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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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odontoidectomy[6]) is a potential surgical solution. Appropriate patient selection is crucial for this procedure, and prior studies have focused on improving selection criteria by investigating clinical and morphometric risk factors, such as CM 1.5, clivoaxial angle (CXA) $<$125–135°, and pB-C2 (Grabb-Oaks measurement; perpendicular line to the line between the basion and C2) $>$9 mm.[3,7–9] However, many of these studies had limited sample sizes, especially those including VD. Also, although independent changes in symptomatology and skull base morphometrics after surgery have been described in the literature, the actual relationship between postoperative clinical outcomes and degrees of morphometric correction has not been studied rigorously.

Here we report on a series of 33 adult OCF cases. Our primary objectives involved investigating (1) clinical outcomes after OCF; (2) postoperative morphometric changes; and, most important, (3) associations between the two.

Study Description

Once our study was approved by our institutional review board, we retrospectively reviewed all patients with CM 1 and 1.5[10] aged over 21 years who underwent PFD and OCF with or without endoscopic endonasal odontoidectomy at a single-center institution between December 2009 and February 2020. The senior neurosurgeon (J.P.G.) was the cosurgeon in every operation. We collected data on basic demographics, preoperative presentations, morphometric measurements, and postoperative outcomes.

In the absence of any universally accepted algorithm for VD, the decision to add odontoidectomy at our institution involves consideration of a number of clinical and radiographic variables. We generally defer odontoidectomy until clinical and radiographic improvement after reduction and fusion posteriorly has been assessed. In rarer cases, the most severe forms of ventral compression are planned and decompressed in a 2-stage surgery with posterior decompression and fusion followed by VD. These are for irreducible ventral compressions and constitute a minority of cases.

For clinical and morphometric characterization, we first analyzed the entire sample and then stratified it into OCF without VD (i.e., OCF-only cohort) and OCF with VD (i.e., OCF + VD cohort). Postoperative clinical improvement was assessed for overall symptomatology and each symptom category. Specifically, we assessed a comprehensive symptom list before and after surgery and whether these symptoms, as reported by the patient at the clinic, completely resolved, partially resolved, remained unchanged, developed, or worsened after surgery. These clinical data were derived from medical chart review and prior clinic note documentation, which had been written by the surgeons and their clinical staff, including physician assistants and nurse practitioners. In order to analyze associations between clinical and morphometric outcomes, we further stratified the entire sample by overall clinical improvement: improved (completely or partially resolved) and not improved (unchanged or worsened).

Morphometric Measurements

Six morphometric variables were considered and collected on preoperative midsagittal 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 C1 anterior arch and the anterior aspect of the dens. BDI is the distance between the inferior aspect of the basion and the superior 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.).

Statistical Analysis

All statistical analyses were performed using R version 4.0.0 with RStudio version 1.2.5042. Fisher's exact test for count data was used for categorical variables, and the Mann-Whitney *U* test and Wilcoxon signed-rank test were used for unpaired and paired continuous variables, respectively. Statistical significance was defined with an alpha level of 0.05.

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

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	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

a surgical washout for wound infection, and another had screw malplacement 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. Overall, the mean CXA, ADI, and BDI significantly increased after surgery 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 ($p = 0.003$), respectively. The mean canal diameter remained stable.

Between the OCF-only and OCF + VD cohorts, the latter preoperatively had a more acute mean CXA (122.6° versus 141.9°; $p < 0.001$) and a longer mean pB-C2 (7.8 mm versus 5.9 mm; $p = 0.01$). Postoperatively, the mean CXA increased significantly in both cohorts (141.9° to 146.8°; $p = 0.044$ in OCF only; and 122.6° to 133.3°; $p = 0.007$ in OCF + VD). The mean BAI decreased in both cohorts, but only the decrease in the OCF + VD cohort was statistically significant (10.1 mm to 6.5 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 morphometric changes (increases in CXA, ADI, and BDI and decreases in pB-C2 and BAI) were all present and statistically significant in the improved group (N = 26). However, in the nonimproved group (N = 7), these postoperative morphometric parameters were relatively unchanged.

