Increase in clivo-axial angle is associated with clinical improvement in children undergoing occipitocervical fusion for complex Chiari malformation: patient series

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BACKGROUND The authors analyzed the pre- and postoperative morphometric properties of pediatric patients with complex Chiari malformation undergoing occipitocervical fusion (OCF) to assess clinical outcomes and morphometric properties that might influence postoperative outcomes.

OBSERVATIONS The authors retrospectively reviewed 35 patients younger than 22 years with Chiari malformation who underwent posterior fossa decompression and OCF with or without endoscopic endonasal odontoidectomy at their institution (13 with and 22 without odontoidectomy). Clivo-axial angle (CXA), pB-C2, atlantodental interval, basion-dens interval, basion-axial interval, and canal diameter at the level of C1 were measured on preoperative and approximately 3-month postoperative computed tomography or magnetic resonance imaging. The authors further stratified the patient cohort into three age groups and compared the three cohorts. The most common presenting symptoms were headache, neck/shoulder pain, and dysphagia; 80% of the cohort had improved clinical outcomes. CXA increased significantly after surgery. When stratified into those who showed postoperative improvement and those who did not, only the former showed a significant increase in CXA. After age stratification, the significant changes in CXA were observed in the 7- to 13-year-old and 14- to 21-year-old cohorts.

LESSONS CXA may be the most important morphometric predictor of clinical outcomes after OCF in pediatric patients with complex Chiari malformation.

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KEYWORDS complex Chiari malformation; craniocervical instability; occipitocervical fusion; retroflexed odontoid; odontoidectomy; skull base morphometrics

Chiari 1 and Chiari 1.5 malformations are pathologies of the craniocervical junction commonly encountered in the practice of pediatric neurosurgery.^{1–3} The diagnostic criterion for Chiari malformation 1 is cerebellar tonsillar herniation of at least 5 mm below the level of the foramen magnum.^{1–4} Chiari malformation 1.5 also includes herniation of a portion of the brainstem, usually the obex.⁵ Both symptomatic pathologies are typically treated with posterior fossa decompression (PFD) with or without duraplasty.

Pediatric patients with complex Chiari malformation also usually have other associated pathologies of the craniocervical junction and spine.^{3,6}

These associated anomalies include scoliosis, basilar invagination, caudal regression syndrome, Klippel-Feil syndrome, atlantoaxial assimilation, and odontoid retroflexion.^{3,6} One of the unintended consequences of patients receiving PFD surgery is the creation of craniocervical instability (CCI),^{7,8} which can lead to neck pain with myelopathic symptoms that require surgical intervention in the form of occipitocervical fusion (OCF). CCI may be exacerbated in patients with underlying connective tissue disorders such as Ehlers-Danlos syndrome (EDS).⁹

Recent work by the Park-Reeves Syringomyelia Consortium prompted us to analyze morphometric properties of our own

ABBREVIATIONS ADI = atlantodental interval; BAI = basion-axial interval; BDI = basion-dens interval; CCI = craniocervical instability; CT = computed tomography; CXA = clivo-axial angle; EDS = Ehlers-Danlos syndrome; MRI = magnetic resonance imaging; OCF = occipitocervical fusion; PFD = posterior fossa decompression; VD = ventral decompression.

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patients who received both PFD and OCF.¹⁰ In this report we analyze the pre- and postoperative morphometric properties of 35 pediatric patients with complex Chiari malformation who received PFD surgery and OCF with or without ventral decompression (VD). We also analyze morphometric properties based on age stratification because the normal craniocervical junction changes as pediatric patients age.^{11–13} Our main goals in this study were to answer the following questions: (1) What are the clinical outcomes after OCF in pediatric patients with complex Chiari malformation? (2) Does age stratification make a difference in OCF surgery outcomes? (3) Are there morphometric properties that influence clinical outcomes?

