | 10 (47.6)             | 7 (58.3)          | 0.72    |
| Decreased hearing                           | 9 (27.3)          | 7 (33.3)              | 2 (16.7)          | 0.43    |
| Tinnitus                                    | 19 (57.6)         | 12 (57.1)             | 7 (58.3)          | 1       |
| Paresthesia                                 | 30 (90.9)         | 19 (90.5)             | 11 (91.7)         | 1       |
| Visual symptoms                             | 20 (60.6)         | 13 (61.9)             | 7 (58.3)          | 1       |
| Urinary dysfunction                         | 13 (39.4)         | 9 (42.9)              | 4 (33.3)          | 0.72    |
| Loss of consciousness                       | 6 (18.2)          | 4 (19.0)              | 2 (16.7)          | 1       |
| Cognitive problems                          | 14 (42.4)         | 10 (47.6)             | 4 (33.3)          | 0.49    |
| Dysautonomia                                | 18 (54.5)         | 9 (42.9)              | 9 (75.0)          | 0.15    |
| Functionality loss                          | 29 (87.9)         | 18 (85.7)             | 11 (91.7)         | 1       |
Also, when their absolute changes in morphometrics were compared (Table 4), the improved group had significantly larger changes in CXA ($18.5^\circ$ versus $11.0^\circ$; $p = 0.047$), pB-C2 ($−1.2$ mm versus $+0.2$ mm; $p = 0.018$), and BAI ($−2.6$ mm versus $+0.9$ mm; $p = 0.019$).

### Discussion

**Observations**

In this study, we evaluated the relationship between clinical outcomes and degree of morphometric correction in post-PFD patients. The results indicated a significant correlation between improved morphometric changes and better clinical outcomes.

### Table 2. Changes in symptomatology after surgery

| Postoperative Symptoms         | Improved, n (%) | Nonimproved, n (%) |
|-------------------------------|-----------------|-------------------|
|                               | Complete | Partial | Unchanged | Worse | New |
| Overall symptomatology        | 6 (18.2) | 20 (60.6) | 6 (18.2) | 1 (3.0) | — |
| Headache                      | 5 (15.6) | 16 (50.0) | 10 (31.3) | 1 (3.1) | 0 (0.0) |
| Neck/shoulder pain            | 4 (14.3) | 12 (42.9) | 11 (39.3) | 1 (3.6) | 0 (0.0) |
| Back pain                     | 2 (12.5) | 8 (50.0) | 3 (18.8) | 2 (12.5) | 1 (6.3) |
| Extremity pain                | 2 (16.7) | 8 (66.7) | 2 (16.7) | 0 (0.0) | 0 (0.0) |
| Dysphagia                     | 6 (28.6) | 10 (47.6) | 4 (19.0) | 1 (4.8) | 0 (0.0) |
| Respiratory problems          | 5 (27.8) | 10 (55.6) | 1 (5.6) | 2 (11.1) | 0 (0.0) |
| Balance instability           | 4 (30.8) | 9 (69.2) | 0 (0.0) | 0 (0.0) | 0 (0.0) |
| Dizziness/vertigo             | 4 (16.0) | 12 (48.0) | 8 (32.0) | 1 (4.0) | 0 (0.0) |
| Muscle weakness               | 4 (25.0) | 7 (43.8) | 5 (31.3) | 0 (0.0) | 0 (0.0) |
| Decreased hearing             | 4 (44.4) | 5 (55.6) | 0 (0.0) | 0 (0.0) | 0 (0.0) |
| Tinnitus                      | 7 (36.8) | 9 (47.4) | 2 (10.5) | 1 (5.3) | 0 (0.0) |
| Paresthesia                   | 11 (37.9) | 11 (37.9) | 6 (20.7) | 1 (3.4) | 0 (0.0) |
| Visual symptoms               | 5 (26.3) | 7 (36.8) | 5 (26.3) | 2 (10.5) | 0 (0.0) |
| Urinary dysfunction           | 5 (41.7) | 4 (33.3) | 1 (8.3) | 2 (16.7) | 0 (0.0) |
| Loss of consciousness         | 4 (66.7) | 2 (33.3) | 0 (0.0) | 0 (0.0) | 0 (0.0) |
| Cognitive problems            | 3 (21.4) | 8 (57.1) | 2 (14.3) | 1 (7.1) | 0 (0.0) |
| Dysautonomia                  | 6 (35.3) | 8 (47.1) | 3 (17.6) | 0 (0.0) | 0 (0.0) |
| Functionality loss            | 5 (17.9) | 14 (50.0) | 8 (28.6) | 1 (3.6) | 0 (0.0) |