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

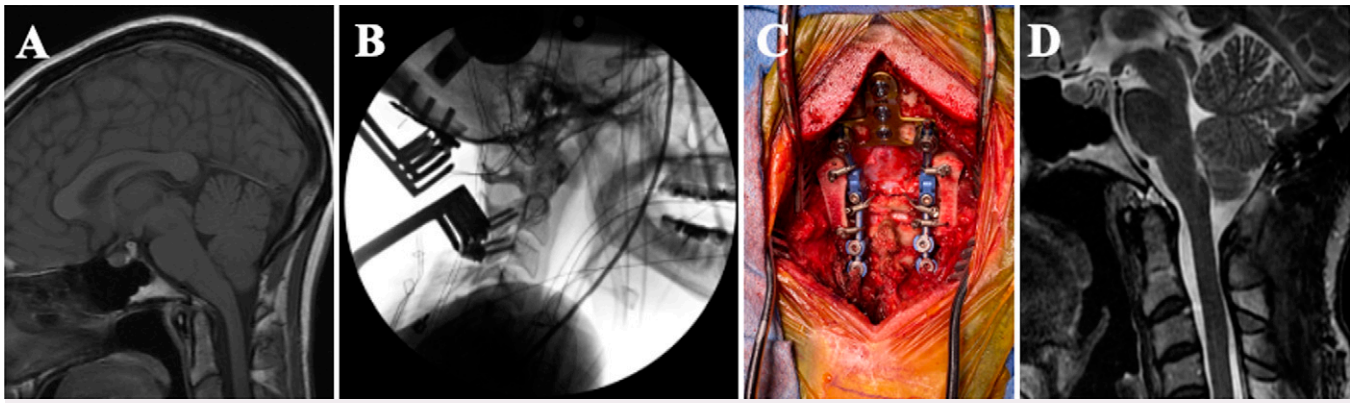

FIG. 1. Illustrative sample case. A 37-year-old woman with a history of CM presented with 3 years of progressive symptomatology, including occipital headaches, neck pain, visual disturbances, vertigo, tinnitus, dysphagia, paresthesias, and problems with thinking. **A:** A preoperative midsagittal T1-weighted MRI scan showed a tonsillar herniation of 7.1 mm, CXA of 126.9°, and pB-C2 of 9.1 mm. She underwent an extradural PFD with OCF (occiput to C3). **B:** An intraoperative radiograph showed correct hardware placement. **C:** An intraoperative photograph showing the occiput-to-C3 construct (taken from a different patient undergoing the same surgery). **D:** A 3-month postoperative midsagittal MRI scan showed adequate decompression and improvements in CXA to 151.2° and pB-C2 to 6.1 mm. The patient reported overall improvement in her preoperative symptoms after surgery.

Also, when their absolute changes in morphometrics were compared (Table 4), the improved group had significantly larger changes in CXA (+8.5° versus +1.0°; 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

TABLE 2. Changes in symptomatology after surgery

	Improved, n (%)		Nonimproved, n (%)		
Postoperative Symptoms	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.

TABLE 3. Preoperative and postoperative morphometrics

	Preoperative Mean (± SD) Measurement, mm or °	Postoperative Mean (± SD) Measurement, mm or °	p Value
All cases (N = 33)			
CXA	134.9 (16.2)	142.0 (13.0)	0.002
pB-C2	6.6 (1.9)	5.1 (1.4)	0.01
ADI	1.5 (1.1)	2.2 (0.9)	0.002
BDI	4.7 (2.8)	6.2 (2.4)	0.02
BAI	8.8 (2.9)	6.9 (1.9)	0.003
Canal diameter	16.8 (2.0)	16.8 (1.9)	0.87
OCF only (n = 21)			
CXA	141.9 (13.2)*	146.8 (12.7)	0.044
pB-C2	5.9 (1.3)†	5.1 (1.4)	0.01
ADI	1.4 (1.0)	2.2 (0.9)	0.002
BDI	4.6 (2.6)	6.2 (2.4)	0.02
BAI	8.1 (2.5)	7.1 (2.0)	0.18
Canal diameter	16.9 (1.7)	16.9 (1.9)	0.51
OCF with VD (n = 12)			
CXA	122.6 (13.8)*	133.3 (8.4)	0.007
pB-C2‡	7.8 (2.3)†	—‡	—‡
ADI‡	1.6 (1.3)	—‡	—‡
BDI‡	4.8 (3.1)	—‡	—‡
BAI	10.1 (3.2)	6.5 (1.8)	0.01
Canal diameter	16.6 (2.4)	16.6 (2.0)	0.52

* The mean preoperative CXAs were significantly different ($P < 0.001$).
† The mean preoperative pB-C2 lengths were significantly different ($P = 0.01$).
‡ These 3 metrics were not obtained postoperatively after VD.

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.[3,11,12] 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 neuraxial stress on the brainstem can cause bulbar symptoms,[12] 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.[3,11,12] 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.[8] 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.[7,9,13] In our study, the mean preoperative pB-C2 in the OCF + VD cohort was 7.8 mm (8.3 mm in improved versus 5.4 mm in nonimproved). In the OCF-only cohort, 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

TABLE 4. Associations between morphometric and clinical outcomes

	Improved, Mean (± SD) Measurement, mm or °			Nonimproved, Mean (± SD) Measurement, mm or °			Mean Change		
	Preoperative	Postoperative	p Value	Preoperative	Postoperative	p Value	Improved	Nonimproved	p Value
All cases		N = 26			N = 7				
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)		+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)		−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)		+0.0	−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.

disruptions.[14] 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.[15] 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°[7,9] and 135°,[3,11,12] 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.

No one morphometric parameter can select a patient for OCF with or without VD. At our institution, in addition to morphometrics and subjective symptomatology, we consider history of prior Chiari surgery, ligament laxity (e.g., EDS), POTS, abnormal movement on flexion-extension imaging, and the subjective response to a cervical collar trial in our evaluation algorithm.

EDS is a rare, heritable group of connective tissue disorders characterized by abnormalities in collagen biosynthesis, leading to tissue fragility and hypermobility of the skin and joints.[16] At the craniovertebral junction, where support provided by the ligaments is crucial for its overall integrity,[17] EDS can cause ligament laxity and, consequently, functional cranial settling and CCI.[3,4] There is a well-recognized association between connective tissue disorders such as EDS and CM, supported by family history data, but the true pathophysiological mechanism has not been completely elucidated.[4] Clinically, patients with EDS also have poor wound healing, so they are predisposed to postoperative complications, as noted in our study.

As part of the decision-making algorithm, we consider the patient's clinical response to a preoperative cervical collar trial, which simulates the effects of OCF without permanent fixation. However, the present study was not designed to investigate the

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.[18] 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 corrections in select skull base morphometrics. 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.

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Disclosures

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Conception and design: Greenfield, Chae, Marianayagam, Baaj. 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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