Study Description

This was a retrospective study approved by our institutional review board. We reviewed the charts of all patients under the age of 22 years who underwent PFD and OCF with or without endoscopic endonasal odontoidectomy¹⁴ at our institution between August 2010 and February 2020. The senior neurosurgeon on the study (J.P.G.) was the lead surgeon involved in every operation. Data were collected on basic patient demographics, preoperative presentation, morphometric measurements, and postoperative outcomes. Postoperative clinical improvement was determined by assessing whether the patient's symptoms completely improved, partially improved, remained relatively unchanged, or worsened.

We first measured and collected all clinical and morphometric parameters across the entire cohort. We then stratified our data into OCF with VD and without VD. To look at the effect of morphometric properties on clinical outcomes, we further subdivided our patients into those who had symptomatic improvement after surgery and those who did not (improved versus not improved). Because of maturation of the craniocervical junction throughout childhood, we further stratified our analysis into three age-defined subgroups: 0 to 6 years old, 7 to 13 years old, and 14 to 21 years old.

We measured five morphometric parameters based on National Institutes of Health/National Institute of Neurological Disorders and Stroke Chiari malformation common data elements registry¹⁵ and analysis of the nonpathological pediatric craniocervical junction carried out by Bapuraj et al.¹¹ These parameters were pB-C2 line, atlantodental interval (ADI), basion-dens interval (BDI), basion-axial interval (BAI), and canal diameter at the level of C1. A sixth parameter (clivovaxial angle [CXA]) was added based on an analysis of pediatric patients with OCF by Bollo et al.¹⁶ The measurements were based on computed tomography (CT) and magnetic resonance imaging (MRI) carried out closest to the date of surgery and on imaging approximately 3 months after surgery. For the OCF and VD cohort, pB-C2, ADI, and BDI were collected only preoperatively because of absence of the rostral dens. Measurement of the CXA in postoperative patients with VD was made utilizing the dural margin of the resected dens. Measurements were made by a team of neurosurgeon (N.J.M.) and senior medical student (J.K.C.) who were blinded, at the time of radiographic measurement, to clinical presentation and outcomes. Both were trained by senior neuroradiology and neurosurgery attending physicians.

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. 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.

Cohort Summary and Preoperative Presentation

A total of 223 PFDs was carried out in the pediatric population at our institution. Of these patients, a total of 35 patients received OCF (22 patients had OCF without VD and 13 received OCF with VD). The demographic characteristics and clinical presentation of these patients are shown in Table 1. An illustrative case is shown in Fig. 1. The mean age was 11.7 ± 5.1 years; 42.9% were girls; and 57.1% carried a formal diagnosis of EDS. Overall, the most common presenting symptoms were headache (91.4%), neck/shoulder pain (65.7%), and dysphagia (60%). There were no statistically significant differences in presenting symptoms in patients with OCF alone or those who also received OCF plus VD. However, there was a statistically significant difference in age between patients receiving OCF only (10.2 ± 5.0) versus those receiving OCF with VD (14.3 ± 4.3).

Clinical Outcomes

The mean follow-up period after surgery was 21.3 ± 13.5 months. The postoperative changes in symptomatology are summarized in Table 2. Nine patients had complete resolution of preoperative symptoms, 19 had improvement without complete resolution, and 7 had no significant change in their symptoms after surgical intervention. None of the patients worsened after surgery. Overall, 80% (28/35) had favorable clinical outcomes.

Complications

A total of six patients, all in the OCF-only cohort, experienced complications. Two patients had superficial wound infections requiring antibiotics, two had wound infections requiring surgical washout, one had wound dehiscence requiring hardware removal, and one had instrumentation failure requiring revision (i.e., screws fixating the occipital portion of the construct into the skull had pulled out). Three of these cases were in the youngest 0- to 6-year-old cohort.

Changes in Morphometrics

The differences in morphometric properties between preoperative and postoperative patients are summarized in Table 3. There were statistically significant changes between the preoperative and postoperative CXA for all cases ($127.9 \pm 18.7^\circ$ versus $137 \pm 16.4^\circ$, $p = 0.003$). A significant difference was also observed in the preoperative and postoperative CXAs for the OCF with VD cohort ($111.3 \pm 14.5^\circ$ versus $125.4 \pm 13.2^\circ$; $p = 0.009$).