Complete = symptom completely resolved; New = new symptom that the patient did not experience preoperatively; Partial = symptom improved but not completely resolved; Unchanged = persistent symptom that either remained unchanged after surgery or transiently improved immediately after surgery, then came back to the baseline; Worse = symptom exacerbated.
with CM undergoing OCF for CCI and/or retroflexed odontoid. On the one hand, our most important finding was that there were significant short-term corrections in skull base morphometrics—namely, CXA, pB-C2, ADI, BDI, and BAI—in those reporting overall clinical improvement. On the other hand, there were no significant morphometric changes in those who did not improve overall.

In our series of 33 cases, there were 2 cohorts: OCF only and OCF + VD. Although our analysis was not adequately powered to detect more subtle differences between these cohorts, those who received VD in the form of endoscopic endonasal odontoidectomy were preoperatively deemed to have worse ventral brainstem compression. This assessment was consistent with our analysis showing that the OCF + VD cohort, on average, had significantly more acute CXA and longer pB-C2. Also, clinically, the latter at presentation had higher proportions of POTS and dysautonomia, both of which could be signs of ventral brainstem compression, though this difference was not statistically significant.

### Relationship Between Morphometrics and Pathophysiology

CXA has received attention in the literature as a key metric for assessing CCI and ventral brainstem compression. Anatomically, an acute CXA represents abnormal angulation between the clivus and the upper cervical spine, often obliterating the normal CSF space anterior to the ventral surface of the brainstem and causing mechanical compression. This neuralaxial stress on the brainstem can cause bulbar symptoms, which were common at presentation in our series. One of the goals of OCF is to increase the CXA, thereby relieving this ventral compression. Prior studies have reported relatively small samples of successful OCF cases showing improved clinical outcomes and corrections in CXA. Here, we observed postoperative clinical improvement associated with statistically significant changes in CXA.

pB-C2 measures the degree of ventral canal encroachment implicated by an acute CXA. When Grabb et al. first described pB-C2 using 40 patients with CM 1, they reported that those with pB-C2 < 9 mm were treated successfully with PFD without VD despite having preoperative signs and symptoms of ventral brainstem compression. Their findings set pB-C2 > 9 mm as a possible morphometric indication for VD, but recent studies have suggested that pB-C2 may not actually predict OCF with or without VD and that the utility of this particular cutoff is debatable. In our study, the mean preoperative pB-C2 in the OCF only cohort was 5.9 mm, though the mean preoperative pB-C2 of 5.9 mm was significantly lower, there was still a significant decrease after the surgery. These results suggest that pB-C2 is indeed an important metric to consider in the pathophysiology of CCI. However, the threshold of 9 mm is likely not absolute, because patients with pB-C2 < 9 mm can certainly benefit from VD if appropriately selected.

Compared with CXA and pB-C2, the other 3 significant parameters in our study—BAI, ADI, and BDI—have received little attention in the Chiari OCF literature. These metrics are used to evaluate for CCI because their abnormal values often reflect ligamentous
disruptions. The upper limits of normal are considered to be 12 mm for BAI, 2–3 mm for ADI, and 8.5–12 mm for BDI. According to these thresholds in the literature, the mean preoperative BAI, ADI, and BDI in our cohort were normal, but they still showed significant postoperative changes within the normal ranges. Without additional studies, it is unclear if significant changes in these parameters observed in our study have clinical relevance.