Morphometric Properties and Clinical Outcomes

The associations between morphometric properties and clinical outcomes are summarized in Table 3 for the various cohorts. Overall, there was a statistically significant change in CXA for patients who showed clinical improvement (127.5 ± 19.6 versus 137.9 ± 16.9 ; $p = 0.003$). There was also a significant difference in CXA for patients who underwent OCF + VD and showed clinical improvement (108.3 ± 12.5 versus 123.6 ± 14.6 , $p = 0.002$).

Age Stratified Changes in Morphometrics

Because of changes that occur in the maturing craniocervical junction, we stratified the data into three age groups. These data are summarized in Table 4. Statistically significant changes in BAI were observed for the 0- to 6-year-old cohort in all types of surgeries (4.6 ± 0.7 versus 7.2 ± 2.9 mm; $p = 0.03$). Also within the all surgeries cohort, there were significant changes between preoperative and

TABLE 1. Preoperative characteristics

Variable	Total N = 35	OCF w/o VD N = 22	OCF w/ VD N = 13	p Value
Mean age ($\pm$ SD)	11.7 (5.1)	10.2 (5.0)	14.3 (4.3)	0.02
Female, N (%)	15 (42.9)	11 (50.0)	4 (30.8)	0.31
Radiographic Chiari type, N (%)				0.59
1	29 (82.9)	19 (86.4)	10 (76.9)	
1.5	5 (14.3)	2 (9.1)	3 (23.1)	
2	1 (2.9)	1 (4.5)	0 (0.0)	
Associated conditions, N (%)				
Syrinx	8 (22.9)	4 (18.2)	4 (30.8)	0.43
EDS	20 (57.1)	14 (63.6)	6 (46.2)	0.48
POTS	8 (22.9)	6 (27.3)	2 (15.4)	0.68
Prior history of Chiari surgery, N (%)	24 (68.6)	15 (68.2)	9 (69.2)	1
Preop symptoms, N (%)				
Headache	32 (91.4)	21 (95.5)	11 (84.6)	0.54
Neck/shoulder pain	23 (65.7)	15 (68.2)	8 (61.5)	0.73
Back pain	16 (45.7)	11 (50.0)	5 (38.5)	0.73
Extremity pain	8 (22.9)	6 (27.3)	2 (15.4)	0.68
Dysphagia	21 (60.0)	13 (59.1)	8 (61.5)	1
Respiratory problems	18 (51.4)	11 (50.0)	7 (53.8)	1
Balance instability	14 (40.0)	9 (40.9)	5 (38.5)	1
Dizziness/vertigo	16 (45.7)	8 (36.4)	8 (61.5)	0.18
Muscle weakness	12 (34.3)	7 (31.8)	5 (38.5)	0.73
Decreased hearing	1 (2.9)	1 (4.5)	0 (0.0)	1
Tinnitus	15 (42.9)	11 (50.0)	4 (30.8)	0.31
Paresthesia	18 (51.4)	12 (54.5)	6 (46.2)	0.49
Visual symptoms	17 (48.6)	9 (40.9)	8 (61.5)	0.31
Urinary dysfunction	11 (31.4)	7 (31.8)	4 (30.8)	1
Loss of consciousness	3 (8.6)	3 (13.6)	0 (0.0)	0.28
Cognitive problems	8 (22.9)	5 (22.7)	3 (23.1)	1
Dysautonomia	13 (37.1)	8 (36.4)	5 (38.5)	1
Functionality loss	21 (60.0)	12 (54.5)	9 (69.2)	0.49

POTS = postural orthostatic tachycardia syndrome.

postoperative CXA in the 7- to 13-year-old cohort ($129.6 \pm 11.6^\circ$ versus $136.9 \pm 14.3^\circ$; $p = 0.02$) and the 14- to 21-year-old cohort ($116.8 \pm 19.2^\circ$ versus $134.2 \pm 18.1^\circ$; $p = 0.003$). In the OCF plus VD group, a significant difference in CXA was observed in the 14- to 21-year-old cohort ($109.3 \pm 16.6^\circ$ versus $126.3 \pm 16.2^\circ$; $p = 0.04$).

Discussion

Observations

In this study we have analyzed morphometric properties of the craniocervical junction in patients with complex Chiari receiving OCF with or without VD. We stratified our cohort of 35 patients into those receiving odontoidectomy and those without odontoidectomy. We also incorporated age stratification to address maturation of craniocervical junction. We found that 80% of all patients in our cohort had postoperative symptom improvement. There was a statistically significant change in CXA between pre- and postoperative patients across the entire cohort. A significant change in pre- and

postoperative CXA was also noted in patients receiving OCF with VD. In all patients who showed postoperative clinical improvement, there was a statistically significant increase in CXA. There was also a significant increase in CXA in patients showing postoperative clinical improvement in the OCF with VD cohort. In our age-stratified analysis, patients in the 0- to 6-year-old group across all types of surgery showed a statistically significant increase in BAI. In the 7- to 13-year-old and 14- to 21-year-old groups there was a significant increase in pre- and postoperative CXA across all types of surgeries.

Changes in CXA

It has been postulated that abnormally acute values for the CXA can lead to bulbar dysfunction and cervical medullary syndrome.^{9,17} The work of Bollo et al. identified various morphometric risk factors that predispose patients with complex Chiari malformation to OCF after receiving PFD surgery.¹⁶ In their multivariate analysis, they found that

FIG. 1. Illustrative sample OCF case. An 11-year-old boy with a history of EDS, central and obstructive sleep apnea (CSA, OSA), and Chiari malformation 1 status post-PFD 2 years ago presented with recurrence of preoperative symptoms (exertional headaches, photophobia, balance instability, blackout spells, and fatigue) and exacerbation of new symptoms (dizziness and breathing difficulty). **A:** A midsagittal T2-weighted MRI showed a CXA of 137° and a pB-C2 of 7.7 mm. For an extended period, the patient wore a hard cervical collar, which dramatically resolved his symptoms, and after much deliberation with the family, we performed a revision of PFD with OCF (occiput to C3). **B:** An intraoperative radiograph demonstrated appropriate hardware placement. **C:** A 3-month postoperative midsagittal T2-weighted MRI showed an increase in CXA to 152° and a decrease in pB-C2 to 6.2 mm. During his 31.3 months of postoperative follow-up, the patient reported clinical improvement and nearly complete resolution of all symptoms.

patients with CXA <125° were likely to need OCF. We corroborate this finding within our cohort of pediatric patients with complex Chiari malformation, which had an average preoperative CXA of 127.9 ± 18.7°. For patients requiring OCF plus VD, the average preoperative CXA

was 111.3 ± 14.5° and for those requiring only OCF it was 125.4 ± 13.2°. It should be noted that the average age of our cohort is slightly older than that reported in the work by Bollo et al. (11.7 versus 8.7 years). Our preoperative CXA values are also in statistical