### Patient Selection for OCF Surgery

There are currently no universally accepted selection criteria for OCF in complex CM cases. There have been varying cutoffs for CXA, such as 125° and 135°, below which it should be deemed pathologic and consideration of OCF is warranted. The average preoperative CXA in our entire series was 134.9°, which fell at the 135° cutoff. However, when stratified by type of surgery, the mean CXAs of OCF only and OCF + VD were 141.9° and 122.6°, respectively. We showed that OCF can be successful both clinically and morphometrically in patients who do not meet these radiographic criteria.

| TABLE 4. Associations between morphometric and clinical outcomes |
|---------------------------------------------------------------|
| **Improved, Mean (± SD) Measurement, mm or °** | **Nonimproved, Mean (± SD) Measurement, mm or °** | **Mean Change** |
| **All cases** | **Preoperative** | **Postoperative** | **p Value** | **Preoperative** | **Postoperative** | **p Value** | **Improved** | **Nonimproved** | **p Value** |
| **CXA** | 134.4 (15.6) | 143.4 (13.0) | <0.001 | 136.5 (19.7) | 137.5 (12.9) | 1 | +8.5 | +1.0 | 0.047 |
| **pB-C2** | 7.0 (2.0) | 5.0 (1.5) | 0.003 | 5.1 (0.5) | 5.3 (1.1) | 0.63 | −1.2 | +0.2 | 0.018 |
| **ADI** | 1.5 (1.2) | 2.2 (1.0) | 0.02 | 1.4 (0.6) | 2.3 (0.8) | 0.06 | +0.8 | +0.9 | 0.86 |
| **BDI** | 4.6 (3.0) | 5.9 (2.2) | 0.04 | 4.9 (1.9) | 7.0 (3.2) | 0.31 | +1.5 | +2.0 | 0.87 |
| **BAI** | 9.3 (3.0) | 6.6 (1.7) | <0.001 | 7.0 (1.6) | 7.9 (2.5) | 0.58 | −2.6 | +0.9 | 0.019 |
| **Canal diameter** | 16.8 (2.1) | 16.8 (2.1) | 0.86 | 16.8 (1.5) | 16.8 (1.4) | 0.94 | +0.0 | +0.0 | 0.89 |
| **OCF only** | **n = 16** | **n = 5** | | | | | | | |
| **CXA** | 141.4 (13.1) | 148.4 (12.9) | 0.02 | 143.4 (14.8) | 142.1 (12.2) | 1 | +6.4 | −1.2 | 0.073 |
| **pB-C2** | 6.2 (1.3) | 5.0 (1.5) | 0.003 | 5.0 (0.4) | 5.3 (1.1) | 0.63 | −1.2 | +0.2 | 0.018 |
| **ADI** | 1.4 (1.1) | 2.2 (1.0) | 0.02 | 1.3 (0.6) | 2.3 (0.8) | 0.06 | +0.8 | +0.9 | 0.86 |
| **BDI** | 4.5 (2.8) | 5.9 (2.2) | 0.04 | 5.0 (2.2) | 7.0 (3.2) | 0.31 | +1.5 | +2.0 | 0.87 |
| **BAI** | 8.5 (2.7) | 6.9 (1.8) | 0.008 | 6.7 (1.1) | 7.6 (2.7) | 0.63 | −1.5 | +1.0 | 0.066 |
| **Canal diameter** | 16.7 (1.9) | 16.7 (2.2) | 0.65 | 17.4 (1.3) | 17.6 (0.3) | 0.63 | +0.0 | +0.2 | 0.97 |
| **OCF + VD** | **n = 10** | **n = 2** | | | | | | | |
| **CXA** | 123.2 (12.7) | 135.0 (8.2) | 0.008 | 119.3 (25.0) | 125.9 (4.9) | 1 | +12.1 | +6.6 | 0.73 |
| **pB-C2** | 8.3 (2.2) | — | — | 5.4 (0.8) | — | — | — | — | — |
| **ADI†** | 1.6 (1.4) | — | — | 1.4 (0.6) | — | — | — | — | — |
| **BDI†** | 4.8 (3.4) | — | — | 4.5 (1.4) | — | — | — | — | — |
| **BAI†** | 10.6 (3.2) | 6.0 (1.4) | 0.004 | 8.0 (2.7) | 8.7 (2.3) | 0.63 | −4.5 | +0.7 | 0.15 |
| **Canal diameter** | 16.9 (2.5) | 17.0 (2.0) | 0.73 | 15.3 (0.6) | 14.8 (0.4) | 0.63 | +0.0 | −0.5 | 1 |

* Pairwise tests were not performed for this subgroup, because there were only 2 cases.
† These 3 metrics were not obtained postoperatively after VD.
utility of these clinical elements in our decision-making algorithm. For such investigation, we would need to broaden the cohort to include nonsurgical patients with CCI and examine all preoperative variables, which could result in creating a useful score. This is the focus of ongoing and future studies.

Implications for the Utility of Intraoperative Imaging

Our results suggest that skull base morphometric parameters may be important to monitor intraoperatively to achieve a more accurate correction. For all of the patients included in this study, we used intraoperative fluoroscopy to guide and confirm appropriate hardware placement. For the past year, and thus for patients not yet included in this report, we have incorporated intraoperative CT both for screw placement and for alignment assessments. Past work suggested that intraoperative CT is reliable and could potentially reduce hardware-related complication rates in posterior cervical spine surgery by ensuring accurate hardware placement. However, the true utility of intraoperative morphometric measurements is unknown and warrants continued investigation.

Limitations

There were a number of limitations in this study. First, there is limited generalizability of our findings due to the retrospective, single-centered nature of the dataset, in which the outcome data ultimately relied on documentation and assessment of patient self-report at the clinic in the absence of any externally validated outcome tool for OCF ± VD in patients with CM. There was consequently room for bias. Second, given our small sample sizes, our study was underpowered in many statistical analyses, especially those involving stratified cohorts. Third, our radiographic data were short-term postoperative data, and they were derived from both CT and MRI with an assumption that measurements from these 2 modalities are not substantially different. Fourth, given the current lack of universally accepted criteria for selecting patients with CM for OCF, our institutional decision-making algorithm might have disproportionately included certain subpopulations—namely, patients with EDS—in our cohort, indirectly influencing the outcomes. Additional studies with larger cohorts are needed to better understand and differentiate changes in dynamic morphometric parameters between EDS and non-EDS patients with CM and CCI.

Lessons

In this study, the majority of patients with CM who were recommended OCF with or without VD reported overall symptomatic relief. Patients reporting improvement had statistically significant decreases in PedsQL score and improvements in Oswestry, neck-related NDI, and neck-related SF-36 components. However, those with no clinical improvement had insignificant changes in these parameters. Preoperative evaluation of potential OCF patients should involve both morphometric and clinical considerations. Although there are different reasons why certain patients with complex CM do not improve after surgery, our findings suggest that smaller degrees of morphometric correction might play a role in restricting maximal symptom amelioration.