TABLE 2. Changes in symptomatology after surgery

Postop Symptoms	Improved		Not Improved		
	Complete (%)	Partial (%)	Unchanged (%)	Worse (%)	New (%)
Overall symptomatology	9 (25.7)	19 (54.3)	7 (20.0)	0 (0.0)	
Headache	10 (31.3)	16 (50.0)	6 (18.8)	0 (0.0)	0 (0.0)
Neck/shoulder pain	7 (29.2)	11 (45.8)	5 (20.8)	0 (0.0)	1 (4.2)
Back pain	2 (10.5)	6 (31.6)	7 (36.8)	1 (5.3)	3 (15.8)
Extremity pain	0 (0.0)	2 (20.0)	5 (50.0)	1 (10.0)	2 (20.0)
Dysphagia	5 (23.8)	14 (66.7)	2 (9.5)	0 (0.0)	0 (0.0)
Respiratory problems	4 (22.2)	11 (61.1)	3 (16.7)	0 (0.0)	0 (0.0)
Balance instability	2 (14.3)	11 (78.6)	1 (7.1)	0 (0.0)	0 (0.0)
Dizziness/vertigo	5 (31.3)	7 (43.8)	4 (25.0)	0 (0.0)	0 (0.0)
Muscle weakness	3 (25.0)	6 (50.0)	2 (16.7)	1 (8.3)	0 (0.0)
Decreased hearing	0 (0.0)	1 (100.0)	0 (0.0)	0 (0.0)	0 (0.0)
Tinnitus	3 (20.0)	9 (60.0)	2 (13.3)	1 (6.7)	0 (0.0)
Paresthesia	6 (33.3)	7 (38.9)	5 (27.8)	0 (0.0)	0 (0.0)
Visual symptoms	4 (22.2)	10 (55.6)	3 (16.7)	0 (0.0)	1 (5.6)
Urinary dysfunction	1 (8.3)	5 (41.7)	4 (33.3)	1 (8.3)	1 (8.3)
Loss of consciousness	1 (33.3)	2 (66.7)	0 (0.0)	0 (0.0)	0 (0.0)
Cognitive problems	2 (25.0)	5 (62.5)	1 (12.5)	0 (0.0)	0 (0.0)
Dysautonomia	0 (0.0)	8 (61.5)	5 (38.5)	0 (0.0)	0 (0.0)
Functionality loss	6 (28.6)	8 (38.1)	6 (28.6)	1 (4.8)	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 and then came back to the baseline; Worse = symptom exacerbated.

Values expressed as number (%).

TABLE 3. Associations between morphometric and clinical outcomes

Parameter	Overall			Improved			Not Improved		
	Preop	Postop	p Value	Preop	Postop	p Value	Preop	Postop	p Value
All cases	N = 35			N = 28			N = 7		
CXA	127.9 (18.7)	137.0 (16.4)	0.003	127.5 (19.6)	137.9 (16.9)	0.003	129.9 (15.3)	134.1 (15.4)	0.58
pB-C2	5.7 (2.0)	5.0 (1.9)	0.87	5.9 (2.2)	5.0 (2.0)	0.74	5.0 (1.1)	4.9 (1.2)	1
ADI	2.5 (1.2)	2.9 (1.3)	0.18	2.5 (1.3)	3.0 (1.4)	0.11	2.5 (1.0)	2.6 (0.9)	0.59
BDI	4.5 (2.3)	6.3 (3.0)	0.18	4.6 (2.4)	6.5 (3.0)	0.23	3.9 (2.1)	5.4 (3.5)	0.63
BAI	7.7 (2.9)	7.5 (3.0)	0.91	8.0 (3.2)	7.5 (3.0)	0.84	6.5 (1.2)	7.6 (3.0)	0.81
Canal diameter	17.2 (2.2)	17.2 (2.0)	0.87	17.2 (2.2)	17.2 (2.1)	0.86	17.4 (2.1)	17.2 (1.8)	0.80
OCF only	N = 22			N = 18			N = 4		
CXA	137.7 (13.1)*	144.0 (14.2)	0.09	138.1 (13.9)	145.9 (12.2)	0.06	136.1 (10.2)	136.4 (20.8)	1
pB-C2	5.1 (1.4)	5.0 (1.9)	0.87	5.2 (1.5)	5.0 (2.0)	0.74	4.8 (1.1)	4.9 (1.2)	1
ADI	2.4 (0.7)	2.9 (1.3)	0.18	2.4 (0.6)	3.0 (1.4)	0.11	2.7 (1.1)	2.6 (0.9)	0.59
BDI	4.9 (2.3)	6.3 (3.0)	0.18	5.0 (2.3)	6.5 (3.0)	0.23	4.6 (2.6)	5.4 (3.5)	0.63
BAI	7.1 (2.1)	7.7 (2.9)	0.60	7.3 (2.3)	7.5 (2.9)	0.88	6.3 (1.3)	8.2 (3.6)	0.88
Canal diameter	17.3 (2.4)	16.9 (1.7)	0.19	17.3 (2.4)	17.0 (1.8)	0.28	17.2 (2.9)	16.7 (1.7)	0.63
OCF + VD	N = 13			N = 10			N = 3		
CXA	111.3 (14.5)*	125.4 (13.2)	0.009	108.3 (12.5)	123.6 (14.6)	0.02	121.5 (19.1)	131.0 (6.4)	0.50
pB-C2†	6.7 (2.5)			7.1 (2.7)			5.3 (1.2)		
ADI†	2.5 (1.8)			2.6 (2.0)			2.2 (1.1)		
BDI†	3.7 (2.2)			4.0 (2.5)			3.0 (1.0)		
BAI	8.7 (3.8)	7.2 (3.1)	0.58	9.3 (4.2)	7.4 (3.5)	0.55	6.7 (1.4)	6.8 (2.4)	1
Canal diameter	17.1 (1.8)	17.8 (2.3)	0.26	16.9 (2.0)	17.7 (2.6)	0.30	17.7 (1.0)	18.0 (1.9)	0.79

Values expressed as mean measurements in mm or degrees ($\pm$ SD).

* The mean preoperative CXAs of OCF and OCF with VD were significantly different ($p < 0.001$).

† These three metrics were not obtained postoperatively after VD.

agreement with those in the Park-Reeves study, with the average patient age in that report being 9.67 years.¹⁰

It is interesting to note that in our study the CXA changed significantly after surgery. It was the only morphometric property to exhibit statistically significant changes after surgery, with the exception of BAI within our age-stratified cohort of 0- to 6-year-old patients. We also observed a statistically significant increase in CXA for our patients who experienced postoperative clinical improvement. Biomechanically, this makes sense because a fusion of the OC junction would have the greatest effect on the CXA. Other authors have also shown that correction of the CXA by OCF in symptomatic patients after PFD is associated with clinical improvement.¹⁷ Indeed, the subset of our patients who exhibited clinical improvement after surgery showed a greater degree of correction in CXA.

Age-Stratified Analysis

Dynamics and morphometrics of the pediatric OC junction changes as a child ages.^{11–13} We felt it was important to look at age-stratified data when considering these properties in relation to CCI in patients with complex Chiari malformation who have received PFD and OCF. Thus, we stratified our cohort into three age groups. We observed a statistically significant increase in postoperative CXA in the all-surgeries group for the 7- to 13-year-old and 14- to 21-year-old cohorts. This was

not identified in the 0- to 6-year-old cohort. A significant increase in CXA was also seen in the 14- to 21-year-old cohort that underwent OCF plus VD.

In the 0- to 6-year-old group, a statistically significant increase in BAI was observed after surgery. A postoperative increase in BAI was not observed in all other cohorts. An analysis of the morphometric properties of the nonpathological craniocervical junction has shown that the BAI increases as a pediatric patient ages.¹¹ In a similarly aged cohort in this work, the normal average value of BAI was 4.6 mm,¹¹ which is the same as our preoperative value in this age range. Postoperatively, the BAI increased significantly to 8.2 mm, which could be attributed to increased mobility of the craniocervical junction in this age group.^{11–13} It has been observed and quantified that the greatest increase in BAI occurs in the first 4 years of life.¹¹ Interestingly, there is no statistically significant change in the CXA in the 0- to 6-year-old cohort that is observed in the other stratified age groups.

Limitations

There were several limitations in our study. First, this was a retrospective study using a single-centered database with a relatively short follow-up period and small sample size, so we were not adequately powered in more nuanced statistical comparisons. This analytical problem was particularly more pronounced when the sample was stratified by surgery type, age cohort, and clinical improvement. Second, in the