References

1. Milhorat TH, Chou MW, Trinidad EM, et al. Chiari I malformation redefined: clinical and radiographic findings for 364 symptomatic patients. *Neurosurgery*. 1999;44(5):1005–1017.
2. Badie B, Mendoza D, Batzdorf U. Posterior fossa volume and response to suboccipital decompression in patients with Chiari I malformation. *Neurosurgery*. 1995;37(2):214–218.
3. Henderson FC Sr, Francomano CA, Koby M, Tuchman K, Adcock J, Patel S. Cervical medullary syndrome secondary to craniovertebral instability and ventral brainstem compression in hereditary hypomobility connective tissue disorders: 5-year follow-up after craniovertebral reduction, fusion, and stabilization. *Neurosurg Rev*. 2019;42(4):915–936.
4. Milhorat TH, Bolognese PA, Nishikawa M, McDonnell NB, Francomano CA. Syndrome of occipitooatlanxial hypomobility, cranial settling, and Chiari malformation type I in patients with hereditary disorders of connective tissue. *J Neurosurg Spine*. 2007;7(6):601–609.
5. Ridder T, Anderson RC, Hankinson TC. Ventral decompression in Chiari malformation, basilar invagination, and related disorders. *Neurosurg Clin N Am*. 2015;26(4):571–578.
6. Alaade AF, Ogando-Rivas E, Forbes J, et al. A dual approach for the management of complex craniovertebral junction abnormalities: endoscopic endonasal odontoidectomy and posterior decompression with fusion. *World Neurosurg*. 2019;2:100010.
7. Bello RJ, Riva-Cambrin J, Brockmeyer MM, Brockmeyer DL. Complex Chiari malformations in children: an analysis of preoperative risk factors for occipitocervical fusion. *J Neurosurg Pediatr*. 2012;10(2):134–141.
8. Grabb PA, Mapstone TB, Oakes WJ. Ventral brainstem compression in pediatric and young adult patients with Chiari I malformations. *Neurosurgery*. 1999;44(3):520–528.
9. CreveCoeur TS, Yahanda AT, Maher CO, et al. Occipital-cervical fusion and ventral decompression in the surgical management of Chiari-1 malformation and syringomyelia: analysis of data from the Park-Reeves Syringomyelia Research Consortium. *Neurosurgery*. 2021;88(2):332–341.
10. Tubbs RS, Iskandar BJ, Bartolucci AA, Oakes WJ. A critical analysis of the Chiari 1.5 malformation. *J Neurosurg*. 2004;101(2(suppl)):179–183.
11. Henderson FC Sr, Henderson FC Jr, Wilson WA 4th, Mark AS, Koby M. Utility of the clivo-axial angle in assessing brainstem deformity: pilot study and literature review. *Neurosurg Rev*. 2018;41(1):149–163.
12. Henderson FC, Wilson WA, Mott S, et al. Deformative stress associated with an abnormal clivo-axial angle: a finite element analysis. *Surg Neurol Int*. 2010;1:30.
13. Ladner TR, Dewan MC, Day MA, et al. Evaluating the relationship of the pB-C2 line to clinical outcomes in a 15-year single-center cohort of pediatric Chiari I malformation. *J Neurosurg Pediatr*. 2015;15(2):178–188.
14. Pang D, Nemzek WR, Zovickian J. Atlanto-occipital dislocation—part 2: the clinical use of (occipital) condyle-C1 interval, comparison with other diagnostic methods, and the manifestation, management, and outcome of atlanto-occipital dislocation in children. *Neurosurgery*. 2007;61(5):995–1015.
15. Rojas CA, Bertozzi JC, Martinez CR, Whitlow J. Reassessment of the craniovertebral junction: normal values on CT. *AJNR Am J Neuroradiol*. 2007;28(9):1819–1823.
16. De Paepe A, Malfait F. The Ehlers-Danlos syndrome, a disorder of connective tissue disorders: 5-year follow-up after craniovertebral reduction, fusion, and stabilization. *Neurosurg Rev*. 2019;42(4):915–936.
17. Raybaud C. Anatomy and development of the craniovertebral junction. *Neural Sci*. 2011;32(suppl 3):S267–S270.
18. Hecht AC, Koehler SM, Laudone JC, Jenkins A, Qureshi S. Is intraoperative CT of posterior cervical spine instrumentation cost-effective and does it reduce complications? *Clin Orthop Relat Res*. 2011;469(4):1035–1041.

Disclosures

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Acquisition of data: Chae, Marianayagam, Baaj. Analysis and interpretation of data: Greenfield, Chae, Marianayagam, Hussain, Baaj. Drafting the article: Chae, Hussain, Baaj. Critically revising the article: Greenfield, Chae, Marianayagam, Hussain, Baaj, Härtl. Reviewed submitted version of manuscript: all authors. Approved the final version of the manuscript on behalf of all authors: Greenfield. Statistical analysis: Chae. Study supervision: Greenfield.

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