TABLE 4. Age-stratified changes in morphometrics

Parameter	0 to 6 Yrs			7 to 13 Yrs			14 to 21 Yrs		
	Preop	Postop	p Value	Preop	Postop	p Value	Preop	Postop	p Value
All cases	N = 7			N = 14			N = 14		
CXA	146.8 (13.1)	142.2 (18.3)	0.58	129.6 (11.6)	136.9 (14.3)	0.02	116.8 (19.2)	134.2 (18.1)	0.003
pB-C2	3.6 (0.8)	4.6 (1.2)	0.44	5.9 (1.4)	5.5 (2.4)	0.28	6.6 (2.3)	4.2 (1.1)	0.25
ADI	2.5 (0.3)	3.0 (0.5)	0.20	2.5 (0.6)	2.7 (1.7)	0.70	2.4 (1.9)	3.2 (0.9)	0.63
BDI	6.5 (2.2)	6.9 (1.4)	1	4.3 (2.1)	5.7 (4.1)	0.56	3.7 (2.2)	6.6 (1.0)	0.13
BAI	4.6 (0.7)	7.2 (2.9)	0.03	8.2 (1.8)	8.2 (3.1)	0.54	8.7 (3.5)	6.9 (3.0)	0.46
Canal diameter	15.8 (2.5)	15.9 (2.2)	0.81	17.6 (2.0)	17.9 (1.4)	0.95	17.6 (2.0)	17.3 (2.1)	0.97
OCF only	N = 6			N = 11			N = 5		
CXA	150.0 (10.7)	146.2 (16.4)	1	134.3 (7.6)	140.3 (14.5)	0.16	130.5 (16.9)	149.8 (9.8)	0.13
pB-C2	3.6 (0.9)	4.6 (1.2)	0.44	5.7 (1.3)	5.5 (2.4)	0.28	5.5 (1.0)	4.2 (1.1)	0.25
ADI	2.6 (0.3)	3.0 (0.5)	0.20	2.5 (0.7)	2.7 (1.7)	0.70	2.2 (1.0)	3.2 (0.9)	0.63
BDI	6.8 (2.2)	6.9 (1.4)	1	4.7 (2.2)	5.7 (4.1)	0.56	3.2 (1.4)	6.6 (1.0)	0.13
BAI	4.7 (0.7)	7.4 (3.1)	0.06	8.3 (1.9)	8.3 (3.4)	0.63	7.4 (1.0)	6.4 (1.4)	0.63
Canal diameter	15.7 (2.7)	15.9 (2.4)	0.84	17.4 (2.2)	17.4 (1.3)	0.38	18.9 (1.0)	17.4 (0.8)	0.13
OCF + VD	N = 1			N = 3			N = 9		
CXA	127.1	118.2		112.3 (5.0)	125.4 (3.4)	0.25	109.3 (16.6)	126.3 (16.2)	0.04
pB-C2*	3.2			6.4 (1.7)			7.2 (2.6)		
ADI*	2.3			2.7 (0.1)			2.5 (2.2)		
BDI*	4.7			3.0 (0.8)			3.9 (2.6)		
BAI	4.2	5.7		8.0 (1.8)	7.9 (2.5)	0.75	9.4 (4.2)	7.1 (3.7)	0.69
Canal diameter	16.4	16.4		18.0 (9.7)	19.4 (0.5)	0.25	16.9 (2.0)	17.3 (2.6)	0.64

Values expressed as mean measurements in mm or degrees ($\pm$ SD).

* These three metrics were not obtained postoperatively after VD.

absence of any validated tool for assessing outcomes after OCF with or without VD in pediatric patients, we relied on our clinical assessment, which inherently had room for bias. Third, our morphometric measurements were short-term, and they were based on both CT and MRI, depending on availability, with an underlying assumption that the two modalities would produce relatively similar measurements.

Lessons

In this study we measured the pre- and postoperative morphometric properties in 35 pediatric patients with complex Chiari malformation treated with PFD subsequently requiring stabilization via OCF. We found a statistically significant increase in CXA after surgery, particularly noted in the older pediatric age ranges. Age stratification showed statistically significant increases in the CXA for our older cohorts (7- to 13-year-old and 14- to 21-year-old groups) but not in our youngest cohort (0 to 6 years old). We also observed that children with the most robust postoperative clinical improvement had the greatest degree of surgical CXA correction. Our data suggest that CXA is crucial to assess and define preoperatively and intraoperatively to maximize clinical success because the postoperative change in CXA was the most important predictor of clinical outcomes after OCF in pediatric patients with complex Chiari malformation.

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Dr. Härtl reported personal fees from Depuy Synthes, Brainlab, and Zimmer Biomet and other from 3D Bio and RealSpine outside the submitted work.

Author Contributions

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

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